Anatomy

Question 10: Anatomy - Blood Supply of Brain/Spinal Cord

Discussion:

The medial medullary syndrome is most commonly associated with infarction in the anterior spinal artery distribution at the level of the medulla. An occlusion at this level may result in ipsilateral CNXII paralysis (fascicle of CNXII), contralateral hemiparesis (pyramid), and contralateral loss of position and vibratory sensation (medial lemniscus). A bilateral lesion in this vascular territory will result in quadriparesis, bilateral loss of proprioception and vibration, and complete paralysis of the tongue.

References:


Question 12: Anatomy - Cranial Nerves

Discussion:

The cavernous sinus, midbrain, pons, and superior orbital fissure do not contain segments of the optic nerve. The intraorbital portion of the optic nerve is contained in the cone formed by the ocular muscles. The apex of this cone is open to the optic foramen and the superior orbital fissure. The ophthalmic artery, ciliary ganglion and nerves, and the fascicles of cranial nerves III, IV, VI, and the ophthalmic branch of cranial nerve V are in close relation to the optic nerve at the orbital apex.

References:


Question 15: Anatomy - Cranial Nerves

Discussion:

Most of the parasympathetic neurons in the ciliary ganglion project fibers to the ciliary muscles to change the shape of the lens for accommodation. A small number of fibers project to the iris sphincter to constrict the pupil. While sympathetic fibers run through the ciliary ganglion, they do not synapse there. Sympathetic fiber activation result in pupillary dilatation.

References:


Question 19: Anatomy - Cortex and Connections

Discussion:

Damage to the fornix can occur with transcallosal surgery to remove a colloid cyst of the third ventricle, which interrupts the Papez circuit and results in loss of the ability to form new memories. The other structures, while important for memory, would not be affected by a transcallosal procedure.

References:

Question 27: Anatomy - Peripheral Nervous System

Discussion:

The posterior cord of the brachial plexus gives off the thoracodorsal and subscapular nerves and terminates by splitting into the axillary and radial nerves. Any muscles innervated by these branches may be weakened with a lesion in the posterior cord of the plexus.

References:


Question 29: Anatomy - Brainstem/Cerebellum

Discussion:

The findings are most consistent with a left dorsolateral medullary syndrome (Wallenberg syndrome) due to ischemia in the distribution of the posterior inferior cerebellar artery. Ipsilateral limb ataxia in this case is due to involvement of the left inferior cerebellar peduncle.

References:


Question 42: Anatomy - Spinal Cord

Discussion:

The lateral vestibulospinal tract facilitates extensor muscle tone, whereas the rubrospinal tract facilitates flexor tone. A lesion between the red nucleus and vestibular nuclei can result in decerebrate posturing.

References:


Question 68: Anatomy - Cortex and Connections

Discussion:

The circumventricular organs, which do not have a blood-brain barrier, are the area postrema, subfornical organ, organum vasculosum, neurohypophysis, median eminence, pineal gland, and subcommisural organ. The area postrema has been implicated as a chemoreceptor trigger zone for vomiting.

References:


Question 77: Anatomy - Basal Ganglia and Thalamus

Discussion:

The thalamus is supplied mainly by the branches of the posterior cerebral, posterior communicating, and posterior choroidal arteries.
Question 79: Anatomy - Blood Supply of Brain/Spinal Cord

Discussion:

A common variant of the circle of Willis is the persistent fetal origin of the posterior cerebral artery (PCA), when the PCA arises directly from the internal carotid artery.

References:


Question 81: Anatomy - Cranial Nerves

Discussion:

The trigeminal nerve supplies the muscles of mastication—the temporalis, masseter, medial and lateral pterygoids, mylohyoid, anterior belly of the digastric, tensor veli palatini, and tensor tympani muscles. The facial nerve supplies the frontalis, corrugator, orbicularis oculi, nasalis, buccinator, orbicularis oris, mentalis, and platysmas.

References:


Question 105: Anatomy - Cortex and Connections

Discussion:

The amygdala lies in the anterior pole of the temporal lobe, just deep to the uncus, and is an important structure in processing the emotional significance of stimuli, including pain. The caudate is the principal basal ganglia nucleus involved in processing oculomotor and prefrontal information. The nucleus accumbens is involved in anticipating reward and habit formation rather than the emotional component of pain. The hippocampus is part of the limbic system and is an essential component of memory formation. The pulvinar is an association nucleus in the thalamus involved in visual processing.

References:


Question 106: Anatomy - Cortex and Connections

Discussion:

The suprachiasmatic nucleus of the hypothalamus is the dominant circadian pacemaker of the mammalian brain. The intergeniculate leaflet and raphe nuclei mediate photic entrainment of the suprachiasmatic nucleus—light is the major entraining stimulus of the circadian system. The pineal gland is more important in seasonal rhythm control. The neurohypophysis does not have a major role in circadian pacemaking.
Question 109: Anatomy - Brainstem/Cerebellum

Discussion:

Axons from neurons in the nucleus gracilis and cuneatus cross the midline as the internal arcuate fibers in the caudal medulla, and project rostrally as the medial lemniscus.

References:


Question 120: Anatomy - Blood Supply of Brain/Spinal Cord

Discussion:

The great vein of Galen enters the dura of the tentorium and is joined by the inferior sagittal sinus to form the straight sinus (sinus rectus).

References:


Question 132: Anatomy - Basal Ganglia and Thalamus

Discussion:

Fibers from the anterior nucleus of the thalamus project to the cingulate gyrus. The anterior nucleus receives projections from the mamillary bodies. This is part of the classic Papez circuit that formed the basis of the concept of the limbic system.

References:


Question 135: Anatomy - Cortex and Connections

Discussion:

Gerstmann syndrome includes agraphia, finger agnosia, right-left disorientation, and acalculia. When all four features are present, the lesion is most often in the dominant (usually left) inferior parietal lobule, which includes the supramarginal and angular gyri.

References:


Question 136: Anatomy - Basal Ganglia and Thalamus

Discussion:
Multi-infarct dementia (MID) was a term replaced by vascular dementia (VaD) due to the recognition of the small strategically placed lesion that is capable of meeting the DSM IV-R criteria of dementia (i.e., a long- and short-term memory impairment, accompanied by a decline in another cognitive domain [i.e., executive, language/praxis, or visuospatial function] that significantly interferes with social and occupational function). The only lesion listed above that, with a size of 0.5 cm, is capable of producing a dementia is one in the left dorsomedial thalamus since it is capable of disrupting the encoding circuit (producing an amnesia) and also the frontal-subcortical circuits (producing profound executive and behavioral dysfunction).

References:


Kalashnikova LA, Gulevskaya TS, Kashina EM. Disorders of higher mental function due to single infarctions in the thalamus and in the area of the thalamofrontal tracts. Neurosci Behav Physiol 1999;29:3.

Question 147: Anatomy - Embryology

Discussion:

Neural crest derivatives include cells of the dorsal root ganglia (involved in transmission of pain) and sympathetic ganglia. These derivatives require nerve growth factor (NGF) for survival. Other neural crest derivatives are Schwann cells and melanocytes.

References:


Question 155: Anatomy - Peripheral Nervous System

Discussion:

The median nerve has motor branches to lumbricales 1 and 2, the abductor pollicis brevis, the opponens pollicis brevis, and the flexor pollicis brevis, distal to the carpal tunnel.

References:


Question 162: Anatomy - Blood Supply of Brain/Spinal Cord

Discussion:

Alexia, without agraphia, follows combined damage to the dominant medial occipital region and the inferior fibers of the splenium of the corpus callosum. This is in the distribution of the left posterior cerebral artery.

References:


Question 170: Anatomy - Peripheral Nervous System
Discussion:

The axillary nerve supplies sensation to the lateral part of the upper arm. Typically, a patient will have an "axillary patch" where sensation is diminished.

References:


Question 179: Anatomy - Basal Ganglia and Thalamus

Discussion:

The basal ganglion modulates motor activity. The indirect loop inhibits thalamocortical neurons, whereas the direct loop disinhibits (excites) thalamocortical projections. In each loop, the globus pallidus interna (GPi) projects inhibitory GABAergic projections to the thalamus. The caudate and putamen primarily project to GPi, globus pallidus externa (GPe), and substantia nigra pars reticulata (SNpr). SNpr also has GABAergic neurons projecting to the thalamus. The subthalamic nucleus has glutamatergic projections to SNpr and GPi.

References:


Question 190: Anatomy - Cortex and Connections

Discussion:

The anterior cingulate gyrus has an inhibitory influence on the micturition reflex and would be the most likely site of impairment.

References:


Question 193: Anatomy - Cranial Nerves

Discussion:

The abducens nerve emerges from the ventral surface of the brainstem at the junction between the pons and medulla. Cranial nerves VII and VIII also emerge at the pontomedullary interface, but they do so much more laterally in the cerebellopontine angle. All of the cranial nerves categorized as general somatic efferent (III, IV, VI, and XII) exit the brainstem medially.

References:


Questions 194 - 198: Anatomy - Peripheral Nervous System
## Discussion

The anterior rami of L2 to L4 give rise to the obturator nerve, whereas the dorsal divisions give rise to the femoral nerve. After all motor branches from the femoral nerve are given off in the thigh, the nerve continues to descend into the lower leg as the saphenous nerve, supplying cutaneous sensation to the medial leg region. Three cutaneous nerves are derived from the L1 dermatome: genitofemoral, ilioinguinal, and iliohypogastric nerves. The iliacus (femoral nerve), psoas (direct branches from Lumbar plexus), and tensor fascia lata (superior gluteal nerve) all flex the hip. The superficial peroneal nerve everts the foot. Only the TFL additionally internally rotates the thigh, and has an attachment to the iliotibial band. Sciatic mononeuropathies can clinically imitate common peroneal mononeuropathies because the peroneal portion of the sciatic tends to be predominantly affected due to fascicular vulnerability. An isolated common peroneal mononeuropathy will not cause hypo- or areflexia of the Achilles deep tendon reflex (DTR).

## References


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### Question 249: Anatomy - Basal Ganglia and Thalamus

## Discussion

The marked structure is the lateral geniculate nucleus, which receives visual input from the optic tract.

## References


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### Question 252: Anatomy - Brainstem/Cerebellum

## Discussion

The photograph shows a cross section of the brain stem at the superior pontine level. The locus ceruleus is identified by arrows bilaterally. The locus ceruleus is the major source of norepinephrine projections in the central nervous system.

## References


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### Question 289: Anatomy - Basal Ganglia and Thalamus

## Discussion

The arrow is pointing to the substantia nigra, which normally functions to facilitate voluntary motor activity originating in the prefrontal and motor cortex, ipsilateral to the substantia nigra. Damage to one substantia nigra results in hemiparkinsonism. Since the upper motor neurons of the motor strip are the motor output elements, and these cross at the medullary decussation, the hemiparkinsonism resulting from a unilateral substantia nigra lesion is contralateral to the lesion.

## References

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Question 316: Anatomy - Basal Ganglia and Thalamus

Discussion:

The ventral posterolateral nucleus projects to areas three, one, and two—the primary somesthetic area of the parietal lobe. The ventroposteromedial nucleus, that conveys facial sensation, also projects to the postcentral gyrus.

References:


Question 343: Anatomy - Spinal Cord

Discussion:

The dorsal and ventral spinocerebellar tracts are the most lateral tracts in the spinal cord and therefore would be expected to be affected first by an extrinsic lateral process. The lateral corticospinal tract lies just medial to the dorsal spinocerebellar tract, while the anterior corticospinal tract is in the anterior midline. The fasciculus gracilis is the medial aspect of the dorsal columns. The tectospinal pathway lies just anterior to the anterior commissure and the reticulospinal tract lies just anterior to the lateral corticospinal tract. The spinothalamic tracts run medial to the ventral spinocerebellar tract.

References:


Question 357: Anatomy - Cortex and Connections

Discussion:

The contralateral cerebellar hemisphere (in this case, the left cerebellar hemisphere) is engaged with motor tasks planned and executed from the opposite cerebral hemisphere. Activation of a cerebral cortical motor strip is accompanied by activation of the contralateral cerebellar hemisphere. Conversely, if the motor areas of a cerebral hemisphere are damaged, the contralateral cerebellar hemisphere shows a reduction in activity.

References:


Question 370: Anatomy - Cortex and Connections

Discussion:
In the retrochiasmatic visual pathways, homonymous hemianopias are most likely to be incongruous when caused by lesions of the optic tracts or lateral geniculate body, and more congruous in the optic radiations and occipital lobe. A lesion in an optic chiasm would cause a bitemporal or junctional visual field defect.

References:

Question 372: Anatomy - Brainstem/Cerebellum

Discussion:
Secondary auditory fibers from the cochlear nuclei form the dorsal, intermediate, and ventral acoustic striae. The dorsal and intermediate striae cross the midline and enter the lateral lemniscus. The fibers of the ventral stria terminate in the superior olivary nuclei and the nucleus of the trapezoid body. These nuclei give rise to tertiary fibers that enter the lateral lemnisci. The lateral lemniscus ascends to the midbrain where most of the fibers terminate in the inferior colliculi.

References:

Question 383: Anatomy - Cortex and Connections

Discussion:
The insular cortex receives visceral nociceptive input via the ventromedial posterior (VMP) thalamic nucleus.

References:

Question 384: Anatomy - Spinal Cord

Discussion:
The layers traversed during a lumbar puncture occur in the following order: skin, subcutaneous fat, supraspinous ligament, intraspinous ligament, ligamentum flavum, epidural fat, subarachnoid space. The pia mater and subpial space are located on the surface of the spinal cord, with the subarachnoid space located between the arachnoid mater and the pia mater.

References:

Question 385: Anatomy - Blood Supply of Brain/Spinal Cord

Discussion:
The middle meningeal artery enters the skull via the foramen spinosum. It is just lateral to the foramen ovale in the sphenoid bone.

References:
Question 388: Anatomy - Spinal Cord

Discussion:

The dorsal spinocerebellar tract arises from the dorsal nucleus of Clarke.

References:


Question 405: Anatomy - Cranial Nerves

Discussion:

Reactivation of the herpes zoster virus is a common neurologic complication in immunocompromised patients. The virus is latent in sensory ganglia, including the sensory ganglia of the facial nerve?the geniculate ganglia. Reactivation of the virus in this location produces facial paralysis, and occasionally hearing loss and vertigo, from viral spread to the 8th nerve. A vesicular rash is typically seen in the external acoustic meatus and lateral tongue.

References:


Question 406: Anatomy - Spinal Cord

Discussion:

The pattern of bilateral upper extremity weakness, affecting distal muscles more than proximal ones, with decreased pinprick sensation with retained fine touch, (ie, a dissociated sensory level in a "cape-like" distribution) is most suggestive of a central cord syndrome. A central cord lesion affecting the anterior commissure will affect the crossing spinothalamic tracts, resulting in sensory deficits only at the level of the lesion. It will not cause more caudal sensory deficits and will spare vibration and light touch, since the dorsal columns are spared.

References:


Question 421: Anatomy - Brainstem/Cerebellum

Discussion:

The patient has a crossed paresis, with right arm and leg weakness, as well as left facial paresis. This localizes to the left pons. The sensory deficits of fine touch on the right arm, trunk, and leg are due to involvement of the left medial lemniscus. The patient's diplopia, with left lateral gaze, is due to involvement of the left abducens nucleus.

References:

**Behavioral/Psychiatry**

**Question 2: Behavioral/Psychiatry - General Psychiatry**

**Discussion:**

Opioid intoxication is the only choice that is associated with constricted pupils. The other choices are associated with dilated pupils.

**References:**


**Question 4: Behavioral/Psychiatry - Neurobehavioral/Neuropsychological Exam**

**Discussion:**

The patient is unable to lay down new information and this is classic for an amnestic disturbance. She has retrograde and anterograde amnesia. While the event had the sudden onset like a stroke or TIA, to get amnesia, bilateral impairment is needed. Acute confusional states are manifested by impaired attention and would be inconsistent with a digit span of 6 forward. Partial complex seizures would also be manifested by poor attention and alertness and would not typically last 5 hours. Wernicke encephalopathy is usually associated with ophthalmoplegia and gait disturbance in the setting of an alcoholic.

**References:**


**Question 6: Behavioral/Psychiatry - Developmental Disorders**

**Discussion:**

Williams syndrome is a neurodevelopmental disorder caused by deletions on chromosome 7. Patients are generally very friendly with excellent social and language skills. Marked impairment is noted on tests of visual processing, affecting both the dorsal and ventral streams. Nonneurologic symptoms include cardiovascular abnormalities, hyperacusis, endocrine disorders, and hypercalcemia.

**References:**


**Question 35: Behavioral/Psychiatry - Dementia**

**Discussion:**

Subcortical dementia is characterized clinically by psychomotor slowing, forgetfulness, cognitive decline, visuospatial impairment, and personality changes, especially in mood.

**References:**
Question 51: Behavioral/Psychiatry - Anatomic syndromes

Discussion:

This syndrome is known as Klüver-Bucy syndrome and occurs as a result of bilateral temporal lesions involving amygdalae. Hypermetamorphosis, a common feature, involves excessive fascination with objects in the patient's environment and the urge to touch them all.

References:


Question 80: Behavioral/Psychiatry - Genes, Biomarkers, & Networks

Discussion:

This patient is presenting with a history that is suggestive of multiple system atrophy (MSA). This condition can manifest with parkinsonism, cerebellar dysfunction, or pyramidal tract signs. Most patients will invariably manifest some symptoms of autonomic dysfunction. Patients with MSA frequently have alpha-synuclein inclusions distributed throughout the cortex.

References:


Question 88: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:

This patient is presenting with the clinical syndrome of Hashimoto encephalitis. In this condition, TSH and T3/T4 levels are frequently normal; however, anti-microsomal antibody antibodies are typically elevated. This condition should be in the differential of any patient with a rapidly progressive dementia, especially in the presence of seizures. Hashimoto encephalitis responds well to intravenous corticosteroid treatment and plasmapheresis.

References:


Question 90: Behavioral/Psychiatry - Psychopharmacology

Discussion:

Patients with obsessive-compulsive disorder have been shown to respond to treatment with certain selective serotonin reuptake inhibitors (SSRIs), including fluoxetine and fluvoxamine, and that is first-line therapy.

References:


Question 91: Behavioral/Psychiatry - Developmental Disorders
Discussion:

The most common cause of inherited mental retardation is fragile X syndrome. Nearly all affected boys manifest attention deficit disorder and have learning disabilities. The most frequent neurocognitive symptoms are difficulty with abstract reasoning, complex problem solving, and expressive language. Many will also show manifestations of autism, with 33% meeting criteria for autism. Female carriers can have a milder form of the disease, with learning disabilities; about 50% of them will manifest attention deficit disorder. Characteristic physical features include a long thin face, prominent forehead and jaw, protuberant ears, hip dislocation, and club feet.

References:


Question 93: Behavioral/Psychiatry - Psychopharmacology

Discussion:

The antipsychotic therapeutic action of first generation antipsychotic agents is blockade of dopamine 2 receptors, specifically in the mesolimbic pathway. Unfortunately, first generation antipsychotics block every D2 receptor, as well as a wide range of other receptors throughout the brain. This includes blockade of dopamine 2 receptors in the mesocortical DA pathway, which can cause worsening negative and cognitive symptoms, the so called neuroleptic-induced deficit syndrome. Blockade of the alpha 1 adrenergic receptor can lead to decreased blood pressure, dizziness, and drowsiness. Blockade of the muscarinic cholinergic receptor can lead to constipation, blurred vision, and dry mouth, as well as drowsiness. Finally, blockade of the histamine 1 receptor can lead to weight gain and drowsiness as well. The first generation antipsychotics do not affect serotonin 1 receptors.

References:


Question 115: Behavioral/Psychiatry - Neurobehavioral/Neuropsychological Exam

Discussion:

The pattern of cognitive deficits, seen in patients with late life schizophrenia, is similar to those of more "typical" schizophrenia. In patients with a schizophrenia diagnosis, deficits are seen in 70% to 80% of these cases; with executive function, processing speed, attention, and working memory being the most commonly affected. The amnestic memory loss and other cortical deficits seen in Alzheimer disease are distinctly unusual and should prompt evaluation for other causes of the cognitive impairment.

References:

Palmer BW, Loughran CI, Meeks TW. Cognitive impairment among older adults with late-life schizophrenia or bipolar disorder. Continuum Lifelong Learning Neurol 2010;16:135-152.

Question 121: Behavioral/Psychiatry - Genes, Biomarkers, & Networks

Discussion:

The 14-3-3 protein is elevated in prion disorders, in contrast to most other dementing disorders. The protein elevation reflects neuronal destruction and so can be elevated in any disease that results in rapid neuronal death, such as trauma, stroke, or encephalitis. Classic findings of prion disorders include hyperekplexia (exaggerated startle), insomnia, encephalopathy, and ataxia.

References:

Question 128: Behavioral/Psychiatry - Anatomic syndromes

Discussion:

The thalamus is a major relay station for most inherent functions of the brain, to include cognition. This nuclear structure can be subdivided into regions based on functional relationships. Lesions in specific subnuclei can lead to different clinical manifestations. For example, lesions in the anterior group are more likely to manifest amnesia, confabulation, anomia and preserved visuospatial function. Paramedian thalamic lesions can manifest with acute decreased consciousness, followed by vertical gaze paresis, disinhibition, and at times amnesia.

References:


Question 131: Behavioral/Psychiatry - Psychopharmacology

Discussion:

The serotonin syndrome results from concomitant administration of medications that enhance serotonin transmission via decreased breakdown, or increased production. Medication combinations to use cautiously include monoamine oxidase inhibitor agents with selective serotonin reuptake inhibitors, tricyclic antidepressants, or dextromethorphan. The serotonin syndrome can be differentiated from neuroleptic malignant syndrome by the presence of shivering and myoclonus in the former.

References:


Question 134: Behavioral/Psychiatry - Genes, Biomarkers, & Networks

Discussion:

The child has manifestations of juvenile Huntington disease (HD). The adult onset form of the disorder is associated with greater than 40 CAG repeats. The gene location is 4p16.3. Like many trinucleotide repeat disorders, earlier onset is often reported in subsequent generations (anticipation), as the repeat expands.

References:


Question 140: Behavioral/Psychiatry - Genes, Biomarkers, & Networks

Discussion:

Presenilin-1 is the most common mutation found in patients with early-onset familial Alzheimer disease. The mutation is 100% penetrant; therefore, all patients who harbor this mutation will develop Alzheimer disease. The APOE protein is a stratifier of risk and does not guarantee development of disease. However, patients with an ε4 allele may be at higher risk and may have lower age of onset of disease.

References:


Question 144: Behavioral/Psychiatry - General Psychiatry

Discussion:

Panic disorder is manifested by recurrent panic attacks. Phobias are strong emotionally charged aversions to specific activities, places or things. If this patient is inordinately worried about having a car crash, it would be appropriate to diagnose her with having a specific phobia--amaxophobia is the fear of riding in cars. However, since her fears center around the possibility of having a panic attack, because she has had them before in a vehicle, she better qualifies for the diagnosis of panic disorder. If this woman is terrified of how other drivers will appraise her driving, then she would fit best under the definition of social phobia, but this is not apparent here. Claustrophobia is the fear of enclosed spaces, but there is no mention of other similar triggers (elevators, for example) for panic attacks in this patient. Generalized anxiety disorder may exist in this patient, but there is not enough information to support this.

References:


Question 151: Behavioral/Psychiatry - Neurobehavioral/Neuropsychological Exam

Discussion:

Apraxia refers to a disorder of learned movement that cannot be explained by deficits of weakness, sensory loss, or attention. There are two principal types of apraxia: (1) ideational, in which the patient cannot correctly pantomime a multistep command and (2) ideomotor, in which the patient cannot perform on command simple actions such as waving good-bye or using a hammer.

References:


Question 159: Behavioral/Psychiatry - Genes, Biomarkers, & Networks

Discussion:

Because of this patient's diagnosis of amnestic mild cognitive impairment (MCI) and his positive family history of Alzheimer disease, he has a high likelihood of progressing to Alzheimer disease. The Alzheimer's Disease Neuroimaging Initiative has established some useful biomarkers that help clinicians predict the chances of conversion from MCI to AD. Based on information obtained from this initiative and prior research, one of the earliest regions to demonstrate hypometabolism is the medial portion of the parietal cortex and the posterior cingulate. As the disease advances, the typical temporal/parietal pattern of hypometabolism is more evident.

References:


Question 166: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:

In patients with systemic lupus erythematosus, an autoimmune process with antineuronal antibodies may be an important cause of neuropsychiatric CNS involvement. Serum antiribosomal-P antibodies have a high specificity for patients with cerebral disease and neuropsychiatric symptoms of psychosis and depression, but do not correlate so well with cognitive symptoms. Correlations have not been identified for the other pairs of behavioral symptoms listed.

References:


Question 172: Behavioral/Psychiatry - General Psychiatry

Discussion:

Delusional disorder is differentiated from schizophrenia by the lack of bizarre delusions, absence of prominent auditory/visual hallucinations, and the absence of significant impairment in psychosocial functioning. The diagnosis of schizoaffective disorder requires the presence of either a depressive, manic, or mixed episode during the illness.

