Anatomy

Question 1: Anatomy - Cortex and Connections

Discussion:

The patient's stereoagnosia and statoagnosia (inability to determine body position in space) both localize to the superior parietal lobule (Brodmann area 7) which lies above the intraparietal sulcus. The inferior parietal lobule includes the supramarginal gyrus (which surrounds the end of the sylvian fissure) and the angular gyrus (which surrounds the end of the superior temporal gyrus). The cuneus is part of the occipital lobe.

References:


Question 2: Anatomy - Peripheral Nervous System

Discussion:

The tibial division of the sciatic nerve supplies the tibialis posterior muscle. The tibialis anterior, peroneus longus, peroneus brevis and extensor digitorum brevis are supplied by the common peroneal nerve.

References:


Question 11: Anatomy - Cranial Nerves

Discussion:

The nervus intermedius is the portion of the facial nerve carrying all the general visceral efferent, general somatic afferent, and special afferent information. The branchial motor fibers going to the muscles of facial expression travel in a separate bundle. Of the potential answers given, an injury to the nervus intermedius would therefore impair taste from the ipsilateral anterior tongue. Facial muscles would not be affected. The parotid gland is innervated by the glossopharyngeal nerve while the striated muscles of the pharynx are innervated by the vagus nerve.

References:


Question 14: Anatomy - Brainstem/Cerebellum

Discussion:

With a right unilateral lesion of the medial longitudinal fasciculus, the patient can abduct the left eye on attempted gaze to the left but the right eye cannot be adducted. Nystagmus occurs in the left (abducting) eye.
Question 16: 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 quadriaparesis, bilateral loss of proprioception and vibration, and complete paralysis of the tongue.

References:

Question 29: Anatomy - Embryology

Discussion:

The diencephalon gives rise to the thalamus, epithalamus, subthalamus and hypothalamus. The telencephalon gives rise to the cerebral hemispheres, the basal ganglia, and the rhinencephalon (hippocampus and piriform lobes)

References:

Question 35: 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 leminsci. The lateral lemniscus ascends to the midbrain where most of the fibers terminate in the inferior colliculi.

References:

Question 41: Anatomy - Cortex and Connections

Discussion:

Homonymous hemianopsia results from lesions occurring in the visual radiations or occipital cortex. As the occipital cortex is approached the visual field defects become increasingly congruent. In addition, occipital cortex lesions characteristically produce macular sparing.

References:
**Question 60: Anatomy - Cranial Nerves**

**Discussion:**

The motor portion of the trigeminal nerve innervates the muscles of mastication: masseter, temporalis, and medial and lateral pterygoids. It also supplies the mylohyoid, anterior digastric, tensor veli palatini, and tensor tympani muscles.

**References:**


**Question 64: Anatomy - Peripheral Nervous System**

**Discussion:**

The Martin-Gruber anastomosis occurs in 15% to 30% of the population. It consists of a communicating branch from the median nerve, usually from the anterior interosseous nerve, to the ulnar nerve in the forearm. It consists of nerve fibers destined for the first dorsal interosseous, most commonly, followed by the adductor pollicis and abductor digiti minimi.

**References:**


**Question 71: Anatomy - Cortex and Connections**

**Discussion:**

Layer IV contains a dense horizontal band of fibers - the external band of Baillarger. This band contains the terminal ramifications of the thalamocortical projections from specific thalamic relay nuclei. It is particularly prominent in the striated or primary visual cortex, and known as the line of Gennari.

**References:**


**Question 93: Anatomy - Peripheral Nervous System**

**Discussion:**

The sciatic nerve passes underneath the piriformis muscle and may be entrapped at that location. Amongst the many muscles which the sciatic innervates is the biceps femoris.

**References:**


**Question 115: Anatomy - Cortex and Connections**

**Discussion:**
The frontal eye fields project to their subcortical targets via the anterior limb of the internal capsule. Fibers originating in the frontal operculum travel via the corticobulbar tract in the genu of the internal capsule. The output of the primary motor cortex (the corticospinal and corticobulbar tracts) travel in the posterior limb and the genu of the internal capsule.

References:


Question 166: Anatomy - Blood Supply of Brain/Spinal Cord

Discussion:

Infarction in the distribution of the anterior inferior cerebellar artery (AICA) involves the cochlea or cochlear nuclei (ipsilateral hearing loss), the vestibular nuclei (vertigo and contralateral beating horizontal/torsional nystagmus), the lateral reticular nucleus (ipsilateral Horner's syndrome), middle cerebellar peduncle (ipsilateral ataxia), the descending tract of V (ipsilateral pain and temperature loss), and lateral spinal thalamic tract (contralateral pain and temperature loss). Infarction of the labyrinthine artery distribution produces acute deafness but not the other findings. Infarction in the posterior inferior cerebellar (PICA) distribution produces a lateral medullary syndrome and does not include hearing loss.

References:


Question 193: Anatomy - Blood Supply of Brain/Spinal Cord

Discussion:

Occlusion of the anterior choroidal artery can result in hemiparesis and hemisensory loss since the posterior limb of the internal capsule is perfused by this artery. Also, the lateral geniculate nucleus of the thalamus is perfused by the anterior choroidal and dysfunction of this nucleus tends to produce a contralateral homonymous hemianopsia with sparing of the horizontal meridian.

References:


Question 204: Anatomy - Cortex and Connections

Discussion:

A group of long association fibers that interconnects the superior and middle frontal gyri with the posterior superior temporal gyrus is the arcuate fasciculus. In the dominant hemisphere, interruption of the arcuate fasciculus, between Wernicke's and Broca's area results in conduction aphasia.

References:


Questions 210 - 214: Anatomy - Basal Ganglia and Thalamus

Discussion:
The globus pallidum has efferent fibers that synapse on the ventral lateral (VL), ventral anterior (VA), and mediodorsal nucleus. Of those, only the mediodorsal projects to the cingulate gyrus. VA projects to the prefrontal cortex, and VL projects primarily to the premotor cortex. The anterior nuclear group also projects to the cingulate gyrus, but its afferents come from the mammillary bodies via the mammillothalamic tract. The reticular nucleus of the thalamus is responsible for the generation of sleep spindles seen on EEG during sleep. The reticular nucleus has no cortical projections, but instead projects to other thalamic nuclei, including the intralaminar nuclei (IL). The intralaminar nuclei are cell groups within the internal medullary lamina, which separates the medial and lateral subdivisions of the thalamus. Its afferents are from nuclei in the brainstem reticular formation, and has diffuse cortical projections. The pulvinar receives fibers from the superior colliculus and has projections to cortical areas 18 and 19, serving as a component in the extragenticulate visual pathway.

References:


Questions 215 - 219: Anatomy - Spinal Cord

Discussion:

Proprioceptive and vibratory loss in the lower extremities, due to a spinal cord lesion, involves the fasciculus gracilis which serves these functions below the level of T6.

A central cord lesion which produces pain and temperature dysfunction in a bilateral "shawl" or "cape" distribution is due to involvement of crossing fibers for these modalities in the anterior (aka ventral) white commissure.

In high cervical cord lesions, ipsilateral diminished pain and temperature sensation in the pre auricular area of the face is due to involvement of cells in the substantia gelatinosa which is the distal continuation of the descending trigeminal nucleus and tract.

The interomediolateral cell column contains pre-ganglionic sympathetic neurons. Involvement of this area in the upper thoracic cord may result in an ipsilateral Horner's syndrome.

Extradural impingement of the lateral cervical cord may cause weakness and upper motor neuron signs in the ipsilateral lower extremity because of the somatotopic organization of sacral and lumbar fibers being most lateral within the lateral corticospinal tract.

References:


Question 276: Anatomy - Brainstem/Cerebellum

Discussion:

Climbing fibers originate in the inferior olivary complex and appear to have glutamate as their neurotransmitter. Each climbing fiber possesses an extensive all - or - none excitatory connection with Purkinje cell dendrites in the cerebellar cortex. The red nucleus projects to inferior olivary complex via the central tegmental tract.

References:
Question 304: Anatomy - Blood Supply of Brain/Spinal Cord

Discussion:

The posterior cerebral artery supplies parts of the inferior temporal lobe, occipital lobe, splenium of the corpus callosum and superior parietal lobule.

References:


Question 331: Anatomy - Peripheral Nervous System

Discussion:

The synapse of the afferent axons for muscle stretch reflexes is located in the anterior (ventral) horn. The muscle stretch reflex is monosynaptic directly from sensory neuron to an alpha motor neuron.

References:


Question 345: 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 346: Anatomy - Cortex and Connections

Discussion:

The imitation of gestures (echopraxia) and mimicking of use of objects in the environment (utilization behavior) are seen after lesions to the lateral orbitofrontal cortex.

References:


Question 357: Anatomy - Cortex and Connections

Discussion:

The primary input to the granule cells of the dentate gyrus comes from the entorhinal cortex via the perforant path. These cells then project to the CA3 pyramidal cells via the mossy fiber pathway.
References:


Question 359: Anatomy - Blood Supply of Brain/Spinal Cord

Discussion:

The internal carotid artery (ICA) can be divided into four segments: cervical, intrapetrosal, intracavernous, and supraclinoid. The ophthalmic artery arises from the ICA just as it emerges from the cavernous sinus, in the supraclinoid segment.

References:


Question 361: Anatomy - Cranial Nerves

Discussion:

The abducens nerve lies immediately adjacent to the internal carotid artery in the cavernous sinus. The oculosympathetic fibers also travel near the abducens nerve while the other cranial nerves lie within the wall.