References:


Question 176: Behavioral/Psychiatry - Psychopharmacology

Discussion:

Anticholinergic side effects are common with tricyclic antidepressants, particularly amitriptyline. Trazodone, while related to amitriptyline chemically, is less likely to induce anticholinergic side effects. Venlafaxine may cause mild anticholinergic effects, but fluoxetine and buspirone are not prone to anticholinergic side effects.

References:


Question 180: Behavioral/Psychiatry - Anatomic syndromes

Discussion:

Misreaching under visual guidance (optic ataxia) and inability to scan and integrate a visual scene or picture (simultanagnosia) are symptoms of Balint syndrome. The third clinical symptom of Balint syndrome is ocular apraxia—the inability to stabilize eye movements. When these symptoms occur together, bilateral occipitoparietal lesions are implicated. Watershed infarction can cause these symptoms, as can multiple embolic strokes. Alzheimer disease can produce this and Balint syndrome has also been described in patients with frontal lesions.

References:


Question 191: Behavioral/Psychiatry - Psychopharmacology

Discussion:

Bromocriptine is a dopamine receptor agonist, which activates postsynaptic dopamine receptors. It is the treatment of choice for neuroleptic malignant syndrome, a life-threatening reaction to neuroleptic medication to which elderly patients have a relative vulnerability.

References:


Questions 204 - 208: Behavioral/Psychiatry - Language Disorders

Discussion:

Basic assessment of language disorders includes fluency, comprehension, repetition, confrontation naming, reading and writing. A characteristic of classic Broca, or expressive aphasia, is speech which is slow and nonfluent with preserved comprehension. In classic Wernicke aphasia or receptive aphasia, speech is spontaneous, but the ability to understand verbal and written information is impaired, and speech output can be senseless. Characteristics of conduction aphasia include fluent speech and good comprehension, but naming and repetition are impaired. Echolalia is a language disturbance seen in patients with transcortical sensory aphasia. Patients with early Alzheimer dementia often present with word-finding difficulty. Poor word deafness has poor verbal comprehension but with preservation of reading.

References:


Question 332: Behavioral/Psychiatry - Anatomic syndromes

Discussion:

Anosognosia, the inability to appreciate one's hemiparetic deficit, along with other neglect syndromes, may be seen with right frontoparietal lesions as this patient seems to have. Right-left confusion and alexia without agraphia generally result from dominant hemisphere lesions. Prosopagnosia results from lesions in occipital and/or temporal lobes. Acalculia results from lesions in the dominant hemisphere.

References:


Question 334: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:

Forced normalization refers to a psychosis occurring after achievement of good clinical seizure control, or resolution of interictal epileptiform discharges.

References:

Question 347: Behavioral/Psychiatry - Dementia

Discussion:

The dementia with Lewy bodies (DLB) consortium has revised criteria for the clinical and pathologic diagnosis of DLB, incorporating new information about the core clinical features and suggesting improved methods to assess them. REM sleep behavior disorder, severe neuroleptic sensitivity, and reduced striatal dopamine transporter activity on functional neuroimaging are given greater diagnostic weighting as features suggestive of a DLB diagnosis. When any of these features are present with one of the primary findings of visual hallucinations, parkinsonism, or fluctuating attention, then the diagnosis of probable DLB is supported.

References:


Question 361: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:

A Medline review found 32 cases of Wernicke encephalopathy after bariatric surgery reported (27 of whom were women), from 2 weeks to 18 months after the procedure. Most patients had vomiting as a risk factor (n = 25) and presented with the triad of Wernicke encephalopathy (confusion, ataxia, and nystagmus; n = 21). Optic neuropathy, papilledema, deafness, seizures, asterixis, weakness, and sensory and motor neuropathy were also reported. Characteristic radiographic findings were hyperintense signals in the periaqueductal gray area and dorsal medial nucleus of the thalamus; radiographs were normal in 15 patients. One series from Brazil reported 4 patients (among 50 patients) with Wernicke encephalopathy; all presented with vomiting and concomitant peripheral neuropathy at a median of 2.5 months (1.5 to 3.0 months) after bariatric surgery. Another series identified 2 of 23 patients (both women) with Wernicke encephalopathy after bariatric surgery. Wernicke encephalopathy after bariatric surgery usually occurs between 4 and 12 weeks postoperatively, especially in young women with vomiting. Atypical neurologic features are common. The diagnosis is mainly clinical because radiographic findings are normal in some patients.

References:


Question 368: Behavioral/Psychiatry - General Psychiatry

Discussion:

Both factitious disorder and malingering are characterized by assuming the sick role and the intentional production of symptoms. The major difference is that the malingerer’s production of symptoms is entirely motivated by external secondary gain, while patients with factitious disorder are motivated only by the emotional comfort of being seen as ill and needy. A history of emotional or other trauma is common in factitious disorder, but it may not be present; nor is this a way to distinguish it from malingering.

References:


Question 373: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease
Discussion:

Multiple sclerosis patients often develop pseudobulbar affect, associated with involuntary emotional displays (typically laughing or crying) that are often out of proportion to the patient's mood. While depression is common, bipolar illness is not. Obsessive-compulsive traits, visual hallucinations, and physical aggression are less often seen.

References:


Question 374: Behavioral/Psychiatry - Neurobehavioral/Neuropsychological Exam

Discussion:

The Wisconsin Card Sort test is a measure of frontal and executive functioning and would be significantly impaired in a patient with multiple sclerosis with prominent executive dysfunction. Demyelinating lesions would result in a subcortical dementia syndrome, which typically is characterized by a poor delayed memory recall, but a relatively preserved recognition memory. Since multiple sclerosis is a white matter disease, it would have less impact on cortical derived cognitive functions like naming objects, calculations, or language abilities.

References:


Question 377: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:

Nutmeg toxicity can lead to hallucinations, headache, nausea and vomiting, tachycardia and sensory changes in the extremities. Symptoms resolve in several hours. None of the other cooking ingredients listed as possible answers are known to cause neuropsychiatric symptoms.

References:


Question 380: Behavioral/Psychiatry - Dementia

Discussion:

The clinical criteria for frontotemporal dementia (FTD) include gradual onset with progressive decline, changes in personal hygiene, loss of social decorum, lack of insight, emotional blunting, and loss of empathy. FTD has been linked to motor neuron disease, as well as parkinsonism, both in the patients with FTD as well as in family members.

References:

Discussion:

Numerous studies have found a serotonergic deficit in patients with frontotemporal dementia (FTD). Experts in the field will often treat these patients with selective serotonin reuptake inhibitors (SSRIs), even in the absence of depression. There is no evidence of a cholinergic deficit in FTD, and studies evaluating the efficacy of cholinesterase inhibitors have been largely neutral or negative. Purported neuroprotective and antioxidant compounds have also not been found to be beneficial.

References:


Question 387: Behavioral/Psychiatry - Dementia

Discussion:

Typical features of early Alzheimer disease include impaired insight, disorientation to day and date, memory loss, and constructional impairments. Frontotemporal dementia patients have poor insight, but memory and especially visual-spatial abilities are not impaired as early as in Alzheimer disease. Dementia of depression, Parkinson disease dementia, and multiple subcortical lacunar strokes causing dementia all have reasonable insight and clue well on memory tasks.

References:


Question 409: Behavioral/Psychiatry - Dementia

Discussion:

The differential diagnosis for a rapidly progressive dementia includes several diseases in addition to prion infection. When considering sporadic Creutzfeldt-Jakob disease, the most sensitive and specific lab finding is the presence of grey matter hyperintensity on the DWI sequence of an MRI, along with an ADC correlate. A FLAIR study may show grey matter high signal; however, this finding can be absent in early stages of the disease. The cerebrospinal fluid studies are highly variable, serve only as markers of accelerated neuronal death, and are not prion specific.

References:


Question 413: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:

Neurologic involvement can occur in 5% to 10% of patients with sarcoidosis. Manifestations can include motor and sensory deficits, progressive visual loss, and cranial nerve palsies. Cognitive impairments can range from mild memory loss to progressive dementia. Sarcoidosis appears to have a predilection for the hypothalamus and basal forebrain resulting in hypopituitarism, hyperphagia, and hypersomnolence.

References:
Question 419: Behavioral/Psychiatry - Dementia

Discussion:

Patients with AIDS dementia generally have a subcortical dementia with psychomotor slowing; difficulty concentrating, especially in conducting serial 7s; impaired reading; and forgetfulness. Familial Alzheimer dementia may present in younger adults but typically presents with cortical dementia features, particularly memory loss. Dementia with Lewy bodies may have subcortical features, but patients are generally much older, gait is almost always abnormal, and there are often other signs of parkinsonism. Primary progressive aphasia presents with predominant language impairment. Frontotemporal dementia may present with psychomotor slowing, but behavioral changes predominate, with relatively fewer language and calculation difficulties.

References:


Question 423: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:

This patient is presenting with encephalitis (fever, mental status change, and abnormal CSF picture). Encephalitis caused by the West Nile virus (WNV) generally happens in older patients and presents with mental status changes and prominent movement disorders, secondary to a predilection for the basal ganglia and thalamus. WNV can also cause a flaccid paralytic syndrome, similar to the polio virus. Rabies virus tends to affect the temporal and limbic lobes, with prominent memory loss and personality changes, followed by seizures and death. Brainstem symptoms are rare. Eastern equine and herpes encephalitis rarely cause movement disorders.

References:


Question 424: Behavioral/Psychiatry - Anatomic syndromes

Discussion:

The medial dorsal nucleus of the thalamus has rich connections with the amygdala, orbitofrontal region, and the temporal lobe. Lesions involving this nucleus typically involve disinhibition, utilization behavior, mania, and memory loss. The lateral dorsal nucleus and pulvinar are more richly connected with brain regions involved in higher order visual functioning, and generally do not yield disinhibition and memory loss. The intralaminar nuclei are part of the relay system within the thalamus and receive input from the somatosensory and the reticular activating system.

References:


Clinical Adult

Question 3: Clinical Adult - Neuromuscular Disorders

Discussion:

The patient described has acute brachial neuritis involving the upper trunk muscles of the plexus. This condition is seen in all age groups, but is also well described in intravenous drug abusers. The presumed mechanism is deposition of immune-mediated complexes directly on nerves of the plexus. There is no evidence for direct trauma, stretch, or compression of the plexus; nor is there sign of infection.

References:


Question 5: Clinical Adult - Neuromuscular Disorders

Discussion:

This patient most likely has seronegative ocular myasthenia gravis. A high percentage of these patients (40% to 50%) have MuSK antibodies directed against the muscle-specific receptor tyrosine kinase.

References:


Question 7: Clinical Adult - Cerebrovascular Disease

Discussion:

Hypoglycemia can present with signs of focal neurologic dysfunction and mimic stroke. Serum glucose should always be checked prior to the administration of IV t-PA. MRI of the brain, echocardiogram and chest x-ray are not required prior to administration of IV t-PA.

References:


Question 23: Clinical Adult - Demyelinating Disease

Discussion:

Radiologically isolated syndrome, sometimes called preclinical MS, is a recent concept used to describe MRI findings suggestive of multiple sclerosis in the absence of any clinical event consistent with the disease. Its significance lies in the fact that several series have shown that up to one-third of patients with this entity will develop a clinical attack consistent with MS, and the majority of patients will show radiographic progression of the disease in a short time.

References:
Question 24: Clinical Adult - Neuro-ophthalmology/Neuro-otology

Discussion:

This patient's symptoms are most consistent with a right fourth nerve palsy. A patient with acquired fourth nerve palsy would use compensatory head positions to decrease the vertical diplopia, which would include: (1) head tilt to opposite shoulder, (2) head turn downward with chin depressed and eyes upward, and (3) face turn to opposite side.

References:


Question 26: Clinical Adult - Movement Disorders

Discussion:

Acute dystonic reactions are often observed after treatment with potent dopamine D2-receptor antagonists, including metoclopramide. Cervical and limb dystonia are most common. Laryngeal dystonia may be a life-threatening form of dystonic reaction in these patients. Anticholinergic medication, including diphenhydramine, is the most appropriate treatment.

References:


Question 28: Clinical Adult - Neuromuscular Disorders

Discussion:

This patient most likely has myasthenia gravis (MG). Anticholinesterase medications produce immediate improvement in muscle weakness in most patients with myasthenia gravis, but have no effect on the immunologic basis of the disorder. Prednisone, azathioprine, and cyclosporine produce immunosuppression and have a beneficial effect on weakness, usually after many weeks, or months, of therapy. Prednisone may transiently worsen weakness in MG in a patient naive to steroids. 3,4-diaminopyridine is used to treat Lambert-Eaton myasthenic syndrome and would not be effective in treating MG.

References:


Question 30: Clinical Adult - Epilepsy

Discussion:

After presentation with a first seizure, clinical risk factors for recurrence include abnormalities on neurologic exam, occurrence of the seizure outside of sleep, and remote symptomatic etiology. Hyponatremia and other electrolyte disturbances can cause symptomatic seizures, but seizures cease after correction of the abnormality. Seizures can be seen with cocaine abuse, as an acute toxic effect, but remote cocaine use of itself does not increase the risk of developing epilepsy. Migraine does not increase the risk of developing epilepsy.

References:

Britton JW. Antiepileptic drug therapy: when to start, when to stop. Continuum Lifelong Learning Neurol 2010;16(3):105-120.
Question 40: Clinical Adult - Dementia

Discussion:

REM sleep behavior disorder (RBD) is characterized by complex nocturnal behaviors involving vocalizations, hitting, punching and gesturing. Dream content is usually recalled and is unpleasant, involving being pursued or attacked. Recent studies confirm a link between RBD with synucleinopathies, most frequently with dementia with Lewy bodies and Parkinson disease.

References:


Question 45: Clinical Adult - Dementia

Discussion:

This patient's symptoms of memory impairment, with normal activities of daily living, are most consistent with amnestic mild cognitive impairment (MCI). MCI may represent the prodromal phase of Alzheimer disease.

References:


Question 54: Clinical Adult - Neurology of Systemic Disease

Discussion:

A femoral neuropathy or high lumbar plexus lesion, associated with retroperitoneal pain in a patient with coagulopathy, suggests a retroperitoneal hemorrhage. Imaging is required to confirm the diagnosis.

References:


Question 55: Clinical Adult - Spinal and Root Disorders

Discussion:

This patient has subacute syndrome of pyramidal tract and dorsal column impairment due to copper deficiency. Alpha-tocopherol transfer protein gene mutation would present typically in childhood with gradual onset. A complete blood count might be abnormal, but not diagnostic. MRI of the spine might show increased T2 signal in the dorsal columns in several acquired nutritional deficiencies, but may be normal. Copper deficiency should be considered in patients with myelopathy following gastric surgery, or after zinc overdose.

References:


Question 58: Clinical Adult - Infectious Disease

Discussion:
Varicella zoster virus can cause a necrotizing myelopathy within 1 to 2 weeks after presentation with a dermatomal rash. This is typically in the thoracic cord and is the result of reactivation of the virus in the dorsal root ganglia, but with centripetal migration into the cord rather than along the nerve. Diagnosis is made by identification of the virus by PCR in the CSF. HSV-1 is associated with Herpes Encephalitis and does not cause myelopathy. HHV6 causes post-transplant acute limbic encephalitis. CMV can cause a transverse myelitis but this is typically in immunocompromised patients. HSV-2 can rarely lead to an ascending myelitis or sacral radiculitis in patients with genital herpes.

References:

Question 59: Clinical Adult - Critical Care/Stroke
Discussion:
Hepatic encephalopathy is characterized by delirium, psychomotor slowing, asterixis, dysarthria and nystagmus. Severe cases are associated with posturing and coma. Laboratory testing can demonstrate abnormalities in liver function tests, ammonia, and clotting. CT head demonstrates generalized cerebral edema and is found in most patients with acute hepatic encephalopathy.

References:

Question 65: Clinical Adult - Movement Disorders
Discussion:
Dopa-responsive dystonia (DRD) presents in childhood as a progressive dystonia in children without a history of cerebral palsy or cognitive delay. It typically starts in a foot and progresses to become generalized; the most common inheritance pattern is autosomal dominant. The disease is unique for its robust and sustained response to low doses of levodopa. Its most notable characteristic is a diurnal variation, with symptoms usually more severe towards the end of the day and improved in the morning.

References:

Question 69: Clinical Adult - Epilepsy
Discussion:
Lamotrigine (LTG) is primarily metabolized by hepatic glucuronidation. Oral hormone contraception (OHC) induces this system and may result in a significant lowering of serum LTG levels. Therefore, the risk of breakthrough seizures is increased and LTG dosing often needs to be concomitantly increased. LTG has no effect on OHC metabolism and does not increase the risk of OHC failure.

References:
Pennel PB. Antiepileptic drugs during pregnancy: What is known and which AED's seem to be safest. Epilepsia 2008;49(suppl 9):43-55

Question 72: Clinical Adult - Cerebrovascular Disease
Discussion:
A round or triangular area of hyperdensity at the posterior aspect of the superior sagittal sinus on a noncontrast head CT is called a delta sign and may be an indicator of a sagittal sinus thrombosis. Risk factors include oral contraceptives, pregnancy, malignancy or a hypercoagulable disorder. Most evidence suggests that the best treatment for cerebral venous thromboses is intravenous heparin or low molecular weight heparins.

References:


Question 73: Clinical Adult - Motor Neuron/Nerve

Discussion:

Most patients with Charcot-Marie-Tooth disease (CMT) present with one of three phenotypes, based on the age at symptom onset. The first, and most common, group presents with a classic phenotype. Affected patients with a classic phenotype begin walking on time, usually by 12 to 15 months of age, and develop weakness or sensory loss during the first two decades of life. Impairment slowly increases thereafter, but patients rarely require ambulation aids beyond ankle-foot orthoses. Most patients with CMT1A and men with CMT1X, the two most common forms of CMT, present with this phenotype. Patients with the second phenotype present with an infantile-onset and do not begin walking until they are at least 15 months of age. These patients are often severely affected and are likely to require above-the-knee bracing, walkers, or wheelchairs for ambulation by 20 years of age. Many patients with early-onset CMT1B or CMT1E fall into this category. The third phenotype is defined as adult-onset, in which patients do not develop symptoms of CMT until adulthood, often not until approximately 40 years of age. An additional large group of patients with CMT1B as well as many women with CMT1X fall into this category. Nerve conduction tests are a useful initial screening test; they help distinguish between demyelinating and axonal forms of CMT.

References:


Question 74: Clinical Adult - Neuro-ophthalmology/Neuro-otology

Discussion:

This patient's symptoms of episodic vertigo, associated with progressive hearing loss and tinnitus, is most compatible with Ménière disease, a syndrome felt to be due to endolymphatic hydrops. Superior semicircular canal dehiscence is a variant of perilymphatic fistula and is characterized by sound, pressure induced vertigo, and oscillopsia. About 50% of patient's have autophony (hearing one's own voice unusually loudly, or even hearing one's heartbeat or eye movements). A diagnosis of vestibular migraine requires that International Headache Society (IHS) criteria for migraine be met, along with episodic or fluctuating symptoms suggestive of a balance disorder.

Migrainous symptoms should occur during, or in temporal relation to, episodes of vertigo or imbalance, and there should not be any other explanation for these symptoms. Vestibular neuronitis typically causes a monophasic prolonged episode of vertigo, and would not be associated with the recurrence and progressive hearing loss of Ménière disease. Vestibular schwannoma should be considered in any patient with progressive unilateral hearing loss, as in this patient, though the episodic vertigo described in this patient would be less typical for vestibular schwannoma.

References:


Question 76: Clinical Adult - Cerebrovascular Disease

Discussion:
This patient has a pupil-sparing third nerve palsy, most likely related to diabetes or small brainstem infarct. A posterior communicating artery aneurysm would usually not spare the pupil. Meningioma would often involve cranial nerves 4 and 6 as well and may be associated with proptosis. The third nerve lesion associated with an extrinsic mass typically would not be pupil-sparing.

References:

Question 84: Clinical Adult - Neuromuscular Disorders

Discussion:

The clinical description of symmetric proximal weakness that improves with repeated testing is most consistent with Lambert-Eaton myasthenic syndrome. This is associated with malignancy, most commonly small cell lung cancer, in two-thirds of patients.

References:

Question 85: Clinical Adult - Neurorehabilitation

Discussion:

This patient has a peroneal nerve palsy most likely related to compression. Additional tests are not required. Observation, avoidance of leg crossing, and an action foot orthotic are the appropriate therapies.

References:

Question 96: Clinical Adult - Infectious Disease

Discussion:

This patient has a syndrome of recurrent, self-limiting, aseptic meningitis, which has also been called Mollaret meningitis. This syndrome is caused by the herpes simplex virus 2 and may benefit from treatment with acyclovir.

References:

Question 97: Clinical Adult - Headache

Discussion:

According to AAN guidelines, there is Class A evidence supporting the use of topiramate for migraine prophylaxis. Botox is indicated for chronic migraine rather than episodic migraine. Frovatriptan can be used as prophylaxis, specifically for menstrual migraines. Indomethacin is used for chronic paroxysmal hemicrania. Gabapentin can be helpful with headaches that have a stabbing type quality. There is currently no Class A evidence for the use of verapamil in this setting.
Question 104: Clinical Adult - Motor Neuron/Nerve

Discussion:

This individual has likely developed a toxic length-dependent sensorimotor polyneuropathy. Metronidazole is associated with a toxic sensorimotor polyneuropathy affecting both large and small fibers. Toxicity is dependent on cumulative dose (usually greater than 30 gm) and recovery is often delayed for 6 to 12 months. The other antibiotics have not been associated with significant adverse peripheral nervous system effects.

References:


Question 118: Clinical Adult - Sleep

Discussion:

The patient has severe, refractory restless legs syndrome (RLS); in this case, in association with end-stage renal disease. He initially responds to a non-ergot dopamine agonist, but then experiences augmentation of symptoms. He is unable to tolerate gabapentin and probably will not tolerate pregabalin. Duloxetine is not indicated for restless legs. This patient has refractory RLS and is therefore most likely to respond to a high-potency opioid.

References:


Question 133: Clinical Adult - Spinal and Root Disorders

Discussion:

This patient's clinical symptoms are most compatible with neurogenic claudication, a syndrome which clinically resembles the symptoms of vascular claudication. Neurogenic claudication typically occurs due to chronic compression of the lumbosacral nerve roots of the cauda equina, within the thecal sac, due to lumbosacral central spinal stenosis. Associated posterior column dysfunction is often seen although the mechanism is unclear.

In contrast, a laterally herniated lumbosacral disc would typically present as an acute lateralized radiculopathy, rather than with the syndrome of bilateral claudication, as in this patient. A process causing diffuse enhancement of nerve roots, such as meningeal carcinomatosis, would usually present more asymmetrically and with pain unrelated to position and activity. A conus lesion or syrinx would be associated with myelopathic features and examination findings.

References:


Question 137: Clinical Adult - Spinal and Root Disorders

Discussion:

Down syndrome can be associated with atlantoaxial instability. Special precautions must be taken prior to any intubation in order to prevent neurologic damage.

References:


Question 141: Clinical Adult - Cerebrovascular Disease

Discussion:

The ECASS 3 trial demonstrated the benefit of IV tPA between 3 and 4.5 hours after symptom onset when compared to placebo. The addition of endovascular treatments including IA tPA and mechanical thrombectomy have not demonstrated superiority to IV tPA alone in several recently published randomized trials (SYNTHESIS, MR RESCUE and IMS III). Current evidence supports the use of IV tPA in eligible patients within 4.5 hours of symptom onset.

References:


Question 145: Clinical Adult - Neuromuscular Disorders

Discussion:

This patient has peroneal compression neuropathy due to significant weight loss and leg crossing. The appropriate management would be to caution him against leg crossing.

References:


Question 150: Clinical Adult - Headache

Discussion:

This patient most likely has orthostatic headache, secondary to spontaneous intracranial hypotension. The usual etiology is a CSF leak located in the spinal meninges. The leak is most readily identified now with MR spine imaging, or a CT myelogram. The other tests listed (meningeal biopsy, serum angiotensin converting enzyme, slit lamp examination, and CSF cytology) would not assist in the diagnosis of this syndrome.

References:


Question 152: Clinical Adult - Headache

Discussion:
This individual presents with a thunderclap headache syndrome in association with multifocal neurologic deficits. After several days of headache, an aneurysmal subarachnoid bleed should be detectable by lumbar puncture, brain CT or brain MRI. Carotid artery dissection is unlikely in the absence of neck pain and in the presence of multifocal deficits not confined to the carotid artery territory. Similarly the multifocal nature of the presentation, and the absence of parenchymal blood, makes a vascular malformation unlikely. Cortical vein thrombosis is typically associated with parenchymal hemorrhage and/or seizure, and acute stroke on MRI, and is therefore also unlikely. This presentation is most consistent with diffuse cerebral vasoconstriction syndrome, which has been associated with the use of SSRIs and vasoactive drugs. The likely finding on cerebral catheter or MR angiography is segmental arterial narrowing, which is usually reversible over time.

References:


Question 153: Clinical Adult - Neurology of Systemic Disease

Discussion:

Trigeminal sensory neuropathy is a common neurologic feature of connective tissue diseases. Dorsal root ganglion involvement, leading to an ataxic sensory neuronopathy, is unique to Sjögren syndrome.

References:


Question 154: Clinical Adult - Movement Disorders

Discussion:

Huntington disease is an autosomal dominant movement disorder characterized by chorea, dementia and behavioral disturbances. The pathologic hallmark of Huntington disease is atrophy of the caudate and this can be visualized with an MRI of the head.

References:


Question 161: Clinical Adult - Dementia

Discussion:

Amnestic mild cognitive impairment (aMCI) is, neuropathologically, most often a precursor state to Alzheimer disease. As such, there is medial temporal lobe pathology demonstrated. This is reflected in MRI studies, which have shown decreased hippocampal volume as an independent predictor for the development of dementia in subjects with aMCI.

References:


Question 171: Clinical Adult - Neuro-oncology
Discussion:

This patient developed hypertension, a seizure, and cortical blindness that is consistent with a reversible posterior leukoencephalopathy while on bevacizumab. Bevacizumab is also associated with an increased incidence of stroke, intracranial hemorrhage, and optic neuropathy. The seizure, somnolence, and normal pupillary responses are most consistent with reversible posterior leukoencephalopathy.

References:


Question 173: Clinical Adult - Neurogenetics

Discussion:

Allelic loss (loss of heterozygosity) of chromosomes 1p and 19q are molecular signatures of grade III (anaplastic) oligodendrogliomas. These tumors generally have increased responsiveness to chemotherapy and increased length of survival compared to grade III astrocytomas and grade IV gliomas. Loss of chromosomal arm 1p is a significant predictor of chemosensitivity, and combined loss of arms 1p and 19q are associated with chemosensitivity, longer recurrence free survival after chemotherapy, and longer overall survival.

References:


Question 174: Clinical Adult - Other Pain Syndromes

Discussion:

This patient's symptoms are most compatible with trigeminal neuralgia (tic douloureux). Most cases of trigeminal neuralgia are felt to be due to compression of the trigeminal nerve by a vascular loop. Imaging is indicated to exclude other, though less common, structural causes such as a meningioma, cerebellopontine angle neoplasm, and aneurysm. Multiple sclerosis can also be a cause of trigeminal neuralgia, but that is less likely in this patient due to her age and lack of other signs and symptoms. In the absence of long tract and cerebellar findings, stroke is extremely unlikely.

References:


Question 175: Clinical Adult - Spinal and Root Disorders

Discussion:

This patient has a C5-C6 disc herniation producing a relatively mild acute C6 radiculopathy. The diagnosis is clear from the information provided, and no additional diagnostic testing (EMG, myelography) should be required. In most cases the symptoms will resolve spontaneously without need for cervical discectomy. Surgery would be required if the patient had significant
radicular weakness, cervical myelopathy, or intractable radicular pain. Epidural steroid injection does not have a defined role in the treatment of radiculopathy, but may improve pain control in patients with radicular pain that is severe or does not respond to oral analgesics.

**References:**


**Question 185: Clinical Adult - Epilepsy**

**Discussion:**

A late posttraumatic seizure is defined as a seizure that occurs more than 1 week after a head injury. Patients with a late posttraumatic seizure are at high risk for recurrent episodes and require long-term treatment with an AED, even after presentation of their first seizure. A short course of treatment with an AED would be more appropriate for a patient with early posttraumatic seizure. There is no significant diagnostic or therapeutic role for long-term video EEG monitoring or epilepsy surgery in this patient.

**References:**


**Question 189: Clinical Adult - Epilepsy**

**Discussion:**

Sudden unexplained death in epilepsy (SUDEP) remains poorly understood. Studies suggest that the most significant risk factors include uncontrolled generalized tonic-clonic convulsions, higher frequency of seizures, and the need for multiple antiepileptic drugs. Absence seizures, nonconvulsive seizures, and etiology of the epilepsy have no known bearing on the risk of SUDEP.

**References:**


**Question 231: Clinical Adult - Dementia**

**Discussion:**

The image depicts the so-called "hummingbird" or "penguin" sign, resulting from midbrain atrophy, without pontine atrophy, seen in progressive supranuclear palsy. Clinically, in this tauopathy, early falls, bradykinesia, and marked axial rigidity, which is levodopa-unresponsive, are present. Supranuclear gaze palsy occurs with initial impairment of down gaze. Limb apraxia is more associated with cortical basal degeneration. REM behavior disorder and tremor are associated with Parkinson disease and alpha-synucleinopathies. Visual hallucinations are a component of the diagnostic criteria for dementia with Lewy bodies.

**References:**


**Question 243: Clinical Adult - Dementia**

**Discussion:**

The patient's presentation is strongly suggestive of Creutzfeldt-Jakob disease (CJD). The otherwise unremarkable MRI and normal routine CSF parameters are consistent with CJD. Diffusion-weighted MRI demonstrates restricted diffusion in the cortical
mantle and basal ganglia, characteristic of this disorder. Of the biomarkers listed, total CSF tau is the most specific and sensitive, followed by neuron-specific enolase and 14-3-3 protein.

References:


**Question 258: Clinical Adult - Cerebrovascular Disease**

**Discussion:**

This is an example of a catastrophic air embolus resulting from cardiopulmonary bypass. The patient has suffered brain death as a result of global cerebral hypoperfusion. This syndrome can occasionally result from chest trauma and neurosurgical procedures, in which the patient is operated with the head above the heart. On the CT, air can be seen diffusely within the cerebral vessels. Despite the coagulopathy associated with all major surgeries, the risks of subarachnoid bleed and cerebral venous sinus thrombosis are not elevated with this type of surgery, or with the use of cardiopulmonary bypass.

References:


**Question 317: Clinical Adult - Demyelinating Disease**

**Discussion:**

Neither pregnancy nor breastfeeding have been shown to adversely affect the course of RRMS using annualized relapse rates. Breastfeeding may in fact be somewhat protective. There is no known contraindication to corticosteroids during pregnancy. MS disease-modifying agents have not been adequately studied during pregnancy. Therefore, reliable data with regards to associated birth defects, or other pregnancy related complications, are not available. The FDA advises discontinuation of these agents, starting about 1 cycle prior to conception, under the justification of "do no harm". RRMS has not been associated with an increased risk of birth defects or pregnancy related complications.

References:


**Question 321: Clinical Adult - Cerebrovascular Disease**

**Discussion:**

This patient has cerebral amyloid angiopathy, which typically presents in elderly persons with spontaneous lobar hemorrhage. Typically patients have a history of dementia and have evidence of multiple tiny old hemorrhages (best seen on gradient-echo MRI imaging).

References:
Question 324: Clinical Adult - Cerebrovascular Disease

Discussion:

This patient most likely suffers from cerebral autosomal dominant arteriopathy, with subcortical infarcts and leukoencephalopathy (CADASIL), due to a mutation in the NOTCH3 gene on chromosome 19. This disorder is often associated with progressive focal neurological deficits. Migraine headaches are a common clinical feature as well.

References:


Question 330: Clinical Adult - Demyelinating Disease

Discussion:

The Kurtzke Expanded Disability Status Scale (EDSS) remains a key assessment tool in most drug trials performed for new multiple sclerosis therapies. Its ratings range from 0 to 10, with half points in between. There are eight sub scales, including pyramidal tract, visual and cognitive assessments; however, the overall ratings are highly dependent upon ambulatory ability.

References:


Question 335: Clinical Adult - Spinal and Root Disorders

Discussion:

Disturbance of the S1 nerve root results in pain radiating down the posterior aspect of the lower extremity, along with sensory disturbance of the fifth digit, lateral foot, and most of the sole of the foot. Weakness is prominent in the gastrocnemius and soleus muscles and there is reduction, or absence, of the Achilles reflex.

References:


Question 337: Clinical Adult - Headache

Discussion:

While the amount of most medications excreted in breast milk is 1% to 2% of the maternal dose, there is some variability depending on both the medication's characteristics and breast milk's characteristics. Barbiturates may cause sedation. Aspirin is considered less safe than acetaminophen. Triptans and tricyclic antidepressants are of unknown risk to the infant, while narcotics are safe.

References:


Question 339: Clinical Adult - Movement Disorders

Discussion:

This individual has restless legs syndrome (RLS). RLS is characterized by 1) an urge to move the legs, usually accompanied by uncomfortable or unpleasant leg sensations, 2) symptoms beginning or worsening during rest or inactivity, 3) symptoms occur only, or are worse, in the evening or night compared to the day, 4) symptoms are partially or totally relieved by movement or stretching for at least as long as the activity continues, and 5) symptoms are not solely accounted for as being primary to another condition, such as leg cramps. Behavioral disorders such as impulse control disorders are not associated with RLS, but can be seen with other movement disorders such as Parkinson disease.

References:


Question 340: Clinical Adult - Neuromuscular Disorders

Discussion:

The subacute history, together with the presence of a non-length-dependent polyneuropathy by examination, is suggestive of an immune mediated demyelinating polyneuropathy. Both CIDP and POEMS (polyneuropathy, organomegaly, endocrinopathy, M protein and skin changes) can have identical neurologic examinations, nerve conduction study and CSF findings. In this case, the presence of organomegaly, endocrinopathy, M protein and skin changes support, but of themselves are not sufficient to confirm, a diagnosis of POEMS. Papilledema would also support a diagnosis of POEMS, but as a minor criterion. Markedly elevated levels of serum VEGF is one of the major diagnostic criteria supporting a diagnosis of POEMS.

References:


Question 344: Clinical Adult - Neuro-ophthalmology/Neuro-otology

Discussion:

The Charles Bonnet syndrome is characterized by complex visual hallucinations in the setting of significant visual deprivation. Most patients are elderly, have normal cognition, and are well aware that the images are not real. Visual hallucinations that localize to cerebral peduncles tend to be hypnagogic. Temporal lobe pathology and the hippocampus can be associated with complex visual hallucinations and seizures, without associated visual loss.

References:


Question 345: Clinical Adult - Headache

Discussion:

Paroxysmal hemicrania is characterized by severe, frequent, brief, unilateral attacks with some associated autonomic features. These headaches can be exquisitely sensitive to indomethacin. Sumatriptan and dihydroergotamine are appropriate for the
abortive therapy of migraine headaches. Inhaled oxygen is used for cluster headaches and intravenous lidocaine can work with SUNCT syndrome.

References:

Question 349: Clinical Adult - Headache

Discussion:
This patient most likely has headaches from daily use of over-the-counter analgesics (medication overuse headache). While work related stress, dietary habits, and menstrual irregularity may contribute to headaches in patients with migraine, their contribution is not likely to be as great as the analgesic overuse.

References:

Question 354: Clinical Adult - Neuromuscular Disorders

Discussion:
Monoclonal gammopathy of unknown significance (MGUS) is associated with a symmetric distal polyneuropathy. Nerve conduction studies characteristically demonstrate decrease in motor conduction velocities that are consistent with demyelination. This can distinguish MGUS as an etiology from many other common causes of neuropathy, which are predominantly axonal.

References:

Question 355: Clinical Adult - Movement Disorders

Discussion:
Anosmia, REM sleep behavior disorder, constipation and depression are commonly described premotor features of idiopathic Parkinson disease and can predate the diagnosis by several years.

References:
Siderowf A, Stern MB. Premotor Parkinson's disease: clinical features, detection, and prospects for treatment.. Ann Neurol. 2008;64: S139-47

Question 356: Clinical Adult - Dementia

Discussion:
Primary progressive aphasia (PPA) is a frontotemporal dementia (frontotemporal degeneration, FTD) that is characterized initially by word-finding trouble, difficulty with naming, and comprehension. These language features are typically a dominant component of the disease in the first 2 years. Patients can later develop dysfunction in other domains, and late stage PPA patients may not appear that different from patients with other late stage dementias.
References:


Question 364: Clinical Adult - Demyelinating Disease

Discussion:

Fingolimod has been shown to cause bradycardia. EKG monitoring is required during the first dose. Some patients become symptomatic and may even experience syncope.

References:


Question 367: Clinical Adult - Neurogenetics

Discussion:

This patient most likely has episodic ataxia type 2, an autosomal dominant disorder, due to a mutation in the CACNA1A gene encoding for a calcium channel protein. Acetazolamide may treat the symptoms of this disorder.

References:


Question 369: Clinical Adult - Critical Care/Stroke

Discussion:

This individual has developed an acute critical illness myopathy. The major risk for this entity is treatment of a critically ill individual with high dose corticosteroids in the ICU setting. Other considerations include critical illness polyneuropathy, disorders of the neuromuscular junction (NMJ), and medication induced myopathy or neuropathy. Acute critical illness peripheral neuropathies, and disorders of the NMJ, are unlikely given the preservation of tendon reflexes, normal nerve conduction study, and normal repetitive stimulation studies. Tacrolimus has been implicated as causing a toxic neuropathy, but not a myopathy. Ciprofloxacin has been implicated as a trigger for myasthenia, but not a myopathy. Statin myopathies tend to be subacute and often associated with CK levels over 10 times normal. Short-term use of propofol has not been associated with acute neuropathy, or a myopathy, per se. Use of high-dose propofol can result in propofol infusion syndrome (metabolic acidosis, heart failure, renal failure, hyperkalemia, hyperlipidemia, and rhabdomyolysis).

References:

Lacomis D, Campellone JV. Continuum Lifelong Learning Neurol 2004;10(2);19-38.


Question 375: Clinical Adult - Dementia

Discussion:
This patient has progressive difficulty with higher level visual processing, including symptoms of Balint syndrome (simultagnosia, oculomotor apraxia and optic ataxia). Of the choices given, her clinical syndrome is most compatible with posterior cortical atrophy. In most cases, posterior cortical atrophy is felt to be a variant early-onset Alzheimer disease, presenting with prominent initial involvement of the occipitoparietal cortex and prominent symptoms of abnormal cortical visual processing.

References:


Question 378: Clinical Adult - Demyelinating Disease

Discussion:

Several new platform therapies have recently become available and are approved for patients with relapsing forms of multiple sclerosis. However, all approved agents have either contraindications or clinical warnings requiring the clinician to tailor therapy precisely for each patient. Fingolimod is associated with probable sudden cardiac death and would be contraindicated in a patient with heart block. Natalizumab, which has black box warnings for development of PML, would not warrant the risk/benefit ratio in a patient with clinically mild disease. Alemtuzumab is not approved for MS, and methotrexate has generally weak data and has been used off-label. Dimethyl fumarate, a novel oral agent approved for relapsing-remitting multiple sclerosis would be an appropriate agent in this setting.

References:


Question 393: Clinical Adult - Epilepsy

Discussion:

According to the recently published findings of the North American Antiepileptic Drug (AED) Pregnancy Registry, lamotrigine is associated with the lowest risk of major congenital malformations during exposure in the first trimester compared to other the AEDs given. The approximate relative risks of the other AEDs listed compared to lamotrigine were 1.2 for levetiracetam; 1.5 for carbamazepine; 1.5 for phenytoin; 2.2 for topiramate; and 5.1 for valproic acid.

References:


Question 397: Clinical Adult - Headache

Discussion:

This patient likely has SUNCT (short-lasting unilateral neuralgiform headache with conjunctival injection and tearing). SUNCT syndrome is characterized by very brief (5 to 240 seconds) stabbing pain in the orbital or temporal region, associated with tearing and redness of the ipsilateral eye. The head pains occur multiple times per day. The headaches are much briefer than those seen in
any other trigeminal autonomic cephalalgias (TAC). The pain is usually not triggered by touching the face, eating, drinking, or talking, as in trigeminal neuralgia.

References:


Question 398: Clinical Adult - Neuro-oncology

Discussion:

A randomized trial published in 2005 demonstrated the benefit of a direct decompressive surgery followed by radiotherapy compared to radiotherapy alone. In patients with acute onset of epidural spinal cord compression, patients who underwent surgery within 24 hours had significant improvement in their ability to walk and the length of time that they were able to continue to walk.

References:


Question 415: Clinical Adult - Neurotoxicology

Discussion:

Nitrous oxide toxicity may cause a progressive myelopathy of the posterior and lateral columns that mimics that of vitamin B₁₂ deficiency. Nitrous oxide irreversibly oxidizes the cobalt moiety of methylcobalamin and interferes with conversion of homocysteine to methionine.

References:


Question 420: Clinical Adult - Critical Care/Stroke

Discussion:

Natalizumab-induced PML can be treated with discontinuation of natalizumab and initiation of plasma exchange. Treatment can be associated with IRIS (immune reconstitution inflammatory syndrome), characterized by acute neurologic deterioration and brain edema that occurs 3 to 6 weeks after stopping natalizumab. The optimal treatment for IRIS is high-dose glucocorticoids followed by a slow taper.

References:


Question 422: Clinical Adult - Epilepsy

Discussion:

Good prognostic features after temporal lobectomy include presence of mesial temporal sclerosis on MRI of the brain, a history of febrile seizures, and mostly unilateral ictal and interictal EEG abnormalities.
Question 425: Clinical Adult - Epilepsy

Discussion:

Juvenile myoclonic epilepsy is the most common form of idiopathic epilepsy and typically presents in adolescence or early adulthood. Patients typically present with generalized seizures precipitated by alcohol or sleep deprivation. Myoclonic jerks and bursts of 4 Hz to 6 Hz spike-and-wave discharges are characteristic. Valproate is very effective at treating both the seizures and myoclonus.

References:


Clinical Pediatrics

Question 9: Clinical Pediatrics - Neurosurgery, Critical Care and Tumors

Discussion:

The Presidents Commission recommended 100% oxygen ventilation for 10 minutes, followed by passive 100% oxygen for a period long enough to achieve a PCO2 of 60. The other tests have high incidence of false-negative results.

References:


Question 13: Clinical Pediatrics - Neurosurgery, Critical Care and Tumors

Discussion:

At birth the median head circumference of a normal term infant is 35 cm. A head circumference of 45 cm is greater than the 98th percentile. The tense fontanelle and eye deviation are concerning for an increase in intracranial pressure. In the absence of other cerebral malformations, congenital aqueductal stenosis is the most common cause of noncommunicating hydrocephalus. A familial X-linked form accounts for 2% of these cases. Increased intracranial pressure in the context of hypoxic-ischemic encephalopathy does not cause increased head circumference at birth. Choroid plexus papillomas are typically located in one lateral ventricle and become symptomatic after the perinatal period, usually by obstructing ventricular outflow. The obstructive hydrocephalus associated with an acute intraventricular hemorrhage at the time of birth takes time to develop. Although Alexander disease can present in the neonatal period with an obstructive hydrocephalus, children with Canavan disease are typically normal at birth with an infantile presentation occurring between 3 and 5 months of age.

References:


Question 21: Clinical Pediatrics - Neuromuscular
Discussion:

Spinal muscular atrophy I usually presents with hypotonia, areflexia, paradoxical respiration with narrow upper chest, tongue fasciculations, and progresses to difficulty swallowing, then respiratory distress. While infants with cerebral palsy may be initially hypotonic, they are generally hyperreflexic. Infants with infantile Gaucher disease generally present with stiffness, not initial hypotonia, and have organomegaly at onset. Muscle stretch reflexes in children with a congenital myopathy are usually present but may be difficult to elicit due to weakness.

References:


Question 38: Clinical Pediatrics - Vascular and Inflammatory Disorders

Discussion:

Cerebrovascular disease occurs in 25% of sickle cell disease. Eighty percent of events occur at or before age 15 and most are caused by a progressive cerebral vasculopathy that can be partially arrested by chronic transfusion therapy to keep hemoglobin S below 30%. Most events are thrombotic.

References:


Question 43: Clinical Pediatrics - Infectious Disease

Discussion:

Group B streptococci causes almost 50% of cases of neonatal bacterial meningitis and is the most common organism. Escherichia coli and Listeria monocytogenes are the second and third most common organisms, respectively.

References:


Question 44: Clinical Pediatrics - Cerebral Palsy

Discussion:

Gliosis of the basal ganglia causes status marmoratus. It is thought to be from poor central perfusion, and when present is highly associated with athetoid or dystonic cerebral palsy. The other abnormalities are associated with cerebral palsy, but are more often seen in spastic forms.

References:


Question 56: Clinical Pediatrics - Neuromuscular

Discussion:
Hallmarks of Friedreich ataxia are a combination of weakness, ataxia, absent tendon jerks, Babinski sign, and abnormal position sense. Romberg sign is the best indication of loss of position sense, not cerebellar functioning. Hammer toes, pes cavus deformities, and scoliosis are frequent findings.

References:


Question 67: Clinical Pediatrics - Neurosurgery, Critical Care and Tumors

Discussion:

Fifteen percent of children with Down syndrome have atlantoaxial instability. Other heritable conditions with similar predisposition are Klippel-Feil, Morquio, and Larsen syndromes, achondroplasia and previous cervical spinal surgery.

References:


Question 87: Clinical Pediatrics - Neuromuscular

Discussion:

Only daily oral corticosteroids have been shown to prolong ambulation in Duchenne muscular dystrophy (DMD). The effect is to induce small amounts of dystrophin, not as an antiinflammatory treatment, so other dosing strategies are not helpful. Intravenous gentamicin has been proposed for one of the rare point mutations causing DMD. Trials of oral carnitine and valproic acid are underway in SMA.

References:


Question 98: Clinical Pediatrics - Movement Disorders

Discussion:

The child's presentation (opsoclonus, myoclonus, ataxia syndrome, alternatively called "OMS", "dancing eyes syndrome" or Kinsbourne syndrome) is most often a paraneoplastic manifestation of occult neuroblastoma in this age group. The most common initial misdiagnosis is acute cerebellar ataxia of childhood, a benign condition. The most helpful test in determining etiology of suspected OMS is CT of the chest, abdomen, and pelvis using oral contrast and fine cut images.

References:

Question 99: Clinical Pediatrics - Hereditary and Metabolic Disorders

Discussion:

The presence of macrocephaly, multiple café au lait spots, and axillary freckling is suggestive of a diagnosis of neurofibromatosis type 1. The ophthalmological finding observed in NF type 1 is Lisch nodules, which represent pigmented hamartomatous nevi affecting the iris. They are present in the vast majority of people with NF1 after the age of 12 years.

References:


Question 100: Clinical Pediatrics - Epilepsy

Discussion:

Typical childhood absence seizures do not have auras or postictal confusion. Automatisms can be seen. The usual EEG shows 3 Hz to 4 Hz generalized spike-and-wave discharges, and imaging studies of the brain are normal. The initial medication of choice is ethosuximide or valproate.

References:


Question 101: Clinical Pediatrics - Epilepsy

Discussion:

Typical childhood absence seizures do not have auras or postictal confusion; automatisms can be seen. The usual EEG shows 3 Hz to 4 Hz generalized spike-and-wave discharges, and imaging studies of the brain are normal. The initial medication of choice is ethosuximide or valproate. Although the trial by Glauser and colleagues demonstrated good efficacy with valproic acid, untoward effects associated with this medication make ethosuximide a better choice.

References:


Question 107: Clinical Pediatrics - Epilepsy

Discussion:

The provocation of astatic seizures in children with generalized epilepsies by carbamazepine was first described by Snead in 1985. Although other anticonvulsive drugs have been associated with similar drug-related provocation, valproic acid, felbamate, ethosuximide, and topiramate are drugs that are among those most likely to reduce the occurrence of disease-related astatic/tonic (drop) seizures.

References:
Question 108: Clinical Pediatrics - Neuromuscular

Discussion:

Nemaline myopathy has a static or slowly progressive course with rod-like inclusions on trichrome studies. Creatine kinase is normal or mildly elevated; EMG may be normal. The most common cause is a mutation in the alpha-actin gene.

References:


Question 116: Clinical Pediatrics - Neonatal

Discussion:

The findings are those of Erb palsy with associated Horner syndrome on the right. Electrodiagnostic testing at this stage is difficult and not particularly helpful. It is unlikely that an intracranial process explains a flaccid, areflexic arm. MRI of cervical spine may be helpful at a later date to define root avulsion injuries if there is no recovery and surgery is planned. Bony injury to the cervical spine would not explain these deficits, and if suspected, contraindicates flexion and extension views. Physical and occupational therapy services, initially for positioning and range of motion and later for active strengthening, are indicated.

References:


Question 117: Clinical Pediatrics - Headache/Other Paroxysmal Disorders

Discussion:

This is a classic presentation of benign paroxysmal vertigo, a syndrome that occurs in children in the first 5 years of life and is a frequent precursor of migraine. Investigations (ie, MRI, EEG) are normal. The episodes usually resolve by the end of the first decade.

References:


Question 129: Clinical Pediatrics - Develop, Learn, Lang, Behav, Psych Disorders

Discussion:
Meador and colleagues found that children at 3 years of age exposed to valproic acid in utero had significantly lower IQ scores than children of the same age who had in utero exposure to to carbamazepine, lamotrigine, and phenytoin.

References:


Question 168: Clinical Pediatrics - Hereditary and Metabolic Disorders

Discussion:

This clinical picture describes a typical presentation and MRI findings of glutaric aciduria type 1. The signs and symptoms of this disorder can have a slow gradual onset or begin acutely following an infectious illness. The macrocephaly may be present at birth. MRI demonstrates widening of the Sylvain fissure, or lack of operculization, and abnormal high T2 signal intensity in the basal ganglia as well as the dentate nucleus, substantia nigra, and pontine medial lemniscus. Subdural hematomas or fluid collections may be present.