References:


Question 370: Anatomy - Peripheral Nervous System

Discussion:

The thoracodorsal nerve branches directly from the posterior cord of the brachial plexus to supply the lattisimus dorsi. The Medial Antebrachial cutaneous nerve branches from the the medial cord, the suprascapular nerve from the upper trunk, and the Lateral pectoral nerve from the lateral cord. The posterior introsseous branches from the radial nerve

References:


Question 375: Anatomy - Basal Ganglia and Thalamus

Discussion:

Fibers from retinal ganglion cells project to the suprachiasmatic nucleus of the hypothalamus. This nucleus serves as the pacemaker for circadian rhythm.

References:

Question 385: Anatomy - Cranial Nerves

Discussion:

The glossopharyngeal nerve exits the skull through the jugular foramen.

References:


Question 391: Anatomy - Basal Ganglia and Thalamus

Discussion:

The pulvinar receives fibers from the superior colliculus and projects to areas 17, 18 and 19. Both the inferior and lateral pulvinar have reciprocal connections with the occipital cortex. The medial geniculate receives input from the inferior colliculus while the ventral anterior nucleus receives input from the globus pallidus pars interna. The ventral posteromedial and posterolateral nuclei receive somatosensory input.

References:


Question 398: Anatomy - Brainstem/Cerebellum

Discussion:

The nucleus of the tractus solitarius (NTS) handles visceral afferents carried by cranial nerves and therefore contains the first central neuron for the baroreceptor afferents. Lesions involving the NTS produce fluctuating hypertension mimicking a pheochromocytoma. The nucleus ambiguus is the source of the branchial motor component of the glossopharyngeal nerve and the vagus. The arcuate nucleus is located in the tuberal region of the hypothalamus. The dorsal motor nucleus of the vagus gives rise to preganglionic parasympathetic fibers. The nucleus prepositus is the largest of the perihypoglossal nuclei and seems to be concerned with horizontal eye movements.

References:


Question 402: Anatomy - Peripheral Nervous System

Discussion:

The pudendal nerve innervates the external anal sphincter and is therefore necessary for the maintenance of voluntary fecal continence. The internal anal sphincter, innervated by the sympathetic fibers of the hypogastric plexus with cell bodies located at L1 and L2, maintains reflex continence. Parasympathetic activity is associated with defecation. The perineal nerve carries sensory information from the scrotum or labia.

References:
Question 409: Anatomy - Peripheral Nervous System

Discussion:

The third order neurons of the oculosympathetic pathway arise from the superior cervical ganglion on the common carotid artery. A dissection of the internal carotid artery may result in an ipsilateral Horner syndrome with ptosis, and a miotic pupil that poorly dilates. The fibers destined for the facial sweat glands are carried on the external carotid artery and therefore would be spared.

References:


Question 418: Anatomy - Brainstem/Cerebellum

Discussion:

Axons of the Purkinje cells are the only ones to emerge from the cerebellar cortex and project to the deep cerebellar nuclei.

References:


Question 428: Anatomy - Cortex and Connections

Discussion:

Alexia without agraphia leads to a deficit in word-reading but not letter-reading. Often there is a right homonymous hemianopsia and a color-naming deficit. This condition is usually associated with damage of the left occipital lobe and the splenium of the corpus callosum, producing a disconnection between the left and right calcarine cortex and the left angular gyrus. The patient is usually able to write spontaneously but not read what has been written. Patients can often identify words spelled orally. Number-reading may be preserved.

References:


Question 435: Anatomy - Cortex and Connections

Discussion:

Seizures arising from the supplementary sensorimotor area (SSMA) are of brief duration, usually lasting 10 to 40 seconds. Rapid onset of asymmetric tonic posturing involving one or more extremities is characteristically observed. SSMA seizures may be frequent and can occur in clusters. They tend to occur primarily in sleep. The clinical picture with involvement of all four extremities and simultaneous preservation of awareness can inadvertently lead to the misdiagnosis of psychogenic seizures.
Discussion:

The vagus nerve derives its fibers from the nucleus ambiguus, the dorsal motor nucleus of the vagus, the spinal trigeminal nucleus, and the nucleus solitarius. The nucleus ambiguous supplies branchial motor fibers that travel in the vagus nerve to the muscles of the pharynx and larynx. The dorsal motor nucleus provides parasympathetic innervation to the heart, lungs and digestive tract.

References:


Behavioral/Psychiatry

Question 9: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:

This patient had nutmeg toxicity. Although a common ingredient in some pies, an excessive amount may be toxic. Nutmeg contains myristicin, which is a hallucinogen. It may produce hallucinations, severe headache and agitation, flushing, palpitations and extremity numbness. The symptoms usually resolve within several hours; however, the patient may require supportive therapy for the headache and autonomic features. A benzodiazepine may also help the agitation.

References:


Question 12: Behavioral/Psychiatry - Neurobiology of Behavior

Discussion:

The majority of structural lesions associated with development of obsessive-compulsive behavior have involved the frontal lobe and/or frontal-basal ganglia network connections.

References:


Question 19: Behavioral/Psychiatry - Psychopharmacology

Discussion:

Amytriptyline has significant anticholinergic effects and would not be ideal for Alzheimer patients. Lorazepam and alprazolam often increase confusion and are too sedating. Valproate is used more for bipolar disorder than depression.

References:
**Question 20: Behavioral/Psychiatry - Psychopharmacology**

**Discussion:**

Habit-forming lorazepam should be avoided in the demented elderly as the effects are often too sedating and increase confusion. Antipsychotics should not be used first line for sleep disturbances due to their significant potential adverse effects in this population. Antihistamines and sedating tertiary tricyclic antidepressants like doxepin have significant anticholinergic effects that would fight the effect of the galantamine.

**References:**


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**Question 21: Behavioral/Psychiatry - Psychopharmacology**

**Discussion:**

If suspiciousness and paranoia are the underlying cause of the aggression, than atypical antipsychotics like quetiapine should be considered first line for treatment. Typical antipsychotics like haloperidol have sedative and extrapyramidal properties that make it less desirable as a daily treatment. In addition, low potency typical antipsychotic agents like thioridazine have significant anticholinergic effects that lead to increased cognitive deficits. Valproate and lorazepam are not particularly good at treating psychosis but can be used for agitation that is not psychotic based.

**References:**


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**Question 31: Behavioral/Psychiatry - Dementia**

**Discussion:**

Parkinsonism with cognitive loss, fluctuating symptoms and no resting tremor points to dementia with Lewy bodies. Asymmetric signs are classical for cortical basal degeneration. Lack of resting tremor and memory loss preceding the Parkinsonism points away from Parkinson disease dementia. Rigidity in the upper extremities and fluctuating symptoms are not characteristic of normal pressure hydrocephalus. Lack of orthostasis or autonomic dysfunction tends to rule out Shy-Drager syndrome.

**References:**


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**Question 32: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease**

**Discussion:**

Recent research suggests that type II diabetes raises the odds of developing Alzheimer disease by 65%. The speed at which diabetics make perceptual comparisons is one of the most sensitive cognitive tests in diabetics with dementia. The speed of this
performance declines by over 40% over time. Treatment of a patient with cognitive decline and type II diabetes calls for aggressive treatment of the disease, including the use of insulin.

References:


Question 38: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:

Both syphilis and Lyme disease present with clinical manifestations based on the stage of the illness. In Lyme disease, most neurologic findings present in stage II which occurs within 6 months of the initial infection. These symptoms can include radiculitis, meningoencephalitis, and cranial neuropathies. The third stage generally occurs up to 9 years after infection and can include arthritis, skin discoloration, and a progressive encephalomyelitis/dementia. MRI in neuroborreliosis typically shows T2 white matter hyperintensities.

References:


Question 54: Behavioral/Psychiatry - General Psychiatry

Discussion:

While anxiety is present in many psychiatric disorders, it is not the major criterion in most. An acute and severe anxiety response is, however, the defining characteristic of a simple phobia.

References:


Question 66: Behavioral/Psychiatry - Dementia

Discussion:

The NINCDS-ADRDA criteria state an individual with probable Alzheimer disease must have an onset between ages 40 and 90 and have progressive cognitive loss with deficits in two or more areas of cognition. Dementia is established by clinical examination and confirmed by neuropsychological tests. The diagnosis of possible Alzheimer disease includes having a dementia syndrome in the absence of other psychiatric or systemic disorders but in the presence of a second systemic or brain disorder sufficient to produce a dementia but is not considered to be the cause of the dementia syndrome after a careful clinical evaluation. Definite Alzheimer disease occurs only when the clinical criteria for probable Alzheimer disease is met and histopathologic evidence of Alzheimer disease is available from an autopsy or biopsy. A non-Alzheimer disease dementia is not correct since there were no psychiatric or systemic disorder found to cause the dementia and the vascular disease was not thought to be the cause of their dementia.

References:


Question 68: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease
Discussion:

This patient had Wernicke encephalopathy, characterized by confusion, extraocular deficits, nystagmus, ataxia, and decreased or absent muscle stretch reflexes. The clue to this case was that he was homeless, suggesting the possibility of nutritional deficiency with or without alcoholism. He was initially administered D50 but no thiamine, a scenario that may precipitate Wernicke's. Because he had been unconscious due to a subdural hematoma, the adverse clinical effects of the D50 were masked until he awoke from anesthesia. The teaching point is that all unconscious patients should receive a dose of thiamine prior to the administration of dextrose solutions.

References:


Question 69: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:

This patient had Wernicke's encephalopathy characterized by confusion, extraocular deficits, nystagmus, ataxia, and decreased or absent muscle stretch reflexes. The clues in this case were that he was homeless suggesting the possibility of nutritional deficiency with or without alcoholism. He was initially administered D50 but no thiamine, a scenario that may precipitate Wernicke's. Because he had been unconscious due to a subdural hematoma, the adverse clinical effects of the D50 were masked until he awoke from anesthesia. The teaching point is that all unconscious patients should receive a dose of thiamine prior to the administration of dextrose solutions.

References:


Question 80: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:

Methyl mercury intoxication may occur through the ingestion of fish that have been exposed to the compound in industrial waste. It causes a subacute syndrome manifested by confusion, apathy, lethargy, dementia, irritability, personality change, constricted visual fields, and a prominent diffuse cerebellar syndrome due to destruction of granule cells of the cerebellum. It can be treated with penicillamine or dimercaptosuccinic acid. BAL should not be used because it causes increased concentration of mercury in the brain.

References:


Question 82: Behavioral/Psychiatry - Language/Speech Abnormalities

Discussion:

The supplementary motor area (SMA) is thought to act as a pacemaker for verbal output. Damage to the SMA or to its connections with Broca area produces the clinical picture of transcortical motor aphasia.

References:


Question 90: Behavioral/Psychiatry - General Psychiatry
Discussion:

Among the defining characteristics of posttraumatic stress disorder are duration of more than a month, the occurrence of flashbacks, reminiscences of the episode of severe stress that the patient suffered, and persistent symptoms of increased arousal, including an exaggerated startle response.

References:


**Question 91: Behavioral/Psychiatry - Occipital Syndromes**

Discussion:

Alexia without agraphia (also known as pure word blindness or acquired pure alexia) is most often produced by left posterior cerebral artery occlusion. A frequent associated finding is a right homonymous hemianopsia.

References:


**Question 100: Behavioral/Psychiatry - Dementia**

Discussion:

Major depression is frequent in Parkinson disease, occurring in 40% to 60% of patients during the course of their illness. It is less common in dementia of the Alzheimer type, Lewy body dementia, frontotemporal dementia, and normal pressure hydrocephalus (NPH).

References:


**Question 107: Behavioral/Psychiatry - Parietal Syndromes**

Discussion:

The Gerstmann's syndrome - agraphia, acalculia, right/left disorientation, finger agnosia - strongly suggests damage in the left (dominant) parietal lobe in particular the angular gyrus.

References:

**Question 110: Behavioral/Psychiatry - Dementia**

Discussion:

The memory disorder of early probable Alzheimer disease involves impairment in word recall (recent memory) with normal digit span (immediate memory), and relatively spared remote memory.

References:

**Question 113: Behavioral/Psychiatry - Temporal-Limbic Syndromes**

**Discussion:**

Topographic disorientation (impaired orientation and navigation in the environment) is most strongly associated with damage in the right posterior parahippocampal region or the infracalcarine cortex. Right parietal damage may cause a milder form of the disorder.

**References:**


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**Question 114: 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 are present with one of the primary findings of visual hallucinations, parkinsonism, or fluctuating attention, then the diagnosis of probable DLB is supported.

**References:**


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**Question 118: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease**

**Discussion:**

After treatment of post-treatment chronic Lyme disease (PTCLD), patients showed improvement in subjective symptoms. The patients also showed slight improvement in objective neuropsychological testing despite their baseline being within normal limits. Baseline neuropsychological testing is usually normal despite patients' complaints of cognitive dysfunction. Patients who received placebo had the exact same clinical outcome. Therefore, retreatment with antibiotics in Lyme disease is not warranted for neurobehavioral symptoms.

**References:**


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**Question 121: Behavioral/Psychiatry - Neurobehavioral/Neuropsychological Exam**

**Discussion:**

Achromatopsia, the loss of the ability to recognize color, will cause total failure of number or pattern recognition on the Ishihara plates. Patients with color anomia will still retain the ability to perceive the patterns on the Ishihara plates.

**References:**

**Question 126: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease**

**Discussion:**

This history depicts a case of herpes simplex virus encephalitis. The treatment of choice is acyclovir.

**References:**


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**Question 132: Behavioral/Psychiatry - Language/Speech Abnormalities**

**Discussion:**

Patients with semantic dementia, a category of frontotemporal dementia, have an insidiously progressive yet relatively focal disease until late in the course of their illness. These patients have fluent yet empty speech, coupled with naming impairment and failure to understand the meaning of words. The latter may eventually manifest as an agnosia and apraxia for the object. It is common for patients to use semantic paraphasias and idiosyncratic words or phrases as substitute fillers for names they cannot recall. They may also develop difficulty identifying familiar people by name. An interesting aspect of this condition is that when words that may be used as either a noun or verb (such as "break") are spoken to the patient, the patient has more difficulty understanding the meaning of the noun than the use of the verb. As the disease progresses, features of social disinhibition such as those seen with frontal lobe dementia begin to emerge. The MRI findings typically demonstrate dominant hemisphere anterior temporal atrophy with relative sparing of the hippocampal formation.

**References:**


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**Question 133: Behavioral/Psychiatry - Language/Speech Abnormalities**

**Discussion:**

Patients with semantic dementia, a category of frontotemporal dementia, have an insidiously progressive yet relatively focal disease until late in the course of their illness. These patients have fluent yet empty speech coupled with naming impairment and failure to understand the meaning of words. The latter may eventually manifest as an agnosia and apraxia for the object. It is common for patients to use semantic paraphasias and idiosyncratic words or phrases as substitute fillers for names they cannot recall. They may also develop difficulty identifying familiar people by name. An interesting aspect of this condition is that when words that may be used as either a noun or verb (such as "break") are spoken to the patient, the patient has more difficulty understanding the meaning of the noun than the use of the verb. As the disease progresses, features of social disinhibition such as those seen with frontal lobe dementia begin to emerge. The MRI findings typically demonstrate dominant hemisphere anterior temporal atrophy with relative sparing of the hippocampal formation.

**References:**


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**Question 140: Behavioral/Psychiatry - Temporal-Limbic Syndromes**

**Discussion:**

A number of brain systems have been implicated in addictive behavior, but none have yet been shown to be necessary for maintaining the addiction to cigarette smoking. Smokers with brain damage involving the insula, a region implicated in conscious urges, are more likely than smokers with brain damage not involving the insula to undergo a disruption of smoking addiction, characterized by the ability to quit smoking easily, immediately, without relapse, and without persistence of the urge to smoke.
Question 141: Behavioral/Psychiatry - Psychopharmacology

Discussion:
Anticholinergic side effects are of considerable significance in the pharmacologic treatment of depression. Among those that are most likely to cause these side effects are the tricyclic antidepressants, especially the tertiary amines such as amitriptyline.

References:

Question 148: Behavioral/Psychiatry - General Psychiatry

Discussion:
Clinical features of catatonia include psychosocial withdrawal, stupor, mutism, immobility, generalized analgesia, posturing, waxy flexibility, mannerisms, rituals, grimacing, and shoulder shrugging.

References:

Question 163: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:
Decreased hemoglobin and elevated mean corpuscular volume are compatible with a macrocytic anemia that may be secondary to vitamin B12 deficiency. Vitamin B12 deficiency, in turn may cause combined system degeneration (affecting posterior columns and lateral corticospinal tracts). Dementia and peripheral neuropathy may also be seen.

References:

Question 173: Behavioral/Psychiatry - Developmental Disorders

Discussion:
A girl with classic Rett syndrome appears normal to her parents for most of the first year of life. She may begin walking or may have mildly delayed motor milestones. Her social development appears normal, and she may begin to use and understand language. Sometime between 1 and 2 years, she experiences a regression of these skills. She stops playing with toys. She stops responding to the spoken word and stops using the few single words she has learned. She becomes withdrawn and loses interest in social interaction. She may stop walking. Purposeful hand use is replaced by stereotypic hand movements. The lost developmental skills are never recovered, and she moves through life at a level of profound mental retardation often accompanied by seizures.

References:

Question 174: Behavioral/Psychiatry - Neurobehavioral/Neuropsychological Exam
Digit span, which involves attentional processes, immediate recall and ability to sequence bits of information, may be reduced following lesions of dorsolateral frontal cortex. Lesions of the fornix, mammillary bodies and medial dorsal nucleus of the thalamus, on the other hand, cause amnesia (impaired secondary memory) without a reduction of digit span.

References:


The 28 year old man in this case has AIDS-Dementia complex. In addition he developed a subacute illness which led to rapid progression over the course of a month and eventually death. This is most consistent with progressive multifocal leukoencephalopathy (PML). Clinically he manifested headache, irritability, inattention, and a host of visual problems consistent with a Balint's syndrome. Although many different areas of the brain may be affected by PML it is common to have lesions involving the subcortical white matter in occipital-parietal locations which correspond to this patient's clinical features.

References:


Akinetic mutism, with somnolence, indicates a disorder affecting the reticular projections from the mesencephalon to the thalamus.

References:


Inability to produce melody in verbal output, amelodia, produces a significant loss of non-verbal affective expression (affective motor aprosodia). The causative lesion is most commonly found in the posterior inferior frontal lobe of the right hemisphere, the region in the right frontal lobe analogous to Broca's area in the left.

References:

Callosal apraxia results from a lesion in the genu of the corpus callosum. This results in a left limb kinetic apraxia. Tactile and auditory input cross the corpus callosum posteriorly and are therefore unaffected by a genu lesion. Alexia without agraphia results from a left occipital splenium of the corpus callosum lesion.

References:

**Question 358: Behavioral/Psychiatry - Language/Speech Abnormalities**

**Discussion:**

The nonfluent cortical aphasias include Broca, global, mixed transcortical, and transcortical motor. Patients with a conduction aphasia are fluent as are patients with any type of sensory aphasia.