References:


Question 169: Clinical Pediatrics - Hereditary and Metabolic Disorders

Discussion:

This clinical picture with dystonia and self-injurious behavior describes a typical presentation for Lesch-Nyhan syndrome which is the result of a deficiency of hypoxanthine-guanine phosphoribosyltransferase. Onset of the extrapyramidal movement disorder is typically prior to 12 months of age. Self-injurious behaviors commence between 2 and 4 years of age.

References:


Question 177: Clinical Pediatrics - Neonatal

Discussion:

Malformations of the vein of Galen present in most neonates with cardiac failure. The remainder present with hydrocephalus or intraventricular hemorrhage. In older infants, hydrocephalus is the most common mode of presentation.

References:


Question 181: Clinical Pediatrics - Epilepsy

Discussion:
Patients with Lennox-Gastaut syndrome have multiple types of seizures, such as tonic, atypical absence, and atonic, with age of onset of 1 to 8 years. The EEG reveals slow spike-and-wave discharges of 1.5 Hz to 2 Hz. Most children are mentally retarded, and approximately 70% have an identifiable cause for the retardation and epilepsy. Tonic seizures are not observed in myoclonic atatic epilepsy (Doose syndrome).

References:


Question 183: Clinical Pediatrics - Infectious Disease

Discussion:

_Bartonella henselae_ (catscratch disease) is usually spread by kittens. A single, enlarged lymph node in the arm axilla or neck is often present. CSF and MRI are usually normal, and the EEG can be normal between convulsions.

References:


Question 186: Clinical Pediatrics - Movement Disorders

Discussion:

Niemann-Pick disease type C is characterized by autosomal recessive inheritance with gradual and progressive neurologic deterioration. Patients have variable hepatosplenomegaly, and progressive neurodegeneration. Findings include vertical supranuclear (usually downgaze) palsy, ataxia, dystonia, seizures, gelastic cataplexy, and dementia.

References:


Questions 209 - 213: Clinical Pediatrics - Headache/Other Paroxysmal Disorders

Discussion:

Night terrors typically occur in deep sleep, often 90 minutes to 3 hours after sleep onset. The child appears awake but is not. Autonomic symptoms are prominent. The child has no memory of the event.

Masturbation by infants is not often recognized immediately, causing excessive diagnostic testing. While most often seen in infant and toddler girls, boys may have similar manifestations. Usually the behavior is specific to the setting (often in highchair or car seat) but may also occur with the child lying on the floor, often with the legs extended or crossed. Facial flushing and sweating is common. Although the behavior can be interrupted, the child typically returns to it as soon as they are not deterred.

Paroxysmal kinesogenic choreoathetosis presents with unilateral choreic or dystonic movements with initiation of voluntary movement such as walking. There is no alteration of consciousness.

Cyanotic breath-holding spells occur in infants after sudden pain or fright. Brief convulsion may be precipitated by the syncope.

Hemiplegic migraine is often dominantly inherited, consists of hemiplegia followed by headache and vomiting. Some are mapped to a calcium channel gene.
Metachromatic leukodystrophy is an autosomal recessive disorder, and in its imaging usually demonstrates involvement of the periventricular and deep white matter with relative sparing of the U-fibers. Neurocognitive symptoms can be the presenting presentation in the late juvenile onset form; with relatively subtle signs on the neurological examination (long tract signs alone are common).

References:

The lesion is a hypothalamic hamartoma. Typical presentation is gelastic epilepsy (seizures beginning with laughter). Endocrine effects are relatively uncommon, but precocious puberty may occur.

References:

X-linked adrenoleukodystrophy frequently presents with abnormal white matter hyperintensities in a parieto-occipital distribution, thus leading to issues in visuo-spatial functioning. Attentional symptoms of later onset than those seen in typical ADHD are common, as is a progressive cognitive decline. Psychiatric symptoms are seen relatively early in the disease. The leading edge of the myelin loss, seen on this image, is also quite characteristic of the disorder. In contrast, for Pelizaeus-Merzbacher Disease, an x-linked disorder, the white matter in the centrum semi-ovale is isodense with the cortical ribbon on T1-weighted imaging. In Canavan disease, there are microcysts on imaging in the setting of macrocephaly. In Sandhoff disease and GM2 gangliosidosis, cortical atrophy may be seen.

References:
Chemotherapy used for leukemia, particularly l-asparaginase, may cause significant hypercoagulability, particularly secondary anti-thrombin III deficiency, which this patient had. Venous sinus thromboses, with infarctions in a venous distribution, are the result.

References:


**Question 247: Clinical Pediatrics - Movement Disorders**

**Discussion:**

The MRI demonstrates a characteristic "eye of the tiger" with low T2 signal centered in a high T2 signal area. The characteristic dystonic movements in PKAN2 may follow an initial period of apparent spasticity. Oromandibular dystonia is prominent. Huntington disease presents with prominent dystonic features in children, rather than chorea, but the MRI usually shows atrophy of caudate as well as cortical atrophy. DYT1 typically has no MRI findings. In glutaric aciduria I, onset of dystonia is earlier, MRI shows necrotic lesions in basal ganglia and bitemporal atrophy. Patients with dopa-responsive dystonia often have early findings suggestive of spasticity, progressing to dystonia, but typically have normal MRI.

References:


**Question 257: Clinical Pediatrics - Hereditary and Metabolic Disorders**

**Discussion:**

Aicardi Syndrome includes agenesis of the corpus callosum and "punched out" retinal colobomas. It is an x-linked dominant condition presumed to be fatal in males. The findings of the characteristic colobomas in the peripheral retina are diagnostic. Although some girls with Aicardi Syndrome have more extensive brain malformation, they do not generally have hypothalamic-pituitary dysfunction. The DCX mutation is associated with laminar heterotopias in girls, not agenesis of the corpus callosum, and lissencephaly in boys. Although mitochondrial disorders may present with prenatal onset CNS migration disorders, agenesis of the corpus callosum would not be expected. Organic acidurias do not generally present with cerebral malformations such as agenesis of the corpus callosum.

References:


**Question 311: Clinical Pediatrics - Hereditary and Metabolic Disorders**

**Discussion:**

This clinical picture describes a typical presentation and MRI findings of Joubert syndrome; in which there is hypoplasia of the cerebellar vermis. The most common features of this syndrome include hyperpnea, hypotonia, oculomotor apraxia, ataxia, and mental retardation. Other neurologic manifestations include seizures. The molar tooth sign, which is the result of the thickening and horizontalization of the superior cerebellar peduncle and a deep interpeduncular fossa, can also be seen in several other disorders including Dekaban-Arima syndrome, Senior-Loken syndrome, and COACH (cerebellar vermis hypoplasia, oligophrenia, ataxia, coloboma, and hepatic fibrosis).
Question 319: Clinical Pediatrics - Neurosurgery, Critical Care and Tumors

Discussion:

Menkes disease, type 1 osteogenesis imperfecta, and glutaric aciduria type 1 are among the heritable conditions that may be associated with clinically significant subdural hemorrhages in the absence of inflicted trauma. Care must be taken to exclude these diagnoses in order to avoid incorrectly diagnosing shaken baby syndrome.

References:


Question 341: Clinical Pediatrics - Cerebral Palsy

Discussion:

Cerebral palsy is a clinical diagnosis that describes a nonprogressive delay in motor development. A diagnosis of cerebral palsy does not define its etiology. This clinical picture describes the classic tetrad of chronic bilirubin encephalopathy (kernicterus): severe athetosis, perceptive high tone deafness, supranuclear palsy of upward gaze, and enamel hypoplasia of the incisor teeth.

References:


Question 348: Clinical Pediatrics - Vascular and Inflammatory Disorders

Discussion:

The presence of a facial angioma in the distribution of the ophthalmic division of the trigeminal nerve, in a child who presents with a unilateral, throbbing headache and neurologic deficits, is suggestive of the diagnosis of Sturge-Weber syndrome. In this syndrome, leptomeningeal angioma may represent an embryonic remnant of the venous plexus that failed to regress. The leptomeningeal angioma is most frequently unilateral, overlying the parietal and occipital lobes, and ipsilateral to the facial angioma. The presence of leptomeningeal angioma confirms the diagnosis of Sturge-Weber syndrome, which is often complicated by headache, stroke-like episodes, hemianopsia, glaucoma, and mental retardation.

References:


Question 350: Clinical Pediatrics - Infectious Disease

Discussion:

Long term sequelae in North American cases of bacterial meningitis occur with hearing loss found in as many as 15% to 30% of cases in various series, while permanent motor deficits or learning disabilities are found in in 5% to 15%; epilepsy or hydrocephalus in less than 5% of cases.

References:


Question 351: Clinical Pediatrics - Epilepsy

Discussion:

Mutations of filamin A have been linked to the presence of bilateral periventricular heterotopias, enlarged cistern magna and thinning or agenesis of the corpus callosum. In adolescent and adult, screening for an aortic aneurysm should be performed. Molar tooth sign is associated with Joubert syndrome; Leptomeningeal angioma is observed with Sturge Weber syndrome. Schizencephaly is also a neuronal migration disorder with expected association of seizures and developmental delay.

References:


Question 358: Clinical Pediatrics - Epilepsy

Discussion:

The combination of tonic spasms and a burst suppression EEG is consistent with the syndromic diagnosis of early infantile epileptic encephalopathy (Ohtahara syndrome). This syndrome, which can also include fragmentary myoclonic jerks and partial seizures, frequently evolves into West syndrome at about 4 to 6 months of age. Etiological causes have included brain malformations as well as metabolic syndromes such as glycine encephalopathy and mitochondrial disorders.

References:


Question 381: Clinical Pediatrics - Develop, Learn, Lang, Behav, Psych Disorders

Discussion:

This child has tuberous sclerosis complex, an autosomal dominant disorder associated with mutations in the tuberin and merlin genes. Intellectual outcome is best correlated with seizure control.

References:
Question 391: Clinical Pediatrics - Headache/Other Paroxysmal Disorders

Discussion:

The combination of dysconjugate, highly variable nystagmus, head nodding and head tilt, without ophthalmologic abnormalities, and with normal neuroimaging, is diagnostic of spasmus nutans. Latent nystagmus is a jerk nystagmus that is evoked or enhanced by covering one eye. Congenital nystagmus is usually conjugate and suppressed by convergence (in contrast to spasmus nutans, which is typically increased by convergence). Opsoclonus describes chaotic, conjugate saccades, seen classically in the paraneoplastic syndrome associated with neuroblastoma. Decompensated strabismus would not produce a paroxysmal movement disorder.

References:


Question 401: Clinical Pediatrics - Develop, Learn, Lang, Behav, Psych Disorders

Discussion:

Lack of indicative pointing is typical of autistic children, while even preverbal children with other causes of delay generally point at objects and "share attention". Late walking is not typical of autism but indicates overall delay in development. Sleep difficulties are common in autistic children but are also common in children with delays, as well as normal toddlers. Stranger anxiety may persist into toddlerhood in otherwise normally developing children.

References:


Contemporary Issues

Question 16: Contemporary Issues - Driving

Discussion:

While the governing principle for determining driving cessation ought to be risk for recurrence, this data is often difficult to estimate because etiology may be uncertain, description may be inaccurate, and treatments may or may not be provided, prove effective, or be heeded. The rules that govern driving privileges after unexplained loss of consciousness associated with a seizure are enacted and enforced by state agencies whose attempts to assign risk on the basis of these various data have generated complex, sometimes unclear, and nationally nonuniform standards. Single unexplained convulsions with loss of consciousness usually necessitate cessation of driving for some interval, although epilepsy is not as yet diagnosed. Some patients with epilepsy are permitted in some jurisdictions to drive if, in the judgment of the treating neurologist, there is high likelihood that all seizures will occur in sleep. Individuals with epilepsy who have remained seizure free for intervals varying from 3 to 12 months (depending on state of residence) may be permitted to resume driving. The responsibilities of the neurologist include clarifying history and diagnosis, providing appropriate treatment, being aware of driving standards within his/her own and possibly other
states, and explaining these standards to the patient, including the necessity of reporting certain events and diagnoses. Neurologists customarily complete portions of the forms patients are mandated to file with state agencies at intervals stipulated by state laws in order to continue driving.

References:


**Question 17: Contemporary Issues - Ethics/Professionalism**

**Discussion:**

Requests to lie to patients about their diagnoses arise from time to time. Some are based on the patient and family's cultural traditions, and some are based on fear of consequences. In this case, the son's request is based on fear of depression and is not a sufficient justification to lie to the patient. Telling the patient her son asked that her diagnosis should be withheld inappropriately undermines trust between the physician and the son, and between the son and his mother. To honor the son's request while later telling the patient her diagnosis by telephone constitutes lying to the son and is inappropriate. The AAN Code of Professional Conduct recommends telling the truth to patients who have decision-making capacity. AAN guidelines on humanistic dimension of professionalism in the practice of neurology state that neurologists have a role in helping patients and families overcome their fears of neurological disorders. Truth-telling allows patients with probable Alzheimer's disease to seek appropriate treatment and to make arrangements for their treatment preferences (eg, end-of-life care planning) and to select their surrogate decision maker. Not all patients want to know everything about their diagnosis and treatment, and some may wish to have their family handle this information and decisions for them. Asking the patient in advance how much information she wishes to receive respects her autonomy by allowing her to make a choice.

References:


**Question 32: Contemporary Issues - Business**

**Discussion:**

The modifier 26 identifies the physician's component and is used to bill for the professional component of a test or report the physician's interpretation of a test.

References:


**Question 61: Contemporary Issues - Ethics/Professionalism**

**Discussion:**

The factors of greatest importance in this case are the fact that the patient has submitted an advanced directive stating that she does not wish to have aggressive treatment and that her husband is her health care proxy (Goldblatt, 2003). Further, two types of written advanced directives are generally recognized: 1) the Living Will and 2) the Durable Power of Attorney. Both have the advantage of providing direction to a surrogate in the event the patient is unable to decide for herself. Both are appropriate as legal instruments for the withdrawal of life-sustaining care (Beauchamp & Childress, 1994).
References:


Question 111: Contemporary Issues - Evidence-based Medicine

Discussion:

Misclassification bias is a form of error that results from systematic inaccuracy in measurement, and it applies to either dichotomous or continuous measures. It is also known as information bias. Interviewer bias and diagnostic bias are two examples of misclassification bias. The radiologists in this example may not be using consistent criteria to assess the degree of stenosis. Lack of standardized criteria could easily lead to diagnostic bias, which is the problem of the cases with stenosis being classified as not having stenosis (and vice versa). Similarly, those with exposure may be classified as unexposed (and vice versa). Selection bias refers to variability in participant enrollment, which is reduced by utilizing a multi-centered trial.

References:

Evidence Based Medicine. 7/24/09 http://www.aan.com/education/ebm/

Question 119: Contemporary Issues - HIPAA

Discussion:

A series of guidelines, including the Belmont Report, the Helsinki Principle and 45 CFR 46, guide ethical conduct for investigators. According to these guidelines, a colleague may not disclose any information regarding a patient's name, contact information, or diagnosis to a researcher without the patient's consent prior to release. Even the ability to opt out of a research study is not permissible if the information above is provided to the researcher without consent.

References:


Question 158: Contemporary Issues - Practice

Discussion:

The diagnosis is Bell palsy. There is Class 1 evidence supporting treatment with steroids to improve recovery. There is weak evidence for antiviral treatment. There is no benefit to combination therapy. The upper and lower facial weakness is unlikely to be a stroke.

References:

Gronseth, G, Peduga, R. Evidence-Based Guideline; steroids and antivirals for Bell's palsy. Neurology 2012;79:115

Question 187: Contemporary Issues - Ethics/Professionalism

Discussion:

Durable power of attorney (DPA) for health care, or proxy directive, is a legal document in which one person assigns another person authority to perform specified actions on behalf of the signer. The power is "durable" because, unlike the usual power of attorney, it continues in effect if the signer of the document becomes incompetent (Beauchamp & Childress, 1994). Living wills which are specific substantive directives regarding medical procedures that should be provided or forgone in specific
circumstances are also valid legal instruments. Both Living Wills and DPA’s protect autonomy interests and may reduce stress for families and health professionals who fear making the wrong decision, but they also generate both practical and moral problems, including the often vague language used in Living Wills, as well as the availability of the assigned surrogate in terms of his or her own ability to make competent decisions at the time of the patient's issues of withdrawal of support. However, both instruments are a positive trend in the ultimate decision making for terminally ill patients when language is clear and surrogates are well informed.

References:

Question 188: Contemporary Issues - Practice

Discussion:
Management of antiplatelets and anticoagulants perioperatively must be done weighing the risk of a thromboembolic event (TE) versus the risk of bleeding. For most dental and many dermatologic procedures, these medications can be continued. When anticoagulants are stopped, the risk of TE increases significantly after 7 days. Bridging with heparin has shown an increase in bleeding complications.

References:

Question 322: Contemporary Issues - Evidence-based Medicine

Discussion:
The sensitivity is the number of true positives over true positives plus false negatives. In this case, that is 95/95+5 or 95%. The false positive rate is 1% (controls with an abnormal EMG) and false negative rate is 5% (normal EMG in patients with the gold standard clinical diagnosis of CTS). Specificity is the number of true negatives over true negatives plus false positives (99/99+1) or 99%.

References:

Question 336: Contemporary Issues - Practice

Discussion:
Organ donation most commonly occurs in the context of severe brain injury and brain death. Public awareness and support of organ donation and transplantation has increased in the last decade. As a result, families often recognize that the patient's circumstances may be such that organ donation is a possibility. The family's request is not illegal, nor does it indicate they are inappropriate decision makers for the patient because the patient has previously indicated the desire to be an organ donor. While it is usually considered improper for physicians to raise the topic of organ donation with a family before brain death has been determined, it is proper to respond to family requests about organ donation whenever they are made. Organ donation can occur even for "coroner's cases" as the coroner can be in the operating room at the time the organs are removed for transplantation. Because patients must be evaluated for medical suitability for organ donation, and because conversations with families about organ donation require considerable skill, physicians should work closely with representatives of organ procurement organizations when organ donation is considered a possibility.

References:
Question 365: Contemporary Issues - Practice

Discussion:

If possible, avoidance of valproate (VPA), phenytoin, and antiepileptic drug (AED) polytherapy during the first trimester of pregnancy should be considered to decrease the risk of major congenital malformations. If possible, avoidance of VPA and AED polytherapy throughout pregnancy should be considered to prevent reduced cognitive outcomes. As the patient refuses coming off valproate, the use of folic acid is important to reduce the risk of fetal malformations.

References:


Question 399: Contemporary Issues - HIPAA

Discussion:

HIPAA regulations regarding patients' protected health information (PHI) are important to safeguard patients' privacy and confidentiality. HIPAA regulations are not intended to interfere with the legitimate exchange of PHI between health professionals caring for patients, as delay of information sharing could result in patient harm. The United States Department of Health and Human Services Office of Civil Rights states that "(physicians) may disclose protected health information for the treatment activities of any health care provider...." For example, a primary care provider may send a copy of an individual’s medical record to a specialist who needs the information to treat the individual, or vice versa. In addition, the circumstance need not be a medical emergency, nor is there a requirement that the request be made in writing as long as copies of the records are being sent for the purpose of patient care.

References:


Question 402: Contemporary Issues - Business

Discussion:

The electronic medical record (EMR) can aid in preventing medication errors by minimizing errors caused by poor handwriting and misinterpretation of written abbreviations. Some EMR systems provide automatic checks for medication interactions and may also prompt the provider with practice guidelines and dosing algorithms; many systems also provide the option for electronically transmitting prescriptions to the patient's pharmacy. The EMR provides a health record that can be coordinated and accessed across systems of care as well as among multiple providers.

References:


Question 404: Contemporary Issues - Practice
Discussion:

Carbamazepine or oxcarbazepine have the highest levels of evidence for pain control in trigeminal neuralgia, while baclofen and lamotrigine may be considered useful. The symptoms are less consistent with temporal arteritis, which would likely be treated with prednisone.

References:


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Neuroimaging

Question 82: Neuroimaging - Developmental/Neurogenetic Disorders

Discussion:

The correct answer is Alexander disease. Defects in the synthesis or maintenance of myelin are characteristic of leukodystrophies, including cerebrotendinous xanthomatosis (predominately cerebellar dysmyelination), Alexander disease (predominately frontal lobe dysmyelination), and X-linked adrenoleukodystrophy (predominately parieto-occipital dysmyelination). Loss of myelin in these conditions tends to be confluent and symmetric, rather than the focal or multifocal lesions seen in neuromyelitis optica (optic nerve, periventricular, and spinal cord) and CADASIL (temporal lobe predominant).

References:


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Question 214: Neuroimaging - Developmental/Neurogenetic Disorders

Discussion:

Arachnoid cysts are congenital space occupying lesions that may result in distortion and displacement of brain tissue. These also may result in deformity of the overlying skull with alteration in contour, and thinning of the calvarium, as in this case. This lesion is extra-axial, excluding the tumor diagnosis. Cortex is preserved, excluding the diagnosis of MCA infarction. The calvarium is not eroded. Arachnoid cysts (correct diagnosis) do not have a high likelihood of causing seizure, and typically are clinically silent, neurologically.

References:


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Question 216: Neuroimaging - Critical Care/Stroke

Discussion:

The CTA of the neck depicted in Figure A demonstrates tapered narrowing at the origin of the right internal carotid artery with no subsequent distal flow. This is further confirmed on the MRA of the head which shows no flow signal involving the intracranial right internal carotid artery. There is cross-filling to the right middle cerebral as a result of anterior cerebral artery and posterior communicating flow. The MRA of the neck and brain on C and D, respectively, demonstrate reestablishment of flow involving the right internal carotid artery, both at the cervical and intracranial levels. The pattern of the initial right internal carotid artery occlusion and decannulation is typical of a carotid dissection. Fiber muscular dysplasia is often associated with a bead-like multisegmental stenosis in the upper internal carotid arteries, not seen on the current study. No carotid aneurysm is
seen. Atrial fibrillation can result in an occluded internal carotid artery which can, in some instances, recannulate. However, the tapered stenosis seen on the CTA is not typical for a thrombotic occlusion.

References:


Question 217: Neuroimaging - Tumors/Cysts

Discussion:

An intra-axial pontine lesion is noted that heterogeneously enhances. Moderate mass effect is present. This is not in a vascular distribution and not the ring-enhancing pattern seen with MS. The most likely diagnosis is a glioma.

References:


Question 218: Neuroimaging - Critical Care/Stroke

Discussion:

The correct answer is the foramen of Luschka. Also known as the lateral apertures of the 4th ventricle, the paired foramen of Luschka allow drainage of CSF from the ventricular system to the extraventricular space. The foramen of Monro connects the two lateral ventricles. The foramen of Magendie, or the medial aperture, is a midline structure which drains CSF from the fourth ventricle into the cisterna magna. The prepontine cistern is a CSF-containing extraventricular space which lies between the basilar bone and the pons. The interpeduncular cistern is a CSF-containing extraventricular space which lies anterior to the brain stem between the cerebral peduncles.

References:


Question 219: Neuroimaging - Spine

Discussion:

The CT scan of the thoracic spine demonstrates a hyperdense mass within the spinal canal with involvement of the adjacent posterior spinous process typical of a lytic lesion. The T2-weighted MRI similarly demonstrates infiltrating of ovoid mass extending into the posterior vertebral body within the spinal canal and within the posterior bony elements of the soft tissue. A hematoma can present as an ovoid hyperintense mass; however, one would not expect lytic destruction within the posterior spinous process or involvement of the posterior vertebral body; nor would these features be seen with an ependymoma or meningioma. Neurofibromas are slow-growing expansile lesions and result in remodeling of bone, particularly neural foramen and not a lytic mass as shown. Of the choices, a metastatic tumor is the most likely.

References:


Question 221: Neuroimaging - Tumors/Cysts

Discussion:

The neuroimaging studies demonstrate a heterogenously enhancing lesion in the left hemisphere with surrounding vasogenic edema and extension into the splenium of the corpus callosum. This appearance is most suggestive of a high-grade glioma.
Cerebral abscesses may look similar, but typically do not have the lacy, fibrillary network of enhancement within the lesions such as the glioma in this case. Primary CNS vasculitis is best demonstrated by segmental stenosis of the intracranial arteries in a "string of beads" appearance. Neurosarcoidosis typically shows strong enhancement of the leptomeninges and granulomas of the parenchyma. Acute intracerebral hemorrhage would be readily apparent on noncontrast CT. Arteriovenous malformation typically shows a network of flow voids that are seen on T2-weighted imaging. Tumefactive demyelination shows an open rim of enhancement, typically directed away from the ventricular surface.

References:


Question 223: Neuroimaging - Critical Care/Stroke

Discussion:

The following figure is a FLAIR-weighted scan sequence at the level of the upper midbrain. There is increased signal within the interpeduncular cistern. While this pattern could be seen as a result of multiple etiologies, including meningitis or subarachnoid hemorrhage, meningitis is not one of the choices listed. Preeclampsia is often associated with features of reversible posterior encephalopathy syndrome which on FLAIR produces regions of increased signal along the juxtacortical or cortical regions not seen on the current study and does not result in fluid level with abnormal signal in the perimesencephalic cistern. The parasagittal sinus shows decreased signal consistent with patency. Pituitary apoplexy can result in a similar clinical presentation; however, the abnormal signal and the perimesencephalic cistern would not be expected. This type of hemorrhage can be non-aneurysmal or less often aneurysmal in etiology.

References:


Question 227: Neuroimaging - Infection

Discussion:

Treatment of HIV patients with combination antiretroviral therapy with transient paradoxical worsening of infectious processes is recognized as immune reconstitution inflammatory syndrome (IRIS). Immune reconstitution inflammatory syndrome is due to rapid, dysregulated restoration of the immune system in HIV patients with resultant worsening of active or subclinical infectious processes. This is often seen in the setting of infection by intracellular pathogens such as mycobacteria (tuberculous and non-tuberculous), cryptococcus, CMV, HSV, VZV and JCV. The inflammatory reaction is usually self-limited but occasionally may be severe enough to result in long-term sequelae and fatal outcomes. As pointed out in the question, the patient has PML, since his CSF was positive for JC virus, the causative agent of PML on polymerase chain reaction testing.

References:


Question 229: Neuroimaging - Critical Care/Stroke

Discussion:

The images demonstrate subarachnoid hemorrhage. This case is most likely due to Call-Fleming syndrome which is a reversible segmental vasoconstriction of cerebral arteries manifested by a thunderclap headache and can be associated with subarachnoid hemorrhage.
hemorrhage, and is associated with antidepressant agents such as SSRIs, vasoactive sympathomimetic drugs, and antimigraine agents. Intracranial hypotension shows increase FLAIR signal along the pachymeninges not subarachnoid. The sagittal sinus is clearly patent. There is no history or imaging findings to suggest trauma. Posterior reversible encephalopathy syndrome results in cortical and juxtacortical white matter edema not present on this study.

References:


Question 230: Neuroimaging - Tumors/Cysts

Discussion:

The MRI images of the thoracic spine demonstrate an intrathecal ovoid lesion posterior to the cord at the T4-5 interspace level down to the upper aspect of the T7 vertebral body which anteriorly displaces the spinal cord with significant cord compression. The cyst has isointense signal with CSF both on the T2 and T1 weighted post-contrast view and shows no enhancement. There are multiple irregular areas of decreased T2 weighted signal posterior to the cord both superior and inferior to the cyst. These segments are due to flow artifact. Flow artifact would not produce displacement or compression of the spinal cord as seen adjacent to the cyst. A epidermoid cyst can have signal characteristics similar to an arachnoid cyst, although it is a less common finding. A intrathecal lipoma would have bright signal in the T1 weighted scan sequence as opposed to the decreased signal as in this instance. A hematoma would no appear isointense to CSF.

References:


Question 234: Neuroimaging - Infection

Discussion:

Toxoplasmosis lesions on MR typically have hypointense to isointense signal and are surrounded by hyperintense T2 and FLAIR signal from prominent vasogenic edema. Post contrast T1 images show multiple nodular or ring enhancing lesions that typically involve the basal ganglia, thalamus and corticomedullary junctions. Toxoplasmosis doesn't affect the ependyma, as lymphoma typically does. Cerebral toxoplasmosis is the most common opportunistic CNS infection in AIDS patients and occurs in 15%-50% of cases. Toxoplasmosis is frequently differentiated on a clinical basis from lymphoma by treatment with anti-Toxo antibiotics. If improvement doesn't occur, then more specific testing to establish or exclude diagnosis of lymphoma is undertaken. Neither HIV encephalopathy nor PML are associated with mass effect and contrast enhancing lesions as seen in the current example. When Coccidiomycosis affects the CNS in HIV+ patients it usually does so as a meningitis.

References:


Question 235: Neuroimaging - Tumors/Cysts

Discussion:

The correct answer is pituitary adenoma. Adenomas are typically homogenously enhancing and exert mass effect on adjacent structures, in this case, the bilateral cavernous sinuses and the optic chiasm. Pituitary adenomas arise from within the sella. Chordomas are heterogenous tumors consisting of notochord remnants which most often occur in the clivus and sacrococcygeal regions. Rathke cleft cysts are non-enhancing. Craniohypophygiomas are more often centered in the suprasellar (rather than intrasellar) compartment and contain cysts and calcification. Meningiomas are dural-based enhancing lesions which can arise from the dura overlying the sella turcica (i.e. the diaphragma sellae) or from the walls of the cavernous sinus. Meningiomas do not arise from the intrasellar compartment.
References:


Question 238: Neuroimaging - Developmental/Neurogenetic Disorders

Discussion:

No lipoma is present. Complete corpus callosal agenesis is demonstrated. There is a high association with other brain anomalies. Patients with callosal complete agenesis have a decreased volume of white matter compared with normals, reflecting the 9% to 10% of brain volume comprised of the corpus. If isolated, there is no significant increase in seizures. The ventricles are near to parallel and relatively straightened compared to the normal shape.

References:


Question 239: Neuroimaging - Epilepsy

Discussion:

The following figure demonstrates a segment of markedly thickened cortical ribbon involving the left frontal lobe characteristic of a cortical migration defect, which is associated with epilepsy. Adjacent to it in the left frontal region is segment of yellow l corresponding to the region of cortical activation from a verb generation task. There is a small mirror image on the right side. A small contralateral activation is a very frequent finding and is not indicative of true bilateral localization. The left frontal lesion does not have an appearance suggestive of a glioma. The relative lack of significant overlap between the migration defect and the language localization which would if similarly confirmed with other verbal task the finding that a surgical resection could be performed on the adjacent abnormality.

References:


Question 240: Neuroimaging - Tumors/Cysts

Discussion:

A large, cystic, rim-enhancing mass is present with a more solid component along the anterior margin. Porencephaly would not enhance, nor would a lipoma. Abscess would be more homogeneously enhancing around the rim and produce more prominent hemiparesis. Hemangioblastomas typically occur in the posterior fossa. The best answer is cystic astrocytoma.

References:


Question 242: Neuroimaging - Infection

Discussion:

The CT and MRI studies demonstrate a cystic defect within the left anterior frontal region. The marked decreased density and lower than water density would be typical of air. Similarly, the marked loss of signal on the T1-weighted study is characteristic of air and lower than signal that is seen due to CSF. A lipoma would have increased signal on the T1-weighted sequence and higher
density than seen on the CT scan. An empyema is an infection within the subdural or epidural space and is not present on this study. A porencephalic cyst would have greater density more typical of CSF and higher signal, as is also the case for an abscess.

References:


Question 244: Neuroimaging - Multiple Sclerosis/Autoimmune Disorders (Non-MS)

Discussion:

Elongated lesion demonstrating hyperintensity on T2-weighted images is most consistent with multiple sclerosis (MS) and represents focal demyelination. Degenerative myelomalacia may have these signal characteristics and ill-defined margins, but there is no adjacent spurring or other degenerative bony disease to have caused this problem. Ependymoma would be seen as a more focal mass with cord enlargement. Syrinx has well-defined cavity margins, although it would have a signal intensity similar to the lesion pictured. The lesion in a B12 deficiency involves the dorsal columns. MS cord lesions are usually one to two segments, or less, in length.

References:


Question 246: Neuroimaging - Spine

Discussion:

Abscess is not the best response because spinal cord abscesses have an acute onset, often with fever, and their course develops rapidly enough that abscesses do not attain the massive size of the lesion shown. Cavernous hemangioma is not the best response because of the very gradual onset this patient suffered that is not typical for cavernous hemangiomas, which typically present with acute or stepwise progression due to mass effect from bleeding. Ependymoma is the best response for this surgically proven case. The presence of a well-defined intramedullary mass with contrast enhancement, T2 hyperintensity and T1 hypointensity in the cord, cystic components adjacent to the solid enhancing portions of the tumor are typical. Also typical is subtle T2 hypointensity along the margins of the well-defined solid portion of the tumor. Metastatic melanoma is not the best choice because intramedullary metastases in the spinal cord are rare, occurring in 1-3% of all patients dying of cancer, and because if a melanoma metastasis had occurred in the spinal cord, the clinical decline would have been more rapid than this patient's slow disease progression. Multiple sclerosis is not the best response because the degree of mass effect seen in this case is not typical for multiple sclerosis, and the bulky well defined mass and the pattern of enhancement are not features seen in multiple sclerosis of the spinal cord.

References:


Question 250: Neuroimaging - Critical Care/Stroke

Discussion:
This child became dehydrated and developed dural and deep cerebral venous thrombosis. The abnormal findings of enlarged arterial flow voids are absent on MR, so arteriovenous malformation (AVM) is not plausible. The findings are not supportive of nonaccidental trauma, as there are no extraaxial collections; nor are there clinical features to suggest this. This child did well with treatment, and the follow-up imaging was normal with recanalization of the thrombosed venous structures. The outcome may be fatal (8% of neonatal cases); up to 50% will develop venous infarct. This is not a normal variant.

References:

Question 253: Neuroimaging - Metabolic, Movement, CSF Circulatory Disorders
Discussion:
The typical features of superficial siderosis of the CNS are deafness and cerebellar ataxia, which is reported in 90% of cases. Hemosiderin deposition in the subpial layers of the brain and spinal cord causes parenchymal damage by chronic slow or repeated bleeding into the subarachnoid space. The MRI demonstrates hemosiderin deposition along the upper cerebellar folia with adjacent cerebellar atrophy.

References:

Question 256: Neuroimaging - Tumors/Cysts
Discussion:
CT and MR images demonstrate infiltration and destructive change in the osseous structures of the orbit, with secondary mass effect on the orbital structures causing proptosis. Thyroid eye disease and idiopathic orbital pseudotumor are diseases that are limited to the soft tissues of the orbit. Ocular melanoma would arise in the ocular globe, and then extend to adjacent tissues. Orbital cellulitis is characterized by infiltration of fat and soft tissue, but not bone destruction. This is a case of metastasis from prostate cancer. Breast, melanoma, and prostate cancer are the leading causes of orbital metastasis.

References:

Question 259: Neuroimaging - Infection
Discussion:
The CT study demonstrates profound cortical atrophy with ventriculomegaly on an ex vacuo basis along with calcifications along the ependymal margin. This is a frequent finding in a setting of congenital cytomegalovirus infections. Fahr disease is an idiopathic disorder with calcifications involving the basal ganglia, the cerebellar, dentate nuclei, and at times, the cortical ribbon, which is not the pattern seen. There is no evidence of an obstruction of the ventricular system seen. Congenital herpes encephalitis results in encephalomalacia involving the mesial temporal, parasagittal, frontal, and insular cortical regions, which is not the pattern seen here. Mitochondrial cytopathies are not associated with ependymal calcifications, as in this case.

References:

Question 261: Neuroimaging - Tumors/Cysts
Discussion:

The images show a cystic cerebellar hemispheric mass with an enhancing mural nodule. T2-weighted images suggest the presence of small vessels within the mural nodule as hypointense flow voids. The mass is intra-axial and infratentorial. The differential diagnosis includes ganglioglioma, medulloblastoma, cystic astrocytoma, and abscess. Demyelinating plaques may present as a mass lesion, but this is unusual and would present with a ring of enhancement, not this pattern. The finding is most consistent with a cerebellar cystic hemangioblastoma.

References:


Question 262: Neuroimaging - Spine

Discussion:

The correct answer is "Type 2 odontoid fracture, spinal immobilization and neurosurgical/orthopedic evaluation". Type 1 odontoid fractures involve the tip of the dens and are stable fractures. Type 2 odontoid fractures occur at the base of the dens and are considered unstable. Type 3 odontoid fractures involve the base of the dens as well as the facets lateral to the odontoid process, and can be either stable or unstable. Spinal immobilization and neurosurgical/orthopedic evaluation is essential for type 2 odontoid fractures given their instability. In this case, the fracture results in slight anterior displacement of the odontoid (seen on the sagittal image). The greater the displacement, the greater the risk of non-union. This patient underwent surgical placement of an odontoid screw for fixation of the fracture.

References:


Question 264: Neuroimaging - Metabolic, Movement, CSF Circulatory Disorders

Discussion:

Intracranial hypotension is the diagnosis. As is typical, there is prominent thickening and enhancement in the dura. Small subdural effusions are visible on the T2 weighted axial image. The patient has the typical clinical symptoms of a postural headache and had spinal needle manipulation for anesthesia during her recent labor and delivery. Eclampsia and posterior reversible encephalopathy syndrome are related in that eclampsia is a particular form of posterior reversible encephalopathy that occurs exclusively in pregnancy. HELLP syndrome is a complication of pregnancy that occurs with hepatic and hematologic abnormalities in a small fraction of cases of eclampsia. The brain imaging findings for these diagnoses are identical and consist of regions of abnormal hyperintense T2 and FLAIR signal in cortical and subcortical tissue, typically in the posterior portions of the cerebral hemispheres. These finding are not present in the images shown. Rather, the abnormality is diffuse non-nodular dural thickening. Meningitis is not a good choice based on the clinical presentation of postural headache, because with meningitis the headache is not postural and is typically associated with nuchal rigidity, not present in the this patient. Also, the pattern of abnormal enhancement is not in the subarachnoid space, which typically is present in about half of meningitis cases; the other may have no abnormal findings on imaging, explaining the importance of CSF analysis in meningitis.

References:


Question 265: Neuroimaging - Spine
Discussion:

This MRI demonstrates an intradural, extramedullary dural-based enhancing mass that most likely represents a cervical meningioma.

References:


Question 268: Neuroimaging - Dementia

Discussion:

The patient is likely to have idiopathic calcification of the basal ganglia, also known as Fahr disease. CT scans demonstrate marked calcification of the basal ganglia and in the cerebellum. Calcification appears hypointensity on T2-weighted MRI. None of the possibilities listed can cause this degree of calcification. Leigh disease causes necrosis of the basal ganglia, particularly putamen, and results in low density of CT and hyperintensity on T2-weighted images. With amyloid angiopathy, hyperparathyroidism, and Wilson disease, there is a known disorder associated with lesser calcifications. In conditions where no such metabolic derangement can be found to explain the calcifications, the diagnosis is idiopathic calcification of the basal ganglia, or Fahr disease.

References:


Question 273: Neuroimaging - Multiple Sclerosis/Autoimmune Disorders (Non-MS)

Discussion:

The small ovoid lesions, many of them with the appearance of Dawson fingers, are typical of multiple sclerosis. On the T1-weighted gadolinium-enhanced images, some of the lesions are old, but some are enhancing, indicating a breakdown of the blood-brain barrier and active disease. As old and recent lesions coexist, a relapsing-remitting form is most likely. Sarcoidosis and CADASIL are much less frequent, as is adrenoleukodystrophy, particularly in a woman of this age.

References:


Question 275: Neuroimaging - Tumors/Cysts

Discussion:

There is a nearly homogenously enhancing mass within the right parietal occipital lobe with adjacent vasogenic edema. The near homogenous enhancement is not the pattern that would be seen in the setting of an abscess and is more typical of lymphoma. Lymphoma is more frequently bright on diffusion-weighted views as compared to glial tumors. Hemangioblastomas are most frequently seen in the posterior fossa. No hemorrhage is seen to suggest a cavernoma.

References:


Question 276: Neuroimaging - Spine

Discussion:
The sagittal figures demonstrate a somewhat homogenously infiltrative process within the C5, C6, and C7 vertebral bodies, with encroachment into the anterior epidural space with cord compression. There is little in the way of adjacent edema or fluid pockets as would be expected with an infectious process. There is epidural infiltration, however, without evidence of calcification of the posterior longitudinal ligament. There are no compression fractures. The patient may have osteoporosis, however, that in itself would not explain the patient's loss of balance. The infiltrative changes, which include the adjacent disc interspaces, are not those of congenital bone deformities and hypoplastic discs seen with a Klippel-Feil deformity, nor is there any evidence of any traumatic injury demonstrated on these views.

References:


Question 277: Neuroimaging - Critical Care/Stroke

Discussion:

A 1.6 x 0.9 centimeter mass is seen that represents a saccular aneurysm of the left internal carotid artery that projects medially. After diagnosis, this aneurysm was successfully treated with endovascular coil.

References:


Question 280: Neuroimaging - Tumors/Cysts

Discussion:

The patient has a history of prostate cancer, which based on MRI sequences has metastasized to the spine. The hypointense (dark) findings in the vertebral bodies are related to edema related to the metastases. Typical hemangiomas have a hyperintense appearance on T1 precontrast views along with a honeycomb appearance. Spondylosis is another name for degenerative disc disease which is not a pertinent finding on these images. Radiation induced changes would cause diffusely hyperintense signal on TI precontrast views due. There is no loss of height at any level to suggest fracture.

References:


Question 281: Neuroimaging - Developmental/Neurogenetic Disorders

Discussion:

The syndrome of septooptic dysplasia (de Morsier syndrome) consists of hypoplasia of the optic nerves, hypoplasia or absence of the septum pellucidum. Clinical presentation may include nystagmus, diminished visual acuity and approximately two-thirds of affected patients will also have hypothalamic-pituitary dysfunction. This MRI scan demonstrates absence of the septum pellucidum, hypoplasia of the corpus callosum and hypotelorism (closely set eyes).

References:


Question 282: Neuroimaging - Spine

Discussion:
The figures demonstrate hyperintensities along the posterior columns. This pattern can be seen in the setting of B12 (cobalamin) deficiency and copper deficiency. Multiple sclerosis and transverse myelitis is less likely to take on this pattern. Iron abnormalities do not produce cord changes.

**References:**


**Question 283: Neuroimaging - Metabolic, Movement, CSF Circulatory Disorders**

**Discussion:**

A. (colloid cyst) is the best response because the culprit lesion for this patient's acute hydrocephalus is the hyper dense lesion that has acutely obstructed outflow from both lateral ventricles, resulting in their acute enlargement and ballooning of third ventricle that causes effacement of the interpeduncular and suprasellar cisterns. E. (subarachnoid hemorrhage is not the best response because there is no visible subarachnoid bleed, and SAH does not explain the dense lesion in the anterior superior third ventricle. B. (bacterial meningitis) is not best response because the patient does not have fever, neck stiffness, and the course has been for several weeks and bacterial meningitis does not explain the dense lesion in the anterior superior third ventricle. D. (pituitary apoplexy) is not best response as the pituitary is not enlarged and this diagnosis does not explain the diffuse cerebral swelling or the hyper dense lesion in the anterior superior third ventricle. C. (Pseudotumor cerebri) is not best response because it does not produce obstructive hydrocephalus.

**References:**


**Question 284: Neuroimaging - Tumors/Cysts**

**Discussion:**

The pituitary lesion shown on the left side of the gland demonstrates classic features of a pituitary microadenoma on the noncontrast MRI, with focal hypointensity less than one centimeter in size. On the image shown, there is less enhancement of the microadenoma as compared to normal pituitary tissue. An epidermoid is a possibility but much less likely. The sella does not appear empty in the images. Rathke cleft cyst would have less signal intensity on T1 and no enhancement. Craniopharyngiomas are suprasellar, whereas this one is within the sella.

**References:**


**Question 286: Neuroimaging - Multiple Sclerosis/Autoimmune Disorders (Non-MS)**

**Discussion:**

The small and medium sized vessels are ragged and demonstrate the segmented pattern of vasculitis. While vasospasm is possible, this typically affects larger vessels than those affected in this case. This appearance is known as "beads on a string".
Question 287: Neuroimaging - Dementia

Discussion:

The morphologic features of a 42-year-old man with short stature, round face, epicanthal folds, and macroglossa, are some of the typical features seen in individuals with Down syndrome. Down syndrome is highly associated with early-onset dementia of the Alzheimer type. The images provided demonstrate moderately severe widening of the cortical sulci and enlargement of the lateral ventricles, particularly the temporal horns indicative of temporal atrophy. This pattern of atrophy is typical of dementia of the Alzheimer type.

References:


Question 290: Neuroimaging - Multiple Sclerosis/Autoimmune Disorders (Non-MS)

Discussion:

The scans show fusiform cord enlargement, best appreciated on T2-weighted images, extending over several spinal levels. Increased intramedullary T2 signal is noted in areas corresponding to cord enlargement. The findings are not entirely specific but are most consistent with acute myelitis as compared to the other choices. The MRI findings in various forms of myelitis are nonspecific, including those of idiopathic myelitis and those of myelitis associated with identifiable causes such as infections and postvaccination, postinfectious, and collagen vascular diseases. Such findings typically include spinal cord enlargement, intramedullary increased T2-signal lesions, and variable enhancement involving several spinal levels. The lack of a hemorrhagic component weighs against a spinal cord contusion. The lack of enhancement could be seen with myelitis but argues strongly against ependymoma. An astrocytoma would be unlikely to present with acutely and would present with cord enlargement.

References:


Question 292: Neuroimaging - Spine

Discussion:

The MRI views demonstrate a mass lesion extending into the anterior spinal canal at the L4-L5 interspace level. The imaging featured is typical of a large disc extrusion, and while a neurofibroma can on occasion look similar to a disc extrusion, this lesion is continuous with the L4-L5 disc, and on that basis is far more likely that etiology. The disc likely impinges the rising L5 root and exiting L4 nerve root. The other choices, including lymphoma and metastatic disease, are highly unlikely to appear continuous with a disc herniation, or have the same signal characteristics as a disc herniation.

References:


Question 293: Neuroimaging - Critical Care/Stroke
Discussion:

A modern CT using multidetector technique and reformatted images achieves greater than 95% sensitivity in the first 24 hours after onset of nontraumatic subarachnoid hemorrhage. CSF circulation and resorption of the blood results in reduced sensitivity to the hemorrhage to about 50% at 1 week.

References:


Question 296: Neuroimaging - Spine

Discussion:

Correct response is extradural mass effect on C5-C6 foramen by disk herniation. The elevated dura is visible as a thin hypointense line on the sagittal T2. This extradural localization makes the intradural responses (d. and e.) incorrect. The herniation lesion is at the C5-C6 level, centered at the disk, and is isointense to disk. Abscess, lymphoma, and metastasis would not be localized in the disk with this appearance.

References:

Hayes CW, Jensen ME, Conway WF. Non-neoplastic lesions of vertebral bodies: Findings in magnetic resonance imaging. Radiographics 1989;9(5):883-903

Question 297: Neuroimaging - Spine

Discussion:

The correct answer is Chiari 1 malformation. There is cerebellar tonsillar ectopia (approximately 1 cm below the foramen magnum) with an associated cervical syrinx, findings consistent with Chiari 1 malformation. The contents of the syrinx correspond to CSF (i.e. T1 hypointense and T2 hyperintense). The lesion has distinct margins and displaces rather than infiltrates adjacent spinal cord. Astrocytomas are infiltrating tumors. Basilar invagination refers to narrowing of the foramen magnum due to upward migration of the top of the C2 vertebrae. Multiple sclerosis causes demyelinating lesions of the brain and spinal cord and is not associated with tonsillar ectopia. Chordomas are tumors of notochord remnants which most often occur in the clivus and sacrococcygeal regions.

References:


Question 299: Neuroimaging - Spine

Discussion:

The correct answer is the traversing S1 nerve roots. The MRI shows a focal right paracentral posterior herniation of the L5-S1 intervertebral disc. The herniated disc material causes mild narrowing of the spinal canal and severe narrowing of the lateral recess at L5-S1. The S1 nerve roots traverses at the L5-S1 level, and is therefore the most likely to be compressed by this disc herniation. The exiting nerve roots at the L5-S1 level (the L5 nerve roots) will already be lateral to the disc herniation and will not be compressed.

References:


Question 303: Neuroimaging - Developmental/Neurogenetic Disorders
**Discussion:**

There is an ovoid extraaxial mass which has a cystic appearance and signal characteristics similar to CSF. There are reticulated linear foci within the lesion and marked high signal on diffusion images. This is the characteristic appearance of an epidermoid cyst. An arachnoid cyst would not have the subtle internal structure seen in this instance and would be isointense to CSF on all sequences, including the diffusion-weighted views. Tumors such as lymphoma or metastases have a higher T1-weighted signal and less hyperintensity as compared to this lesion. A benign bone cyst would not be expected to be bright on the diffusion-weighted views.

**References:**


**Question 304: Neuroimaging - Critical Care/Stroke**

**Discussion:**

The patient's case challenges recognition of the right subdural, left epidural, bilateral brain contusion, right uncal herniation and left scalp hematoma.

**References:**


**Question 305: Neuroimaging - Critical Care/Stroke**

**Discussion:**

The CT scan of the head demonstrates subarachnoid hemorrhage due to trauma. The CTA on the axial and sagittal views demonstrate contrast filling extending along the right cavernous sinus with engorgement of the right ophthalmic vein, extending to the right eye. The pattern is consistent with a cavernous sinus carotid artery fistula with arterial feeding from the cavernous carotid artery into the adjacent venous structure, resulting from the trauma. While there is a subarachnoid hemorrhage seen on this study, there are no imaging findings to suggest vasospasm, and while there are likely fractures within the calvaria and possibly the orbit, that is not the etiology of the patient's right ophthalmologic process. A posterior communicating artery aneurysm rupture would be unlikely to result in a fistula extending into the right eye, as seen. There is no evidence of optic nerve compression.