References:


**Question 366: Behavioral/Psychiatry - General Psychiatry**

**Discussion:**

Problems with interpersonal relationships, fear of abandonment, waxing and waning between idealizing and devaluing people close to them, impulsive outbursts of anger, suicidal gestures, associated depression, and often dissociative states are all commonly seen in the patient with borderline personality disorder.

References:


**Question 367: 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 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 372: Behavioral/Psychiatry - Dementia**

**Discussion:**

Spatial orientation is preserved in frontotemporal dementia even late into disease progression.

References:

Question 382: Behavioral/Psychiatry - Frontal Systems Syndromes

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 patients mood. While depression is common, manic-depressive illness is not. Obsessive-compulsive traits, visual hallucinations, and physical aggression are less often seen.

References:


Question 383: 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 dysfunctioning. 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 387: Behavioral/Psychiatry - Developmental Disorders

Discussion:

Attachment theory investigates secure and insecure parent-child bonding in early life and its consequences for later development.

References:

Question 400: Behavioral/Psychiatry - Occipital Syndromes

Discussion:

Alexia without agraphia is often seen with lesions of the left occipital cortex that frequently are seen to extend into the splenium of the ipsilateral corpus callosum. Because of the occipital cortical involvement and splenial involvement, there may be associated color anomia and right homonomous hemianopia.

References:


Question 401: Behavioral/Psychiatry - Parietal Syndromes

Discussion:
Damage to the nondominant parietal lobe results in prominent neglect or denial of the contralateral half of the body and space and dressing apraxia. Right-left confusion occurs with dominant parietal lobe lesions.

References:


Question 431: Behavioral/Psychiatry - Dementia

Discussion:

Since the instrumental activities of daily living are performed normally, she cannot have a dementia condition or Alzheimers disease. Age associated memory impairment and amnestic MCI refer to individuals with only memory impairments.

References:


Question 432: Behavioral/Psychiatry - Dementia

Discussion:

Apolipoprotein E is a risk factor for Alzheimer disease but can not predict progression to Alzheimers disease. Serial volumetric MRIs looking at hippocampal size may be able to predict a progressive course but a single routine MRI could not. LDL and CSF tau do not have predictive abilities for progression. PET early on may show bilateral parietal and temporal hypometabolism that has been shown to predict a progressive declining course.

References:

Minoshima et al.. PET. Ann Neurol 1997;42:85-94

Question 436: Behavioral/Psychiatry - Developmental Disorders

Discussion:

There are association between ADHD and polymorphisms in DRD4, DRD5 and SLC6A3 which encode dopamine D4 and D5 receptors and the dopamine transporter, respectively. A weaker, but nevertheless replicated, body of evidence also supports associations with SNAP-25 (synaptosomal-associated protein, 25 kDa) and SLC6A4 (serotonin transporter).

References:


Question 440: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:

Ischemia of the midbrain tectum can result in visual hallucinations known as peduncular hallucinosis. Bilateral basilar midbrain ischemia can result in quadraparesis. Medullary infarcts may cause quadraparesis and anterior cerebral infarcts may cause paraparesis but neither will cause visual hallucinations. Posterior cerebral artery infarcts may cause visual hallucinations (usually not formed) but will not cause paresis. Temporal lobe infarct may cause psychosis but not quadraparesis.

References:
**Question 443: Behavioral/Psychiatry - Neurobiology of Behavior**

**Discussion:**

Dorsolateral association cortex monitors, judges, facilitates and inhibits activity of distributed complex cortical networks. This is referred to as the "executive function" of the frontal lobes.

**References:**


**Clinical Adult**

**Question 5: Clinical Adult - Neuro-ophthalmology/Neuro-otology**

**Discussion:**

Symptoms of perilymph fistula are variable, but may include recurrent vestibulopathy. Characteristic precipitating factors include cough, sneeze, straining, and exercise.

**References:**


**Question 8: Clinical Adult - Neuro-oncology**

**Discussion:**

This patient has clinical features indicating Foster Kennedy syndrome, classically caused by an olfactory groove meningioma. This syndrome is characterized by a lesion involving one or both olfactory nerves and one optic nerve. This results in unilateral or bilateral reduction of olfaction, reduced visual acuity and color vision in the affected eye, and papilledema in the other eye, due to raised intracranial pressure.

**References:**


**Question 15: Clinical Adult - Cerebrovascular Disease**

**Discussion:**

This patient has most likely had an embolic stroke from a mural thrombus. This occurs in 2% to 6% of patients with anterior myocardial infarction (MI), usually within the first 2 weeks of the MI.

**References:**


**Question 17: Clinical Adult - Movement Disorders**
**Discussion:**

This patient's symptoms are most consistent with restless legs syndrome. Iron deficiency is commonly associated with restless legs syndrome, necessitating a serum ferritin level in this patient.

**References:**


**Question 23: Clinical Adult - Critical Care/Trauma**

**Discussion:**

The patient has a grade II concussion requiring a 1-week withdrawal from contact sports. Following this she should be able to return to contact sports without difficulty unless a neurologic deficit has emerged.

**References:**


**Question 28: Clinical Adult - Spinal and Root Disorders**

**Discussion:**

History and examination is consistent with spinal stenosis in the lumbar area. The examination and the fact that he can ride a bike implies peripheral vascular disease and deep venous thrombosis are less likely.

**References:**


**Question 30: Clinical Adult - Dementia**

**Discussion:**

The clinical history is most consistent with Lewy body dementia. The presence of parkinsonism, dementia, visual hallucinations and REM behavior disorder is characteristic of that disorder. The other disorders would be characterized by other clinical features, including autonomic dysfunction, ataxia, and gaze abnormalities.

**References:**


**Question 37: Clinical Adult - Spinal and Root Disorders**

**Discussion:**

This patient most likely has epidural cord compression from metastatic lung cancer, a neurologic emergency. Urgent treatment with high dose IV corticosteroids is indicated in addition to immediate spinal imaging with MRI.
Discussion:

Ciguatera poisoning results from consuming large reef fish (i.e., grouper, barracuda, and red snapper), which may contain ciguatoxin (CTX). CTX is a heat-stable toxin produced by marine microorganisms (dinoflagellates) in coral reefs and then passed up the food chain where it becomes concentrated in large reef fish like grouper and barracuda. CTX opens voltage-gated sodium channels in peripheral nerves at the nodes of Ranvier causing spontaneous depolarization. The clinical manifestations are usually sensory. Paradoxical sensory disturbances are particularly common (i.e., hot feels cold and vice versa). Maitotoxin, also in reef fish, activates voltage-dependent calcium channels and usually produces bradycardia and hypotension. Tetrodotoxin, saxitoxin, and conotoxin are found in puffer fish, shellfish, and sea snails respectively and present with paralysis not sensory phenomena.

References:


Question 50: Clinical Adult - Motor Neuron/Nerve

Discussion:

The history and examination are suggestive of a rapidly progressive parainfectious polyneuropathy. The CSF is consistent with acute inflammatory demyelinating polyneuropathy (AIDP). His prior history, CSF data, pain, and other features make the other diagnoses less likely than AIDP.

References:


Question 52: Clinical Adult - Movement Disorders

Discussion:

Dopamine agonists have been reported to produce a behavioral dyscontrol syndrome in patients with Parkinson disease.

References:


Question 58: Clinical Adult - Dementia

Discussion:

This patient presents with abnormal memory, but maintains a high level of functioning, precluding a diagnosis of dementia. According to current research, mild cognitive impairment (MCI) is a definite risk factor for the development of Alzheimer disease (AD). The conversion rate from MCI to AD is approximately 10% to 15% per year.

References:
Question 62: Clinical Adult - Spinal and Root Disorders

Discussion:

This man presents with a peripheral sensory syndrome, followed by upper motor neuron involvement and optic nerve involvement. He then has diffuse cognitive abnormalities, making adrenal leukodystrophy the most likely diagnosis. Since his course extends over almost 10 years, HIV, also multifocal, is unlikely and is more acute. Progressive multifocal leukoencephalopathy (PML) from JC virus is much more rapidly fatal and would not have peripheral nerve disease. Expanded trinucleotide repeat in frataxin gene is the most common genetic mutation found in Friedreich ataxia, but it involves both corticospinal tracts and peripheral nerve. Cognition is spared and dorsal column involvement is prominent. Accumulation of long-chain fatty acids is correct, with the clinical course typical of an X-linked disease with diffuse central and peripheral nervous system involvement, characteristic for adrenomyeloneuropathy.

References:


Question 65: Clinical Adult - Headache

Discussion:

This patient's symptoms are most compatible with migraine. There are no features of her history or examination to suggest a more serious underlying disorder such as meningitis, subarachnoid hemorrhage, or a space occupying lesion. Parenteral symptomatic therapy is warranted of which sumatriptan would likely be most effective.

References:


Question 70: Clinical Adult - Epilepsy

Discussion:

In a patient with status epilepticus, hypoglycemia should always be immediately considered as an underlying etiology, particularly in a patient with diabetes who is on insulin. If serum glucose is not immediately available or the value is uncertain, 50 cc of 50% dextrose should be administered intravenously concurrently with 100 mg of intravenous thiamine.

References:


Question 72: Clinical Adult - Critical Care/Trauma

Discussion:

The clinical scenario suggests an epidural hemorrhage involving the left hemisphere and causing uncal herniation. The other lesions may be caused by trauma, but localization of the patient's deficits and delay in onset of symptoms would be inconsistent with the diagnosis.

References:

Question 78: Clinical Adult - Neuro-ophthalmology/Neuro-otology

Discussion:

This patient's symptoms are most consistent with a right fourth nerve palsy. The compensatory head positions that a patient with acquired fourth nerve palsy would use to decrease the vertical diplopia 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 87: Clinical Adult - Epilepsy

Discussion:

This patient's symptoms and EEG findings are most consistent with juvenile myoclonic epilepsy (JME). JME is a generalized epilepsy that typically presents during adolescence or early adulthood. Patients with JME typically describe a history of morning myoclonic jerks, and both the generalized seizures and the myoclonic jerks commonly are precipitated by sleep deprivation and alcohol.

References:


Question 94: Clinical Adult - Critical Care/Trauma

Discussion:

Herpes simplex encephalitis is the most common form of acute sporadic encephalitis. It is characterized by lymphocytic pleocytosis and evidence of mild hemorrhage in the CSF.

References:


Question 101: Clinical Adult - Neuro-oncology

Discussion:

Patients with sensory neuronopathy and associated lung carcinoma frequently have anti-Hu antibodies. Anti-Jo1 antibodies are associated with polymyositis, acetylcholine receptor antibodies with myasthenia gravis, calcium channel antibodies with Lambert-Eaton myasthenic syndrome, and anti-amphiphysin antibodies with stiff person syndrome.

References:


Question 102: Clinical Adult - Spinal and Root Disorders
Discussion:

This patient had anterior spinal artery syndrome based upon the clinical findings of acute paraplegia with sparing of dorsal column function in patient with clear vascular risk factors. Complete transection of the cord would present with flaccid paralysis, complete sensory loss and autonomic dysfunction. Brown-Sequard syndrome involves contralateral loss of pain and temperature sensation due to the spinothalamic pathway. There will be ipsilateral weakness and loss of proprioception due to hemicord involvement. Central cord syndrome involves dissociated sensory loss with diminished pain and temperature but preserved proprioception such as seen in syrinx.

References:


Question 103: Clinical Adult - Headache

Discussion:

Triptans are contraindicated in the presence of known coronary artery disease due to the risk of precipitating vasospasm and coronary ischemia.

References:


Question 105: Clinical Adult - Neuro-ophthalmology/Neuro-otology

Discussion:

This is a patient with a swollen, painless optic nerve. The lack of pain and age of the patient eliminates optic neuritis and makes anterior ischemic optic neuropathy the most logical diagnosis. Neuroretinitis would show inflammation of the macula. Central retinal vein occlusion would produce retinal hemorrhages. Central retinal artery occlusion would produce diffuse pallor of the retina rather than disc edema.

References:


Question 116: Clinical Adult - Headache

Discussion:

This patient's symptoms are most consistent with cluster headaches. Patients with cluster headaches often describe ipsilateral rhinorrhea and tearing during the severe paroxysms of pain. A history of neck trauma would be important in the history of a patient with possible large artery dissection, which could cause retro-orbital pain but not the "clusters" of episodes of brief severe retro-orbital pain separated by years described in this patient. This patient's symptoms are not suggestive of trigeminal neuralgia, which can be triggered by touching of the cheek or brushing the teeth. In addition, this patient's symptoms are not highly suggestive of migraine which can be associated with photophobia and sonophobia and sometimes preceded by a visual aura.

References:


Question 120: Clinical Adult - Dementia
Discussion:

This patient most likely has Creutzfeldt-Jakob disease. Creutzfeldt-Jakob disease is characterized by a rapidly progressive dementia, focal neurologic deficits, and myoclonus. Neuroimaging may be normal or reveal subtle changes on FLAIR and DWI (diffusion-weighted imaging) in the basal ganglia or cortical mantle. The CSF examination shows no evidence of inflammation. An elevated 14-3-3 protein level or neuron-specific enolase may indicate acute widespread neuronal injury.

References:


Question 131: Clinical Adult - Demyelinating Disease

Discussion:

This patient's history and findings, including a history of severe optic neuritis in the past and a new episode of acute longitudinal myelitis, are most compatible with neuromyelitis optica (Devic's disease). Antibodies to aquaporin-4 are a sensitive and very specific marker for the diagnosis of neuromyelitis optica.

References:


Question 134: Clinical Adult - Neurogenetics

Discussion:

This patient has chronic progressive external ophthalmoplegia which is characteristic for Kearns-Sayre syndrome (KSS). KSS and oculopharyngeal muscular dystrophy (OPMD) both have ptosis, ophthalmoplegia, dysphagia, and proximal weakness. Hearing loss, cerebellar ataxia, and cardiac conduction defects are not features of OPMD but are seen with KSS. MELAS would have a history of migraine and stroke-like episodes.

References:


Question 137: 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, azathiaprine, 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 139: Clinical Adult - Demyelinating Disease

Discussion:
Discussion:

There is evidence that neutralizing antibodies (Nabs) affect interferon efficacy and have an effect on disease progression. Changing interferon preparations or route of administration would have no effect as there is cross-reactivity of the antibodies. Moreover, subcutaneous administration is more likely to be associated with Nab production. Natalizumab carries the risk of opportunistic infection with JC virus and is usually reserved for patients who have failed other, safer disease modifying agents. At this stage, the most prudent step would be to repeat antibody testing in 3 months. If the Nab titer remains elevated, then switching to a non-interferon alternate agent (glatiramer acetate) is appropriate.

References:


Question 149: Clinical Adult - Headache

Discussion:

This patient's history of brief (minutes), frequent, unilateral headaches with autonomic features is consistent with paroxysmal hemicrania. A trial of indomethacin is indicated as paroxysmal hemicrania is one of the indomethacin-responsive headache syndromes.

References:


Question 154: Clinical Adult - Neuromuscular Disorders

Discussion:

This patient most likely has idiopathic brachial plexitis (neuralgic amyotrophy, or Parsonage-Turner syndrome). This disorder is more common in young adults and often follows physical exertion or other physical stress, including upper respiratory tract infection, vaccination, surgery, and childbirth. A small percentage of patients have an autosomal dominantly inherited form of the disorder. Typically, idiopathic brachial plexitis presents with severe pain followed by weakness and sensory loss in the upper extremity. The prognosis for eventual recovery is good. The role of corticosteroids is not certain, although they often help pain considerably.

References:


Question 157: Clinical Adult - Neurology of Systemic Disease

Discussion:

The most likely diagnosis in this patient with rheumatoid arthritis is atlanto-axial subluxation, a serious complication due to damage to the transverse ligament or erosion of the odontoid process. It typically presents with cervical or occipital pain worse with movement and may produce a progressive high cervical myelopathy and sometimes symptoms of vertebrobasilar insufficiency.
Question 160: Clinical Adult - Epilepsy

Discussion:

While any of the listed drugs may be helpful in controlling her complex partial seizures, phenytoin, carbamazepine, and oxcarbazepine are inducers of the hepatic cytochrome P450 system which speeds the metabolism of oral contraceptives (OCs) and may contribute to contraceptive failure. Valproic acid does not interfere with OC metabolism but may cause weight gain, alopecia, tremor, and a high risk of teratogenesis. Therefore, levetiracetam would be the best drug to try first.

References:


Question 167: Clinical Adult - Headache

Discussion:

This patient's symptoms are most suggestive of temporal arteritis. An erythrocyte sedimentation rate (ESR) should be sent and reviewed while the patient is in the emergency department. Elevation of the ESR should lead to urgent steroid therapy to avoid irreversible visual loss, with a temporal artery biopsy to follow as soon as possible to provide histologic proof of the diagnosis.

References:


Question 169: Clinical Adult - Dementia

Discussion:

Predicting the chances of a good outcome is notoriously inaccurate, but duration of dementia is one of the more important considerations. Depressive symptoms may improve with shunting, but this may occur whether or not the patient's dementia syndrome improves. If the patient's shunt results in a decrease in ventricular size, that is indeed favorable, but the duration of dementia is a more critical variable. Persons with chronic normal pressure hydrocephalus (NPH) may not respond to the shunt, even if follow-up CT scans suggest that the hydrocephalus is improved. The duration of urinary incontinence reflects the degree of damage to the fibers of the medial frontal lobe micturition centers. The bilateral Babinski signs, severe gait abnormality, and spasticity all suggest the same type of damage. The duration of incontinence has little predictive value. Finally, subcortical types of dementia are more likely to be treatable than cortical varieties. This by itself is no guarantee that the shunt will produce clinical improvement in the dementia syndrome.

References:


Question 181: Clinical Adult - Neuromuscular Disorders

Discussion:

The correct answer is proximal myotonic myopathy (DM2). The predominantly proximal weakness would be unusual for myotonic dystrophy and the significant weakness suggests that this is not myotonia congenita or paramyotonia congenita.
Myotonic dystrophy (DM1) undergoes genetic amplification and characteristically will have earlier onset with each generation, including risk of the congenital form. Patients with DM1 have characteristic facial weakness and wasting.

References:

Question 192: Clinical Adult - Neuromuscular Disorders

Discussion:
This patient's symptoms are most consistent with a mild left ulnar neuropathy at the elbow due to neuropraxia from resting his arm on the car armrest. Given the mild, purely sensory signs and symptoms, it is most appropriate at this time to observe for possible resolution of his symptoms with avoidance of continued pressure on his ulnar nerve at the elbow. If symptoms persist or worsen, further investigation (e.g. EMG/NCV) would be indicated.

References:

Question 194: Clinical Adult - Cerebrovascular Disease

Discussion:
Intravenous TPA is FDA approved for the treatment of acute stroke within a 3-hour window from symptom onset. Contraindications to TPA include hemorrhagic stroke, coagulopathy, recent surgery or stroke. Intra-arterial thrombolysis is still considered investigational.