**References:**


**Question 307: Neuroimaging - Metabolic, Movement, CSF Circulatory Disorders**

**Discussion:**

Congenital aqueductal stenosis is the best answer because the caudal aqueduct is visibly obstructed with typical widening of the rostral portion. Third ventricle is dilated with "ballooning" of third ventricle so that it fills the suprasellar cistern, inferiorly displacing anterior visual pathway (optic tracts, chiasm, nerves). Chemical ventriculitis from ruptured dermoid is not the best response because the clinical presentation of ruptured dermoid of headache and seizure is not present and imaging findings of very hypodense fat in the ventricles and subarachnoid space are not seen.

**References:**

Question 308: Neuroimaging - Metabolic, Movement, CSF Circulatory Disorders

Discussion:

Thyroid associated ophthalmopathy (Grave's ophthalmopathy) is an autoimmune inflammatory condition of the orbit associated with hyper- or hypothyroidism. The mnemonic "I'M SLOW" (inferior > medial > superior > lateral > oblique) describes the order of predilection for involvement of the extraocular muscles. This MRI scan demonstrates enlargement of the extraocular muscles of the left orbit, particularly the inferior rectus. Treatment involves corticosteroids as well as correction of the underlying thyroid dysfunction.

References:


Question 309: Neuroimaging - Infection

Discussion:

Axial CT, T2, and post contrast T1 weighted images show an asymmetric nonenhancing lesion in the right frontal lobe with no mass effect. The lesion primarily affects the white matter. This is typical for PML.

References:


Question 310: Neuroimaging - Epilepsy

Discussion:

The figures demonstrate enlargement of the region adjacent to the choroidal fissure as a result of atrophy within the mesial temporal region, with associated segments of abnormal T2-weighted signal, characteristic of gliosis. The combination of these features is typical of mesial temporal sclerosis, which in the setting of atrophy is associated with epilepsy.

References:


Question 312: Neuroimaging - Multiple Sclerosis/Autoimmune Disorders (Non-MS)

Discussion:

The figures demonstrate enhancement of the left optic nerve. These are the imaging features seen with acute optic neuritis. Bartonella infection (catscratch disease) is frequently associated with a small focus of enhancement at the disc optic nerve, at the junction of the globe and not the long segment of enhancement seen on this study. An orbital cavernoma can present as an intra or extra conal mass; it would not be expected to result in a diffuse pattern of enhancement of the optic nerve. Similarly, orbital pseudotumor is associated with intra or extra corneal inflammatory tissues and not isolated enhancement of the optic nerve.

References:

Question 313: Neuroimaging - Critical Care/Stroke

Discussion:

Epidural hematoma is not the best response because the epidural fat surrounding the thecal sac is not displaced, and is peripheral with respect to the modestly T1 hyperintense subdural hematoma distorting the contents of the spinal canal. Epidural abscess is not the best response because signal intensity in the adjacent vertebrae and intervertebral disks is normal. Epidural abscess is usually a sequela of diskitis and osteomyelitis, diseases which are clearly not present. Spinal AVM can result in intraspinal hemorrhage in subarachnoid, intramedullary, epidural and subdural locations, but the cardinal finding for spinal AVM is the visible vascular malformation with flow signal voids and it is not present in this case. Subarachnoid hemorrhage is not the best response because the blood would be in the subarachnoid space, not distorting the subarachnoid space from outside as in this case of subdural hematoma. Note that this subdural hematoma extending from T11 through sacral levels is deep to the thin hypointense dark line of the dura, as well as deep to the epidural fat that is the useful marker of the epidural space. Spinal subdural hematoma may be spontaneous, may be related to anticoagulation use or bleeding disorders, infrequently with spinal trauma, and rarely with spinal vascular malformations. Spinal subdural hematoma presents clinically with acute onset of signs of spinal cord or cauda equina compression preceded with back or radicular pain. Occasionally, only isolated pain presents.

References:


Question 314: Neuroimaging - Dementia

Discussion:

The patient has amyloid angiopathy, most commonly seen in patients with Alzheimer disease. The other choices have no correlation with intraparenchymal microhemorrhages.

References:


Pathology

Question 22: Pathology - Neuromuscular Disease

Discussion:

Thickened blood vessels with basement membrane reduplication are quite typical of diabetic neuropathy. Apple-green birefringence on congo red stain indicated the presence of amyloid, whereas lymphocytic infiltrate would indicate an inflammatory neuropathy. Onion bulbs are seen in various neuropathies with repeated episodes of demyelination and remyelination, while macrophage mediated demyelination is typical of Guillain-Barre Syndrome.

References:


Question 36: Pathology - Neurodegenerative Disease

Discussion:
The clinical scenario suggests dementia with Lewy bodies. In addition to cortical Lewy bodies, striking spongiform change that is typically localized to the temporal cortex may be present. Spongiform change of the cerebral cortex may be found in conditions other than prion disease. These include corticobasal degeneration, Pick disease, dementia with motor neuron disease, and frontal lobe dementias. However, the spongiform change in these disorders is typically found in layer 2 of the frontal and temporal cortex. Spongiform-like changes can also be seen in cerebral hypoxia and edema.

References:

Question 52: Pathology - Demyelinating Disease

Discussion:
The genetic basis and pathophysiology of most of the leukodystrophies have been known for some time. Light has recently been shed on one of the last holdouts: Alexander disease. The morphologic hallmark of Alexander disease is the presence of profuse numbers of Rosenthal fibers. Rosenthal fibers are composed of densely compacted glial intermediate filaments made of glial fibrillary acidic protein (GFAP). Recent studies have demonstrated that a large percentage of Alexander disease cases are associated with a mutation in the GFAP gene.

References:

Question 63: Pathology - Developmental

Discussion:
Of those disorders listed, only polymicrogyria is considered a neuronal migration disorder. Anencephaly is due to failure of closure of the anterior neuropore, while hydranencephaly results from early neonatal hypoxic / ischemic injury. Dandy-Walker malformation is a posterior fossa disorder in which some or all of the cerebellar vermis is absent together with cystic dilatation of the 4th ventricle. Agenesis of the corpus callosum represents a midline pattern defect that can occur alone or in the context of other developmental abnormalities.

References:

Question 78: Pathology - Neuromuscular Disease

Discussion:
Malignant hyperthermia is a potential complication seen in patients with central core myopathy associated with the administration of certain anesthetic drugs such as succinylcholine and halothane. Screening for susceptibility to malignant hyperthermia is now done by testing for mutations in the ryanodine receptor RYR1 gene.

References:
Question 86: Pathology - Toxic/Metabolic Disease

Discussion:

The vehicle was no longer running, because it ran out of gasoline. The bilateral hemorrhagic lesions in globus pallidus are characteristic of carbon monoxide poisoning.

References:


Question 110: Pathology - Neurodegenerative Disease

Discussion:

The nucleus of Onufrowicz of the sacral cord is spared in motor neuron disease. The remaining structures or cells contain motor nuclei and are involved to varying degrees in amyotrophic lateral sclerosis.

References:


Question 125: Pathology - Infectious Disease

Discussion:

Most CNS infections are the result of spread of organisms from other sites in the body. Local extension from either the paranasal sinuses or the skull is unusual. Direct implantation of organisms can occur as the result of trauma or as a complication of surgery. Infection via the peripheral nervous system occurs in rabies.

References:


Question 138: Pathology - Tumors

Discussion:

The clinical and imaging findings depict an aggressive pediatric brain tumor, arising within the cerebellum and producing "drop metastases" along the spinal axis. Hemangioblastoma often arises within the cerebellum, but is more typically seen in adults. All of the other tumors listed may arise in children, however only medulloblastoma exhibits the biologically aggressive behavior (rapid growth and metastases) described in the current case.

References:


Question 163: Pathology - Epilepsy
Discussion:

Ammon's horn (hippocampal) sclerosis is the most common neuropathologic change associated with the complex partial seizures (described in the vignette) of intractable temporal lobe epilepsy. MRI studies typically reveal a unilateral atrophic hippocampus, which has a bright T2 signal. The affected hippocampus will show gliosis and neuronal loss of the CA1 pyramidal cell layer with CA3 and CA4 somewhat less involved. Area CA2 is relatively spared while the subiculum is typically uninvolved.

A history of prolonged severe febrile seizures is often identified in patients with intractable temporal lobe epilepsy (TLE). The etiological relationship of such seizures to eventual chronic TLE is unclear.

References:


Question 164: Pathology - Epilepsy

Discussion:

Ammon's horn (hippocampal) sclerosis is the most common neuropathological change associated with the complex partial seizures (described in the vignette) of intractable temporal lobe epilepsy. MRI studies typically reveal a unilateral atrophic hippocampus, which has a bright T2 signal. The affected hippocampus will show gliosis and neuronal loss of the CA1 pyramidal cell layer with CA3 and CA4 somewhat less involved. Area CA2 is relatively spared while the subiculum is typically uninvolved.

A history of prolonged severe febrile seizures is often identified in patients with intractable temporal lobe epilepsy (TLE). The etiological relationship of such seizures to eventual chronic TLE is unclear.

References:


Question 165: Pathology - Neurodegenerative Disease

Discussion:

The epsilon 4 variant of APOE is the only established risk factor for late-onset Alzheimer disease (AD). Mutations in genes for amyloid precursor protein and presenilin 1 and 2 are associated with early-onset familial AD. Hyperphosphorylated forms of tau accumulate in many forms of frontotemporal dementia but genetic variants of tau have not been identified as risk factors for late-onset AD.

References:


Question 178: Pathology - Neurodegenerative Disease

Discussion:

The combination of postural instability and falls, bradykinesia, dysarthria, gaze abnormalities, and a frontal-subcortical type of dementia points to a Parkinsonian disorder, but Parkinson disease is unlikely given the absence of response to L-DOPA. This combination of symptoms is most consistent with progressive supranuclear palsy (PSP), a sporadic, male predominant disorder.
Supranuclear gaze abnormalities, postural instability with frequent, serious falls, and lack of response to L-DOPA are all characteristic of PSP. PSP is a tauopathy characterized neuropathologically by neurofibrillary tangles within brainstem and deep grey nuclei. Lewy bodies are not a feature of PSP. Pick disease (with Pick bodies in the dentate fascia) is a form of frontotemporal degeneration and is not typically associated with movement disorder. Selective loss of spiny neurons in the caudate nucleus is a key finding in Huntington disease. It is not a feature of PSP. Cortical plaques and tangles are the essential neuropathologic findings in Alzheimer disease. Plaques are not characteristic of PSP. The neurofibrillary tangles of PSP are predominantly subcortical and have a different morphology (globose) than those usually seen in AD, although both are composed of tau filaments.

References:

Question 182: Pathology - Demyelinating Disease

Discussion:
This is a case of progressive multifocal leukoencephalopathy which is caused by JC virus; glassy plumb-colored intranuclear inclusions are found in oligodendrocytes in these cases, along with enlarged bizarre appearing astrocytes and areas of demyelination.

References:

Question 222: Pathology - Neuromuscular Disease

Discussion:
Nemaline myopathy is one of the most common congenital myopathies. There is a wide clinical spectrum of disease with the common scenario of infantile hypotonia. The defining pathologic feature of the disease is the finding of rod- or thread-like structures on Gomori trichrome stain. These structures are well demonstrated on toluidine-blue-stained plastic sections (shown in this case). Ultrastructure shows rod-like or oval, electron-dense bodies derived from the sarcomeric Z-line. At least five different mutations of genes may be responsible for the disease, genes that encode Z-line proteins or proteins attached to the Z-line such as sarcomeric skeletal muscle actin (ACTA1). The latter mutation was observed in this case. None of the other myopathies listed would have such striking Nemaline rods.

References:


Question 226: Pathology - Tumors

Discussion:
Pineocytomas are pineal tumors of adults. They often present with symptoms of CSF flow obstruction or mass effect. These are contrast-enhancing midline lesions which may invade adjacent tissue and seed CSF pathways. Pineocytomas resemble normal pineal, but have larger nuclei and more open chromatin. Mitoses are infrequent and there is no necrosis. There are frequently large anuclear areas termed pineocytomatous rosettes. This is a low-grade (WHO grade I) tumor for which gross total resection may result in no recurrence.

References:
**Question 232: Pathology - Neuromuscular Disease**

**Discussion:**

This muscle biopsy shows a predominance of tiny rounded atrophic fibers with few hypertrophic fibers, typical of SMA. ALS, another motor neuron disease, is more typical of adults and the muscle biopsy would contain numerous angulated atrophic fibers. Muscle fibers with multiple internal nuclei are seen with myotonic dystrophy, while small fibers with singular central nuclei are diagnostic of myotubular myopathy. Muscle samples from patients with Duchenne muscular dystrophy show variable numbers of degenerating and regenerating muscle fibers, fiber size variation, and interstitial fibrosis / fatty replacement, depending upon the stage of the disease.

**References:**


**Question 233: Pathology - Cerebrovascular Disease**

**Discussion:**

Giant cell (temporal) arteritis is a granulomatous vasculitis affecting chiefly the extracranial arteries of the head (superficial temporal, ophthalmic, carotid and vertebral); usually sparing intracranial vessels. It may also affect the aorta and its branches. Patients are usually over 55 years of age, with a mean age of 75 years. The central symptom is frequently headache. Other symptoms/signs include fever, visual impairment, scalp tenderness, malaise, myalgia, weight loss, anemia, jaw claudication, neck pain and tenderness of the temporal artery to palpation with decreased arterial pulsation. The etiology is not clear, although apparently cell mediated. Inflammatory cells are generally present in the media. Multinucleate giant cells are the hallmark (hence the name), but are not obligatory for the diagnosis. If present, they are usually seen along the internal elastic membrane and may engulf fragments of elastica. Fibrinoid necrosis may be present. Treatment with high dose steroids prior to the biopsy may result in a lack of inflammation and only scar formation to serve as proof of injury.

**References:**


**Question 237: Pathology - Neurodegenerative Disease**

**Discussion:**

The gross photograph of the brain shows lobar atrophy. Lobar atrophy of the frontal and temporal lobes is typical of Pick disease. Microscopically, intracytoplasmic argyrophilic neuronal inclusions and gliosis are observed in Pick disease. The patterns of atrophy in the other listed diseases would be in different distributions and much less by comparison.

**References:**


**Question 245: Pathology - Critical Care/Stroke**

**Discussion:**

The image depicts axonal spheroids which are indicative of diffuse axonal injury. Neurofibrillary tangles, neuritic plaques, granulovacuolar degeneration, and Hirano bodies are microscopic features of Alzheimer disease, whereas Pick and Lewy bodies
are typical of Pick disease and Diffuse Lewy body disease, respectively. Axonal spheroids have a superficial resemblance to fungal organisms, however one would expect to see an inflammatory response to the latter as well as positivity for PAS.

References:


Question 248: Pathology - Hypothalamus/Pituitary

Discussion:

A hypothalamic hamartoma is an unusual lesion of infancy and childhood that produces a discrete mass in the floor of the third ventricle. The lesion is composed of a disorganized hypocellular collection of mature neurons and glia. Hypothalamic hamartomas are associated clinically with precocious puberty and peculiar laughing seizures (gelastic epilepsy). These lesions can sometimes be endocrinologically active and have been associated with acromegaly, among other syndromes; thus simulating a pituitary adenoma. Pilocytic astrocytomas, craniopharyngiomas, and mature teratomas may also arise in this area, but such tumors usually have a cystic component.

References:


Question 251: Pathology - Cerebrovascular Disease

Discussion:

The myelin-stained section at the level of the inferior olivary nuclei shows infarction (pallor on this myelin stain) of the dorsolateral portion of the medulla in the distribution of the posterior inferior cerebellar artery. This usually results from occlusion of the vertebral artery and can present clinically as "lateral medullary syndrome".

References:


Question 254: Pathology - Demyelinating Disease

Discussion:

The periventricular translucent grey areas are subacute to chronic plaques with very sharp appearing borders. In conjunction with her current symptoms suggesting acute changes somewhere in her visual system, this would imply multiple sclerosis (MS). Young females are at greatest risk for MS. Progressive multifocal leukoencephalopathy (PML) is also a white matter disease, for which she has no obvious risk factors. None of the other choices give the white matter appearance seen here.

References:


Question 255: Pathology - Epilepsy

Discussion:

The sections show a neoplasm containing numerous neoplastic neurons which show immunoreactivity for synaptophysin. The tumor is low grade and lacks the cytological monotony of neurocytoma or medulloblastoma. Synaptophysin immunoreactivity
would be expected in ganglioglioma, medulloblastoma and neurocytoma but not craniopharyngioma. Cortical dysplasia would not present with a cystic mass. Medulloblastomas, by definition, arise in the cerebellum.

References:

Question 260: Pathology - Cerebrovascular Disease
Discussion:
Cerebral amyloid angiopathy is seen in patients who are usually over the age of 60 years and characterized by hemorrhages which tend to be superficial (lobar) with extension into the subarachnoid space. The amyloid is deposited in vessels in the cortex and leptomeninges and appears histologically as waxy, pink material in the vessel walls. This material may resemble the hyaline change of arteriolosclerosis. Congo red staining with apple green birefringence on polarization confirms that the material is amyloid. Aneurysmal dilatation and splitting (double-barrel?appearance) of the vessel walls is sometimes seen. Granulomatous reactions occur occasionally resulting in amyloid beta related angiitis. There is currently no treatment and patients are at risk for additional hemorrhage and infarction.

References:

Question 263: Pathology - Critical Care/Stroke
Discussion:
Subdural hematomas can occur in individuals with atrophy with minor trauma. The image shows a unilateral subdural hematoma with ipsilateral cingulate herniation. Epidural hematomas are often associated with a skull fracture with laceration of the middle meningeal artery. Coup contusions are unusual in cases where the head is in motion. Extensive contrecoup contusions would be unlikely in the setting of a minor fall.

References:

Question 266: Pathology - Basic Reactions
Discussion:
The surface of the brain is apparent (so the dura is attached to the skull) in this picture. The blood is therefore between dura and brain (subdural). As the brain atrophies with age, the dural bridging vessels become more prone to tearing while traversing the subdural space. The space inside her ventricles was decreased (by shunt), which increased the size of the subdural space, and she developed a hemiparesis from compression (mass effect).

References:

Question 267: Pathology - Cerebrovascular Disease
Discussion:
Hypertension is the most common cause of non-traumatic intraparenchymal central nervous system (CNS) hemorrhage. While intracerebral deep grey matter is the most common site for rupture of vessels secondary to hypertension, pons and rarely cerebellum also account for some cases. No obvious malformation or tumor is seen in this picture, and while they may be difficult in some case to find, the gross lack of other features increases the likelihood that the ?pure? clot is from vessel rupture. Some fungal infections are angioinvasive (possibly leading to vessel rupture) and he is diabetic (a risk factor). These infections usually directly enter the CNS from the sinuses in diabetics and affect dural/leptomeningeal vessels.

References:


Discussion:

Neurosarcoïd typically affects the basal cranial meninges; the spinal meninges may also be involved. Although many people with neurosarcoïd have no neurologic symptoms, symptomatic patients often have cranial neuropathies (50-75%). The facial nerve is the most frequent cranial nerve involved, sometimes bilaterally. Pituitary stalk involvement is common, so hypothalamic and pituitary dysfunction may occur. Half of patients also develop parenchymal lesions; the diencephalon being most frequently affected. Aseptic meningitis, nodular dural masses, and hydrocephalus may occur; peripheral nerve and muscle may be involved. The gold standard for diagnosis of neurosarcoïd is biopsy, although a presumptive diagnosis may be made of neurosarcoïd in someone with systemic disease and characteristic findings on scans. Often biopsies of neurosarcoïd are done because of a lack of systemic diagnosis and/or mass like lesions on scans. The pathology of neurosarcoïd demonstrates compact granulomas, which are all generally small and vary little, unlike those in other granulomatous diseases. When the sarcoïd granulomas coalesce, larger masses may be formed. Although the granulomas are usually non-necrotizing, they may demonstrate necrosis. The tightly packed epithelioid histiocytes may have associated multinucleated giant cells. This is a diagnosis of exclusion, and special stains for fungal and mycobacterial organisms are, of course, negative.

References:


Discussion:

The biopsy shows a severe vacuolar myopathy due to extensive glycogen storage. These findings are consistent with Pompe disease (glycogen storage disease type II), which is caused by a genetic deficiency of acid-alpha-glucosidase (acid maltase). In it's most severe form, the disease presents in infancy with cardiomegaly, hepatomegaly, progressive muscle weakness, macroglossia, and hypotonia ("floppy baby"). Of the choices provided, Pompe disease is the only disorder that presents findings of vacuolar myopathy on a muscle biopsy. Pathologic findings of Haltia-Santavuori disease, (infantile neuronal ceroid lipofuscinosis), and Alexander disease primarily affect the brain. Krabbe disease is a leukodystrophy affecting CNS white matter. Kufs disease is an adult form of neuronal ceroid lipofuscinosis.

References:


Discussion:

A coronal section of the brain at autopsy shows mammillary bodies that are congested and discolored, which is a characteristic finding in thiamine deficiency manifesting as Wernicke encephalopathy. Few other disorders affect the mamillary bodies.
Vitamin B12 deficiency can cause subacute combined degeneration, a spinal cord lesion. Methanol poisoning classically causes hemorrhagic putaminal lesions.

References:

**Question 291: Pathology - Infectious Disease**

**Discussion:**
Cerebral toxoplasmosis is characterized by microglial nodules with associated encysted bradyzoites. It should be distinguished from opportunistic fungal infections, which often produce granulomatous inflammation, and from CNS lymphomas, which are characterized by large lymphoid cells which typically surround blood vessels.

References:

**Question 294: Pathology - Tumors**

**Discussion:**
All of the tumors listed may present in the posterior fossa, however this image depicts the typical microscopic findings of hemangioblastoma (vacuolated stromal cells and variably sized vascular structures). The differential diagnosis for a cyst with mural nodule within the cerebellum includes pilocytic astrocytoma and hemangioblastoma, the former of which is predominantly pediatric and contains glial processes and Rosenthal fibers on histology. Dysplastic gangliocytoma (Lhermitte-Duclos) represents a WHO grade I lesion of the cerebellum containing dysmorphic ganglion cells resembling Purkinje cells, arising in the context of Cowden syndrome.

References:

**Question 300: Pathology - Tumors**

**Discussion:**
The image shows a colloid cyst of the third ventricle. Patients can occasionally manifest sudden death from this otherwise histologically benign lesion. Hypothalamic hamartoma would be within the hypothalamus, not the third ventricle. Lipomas classically are seen near the corpus callosum. Subependymomas only occur rarely in the third ventricle.

References:

**Question 301: Pathology - Tumors**

**Discussion:**
The subependymoma is an intraventricular, WHO grade I lesion that occurs most commonly as an incidental finding in middle-aged and elderly patients with a male predominance. It is a variant of ependymoma, characterized by clusters of bland nuclei in abundant, fibrillary stroma, albeit of lower grade. Symptomatic lesions may occur due to ventricular obstruction, but surgical removal is generally curative.
**Question 302: Pathology - Cerebrovascular Disease**

**Discussion:**

The photomicrograph shows a cavernous malformation with back-to-back vascular channels. This condition can occur as an autosomal dominant disorder and unfortunately is a frequent cause of morbidity and even mortality in affected individuals.

**References:**


**Question 306: Pathology - Neurodegenerative Disease**

**Discussion:**

The histologic hallmarks of Alzheimer disease are neuritic plaques, neurofibrillary tangles, Hirano bodies, neuronal granulovacuolar degeneration, and the deposition of amyloid in the walls of blood vessels. The pyramidal neuron illustrated showed granulovacuolar degeneration, a neurofibrillary tangle, and a Hirano body.

**References:**


**Question 333: Pathology - Developmental**

**Discussion:**

The oligodendrogliomas are susceptible to hypoxia because of their metabolic state. Microglia, astrocytes, and ependyma are relatively resistant to hypoxia. Neurons are resistant in young children because they have not yet been myelinated and are not metabolically active.

**References:**


**Question 338: Pathology - Cerebrovascular Disease**

**Discussion:**

The presenting signs and symptoms strongly suggest that this is a ruptured saccular or berry aneurysm. They frequently occur on the anterior communicating artery. Fusiform aneurysms of the basilar artery typically become atherosclerotic and are much less likely to bleed. Microscopic Charcot-Bouchard aneurysms of deep penetrating arteries and arterioles are primarily associated with hypertensive brain intraparenchymal hemorrhages. Congophilic material accumulates in cortical and leptomeningeal arteries in amyloid angiopathy, which most frequently cause lobar intraparenchymal hemorrhage. This is primarily a disorder of the elderly, and would be unusual in a young person. Laminar cortical necrosis results from hypoxic / ischemic injury to pyramidal cortical neurons and is not related to subarachnoid hemorrhage.

**References:**
**Question 342: Pathology - Neurodegenerative Disease**

**Discussion:**

Familial frontotemporal dementia that arises from a mutation on chromosome 17 results in a mutation of the gene encoding for tau protein.