References:

Question 199: Clinical Adult - Motor Neuron/Nerve

Discussion:
Midodrine is a prodrug that is metabolized in the liver to des-glymidodrine, a potent alpha-1 adrenergic agonist that produces arterial and venous vasoconstriction. Unlike other sympathomimetics, midodrine has a predictable absorption and plasma half life. It is currently the drug of second choice after fludrocortisone for management of neurogenic orthostatic hypotension. The dose is 10-40 mg/day. It should not be administered in the evening given the risk of supine hypertension. Scalp pruritus is a common side effect.

References:

Question 201: Clinical Adult - Epilepsy

Discussion:
Tramadol is a synthetic, centrally acting analgesic with weak mu receptor activity. Tramadol also inhibits reuptake of norepinephrine and serotonin. Seizures have been reported in patients receiving the drug in overdose and, rarely, at the recommended dose. It is important to consider tramadol as a possible cause of seizures even when used at recommended doses.
Question 300: Clinical Adult - Headache

Discussion:

This patient has a spontaneous low-pressure headache that is likely due to a dural tear that occurred during her fall. The radiographic appearance of diffuse dural enhancement is typical.

References:


Question 320: Clinical Adult - Demyelinating Disease

Discussion:

This patient's signs and symptoms are most compatible with retrobulbar optic neuritis. Intravenous corticosteroids are beneficial in hastening resolution of visual dysfunction in patients with retrobulbar optic neuritis and are appropriate to recommend to this patient given the severity of his visual dysfunction.

References:


Question 324: Clinical Adult - Neuromuscular Disorders

Discussion:

This patient's signs, symptoms, and laboratory findings are most consistent with Miyoshi myopathy. Miyoshi myopathy is an autosomal recessive muscular dystrophy that characteristically presents with weakness in the posterior calves, marked elevation in serum CK, and typically has onset between the ages of 12 and 30. Miyoshi myopathy is caused by mutations in the gene encoding dysferlin, a muscle surface membrane protein.

References:


Question 325: Clinical Adult - Neuromuscular Disorders

Discussion:

This patient's signs and symptoms of sensory loss to pinprick with preservation of vibratory sense and reflexes, is most consistent with a small fiber neuropathy. His symptoms are compatible with leprosy, which occurs due to Mycobacterium leprae, a neurotropic bacterium. This diagnosis is supported by the distribution of this patient's sensory loss, which includes his ears and nose, and the fact that he has emigrated from a country endemic for leprosy.

References:
**Question 330: Clinical Adult - Movement Disorders**

**Discussion:**

This patient has an acute dystonic reaction caused by dopamine receptor blockade within the CNS by metoclopramide. This adverse effect is most likely to occur in younger patients within the first 48 hours of starting antidopaminergic medications. The appropriate therapy is an anticholinergic agent such as benztropine or diphenhydramine.

**References:**


**Question 332: 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 gastric surgery and zinc overdose.

**References:**


**Question 334: Clinical Adult - Neurology of Systemic Disease**

**Discussion:**

This patient has Wernicke encephalopathy. Thiamine is involved in function of pyruvate dehydrogenase. Deficiency of thiamine produces activity in a shunt pathway which in turn produces hemorrhagic necrosis in deep brain structures.

**References:**


**Question 341: Clinical Adult - Sleep**

**Discussion:**

This patient gives a classic history for narcolepsy. She has three of the typical clinical features of narcolepsy: excessive daytime sleepiness, hypnagogic hallucinations, and sleep paralysis. She should be questioned about the presence of cataplexy, which is also often present in these patients.

**References:**


**Question 342: Clinical Adult - Neuromuscular Disorders**
Discussion:

This patient most likely has Lambert-Eaton myasthenic syndrome (LEMS). LEMS is an autoimmune disorder resulting from antibodies directed against the voltage-gated calcium channel of cholinergic synapses. The muscle weakness is usually proximal and is improved with repeated contraction. The nerve conduction studies typically show an increment with 50 Hz repetitive stimulation.

References:


Question 343: Clinical Adult - Epilepsy

Discussion:

This patient most likely has psychogenic seizures. Characteristic features include the back-arching behavior which is rarely seen in epileptic events. The pelvic thrusting is also unusual in epilepsy but can be seen in some frontal lobe seizures. Other motor phenomena commonly associated with psychogenic seizures includes irregular, asynchronous movements of the extremities, side-to-side head movements, stuttering, weeping and preserved awareness during bilateral motor activity.

References:


Question 350: Clinical Adult - Neurorehabilitation

Discussion:

This patient's signs and symptoms are most consistent with a neuropraxic radial nerve palsy from mechanical compression. Further diagnostic testing is not necessary in this patient. This patient should be treated with wrist splinting to avoid a flexion contracture of the wrist and improve hand function. He will most likely experience spontaneous recovery of function.

References:


Question 353: 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 usually represents the prodromal phase of Alzheimer disease.

References:


Question 355: Clinical Adult - Movement Disorders

Discussion:

This patient most likely has fragile X-associated tremor ataxia syndrome (FXTAS). He is not likely to have Friedreich ataxia, dentatorubral-pallidoluysian atrophy, or spinocerebellar ataxia type 3 given his age, absence of other clinical features, and the
history of mental retardation in his daughter's son. Patients with Charcot-Marie-Tooth disease may have postural and action tremor but should not have cerebellar ataxia.

References:


Question 356: Clinical Adult - Other Pain Syndromes

Discussion:

This patient's symptoms are most consistent with trigeminal neuralgia. Since her symptoms have been unresponsive to carbamazepine and baclofen, consideration of microvascular decompression is appropriate. Microvascular decompression is effective in many cases of trigeminal neuralgia, supporting the hypothesis that the disorder may be due to compression of the nerve by a vascular loop.

References:


Question 365: Clinical Adult - Dementia

Discussion:

The history, time course of the progressive dementia, and hallucinations are consistent with Lewy body dementia. Dementia of the Alzheimer type typically does not cause vivid hallucinations. Creutzfeld-Jakob disease is typically more rapid in its tempo of decline. A pseudodementia due to depression should not present in the fashion outlined. Although the rigidity of limbs and slow gait may be suggestive of Parkinson disease, the combination of these findings with vivid visual hallucinations, fluctuating cognition and attention, and early dementia, is more consistent with Lewy body dementia.

References:


Question 373: 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 deficit. Migraine headaches are a common clinical feature as well.

References:


Question 374: Clinical Adult - Neurogenetics

Discussion:
Axillary and inguinal freckling, café au lait spots, and cutaneous neurofibromas are characteristic skin findings in patients with neurofibromatosis type 1.

References:

Question 380: Clinical Adult - Demyelinating Disease

Discussion:
None of the disease modifying therapies for MS are considered safe during pregnancy. There is a higher risk of miscarriage in patients on interferons. It is recommended that the drugs be discontinued prior to conception. Usually MS activity is reduced during pregnancy.

References:

Question 384: Clinical Adult - Movement Disorders

Discussion:
Orthostatic tremor or "shaky leg" syndrome is a disorder of middle-aged or elderly people and is characterized by feelings of unsteadiness in the legs and a fear of falling when standing. Other associated symptoms include difficulty in initiating walking and leg discomfort when standing. The symptoms are attenuated by walking, abolished by sitting, and due to a high frequency tremor in the weight-bearing muscles.

References:

Question 392: Clinical Adult - Infectious Disease

Discussion:
Cytomegalovirus infection has been associated with a lumbosacral radiculitis in patients with HIV infection.

References:

Question 396: Clinical Adult - Dementia

Discussion:
This patient most likely has hemorrhage or ischemia of a hyperplastic pituitary gland, a not infrequent occurrence during pregnancy and immediately after. Sagittal sinus thrombosis and pseudotumor cerebri would be unlikely to produce a third nerve palsy. She is not at increased risk of aneurysmal rupture and eclampsia would not occur post-partum.

References:
Question 406: Clinical Adult - Infectious Disease

Discussion:

Oral doxycycline has been found on evidence-based review through the AAN Practice Parameters to be a safe and effective treatment for peripheral nervous system Lyme disease and for CNS Lyme disease without parenchymal involvement.

References:


Question 410: Clinical Adult - Cerebrovascular Disease

Discussion:

Postpartum women are at increased risk for sagittal sinus thrombosis due to their hypercoagulable state. Headache, delirium, and focal seizures are frequent early symptoms. CT may also show hemorrhagic infarction and edema in a non-arterial distribution.

References:


Question 412: Clinical Adult - Cerebrovascular Disease

Discussion:

A hemorrhagic lesion of the pituitary involving the chiasm would explain the sudden visual field deficit, somnolence, and hypotension. Elevated intracranial pressure or bilateral cavernous sinus lesions could explain the bilateral sixth nerve palsies with pituitary apoplexy. The other diagnoses listed are not associated with a bitemporal visual field deficit and the clinical description presented here.

References:


Question 414: Clinical Adult - Demyelinating Disease

Discussion:

The patient described has a clinical isolated syndrome of optic neuritis with an abnormal MRI scan of the brain. According to the 10-year follow up data of the Optic Neuritis Treatment Trial (2003), subjects with optic neuritis (of otherwise indeterminate cause) and abnormal brain MRIs at presentation had a 56% chance of being diagnosed with MS in 10 years. Those with a normal brain MRI had a 22% chance. Although the criteria for diagnosing MS have undergone several revisions, by current McDonald Criteria, this patient would not have met the requirement for dissemination in time. Although poor recovery of vision has an association with Neuromyelitis Optica (NMO), there is nothing else to suggest that diagnosis in this patient.

References:

Question 423: Clinical Adult - Epilepsy

Discussion:

This patient's history, including the description of the witnessed event with pelvic thrusting in the emergency department, and her unresponsiveness to multiple medication trials, is suggestive of psychogenic seizures. Although she reports that previous EEGs have shown abnormalities, this does not in itself imply the diagnosis of epilepsy. The diagnostic procedure that is most likely to be beneficial in differentiating between epilepsy and psychogenic seizures is capturing an episode during video-EEG monitoring, which should be performed on this patient.

References:


Question 424: 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 425: Clinical Adult - Cerebrovascular Disease

Discussion:

Cerebral vasospasm is a serious complication of subarachnoid hemorrhage typically occurring between the 4th and 14th day after hemorrhage. It often presents with focal neurologic deficits and drowsiness.

References:


Question 426: Clinical Adult - Cerebrovascular Disease

Discussion:

This patient has color agnosia and prosopagnosia most likely due to bilateral ischemia in the mesial inferior occipitotemporal cortex.

References:


Question 445: 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.

References:

Clinical Pediatrics

Question 6: Clinical Pediatrics - Learning/Language Disorders

Discussion:
Most children with Angelman syndrome have little or no spoken language. Sign language may amplify communication. The other conditions tend to be associated with milder disorders or peculiarities of language.

References:

Question 18: 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 51: Clinical Pediatrics - Developmental Disorders

Discussion:
Although normal at birth, the head circumference of infants with Tay-Sachs disease characteristically increases during the second year of life, while all of the other conditions are associated with progressive microcephaly.

References:

Question 55: Clinical Pediatrics - Vascular Disorders

Discussion:
Acute arterial ischemic stroke in the newborn quite characteristically manifests transient focal seizures without associated hemiparesis. The appearance of hemiparesis is often delayed for a number of weeks. Encephalopathy is characteristic of other listed conditions and a greater likelihood of variously distributed weakness and more persistent seizures.

References:


Question 57: Clinical Pediatrics - Behavioral/Psychiatric

Discussion:

The diagnostic and statistical manual of mental disorder's (DSM IV [4th edition]), criteria for the diagnosis of Asperger syndrome include: A. Qualitative impairment in social interaction as manifested by at least two of the following: (1) marked impairment in the use of multiple nonverbal behaviors, such as eye-to-eye gaze, facial expression, body postures, and gestures to regulate social interaction; (2) failure to develop peer relationships appropriate to developmental level; (3) a lack of spontaneous seeking to share enjoyment, interests, or achievements with other people; (4) lack of social or emotional reciprocity. B. Restricted repetitive and stereotyped patterns of behavior, interests, and activities, as manifested by at least one of the following: (1) encompassing preoccupation with one or more stereotyped and restricted patterns of interest that is abnormal either in intensity or focus; (2) apparently inflexible adherence to specific, nonfunctional routines or rituals; (3) stereotyped and repetitive motor mannerisms (eg, hand or finger flapping or twisting or complex whole body movements); (4) persistent preoccupation with parts of objects. C. The disturbance causes clinically significant impairment in social, occupational, or other important areas of functioning. D. There is no clinically significant general delay in language (ie, single words used by age 2 years, communicative phrases used by age 3 years). E. There is no clinical delay in cognitive development or in the development of age-appropriate self-help skills, adaptive behavior (other than in social interaction), and curiosity about the environment in childhood. F. Criteria are not met for another specific pervasive developmental disorder or schizophrenia. The criteria do not include delay in cognitive development, language acquisition, or in development of self-help adaptive skills. Stereotyped and repetitive use of language is not a criterion for Asperger syndrome. These are all differences that distinguish Asperger syndrome from autistic disorder.

References:


Question 73: Clinical Pediatrics - Learning/Language Disorders

Discussion:

Semantic pragmatic syndrome is a condition where language elements are accurately repeated within a vapid context with impaired prosody. It is a common finding in high-functioning autism.

References:


Question 75: Clinical Pediatrics - Spine

Discussion:

A pediatric neurologist is frequently asked to evaluate a child that refuses to walk. The causes can be benign behavioral problems, acute neurologic problems, but also non-neurologic abnormalities simulating a neurologic deficit but requiring immediate therapeutic intervention. Childhood diskitis may occur in the thoracic, lumbar, or sacral spine and can affect children of all ages, but it is most common in the lumbar region in children younger than 5 years. Physical examination, laboratory tests, and radiological studies all aid in the diagnosis of this clinical syndrome, and proper use can prevent unnecessary invasive intervention. Presentation varies with age; the child may refuse to bear weight on the lower extremities or may present with back
pain, abdominal pain, a limp, or, if an infant or toddler, with irritability. The etiology appears to be a bacterial infection, usually caused by *Staphylococcus aureus*. Most children improve rapidly with a 4- to 6-week course of antibiotics. Although not routinely necessary, immobilization decreases symptoms and, in the case of osseous destruction, prevents progression of spinal deformity. Biopsy of the infected disk space is reserved for children refractory to intravenous antibiotics. Follow-up should include plain radiographs at regular intervals for 12 to 18 months to ensure resolution of the destructive process.

References:


Question 96: Clinical Pediatrics - Critical Care/Trauma

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 108: Clinical Pediatrics - Disturbances of Consciousness

Discussion:

Although any of these conditions may result in neurologic deterioration, epilepsy is not a prominent element of any but glucose transporter deficiency (De Vivo disease). Persistent and worsening seizures and encephalopathy are hallmarks of De Vivo disease due to energy failure, which can effectively be treated with the ketogenic diet.

References:


Question 111: Clinical Pediatrics - Motor Neuron/Neuromuscular

Discussion:

Loss of ambulation in Duchenne usually occurs in the second decade of life. Appropriate treatment with daily prednisone has been shown to prolong anticipated ambulation by approximately 2-3 years. This benefit is lost if patients gain too much weight on this therapy.

References:


Question 117: Clinical Pediatrics - Infectious Disease

Discussion:
Seizures occur in 20% to 50% of children with meningitis and although they may be associated with vasculitis, infarction, or extra-axial fluid collections, they are not strongly indicative of any these, perhaps resulting more often from cytokine release or fever. Chronic seizures are unusual after recovery from meningitis.

References:

**Question 122: Clinical Pediatrics - Movement Disorders**

Discussion:
GTP cyclohydrolase deficiency may produce abnormalities of tone and movement that result in a parkinsonian syndrome. An additional diagnostic clue is the occurrence of repeated bouts of otherwise unexplained hyperthermia.

References:

**Question 123: Clinical Pediatrics - Neonatal**

Discussion:
Smoking during pregnancy and after delivery represent independent risk factors for SIDS death, together possibly accounting for as many as 30% of SIDS deaths.

References:

**Question 127: Clinical Pediatrics - Heritable Metabolic or Degenerative**

Discussion:
The findings are most suggestive of pantothenate kinase-associated neurologic disorder, associated with mutations in PANK 2. The disorder formerly called Hallervorden-Spatz Syndrome is now referred to as PANK 2-associated disorder due to Hallervorden's association with Nazi experimentation. Frataxin is associated with Friedreich ataxia, superoxide dismutase (SOD) with familial ALS, pyruvate dehydrogenase (PDH) with a syndrome of lactic acidosis, seizures and neurologic deterioration, and ATM with ataxia telangiectasia.

References:

**Question 128: Clinical Pediatrics - Neonatal**

Discussion:
Neonatal herpes simplex virus (HSV) encephalitis is associated with acute multisystem invasive herpetic disease in approximately two-thirds of all cases, while most adult cases occur as reactivation of latent herpetic infection with manifestations limited to the nervous system. Polymerase chain reactions are positive in 90% to 95% of cases, and are considered standard of
care. Acyclovir is well tolerated by neonates; however, treatment results in neonates have proved disappointing as compared to older children, with most neonatal cases showing poor outcome.

References:


Question 136: 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 142: Clinical Pediatrics - Developmental Disorders

Discussion:

Most children, and many adults, engage transiently in repetitive, purposeless movements. They have been variously described as motor rhythmias, rhythmic habit patterns, rhythmic stereotypies, habit spasms, mannerisms, and automatisms. The movements described above are common in normal children, except for hand-wrangling and knitting stereotypies, which are frequently seen in Rett syndrome.

References:


Question 150: Clinical Pediatrics - Infectious Disease

Discussion:

Gait ataxia is a universal manifestation of acute cerebellar ataxia (ACA) and the most severe finding. Finger-nose-finger dysmetria is found in approximately two-thirds of patients with ACA and is usually quite mild. Mental status is characteristically normal, meningismus is quite rare, and seizures do not occur.

References:


Question 152: Clinical Pediatrics - Neurosurgery

Discussion:

Toxoplasmosis may manifest after delivery with liver abnormalities and obstructive hydrocephalus. Physical findings of hydrocephalus include prominent veins and "sundown" eyes. Cytomegalovirus and rubella do not typically cause hydrocephalus.
Question 164: Clinical Pediatrics - Epilepsy

Discussion:

Typical childhood absence seizures do not have auras or post-ictal 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 drug of choice is ethosuximide or valproate.

References:


Question 170: Clinical Pediatrics - Motor Neuron/Neuromuscular

Discussion:

Involvement of muscle is a characteristic feature of mitochondriopathies such as cytochrome oxidase type 1 deficiency, as is neuropathy.

References:


Question 175: Clinical Pediatrics - Inflammatory Disease

Discussion:

The combination of findings represents classic multiple sclerosis. Posterior column findings are particularly important diagnostically. Although acute disseminated encephalomyelitis may occur in adolescents and adults, encephalopathy is characteristically present. Neuromyelitis optica spares the brain and spinal cord changes are typically more extensive. Combined systems degeneration spares the eyes and does not produce the imaging changes noted. Adrenoleukodystrophy with adolescent onset is usually myelitic, but is almost always a disease of young men and does not manifest the additional findings or imaging changes noted here.

References:


Question 180: Clinical Pediatrics - Vascular Disorders

Discussion:

The distribution of lesions is in the posterior vascular distribution, which should raise concerns for a vasculitis. The mildly elevated WBC and protein are typical for primary CNS vasculitis, which has about 90% mortality if not detected and treated. ADEM does not follow vascular distribution and is usually more disseminated. TB and Fungal infections usually have higher protein and lower glucose. MELAS usually has a more stuttering onset with calcification of the basal ganglia with normal cell count and protein.
Question 203: Clinical Pediatrics - Behavioral/Psychiatric

Discussion:

Fragile X syndrome is the most commonly detected genetic etiology for autistic spectrum disorder, and often has milder presentations. MECP2 abnormalities, inborn errors, epilepsy syndromes and cortical migrational defects can have autistic symptomology, but routine screening is not advised unless other symptoms point to these disorders.

References:


Questions 205 - 209: Clinical Pediatrics - Headache

Discussion:

Childhood periodic syndromes are believed to be migraine variants, although often do not involve headache at the time of the major symptoms. When the history is clear, further workup may not be needed. Periodic syndromes include cyclic vomiting, abdominal migraine, benign paroxysmal vertigo, alternating hemiplegia of childhood and paroxysmal torticollis. Basilar-type migraines feature predominant visual and cerebellar symptoms often with altered consciousness.

References:


Question 258: Clinical Pediatrics - Heritable Metabolic or Degenerative

Discussion:

Tuberous sclerosis (TS) is characterized by skin lesions including depigmented macules (so called "ash leaf spots"), shagreen patches, periungual fibromas, and facial lesions termed adenoma sebaceum. Cardiac rhabdomyomas are seen prenatally and at birth but generally regress. About 50% of patients with TS are mentally retarded, but 50% are normal intellectually. Although the gene is transmitted as an autosomal dominant, over 25% represent new mutations. MRI should be done at diagnosis then periodically. Lesions at the foramen of Monro may progress but generally do so very slowly. Infantile spasms are quite common as a presenting feature of TS, and TS is said to be the most common single identifiable cause of symptomatic infantile spasms.

References:


Question 335: Clinical Pediatrics - Movement Disorders

Discussion:
Opsoclonus-myoclonus syndrome features chaotic random eye movements with myoclonic jerks and is an immune-mediated disorder. A significant number will also present with ataxia and truncal hypotonia. Most will have a neuroblastoma but some patients will have no identifiable etiology.

References:

Question 336: Clinical Pediatrics - Developmental Disorders
Discussion:
Unilateral closed-lip schizencephaly carries nearly a 70% risk for epilepsy.

References:

Question 351: Clinical Pediatrics - Neonatal
Discussion:
Phenobarbital may result in vitamin K deficiency that may be difficult to treat with parenteral vitamin K postnatally. This elevates the risk of the neonate for intraparenchymal brain hemorrhage.

References:

Question 352: Clinical Pediatrics - Epilepsy
Discussion:
Benign myoclonic epilepsy of infancy (BMEI) is a generally benign generalized myoclonic seizure disorder occurring in otherwise healthy infants, some of whom (about 25%) have a history of isolated febrile convulsions. They do not occur in clusters, and are not associated with tonic-clonic seizures

References:

Question 368: Clinical Pediatrics - Headache
Discussion:
Chronic daily headache due to overuse of analgesics (medication overuse headache) should be treated by withdrawal of all minor analgesics other than naproxen. Brief courses of dihydroergotamine or steroids may be helpful. Institution of prophylactic medication may improve overall headache control.

References:
Question 371: Clinical Pediatrics - Disturbances of Consciousness

Discussion:

Night terrors (pavor nocturnus) in children are an arousal during slow wave sleep and characteristically occur during the first half of the night, 30 minutes after onset of sleep. The child often cries out and is agitated and uncommunicative. Treatment is not necessary, but often the episodes are confused with nocturnal complex partial seizures.

References:


Question 378: 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 379: Clinical Pediatrics - Disturbances of Consciousness

Discussion:

Narcolepsy consists of a diagnostic tetrad that includes excessive daytime sleepiness, cataplexy, hypnagogic or hypnopompic hallucinations, and sleep paralysis. The most common presenting symptom in children is excessive daytime sleepiness, which in children and young adults may precede the development of cataplexy by several years. Diagnosis of narcolepsy is usually made when a positive multiple sleep latency test (MSLT) is found in individuals who experience both excessive daytime weakness and cataplexy.

References:


Question 386: Clinical Pediatrics - Motor Neuron/Neuromuscular

Discussion:

The most common presentation of nemaline myopathy is congenital hypotonia. Affected children are usually quite bright but exhibit a characteristic open-mouth appearance. Extra-ocular muscles are not affected and muscle weakness is static throughout life.
Question 393: Clinical Pediatrics - Tumors

Discussion:

These findings are highly suggestive of a brainstem tumor. Most but not all brainstem tumors in children present with progressive brainstem signs without headache or encephalopathy. However, the brainstem signs in this case are not consistent with the other age-appropriate differential considerations listed here.

References:


Question 419: Clinical Pediatrics - Infectious Disease

Discussion:

Bartonella is generally spread by cats, particularly feral kittens that live outdoors or in barns. Status epilepticus is a rare complication and can be difficult to treat. Occasionally demyelinating lesions are caused by Bartonella. Neuroborreliosis and babesiosis do not usually present with convulsive status epilepticus. Tuberculous meningitis presents with elevated CSF protein. Malaria is not endemic in the United States.

References:


Question 422: Clinical Pediatrics - Epilepsy

Discussion:

Levetiracetam, the antiseizure mechanism of which is uncertain, does not pose the risk the other choices have for worsening acidemia.

References:


Question 427: Clinical Pediatrics - Disturbances of Consciousness

Discussion:

Same-day return to play after a first concussion is recommended only if symptoms persist for more than 15 minutes. If 30 minutes, play should not be resumed for 1 week. Symptom duration tends to be longer in younger rather than older athletes. Symptom duration also tends to be longer with multiple concussions. Headache, nausea, and vomiting are more common after concussion in children than in adults and are not sensitive indicators of intracranial pathology. The multiply-concussed athlete who experiences an additional loss of consciousness should not return to play for more than 1 month regardless of the duration of that loss of consciousness.

References:


**Question 437: Clinical Pediatrics - Motor Neuron/Neuromuscular**

**Discussion:**

Myotonic dystrophy inherited from the mother can produce severe weakness in a newborn, and can often be reliably diagnosed by examining the mother. Because some neonates are so weak, EMG can be unreliable. Muscle biopsy findings are non-specific.

**References:**


**Question 444: Clinical Pediatrics - Epilepsy**

**Discussion:**

Familial nocturnal frontal lobe epilepsy is an autosomal dominant condition usually caused by a mutation in the nicotinic receptor. It is often mistaken for a parasomnia or pseudoseizures because of partial awareness. Fear can be prominent and some patients are afraid to fall asleep. Panayiotopoulos syndrome is another primarily nocturnal seizure disorder with prominent autonomic features but without the hyperkinetic motor symptoms.

**References:**


**Contemporary Issues**

**Question 7: Contemporary Issues - HIPAA**

**Discussion:**

Per recent privacy rules, appropriate release should be obtained from the parents or custodian of a minor before divulging clinical information. The Health Insurance Portability and Accountability Act (HIPAA) was signed into law August 21, 1996. The medical records of a minor may not be released or discussed with other family members without the written consent of the child's legal guardian, most often the parent, but not always (USDHHS, 2008).

**References:**


**Question 77: Contemporary Issues - Patient Safety**

**Discussion:**
Patients with neurologic or general conditions associated with an increased risk of falling should be asked about recent falls and further examined for the presence of specific neurologic deficits that predict falls, which include gait and balance disorders; deficits of lower extremity strength, sensation, and coordination; and cognitive impairments. Each year, unintentional falls in the United States account for more than 16,000 deaths, of which three quarters occur among persons over 64 years of age. As a result of falls, seniors experience nonfatal injuries resulting in nearly 500,000 hospitalizations and more than 1.8 million emergency department visits annually. In addition to advanced age, several specific risk factors have been identified, including muscle weakness, deficits in gait or balance, visual deficits, arthritis, impairments in activities of daily living, depression, and cognitive impairment.

References:


Question 97: Contemporary Issues - Ethics

Discussion:

It is not appropriate to order genetic testing on either a minor child or an adult in the absence of symptoms. Performing genetic testing on an asymptomatic individual, regardless of legal status of minor, mature minor, or adult age, represents predictive testing. Predictive testing should be preceded by pretest counseling that includes a discussion of the genetic aspects of Huntington disease (HD), the potential for nondefinitive results, as well as the potential social and psychological risks and benefits of the test. Also, patients should provide informed consent prior to testing. Most physicians and genetic counselors believe that it is inappropriate to test children under 18 who are not legally able to provide informed consent. It is not appropriate to test the boy, who cannot provide informed consent for many years.

References:


Question 106: Contemporary Issues - End-of-Life/Palliative Care

Discussion:

When medication to help alleviate pain, suffering, and anxiety is administered as part of palliative care to a patient with a terminal disease, who does not wish to be placed on respiratory support, and the patient dies during the process of receiving the medication this is referred