**References:**


**Question 360: Pathology - Neuromuscular Disease**

**Discussion:**

In males suspected of having Duchenne's or Becker's muscular dystrophy, dystrophin analysis may reveal the presence of a mutation in affected individuals. Carrier analysis can also be performed. The precise location and size of the dystrophin mutation determines whether the more severe Duchenne's phenotype or the milder Becker's phenotype will be seen.

**References:**


**Question 394: Pathology - Developmental**

**Discussion:**

The cyst wall is the roof of the fourth ventricle. The vermis may be either absent entirely or hypoplastic in Dandy-Walker malformation, in association with an enlarged posterior fossa and frequent brainstem microscopic developmental abnormalities. The Chiari malformations have a small posterior fossa. Craniorachischisis is a neural tube which is open for the most part from one end to the other. Joubert syndrome shows the ?molar tooth? abnormality of brainstem.

**References:**


**Question 396: Pathology - Demyelinating Disease**

**Discussion:**

Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) results from missense mutations of the Notch 3 gene on chromosome 19. The small deep white matter vessels show replacement of the media by eosinophilic, periodic acid-Schiff positive, Congo red negative, granular material. Ultrastructurally, there is compact electron-dense material known as granular osmophilic material surrounding myocytes in the arterial media. Although the predominant effects of CADASIL are on the central nervous system, it is a systemic vasculopathy that can be appreciated on skin or nerve biopsy. Vasculitis is not seen in CADASIL.

**References:**

Question 400: Pathology - Neuromuscular Disease

Discussion:

Many histological findings are characteristic of some conditions; for example atrophic angular fibers, target fibers, fiber type grouping and group atrophy are indicative of neurogenic disorder.

References:


Question 411: Pathology - Demyelinating Disease

Discussion:

In neuromyelitis optica, women are affected up to four times more often than men and the clinical course of the disease is frequently rapidly progressive. Devic disease is much more common in Asia, with up to 8% of multiple sclerosis cases in Japan being of the Devic type. Oligoclonal bands are absent from the CSF in most cases. The presence of necrosis in spinal cord virtually defines the entity. The disease is now recognized to be related to antibodies to aquaporin 4 in the cell membranes of astrocytes which acts as a channel for the transport of water across the cell membrane. Demyelinating lesions are not seen in other areas of the brain as would be more typical of multiple sclerosis.

References:


Pharmacology/Chemistry

Question 8: Pharmacology/Chemistry - Movement Disorders

Discussion:

Because gastrointestinal monoamine oxidase (MAO) effectively prevents dietary pressor amines, typically tyramine, from entering the tissues, a marked hypertensive response (the "cheese reaction") can occur when subjects treated with antidepressant MAO inhibitors ingest foods or beverages rich in such amines. This is due to the potentiation of sympathomimetic activity of ingested tyramine present in cheese and other food stuff, resulting from its ability to release noradrenaline, when prevented from metabolism by MAO. Selegiline (a preferential MAO-B inhibitor) has no MAO-A inhibition at 10 mg/d or less, but at 20 mg/D or above it loses this specificity.

References:


Question 11: Pharmacology/Chemistry - Demyelinating Disorders

Discussion:
Animal studies suggest that a fall in post-partum estrogen levels may contribute to multiple sclerosis exacerbations. Progesterone has immunosuppressive effects and vitamin D may have a protective effect in this disorder. Prolactin may stimulate immune responses and can theoretically promote an exacerbation.

References:

Question 18: Pharmacology/Chemistry - Demyelinating Disorders

Discussion:

Fingolimod is a sphingosine-1 phosphate-receptor modulator approved for oral therapy for relapsing remitting multiple sclerosis. Activating the first subtype of the sphingosine-1 phosphate-receptor reduces lymphocyte recirculation from the lymph nodes resulting in functional immunosuppression. Activating the third subtype of the sphingosine-1 phosphate-receptor reduces heart rate and prolongs the PR interval. The cardiac effects of fingolimod are maximal after the first dose but persist for about 14 days. In April, 2012, the FDA and the European Medicines Agency revised the prescribing information for Gilenya based on independent safety reviews initiated by the agencies after deaths had been reported among patients taking Gilenya.

References:

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Schmouder R. Serra D. Wang Y. Kovarik JM. DiMarco J. Hunt TL. Bastien MC.. FTY720: placebo-controlled study of the effect on cardiac rate and rhythm in healthy subjects.. Journal of Clinical Pharmacology; 2006; 46(8):895-904,


Question 25: Pharmacology/Chemistry - Other Pain Syndromes

Discussion:

Daily doses of 400 mg or more of tramadol may provoke seizure activity.

References:


Question 33: Pharmacology/Chemistry - Epilepsy

Discussion:

Oxcarbazepine (and its chemical analogue carbamazepine) has been reported to produce hyponatremia due to the syndrome of inappropriate antidiuretic hormone secretion (SIADH). This is reversible after the drug is withdrawn.

References:


Question 34: Pharmacology/Chemistry - Neurogenetics

Discussion:
Fabry's disease is an X-linked defect in alpha-galactosidase. It is characterized by painful peripheral neuropathy with autonomic manifestations, a typical rash in the lower half of the body, and accumulation of glycolipids in the endothelium of cerebral vessels and renal glomerular arterioles. Patients have a higher risk of cardiovascular events in adulthood.

References:


Question 39: Pharmacology/Chemistry - Epilepsy

Discussion:

Topiramate increases the risk for calcium kidney stones by making it easier for calcium to bind with phosphate (forming calcium phosphate stones). Of note, when people talk about calcium kidney stones, they usually mean the most common calcium oxalate stones. Although they are often treated similarly, there are important differences. There is no contraindication to administer topiramate to a patient with history of calcium oxalate stones.

References:


Question 48: Pharmacology/Chemistry - Neuromuscular Disorders

Discussion:

Fifteen percent of all myasthenia gravis patients are seronegative for the acetylcholine antibodies. A percentage of these patients have detectable antibodies to the muscle specific receptor tyrosine kinase (MuSK), which is localized to the NMJ. MuSK mediates clustering of Ach receptors in the NMJ via signalling from agrin, which is secreted by the motor neuron. Stimulation with agrin activates the muscle-specific receptor tyrosine kinase (MuSK) which mediates clustering of Ach receptors by several signaling pathways.

References:


Question 49: Pharmacology/Chemistry - Neuromuscular Disorders

Discussion:

This patient's EMG is consistent with a pure sensory disorder since there is loss of sensory nerve conduction responses with sparing of motor nerve conduction responses and normal motor unit action potentials on needle EMG examination. The global loss of sensory nerve conduction responses suggests a neuronopathy instead of a length dependent axonal sensory neuropathy. Causes of a sensory neuronopathy include Sjögren syndrome, paraneoplastic syndrome usually with an anti-HU antibody, pyridoxine intoxication (at least 200 mg per day), a variant of Guillain-Bare syndrome, and with platinum-based chemotherapies.

References:
**Question 50: Pharmacology/Chemistry - Headache**

**Discussion:**

The pathophysiology of migraine is complex. The first phase of a migraine attack involves activation of the trigeminovascular reflex, resulting in release of vasoactive peptides (calcitonin gene-related peptide, neurokinin A, substance P) from trigeminal afferents supplying dural blood vessels. This produces vasodilation and sterile inflammation in dural vessels, leading to activation of first-order trigeminal afferents (peripheral sensitization) and manifested clinically by throbbing head (and neck) pain. As the attack progresses, second- and third-order trigeminothalamic and thalamocortical neurons become activated, mediated primarily by nitric oxide and glutamate transmission, resulting in central sensitization. This is clinically reflected by cutaneous allodynia.

**References:**


**Question 53: Pharmacology/Chemistry - Dementia**

**Discussion:**

This patient has clinical features and findings of early probable Alzheimer's disease (AD), characterized by progressive short-term memory impairment and cognitive dysfunction in multiple domains (language, visuospatial, and executive). Definite AD requires the demonstration of pathologic hallmarks on autopsy: global neuronal atrophy and loss of synapses, deposition of extracellular amyloid protein (A1-42), and accumulations of intracellular neurofibrillary tangles (NFT). NFTs are composed of paired filaments of phosphorylated tau protein. Clinical progression of dementia in AD correlates with NFT tissue burden. Studies of CSF in AD have shown significantly decreased levels of A beta1-42 and elevated levels of tau and phosphorylated-tau compared with age-matched controls. However, these changes are not specific for AD, and can be found in individuals with other neurodegenerative disorders and in normal aging. Thus, the utility of CSF testing in the evaluation of AD and dementia is unsettled. Cognitive symptoms of AD in patients include centrally-acting cholinesterase inhibitors (donepezil, rivastigmine, galantamine) and the NMDA-antagonist memantine.

**References:**


**Question 60: Pharmacology/Chemistry - Headache**

**Discussion:**

Giant cell temporal arteritis is a vasculitis that can lead to devastating ophthalmic and systemic complications. Jaw claudication, superficial temporal artery tenderness, fever, and visual loss are seen. Treatment primarily to prevent visual loss requires extended management with corticosteroids - initially at a high dose (eg, Prednisone 80-100 mg/d).

**References:**


**Question 62: Pharmacology/Chemistry - Other Pain Syndromes**

**Discussion:**


Capsaicin produces a release (and subsequent depletion with repeated applications) of substance P at the afferent sensory neurons.

References:


Question 64: Pharmacology/Chemistry - Movement Disorders

Discussion:

Paraneoplastic cerebellar degenerations are disorders of the cerebellum, the part of the brain responsible for coordination, and are associated with tumors (neoplasms). They arise when tumors express proteins that are normally found only in neurons, and it is believed that the immune system, in its attempt to kill the tumor, also damages the cerebellum. Certain tumors are more common than others. These include cancer of the ovary, uterus, or adnexa, cancer of the breast, and especially small cell carcinoma of the lung. The condition frequently begins before the tumor is diagnosed.

Pathological examination often reveals profound loss of Purkinje cells, which are the output cells for the cerebellum. The MRI scan of the brain may be normal. Anti-Yo is the anti-Purkinje cell antibody and reacts only with Purkinje cells. Patients with anti-Yo have a gynecological cancer, ovarian or breast 90% of the time. Anti-Hu reacts with nearly all neurons and is commonly associated with sensory neuropathy and encephalomyeloneuropathy.

References:


Question 70: Pharmacology/Chemistry - Other Pain Syndromes

Discussion:

The analgesic effects of pregabalin is primarily related to the decrease in central sensitization and nociceptive transmission to the action on the alpha-2-delta subunit of the calcium channel

References:


Question 83: Pharmacology/Chemistry - Movement Disorders

Discussion:

The main site of disposal of manganese in the body is biliary excretion. Patients with biliary atresia, chronic liver disease, or exposure to high dose of manganese during prolonged parenteral nutrition, are prone to develop manganese intoxication. Clinically, it is characterized by parkinsonism and dystonia, which do not respond to levodopa. T1-weighted MRI shows hyperintensity in the globus pallidus, striatum, and midbrain. The primary site of damage is the globus pallidus.

References:


Question 92: Pharmacology/Chemistry - Dementia

Discussion:

REM sleep behavior disorder (RBD) is characterized by complex behavioral manifestations in response to dream content that may cause sleep disruption or injuries to the patient or the bed partner. In this case, the patients need treatment to control their symptoms. Pharmacologic agents have been reported to have positive and negative impacts on REM sleep muscle atonia and the motor behaviors associated with RBD. Clonazepam is efficacious and well tolerated by the majority of patients afflicted by RBD and should be considered as initial treatment. In patients at risk of falls who have cognitive impairment or who have obstructive sleep apneas, melatonin may be a good alternative to clonazepam. Anticholinesterase inhibitors and dopaminergic agents are not of clear benefit. Monoamine oxidase inhibitors, tricyclic antidepressants, serotonergic synaptic reuptake inhibitors, and noradrenergic antagonists can induce or aggravate RBD symptoms and should be avoided in patients with RBD.

References:


Question 94: Pharmacology/Chemistry - Epilepsy

Discussion:

Drugs that can raise carbamazepine levels include isoniazid, erythromycin, cimetidine, calcium channel blockers (such as verapamil), and propoxyphene. Carbamazepine levels are lowered by phenobarbital, phenytoin, and primidone. Warfarin, chlorpromazine, digoxin, and gabapentin have no significant effect on carbamazepine levels.

References:


Question 102: Pharmacology/Chemistry - Cerebrovascular Disease

Discussion:

Dabigatran, an FDA-approved oral anticoagulant, is a direct thrombin inhibitor (DIT) that was demonstrated to reduce the incidence of stroke and systemic emboli compared with dose-adjusted warfarin in patients with non-valvular atrial fibrillation (AF), while concomitantly decreases the risk of bleeding in a pivotal study (Randomized Evaluation of Long-term Anticoagulant Therapy, or RE-LY). Two other new oral anticoagulants, apixaban and rivaroxaban, inhibit factor Xa, and have also demonstrated non-inferiority with warfarin in patients with AF in prevention of stroke, with a lower bleeding risk.

References:


Question 122: Pharmacology/Chemistry - Movement Disorders

Discussion:

Inhibition of catechol-O-methyltransferase (COMT) prolongs the action of dopamine by inhibiting its metabolism. Clinical trials have shown reduction of the wearing off effect in parkinsonian patients treated with tolcapne and entacapone. The other agents listed are dopamine receptor agonists.
References:


Question 139: Pharmacology/Chemistry - Movement Disorders

Discussion:

Up to 1% percent of patients treated with the antipsychotic drug clozapine for several months experience bone marrow suppression.

References:


Question 142: Pharmacology/Chemistry - Demyelinating Disorders

Discussion:

Dalfampridine is a potassium-channel blocker that improves nerve conduction along centrally demyelinated axons, and has been demonstrated to improve ambulation speed in approximately one-third of patients with multiple sclerosis. The drug carries a 0.2% risk of seizures, which is usually associated with toxic blood levels. Fampridine is cleared unchanged by the kidneys and is contraindicated in patients with creatinine clearance (GFR) of less than 50 cc/min because of the resultant risk of developing seizures. The risk of seizures in patients with GFR of 51-80 is uncertain.

References:


Question 143: Pharmacology/Chemistry - Movement Disorders

Discussion:

Dopamine agonist treatment in Parkinson disease has been associated with several impulse control disorders, including pathologic gambling, hypersexuality, compulsive eating, and compulsive shopping.

References:


Question 146: Pharmacology/Chemistry - Epilepsy

Discussion:

The HLA-B*1502 allele increases the risk of Stevens-Johnson syndrome after carbamazepine exposure by 10 times. This allele is commonly found in patients from Southeast Asia, specifically China, Malaysia, and Singapore. The FDA recommends genotyping all Asians for the allele. Carbamazepine directly binds to HLA-B molecules on antigen presenting T cells and contributes to cell death mediated by cytotoxic T cells in persons with Stevens-Johnson syndrome.
References:


Question 148: Pharmacology/Chemistry - Demyelinating Disorders

Discussion:

Teriflunomide (TF) is a new oral immunomodulating agent approved for relapsing-remitting multiple sclerosis. TF reversibly inhibits dihydroorotate dehydrogenase, a key mitochondrial enzyme involved in new pyrimidine synthesis for DNA replication. Consequently, the drug reduces T- and B-cell activation, proliferation, and function in response to autoantigens. However, teriflunomide preserves the replication and function of slowly dividing cells that use exogenous supplies of pyrimidine nucleotides through the so-called salvage pathway.

References:


Question 149: Pharmacology/Chemistry - Aging, Degenerative Diseases

Discussion:

The pedunculopontine nucleus (PPN) receives inputs from the globus pallidus and subthalamic nucleus, and projects to the substantia nigra, thalamus, and medullary reticulospinal neurons. The PPN contains one population of cholinergic and another population of glutamatergic neurons. It has been implicated as a component of the mesencephalic locomotor region. There is loss of PPN neurons in Parkinson's disease, and this has been correlated to the gait abnormalities in this disorder.

References:

Question 167: Pharmacology/Chemistry - Movement Disorders

Discussion:

Tardive dyskinesia results from chronic treatment with dopaminergic D2 receptor blockers. These include the classical high-potency neuroleptics, but also antiemetic agents such as prochlorperazine or metoclopramide.

References:


Question 192: Pharmacology/Chemistry - Headache

Discussion:

Blockade of histamine (H-1 and H-2) receptors can produce sedation, drowsiness; potentiation of central depressant drugs; weight gain. Blockade of muscarinic acetylcholine receptors can cause dry mouth, blurred vision, sinus tachycardia, constipation, urinary retention, memory impairment. Blockade of norepinephrine uptake at nerve endings may produce tremors, jitteriness, tachycardia, and diaphoresis, erectile and ejaculatory dysfunction. Blockade of serotonin uptake at nerve endings can cause sexual dysfunction, nausea, vomiting, diarrhea, anorexia, increase or decrease in anxiety (dose-dependent), asthenia (tiredness), insomnia, and extrapyramidal side effects. Blockade of serotonin-2 (5-HT2) receptors produces ejaculatory dysfunction,
hypotension, alleviation of migraine headaches, decrease in anxiety, and decrease motor restlessness. Blockade of α1-adrenergic receptors can cause postural hypotension, dizziness which predisposes to falls possibly resulting in broken bones and subdural hematomas. Blockade of α2-adrenergic receptors may produce priapism.

References:

Question 315: Pharmacology/Chemistry - Cerebrovascular Disease

Discussion:
The aim of treatment of severe hypertension in neurological catastrophes associated with increased intracranial pressure is to reduce the blood pressure to safe levels avoiding further increase in intracranial pressure. The drugs of choice include labetalol, esmolol, and enalapril. Clonidine may be used in some cases. Vasodilators, such as calcium channel blockers or nitrates, are best avoided as they may increase intracranial pressure in the setting of reduced systemic blood pressure, thus reducing cerebral perfusion pressure in areas with poor cerebral autoregulation.

References:

Question 323: Pharmacology/Chemistry - Neuromuscular Disorders

Discussion:
Reduced concentrations of coenzyme Q-10 (CoQ-10) have been found in patients with myopathy complicating the use of statins. Primary CoQ-10 deficiency produces either a myopathy or progressive cerebellar degeneration, reflecting the key role of CoQ-10 as an electron shuttle between complexes I or II and complex III in the respiratory chain.

References:

Question 325: Pharmacology/Chemistry - Neurogenetics

Discussion:
Point mutations of the peripheral myelin protein-22 gene in chromosome 17p are responsible for Charcot-Marie-Tooth disease type 1A. Charcot-Marie-Tooth disease type 1B, linked to chromosome 1, is associated with point mutations of the P0 gene that encodes for myelin protein zero.

References:

Question 328: Pharmacology/Chemistry - Neuromuscular Disorders

Discussion:
Muscle degeneration and myotonia are clinical hallmarks of myotonic dystrophy type 1 (DM1), an autosomal dominant multisystemic disorder also associated with endocrine abnormalities, cataracts, sleep disturbances, and cardiac conduction defects. It is caused by a CTG repeat expansion in the 3' untranslated region of the myotonic dystrophy protein kinase (DMPK) gene. Transgenic mice engineered to express mRNA with expanded (CUG)250 repeats (HSALR mice) exhibit prominent myotonia and altered splicing of muscle chloride channel gene (Clcn1) transcripts. Similarly, results were observed for Chloride channel activity in knockout mice for muscleblind-like 1, a protein necessary for pre-mRNA muscle chloride channel splicing and normal function. This a second murine model of DM1 that exhibits prominent myotonia and altered Chloride channel (ClC-1) splicing. These results support a molecular mechanism for myotonia in DM1 in which a reduction in both the number of functional sarcolemmal ClC-1 and due to CUG repeat-containing mRNA molecules sequestering muscle-blind like 1 (Mbnl1) proteins required for proper CLCN1 pre-mRNA splicing and chloride channel function.

References:


Question 346: Pharmacology/Chemistry - Movement Disorders

Discussion:

Suboptimal doses of carbidopa are a frequent cause of nausea/vomiting at the initiation of levodopa therapy. It takes 75 mg to 150 mg of carbidopa per day to saturate the peripheral aromatic amino acid decarboxylase enzyme to prevent peripheral side effects of levodopa. While decreasing the levodopa dose may reduce nausea and vomiting, it may result in less symptomatic improvement and thus would not be the optimal choice.

References:


Question 353: Pharmacology/Chemistry - Headache

Discussion:

Tramadol, a centrally acting analgesic structurally related to codeine and morphine, consists of two enantiomers, both of which contribute to analgesic activity via different mechanisms. (+)-Tramadol and the metabolite (+)-O-desmethyl-tramadol (M1) are agonists of the mu opioid receptor. (+)-Tramadol inhibits serotonin reuptake and (-)-tramadol inhibits norepinephrine reuptake, enhancing inhibitory effects on pain transmission in the spinal cord. The complementary and synergistic actions of the two enantiomers improve the analgesic efficacy and tolerability profile of the racemate.

References:


Question 363: Pharmacology/Chemistry - Toxic/Metabolic Disease

Discussion:

Excessive zinc ingestion is a well-recognized cause of copper deficiency. In addition to the common use of zinc in the prevention or treatment of common colds and sinusitis, zinc therapy has been used for conditions such as acrodermatitis enteropathica, decubitus ulcers, sickle cell disease, celiac disease, memory impairment, and acne. Unusual sources of excess zinc have included a patient who consumed an entire tube of a denture cream that contained zinc daily for 5 years and patients swallowing coins containing zinc. Zinc causes an upregulation of metallothionein production in the enterocytes. Metallothionein is an intracellular ligand, and copper has a higher affinity for metallothionein than zinc. Copper displaces zinc from metallothionein, binds preferentially to the metallothionein, remains in the enterocytes, and is lost in the feces as the intestinal cells are sloughed.
Subsequent copper deficiency is recognized caused of subacute combined degeneration, similar to that associated with B12 deficiency.

References:

Hedera P. Peltier A. Fink JK. Wilcock S. London Z. Brewer GJ. Myelopolyneuropathy and pancytopenia due to copper deficiency and high zinc levels of unknown origin II. The denture cream is a primary source of excessive zinc. Neurotoxicology; 2009; 30(6):996-9,


Question 371: Pharmacology/Chemistry - Epilepsy

Discussion:

Lamotrigine is metabolized in the liver predominantly by glucuronic acid conjugation; the major metabolite is an inactive 2-N-glucuronide conjugate. Carbamazepine, phenytoin, and phenobarbital induce glucuronic acid conjugation of lamotrigine resulting in a 40% reduction of the lamotrigine level. Valproic acid inhibits glucuronic acid conjugation of lamotrigine doubling the lamotrigine level. Levetiracetam does not induce or inhibit glucuronic acid conjugation of lamotrigine.

References:


Question 376: Pharmacology/Chemistry - Neuromuscular Disorders

Discussion:

Myotonia congenita is an inherited neuromuscular disorder characterized by the inability of muscles to quickly relax after a voluntary contraction. The condition is present since early childhood, but symptoms can be mild. Most children will be 2 or 3 years old when parents first notice their muscle stiffness, particularly in the legs and often provoked by sudden activity after rest. The disease does not cause muscle wasting; in fact, it may cause muscle enlargement. Muscle strength is increased. There is an autosomal dominant form (Thomsen) and an autosomal recessive form (Becker), which are both associated with mutations of the gene that codes for the muscle chloride channel CLCN1 on chromosome 7q35. CLCN1 is necessary in order to stabilize the high resting membrane potential of skeletal muscle. Dysfunction of this channel as a result of genetic mutation, causes partial depolarization of the membrane and allows a hyperexcitable state to exist, resulting in myotonia. It is postulated that permanent excitability gives rise to constant mild muscle activity, resulting in muscle hypertrophy. Muscle stiffness responds well to drugs that reduce the associated hyperexcitability of the sarcolemma by interfering with sodium channels located on it. These drugs theoretically reduce spontaneous discharges of electrical myotonia by decreasing the number of available sodium channels, but they have no known effects on chloride channels. One such drug is mexiletine, which is able to reduce myotonia with doses of 200 mg 2 or 3 times a day. Phenytoin 200 mg/d to 300 mg/d can also be used to treat myotonia. A number of overlapping inherited nondystrophic myotonia syndromes result from mutations to the gene encoding the voltage-gated sodium channel type IV alpha subunit (SCN4A). Paradoxical myotonia, in which there is an increase in myotonia with physical activity, and aggravation of myotonia with cold are both indicative of paramyotonia congenita. Transient paralysis indicates hyperkalemic periodic paralysis, and fluctuating myotonia indicates potassium-aggravating myotonia.

References:


Question 389: Pharmacology/Chemistry - Epilepsy

Discussion:
Hypohidrosis is an infrequent side effect of zonisamide, particularly in children. Hypohidrosis can lead to symptoms of heat stroke due to impaired heat-dissipation mechanisms and produce hyperthermia.

References:


Question 390: Pharmacology/Chemistry - Sleep Disorders

Discussion:

In humans, where narcolepsy is associated with human leucocyte antigen (HLA) abnormalities, recent studies have shown that narcolepsy with cataplexy is usually caused (>90%) by the lack of two related brain chemicals called hypocretin-1 and hypocretin-2. The cause of narcolepsy without cataplexy is still under investigation. Over 90% of patients with narcolepsy-cataplexy carry HLA-DQB1*0602. This marker is more specific and sensitive than the old marker HLA-DR2, and so it is speculated that patients with narcolepsy-cataplexy probably have an autoimmune disorder.

References:


Question 392: Pharmacology/Chemistry - Epilepsy

Discussion:

The symptoms are common in patients with hyperammonemia. Disorders of urea cycle are common in mentally retarded or developmentally abnormal persons with epilepsy. VPA increases levels of gamma-aminobutyric acid and prolongs the recovery of inactivated sodium channels. These properties may be responsible for its action as a CNS depressant. VPA may also cause impairments in fatty-acid metabolism and disrupt the urea cycle, leading to hyperammonemia. VPA also alters fatty-acid metabolism, impairs beta-oxidation (a mitochondrial process), and disrupts the urea cycle. Hyperammonemia and other metabolic effects, as well as end-organ effects (hepatitis, pancreatitis, hemodynamic compromise), may be the result of severe toxicity due to these impaired metabolic processes.

There are no known drug-drug interactions between valproate and levetiracetam. No reports of levetiracetam-induced hepatitis are in the literature.

References:


Question 395: Pharmacology/Chemistry - Aging, Degenerative Diseases

Discussion:

Midodrine is a prodrug that is converted in the liver to an alpha agonist. Its predictable absorption and pharmacokinetics makes it the drug of choice for treatment of orthostatic hypotension unresponsive to fludrocortisone.

References:


Question 412: Pharmacology/Chemistry - Other Pain Syndromes

Discussion:

Codeine, used to treat mild and moderate pain, acts through metabolically formed morphine. It is mainly metabolized by glucuronidation, but minor pathways are N-demethylation to norcodeine and O-demethylation to morphine. The latter pathway depends on the genetically polymorphic CYP2D6, which is absent in 7% of the white population. Dependence of codeine hypoalgesia on morphine formation via CYP2D6 makes this effect liable to interaction with drugs that are inhibitors of CYP2D6 such as quinidine, some selective serotonin reuptake inhibitors, and some neuroleptics.

References:


Question 414: Pharmacology/Chemistry - Toxic/Metabolic Disease

Discussion:

Paralytic shell-fish poison (saxitoxin) resembles tetrodotoxin in its ability to reversibly bind to membrane voltage-gated sodium channels.

References:


Question 417: Pharmacology/Chemistry - Epilepsy

Discussion:

Fosphenytoin is a phosphate ester prodrug developed as an alternative to intravenous phenytoin for acute treatment of seizures. Advantages include more convenient and rapid intravenous administration, availability for intramuscular injection, and low potential for adverse local reactions at injection sites. Drawbacks include the occurrence of transient paraesthesias and pruritus at rapid infusion rates, and cost. Fosphenytoin is highly bound (93-98%) to plasma proteins. Saturable binding at higher plasma concentrations accounts for an increase in its distribution volume and clearance with increasing dose and infusion rate. Fosphenytoin is entirely eliminated through metabolism to phenytoin by blood and tissue phosphatases. The bioavailability of the derived phenytoin relative to intravenous phenytoin is approximately 100% following intravenous or intramuscular administration. The half-life for conversion of fosphenytoin to phenytoin ranges from 7-15 minutes. Faster intravenous infusion rates and competitive displacement of derived phenytoin from plasma protein binding sites by fosphenytoin compensate for the expected conversion-related delay in appearance of phenytoin in the plasma. Unbound phenytoin plasma concentrations achieved with intravenous fosphenytoin loading doses of 100-150 or 50-100mg phenytoin sodium equivalents/min are comparable, and achieved at similar times, to those with equimolar doses of intravenous phenytoin at 50 (maximum recommended rate) or 20-40 mg/min, respectively. The rapid achievement of effective concentrations permits the use of fosphenytoin in emergency situations, such as status epilepticus. Following intramuscular administration, therapeutic phenytoin plasma concentrations are observed within 30 minutes and maximum plasma concentrations occur at approximately 30 minutes for fosphenytoin and at 2-4 hours for derived phenytoin. Diazepam may be given by intravenous (IV) or intramuscular (IM) injection, but absorption following intramuscular administration may be slow and erratic depending on the muscle mass used and other factors. The other medications cannot be administered IM.

References:


Question 418: Pharmacology/Chemistry - Epilepsy
Discussion:

Topiramate has the broadest spectrum of action for multiple seizure types of the drugs listed. Some of the newer AEDs are considered broad spectrum, meaning that they work in idiopathic generalized epilepsy and focal epilepsies. These newer AEDs include lamotrigine, topiramate, levetiracetam, and zonisamide.

References:


Physiology

Question 1: Physiology - Basic Physiology

Discussion:

Botulinum toxin causes inhibition of release of acetylcholine vesicles from the presynaptic terminals, causing reduction in the size of motor nerve compound muscle action potentials.

References:


Question 14: Physiology - EMG

Discussion:

The L5 root innervates tibialis anterior, tibialis posterior, and gluteus medius muscles. The L4 root innervates tibialis anterior, quadriceps, and adductor muscles. The peroneal nerve innervates the muscles of the anterior and lateral compartments of the leg.

References:


Question 20: Physiology - EMG

Discussion:

The femoral nerve innervates muscles involved in hip flexion and knee extension. Its sensory territory includes the territory of the saphenous nerve below the knee.

References:

**Question 31: Physiology - EEG**

**Discussion:**

Frontal sharp wave transients are a normal waveform seen during sleep in near-term infants. Tracé discontinu is a discontinuous background pattern seen in very preterm infants and which resembles suppression burst. Temporal alpha bursts are not limited to sleep and are seen around 33 weeks for only a brief period. Sleep spindles are not seen until at least 44 weeks and would not be present in a pre-term infant. Vertex transients are first seen around 46 weeks.

**References:**


**Question 37: Physiology - EEG**

**Discussion:**

Periodic waves of smooth or triangular shaped may be picked up by an electrode on or near a scalp artery as a result of pulse waves in the frontal and temporal areas than with electrodes in the posterior head regions.

**References:**


**Question 41: Physiology - EEG**

**Discussion:**

Periodic lateralized sharp wave discharges in a patient with a several day history of fever, seizures and obtundation would most likely suggest herpes simplex encephalitis.

**References:**


**Question 46: Physiology - EEG**

**Discussion:**

Electrocerebral inactivity (ECI) is defined as "no EEG activity over 2 microvolts."

**References:**


**Question 47: Physiology - EMG**

**Discussion:**
Single fiber EMG (SFEMG) is the most sensitive diagnostic test for myasthenia gravis. In generalized disease, SFEMG examination of a weak muscle demonstrates increased jitter in more than 90% of cases. Slow repetitive stimulation of a distal muscle is approximately 50% sensitive. Repetitive stimulation of a proximal muscle has a higher yield (approximately 75%). Rapid repetitive stimulation (more than 10 hz) is used in presynaptic disorders of neuromuscular transmission.

References:


Question 57: Physiology - EEG

Discussion:

Favorable prognostic factors on EEG are variability, reactivity to external stimuli, varying sleep patterns, and increase in background frequencies. Poor prognostic factors are invariant pattern, no reactivity, monorhythmic pattern, burst suppression, generalized periodic discharges, very low voltage tracing, and generalized suppression. Over 96% of the patients with poor prognostic findings on the EEG following a cardiac arrest either die within a few days after the cardiopulmonary arrest, or if they survive, do so in a persistent vegetative state.

References:


Question 66: Physiology - EMG

Discussion:

The proximity of the sciatic nerve explains its vulnerability during hip replacement surgery. The peroneal division of this nerve is often more severely affected. The abnormalities of the superficial peroneal and sural sensory nerve action potentials serve to localize the lesion distal to the dorsal root ganglion and argue against a lumbosacral root lesion. The innervation of the short head of the biceps femoris and anterior tibialis muscles both come from the peroneal division of the sciatic nerve.

References:


Question 71: Physiology - Evoked Potentials

Discussion:

Somatosensory evoked potentials record activity in the dorsal column/medial lemniscal sensory pathways. Neurons in the spinothalamic tracts and spinoreticulothalamic systems have small diameter axons and are not recordable with standard SEP techniques. Propriospinal neurons interconnect adjacent regions of the spinal cord and do not contribute to the SEP. The ventral spinocerebellar tract ascends to the midbrain then enters through the superior cerebellar peduncle. It does not contribute to the SEP signal.

References:


Question 75: Physiology - EMG
Discussion:

Sensory responses are affected in lesions distal to the dorsal root ganglion (ie, plexopathies) while they are spared in lesions proximal to the dorsal root ganglion (ie, radiculopathies). Thus, they are quite useful in distinguishing between root and plexus lesions.

References:


Question 89: Physiology - Sleep

Discussion:

The late sleep-onset times and the late rise time on weekends in a teenager suggests delayed sleep phase syndrome. Wrist actigraphy is particularly helpful in diagnosing disorders of circadian rhythm.

References:


Question 95: Physiology - Sleep

Discussion:

EEG findings during REM sleep are comprised of mixed frequency activity, similar to stage 1 of NREM (non-REM) sleep, with markedly decreased tone in chin EMG activity with bursts of REM. Sleep spindles are seen during stage 2 of NREM sleep while stage 4 of NREM sleep comprises more than 50% of delta activity on an epoch.

References:


Question 103: Physiology - EEG

Discussion:

Patients with increased intracranial pressure, such as that caused by ventricular outflow obstruction, typically have EEGs that show rhythmic slow activity in the theta-delta frequency range. The site is often distant from the site of obstruction. These EEG findings are not specific.

References:


Question 112: Physiology - EEG

Discussion:

EEG measures the potential difference between two electrodes. Another way to say this is that inputs to the two electrodes which are common to both are excluded (not amplified). Electrical interference is by far the most common input to all electrodes but, because it is shared by all electrodes, it is rejected allowing the underlying cerebral signal to be seen.
References:


**Question 113: Physiology - EEG**

**Discussion:**

A highly epileptogenic EEG pattern that has a high incidence of associated seizures is the 3-Hz spike-and-wave. Benign EEG patterns that are unassociated with seizures include 6-Hz spike-and-wave, small sharp spikes, and wicket spikes. Triphasic sharp waves are not considered epileptiform in character and are, instead, indicative of an encephalopathy.

References:


**Question 114: Physiology - Basic Physiology**

**Discussion:**

Fusimotor fibers transmit impulses from gamma motor neurons in the ventral horn of the spinal cord to muscle spindles. This system allows muscle spindles to remain sensitive over a broad range of muscle size by shortening the spindle as the muscle shortens.

References:


**Question 123: Physiology - EMG**

**Discussion:**

Spinal muscular atrophy is the most common cause of progressive motor neuron disease in infancy. Mutations of the survival motor neuron gene is present in 95% of children with spinal muscular atrophy type 1.

References:


**Question 124: Physiology - EEG**

**Discussion:**

Periodic lateralized epileptiform discharges (PLEDs) are seen after an acute or subacute cerebral insult such as a stroke and are often associated with focal seizures. Burst suppression represents a severe disturbance of cerebral function and may be seen in a variety of contexts including anesthesia and global anoxic injury. Rhythmic midtemporal discharges and small sharp spikes are benign variants which are commonly misinterpreted as epileptiform abnormalities.
References:


Question 126: Physiology - EMG

Discussion:

Limb onset amyotrophic lateral sclerosis can appear to affect only one limb with lower motor neuron signs. The presence of upper motor neuron signs, even when not below the lower motor neuron signs, plus evidence on EMG of diffuse active denervation and reinnervation (large motor units) at multiple levels (cervical, thoracic, and lumbar) is consistent with this diagnosis.

References:


Question 127: Physiology - EEG

Discussion:

Light sensitivity manifested by photomyoclonus induced by photic stimulation can occur with abrupt alcohol withdrawal. The other epileptic events are no more likely to happen in this context than in the general population. Photic driving would not be expected to be affected by alcohol withdrawal.

References:


Question 130: Physiology - EMG

Discussion:

The posterior interosseous nerve, a distal branch of the radial nerve, supplies the extensor muscles in the forearm, with the exception of the ECRL. The radial nerve supplies the triceps and extensor carpi radialis longus muscles proximal to the branchpoint of the posterior interosseous nerve (PIN). As such the EMG findings are limited to the distribution of the PIN, sparing more proximal radial-innervated muscles.

References:


Question 156: Physiology - EMG

Discussion:
Median neuropathy at the wrist (carpal tunnel syndrome) presents with hand and upper limb pain with nocturnal exacerbations. The hallmark of diagnosis is the presence of slowing sensory and motor axons of median in the segment of nerve traversing the carpal tunnel. The ulnar and radial sensory and motor axons are spared.

References:


Question 157: Physiology - Autonomic Studies

Discussion:

The quantitative sudomotor axon reflex test (QSART) is abnormal in 80% of patients with distal small fiber neuropathy. The EMG/nerve conduction studies are normal.

References:


Question 160: Physiology - EMG

Discussion:

Hypokalemic periodic paralysis usually presents in the 2nd decade of life. Low K⁺ in the serum may be found during the paralysis. Respiratory muscles are usually spared. Paralysis is often precipitated by a carbohydrate load the night before or excessive exercise. This is a channelopathy, with the L-type Ca++ channel abnormal.

References:


Question 184: Physiology - EMG

Discussion:

The clinical history of stiffness made worse with continuous activity, worse in the cold, without weakness is typical of paramyotonia congenita. PMC is caused by a mutation in the voltage-gated sodium channel encoded by the SCN4A gene. Myotonia congenita is caused by a chloride channel mutation and, in this disorder, stiffness improves with activity. Also, legs are more affected than arms. Hypokalemic periodic paralysis is caused by a calcium channel mutation, does not demonstrate myotonic and is characterized by attacks of weakness. Andersen-Tawil syndrome is caused by a potassium channel mutation and includes attacks of weakness, cardiac arrhythmias and dysmorphic features.

References:


Questions 199 - 203: Physiology - EMG

Discussion:

Abductor pollicis brevis is innervated by the median nerve with radicular derivation from C8 and T1. The abductor digiti minimi is innervated by the ulnar nerve but also has radicular contributions from C8 and T1. Presence of fibrillation potentials indicating
denervation in the abductor digiti minimi coupled with the absence of fibrillation potentials in the abductor pollicis brevis indicate that an ulnar mononeuropathy is much more likely in this case than a radiculopathy.

The serratus anterior muscle is innervated by the long thoracic nerve and stabilizes the scapula for forward pushing and arm elevation. No sensory loss is evident, and on examination scapular winging is evident.

Radial mononeuropathy, as in this case, most commonly occurs at the midhumeral level near the spiral groove. The triceps is spared because branches innervating this muscle originate proximal to the spiral groove. Radial mononeuropathy presents predominantly with wrist and finger drop with little or no pain.

The serratus anterior muscle is innervated by the long thoracic nerve, which is spared even in large brachial plexus lesions.

The deltoid muscle is innervated by the axillary nerve, and the biceps muscle is innervated by the musculocutaneous nerve. Both have C5 root input.

References:


Question 215: Physiology - EEG

Discussion:

Electropositive potentials from occipital leads (O1, 2) may be produced by the R-wave of the ECG in patients. Rhythmic delta activity confined to a single electrode position likely represents pulse artifact. Pulse artifact may appear as periodic sharply contoured potentials which are time-locked to ECG. Defects in electrical and mechanical continuity of one electrode can produce bizarre, often sudden electrode potentials which differ markedly from ongoing background activity, do not blend with other simultaneously recorded activity, and appear only in derivations involving one electrode. Movement of the tongue, whose tip is electrically negative with respect to its base, may produce widely distributed, low frequency intermittent potentials that may resemble "projected rhythms". A burst of muscle potentials may precede such low frequency waves, serving to differentiate glossokinetic potentials from "projected" activity.

References:


Question 228: Physiology - EEG

Discussion:

The fully developed triphasic waves in this tracing begin with an initial brief negative wave (upward pen deflection), followed by a prominent electropositive sharply contoured wave, and then a broader negative wave. The duration of each succeeding wave of the complex is greater. Most of these waves have no anterior-posterior lag. Triphasic waves are bilaterally synchronous and usually frontally predominant. In metabolic encephalopathies, they usually appear in groups.

References:

Question 236: Physiology - EEG

Discussion:

Panayiotopoulos syndrome is associated with occipital spikes. Juvenile absence epilepsy (JAE) has 3-5 Hz generalized spike and polyspike and slow wave bursts. Benign neonatal convulsions may be associated with a range of paroxysmal abnormalities superimposed upon a normal background. Rasmussen encephalitis usually has unilateral slowing and spikes.

References:


Question 270: Physiology - EEG

Discussion:

The 14 and 6 phantom spikes occur bilaterally synchronous and diffuse fashion. The bursts are brief and last 1 to 2 seconds. At times the 14 Hz or, more commonly, the 6 Hz pattern may be seen in isolation. The pattern is considered a normal variant but can easily be confused with epileptiform discharges.

References:


Question 271: Physiology - EEG

Discussion:

The EEG sample of a 17-month-old infant shows sleep spindles.

References:


Question 272: Physiology - EEG

Discussion:

LGS is associated with slow background activities of wakefulness with interictal generalized slow spike wave discharges. Landau-Kleffner syndrome consists of language regression, seizures, and the aforementioned sleep-activated spikes discharges; CAE/pyknolepsy with 3Hz spike wave; JME with polyspike wave. Infantile spasms is associated with hypsarrhythmia.

References:

Juvenile myoclonic epilepsy is characterized by generalized polyspike wave discharges. These findings are often brought out by photic stimulation. Studies demonstrate that "photoparoxysmal discharges" - polyspike wave discharges evoked by photic stimulations - that outlast stimuli are often associated with epilepsy, whereas those that do not outlast stimuli can be seen in asymptomatic individuals.

References:


Question 279: Physiology - EEG

Discussion:

Hypsarrhythmia is a characteristic pattern seen in a majority of infants with infantile spasms. It is characterized by high-voltage and irregular slow waves with multiple sharp and spike discharges.

References:


Question 285: Physiology - EMG

Discussion:

The figure demonstrates a normal distal ulnar-ADM CMAP with temporal dispersion and conduction block found on proximal stimulation. This phenomenon is an acquired demyelinating process. The patient has progressive weakness with fasciculations supporting a lower motor neuron disorder. ALS is a motor neuronopathy that results in loss of motor neurons; demyelination is not a feature. The predominant pathology in diabetic sensorimotor polyneuropathy is axonal, which results in low-amplitude sensory and motor potentials, but not demyelination/conduction block. Myasthenia Gravis, as a disorder of the neuromuscular junction, typically demonstrates normal CMAPs; conduction block/demyelination is not seen. Likewise, metabolic myopathy does not demonstrate conduction block/demyelination in motor nerves.

References:

Question 295: Physiology - EEG

Discussion:

This is a normal awake EEG from a 10 month old infant. The posterior dominant rhythm is 5-5.5 Hz which is normal for patients in this age range. The presence of a posterior dominant rhythm excludes sleep as a possible interpretation. There are no epileptiform changes and there is no evidence of a post-ictal process. Drowsiness in children is often associated with hypnogogic hypersynchrony - a finding not seen in this tracing.

References:


Question 298: Physiology - EEG

Discussion:
Subacute sclerosing panencephalitis is associated with generalized periodic long-interval diffuse discharges in the EEG that recur every 4 to 15 seconds. Spongiform encephalopathies due to prion disease presents with PEDs, but intervals are typically shorter than those of SSPE. HSV encephalitis presents with lateralized periodic epileptiform discharges. Patient's with Tay Sachs disease tend to have a slow background with or without multifocal epileptiform transients. Alzheimer's disease patients usually have normal EEGs or sometimes having excessive theta activities.

References:

Question 318: Physiology - EMG

Discussion:
The amplitude of the facial nerve compound muscle action potential (CMAP) is correlated with the number of functional axons in the facial nerve. The lower the CMAP amplitude, the more axonal degeneration and the poorer the prognosis.

References:

Question 320: Physiology - EMG

Discussion:
Friedreich's ataxia is an autosomal recessive spinocerebellar syndrome. Progressive ataxia with abnormal sensory NCS and normal motor NCS is consistent with this diagnosis. Metachromatic leukodystrophy, CMT1 and Cockayne's syndrome show motor and sensory nerve responses with marked slowing consistent with a demyelinating disease. AIDP has a different time course and shows prolonged latencies and slow conduction velocities.

References:

Question 326: Physiology - Basic Physiology

Discussion:
Influx of calcium into the presynaptic nerve terminal may increase the probability of neurotransmitter release by binding with the calcium/calmodulin-dependent protein kinase which phosphorylates synapsin 1, allowing vesicles to be released from actin filaments and move towards release sites. Calcium also binds to synaptotagmin which allows docking and fusion of vesicles.

References:

Question 327: Physiology - EMG

Discussion:
An incremental response to tetanic stimulation indicates a presynaptic defect at the neuromuscular junction. In an infant, the most likely reason for developing this is from intestinal botulism.
Discussion:
The decremental response to 2 to 3 Hz repetitive nerve stimulation in patients with myasthenia gravis is due to failure of neuromuscular transmission at a number of endplates resulting in fewer muscle fiber action potentials contributing to the compound muscle action potential. Decreased presynaptic calcium influx occurs in Lambert-Eaton myasthenic syndrome. Impaired fusion of Ach-containing vesicles to the pre-synaptic membrane occurs in botulism.

References:

Discussion:
Triphasic waves, though non-diagnostic, are most commonly seen in the context of a metabolic disturbance and particularly with hepatic or renal dysfunction.

References:

Discussion:
Increased amplitude and duration of a motor unit potential generally suggests disorders of the lower motor neuron such as motor neuron disease (ALS), poliomyelitis, syringomyelia or chronic neuropathies or radiculopathies. Reduction in amplitude and duration of the motor unit potential suggests primarily myopathic disorders such as muscular dystrophy, congenital or other myopathies, myositis and disorders of neuromuscular transmission. Damaged axon terminals may result in random loss of muscle fibers within a motor unit. Similarly during early reinnervation, immature motor units consist of only a few muscle fibers. Motor unit potentials may then become polyphasic, of low amplitude and of short duration.

References:


Discussion:
The clinical presentation is typical for hereditary neuropathy with liability to pressure palsies (HNPP). This disorder is caused by a deletion or point mutation in the PMP22 gene. Motor and sensory nerve conduction velocities are reduced in affected patients as well as asymptomatic gene carriers. Since nerve conduction studies were delayed until there was full recovery in this patient, they are unlikely to show a focal abnormality in conduction in the left peroneal nerve.
References:


**Question 362: Physiology - EEG**

**Discussion:**

Nonconvulsive status epilepticus can be prolonged and is associated with fairly abrupt deterioration in mental function and a paroxysmal EEG. These features differentiate nonconvulsive status from the dementias.

**References:**


**Question 366: Physiology - Sleep**

**Discussion:**

Patients with excessive daytime sleepiness must be evaluated for sleep apnea, narcolepsy, idiopathic hypersomnia, restless legs syndrome, or parasomnias. The absence of any abnormality on the overnight polysomnography and multiple sleep latency test other than less-than-5-minute sleep latency is consistent with idiopathic hypersomnia.

**References:**


**Question 379: Physiology - EEG**

**Discussion:**

The location of the ictal focus is the most important factor in determining seizure onset during chronic intracranial EEG monitoring and this is best represented by ictal onset. The subsequent expression of the ictal discharge, while important, is less useful in localizing the epileptogenic zone.

**References:**


**Question 386: Physiology - EMG**

**Discussion:**

Fifty percent decrease in compound muscle action potential amplitude at the elbow compared with the wrist is consistent with conduction block below the proximal point of stimulation and above the distal point of stimulation. Conduction block is characteristic of acquired demyelination nerve lesions.
Question 403: Physiology - Basic Physiology

Discussion:

Patients with demyelinating neuropathies develop paralysis primarily because of conduction block, rather than slow conduction velocity.

References:


Question 407: Physiology - EMG

Discussion:

The diffuse reduction in motor amplitudes, normal sensory responses and denervation all support a motor disorder. This patient suffers from an acute motor neuron disorder with CSF pleocytosis. A variety of acute viral myelitis including poliomyelitis, West Nile virus and enteroviruses can produce an infectious motor neuron disorder.

References:


Question 408: Physiology - EEG

Discussion:

Repetitive spikes that increase in amplitude and decrease in frequency is the usual EEG pattern at the onset of a generalized tonic clonic seizure. Evolution of a discharge is one of the most important features that defines it as an ictal event. Generalized high amplitude spike and slow wave discharges are more characteristic of the later, clonic phase of GTC seizures. Periodic generalized sharp waves are pathognomonic of Creutzfeldt-Jacob disease.

References:


Question 410: Physiology - EEG

Discussion:

Medial temporal lobe epilepsy is the most common partial seizure disorder. Secondary generalization can occur. EEG in such patients usually shows focal temporal spikes.

References:


Question 416: Physiology - Evoked Potentials
Discussion:

In brainstem auditory evoked potentials, absence of wave I with intact wave V is most commonly due to peripheral hearing loss. Wave I is generated by cranial nerve 8. Central auditory pathways are highly crossed so later waveforms may be preserved with unilateral auditory nerve pathology.

References: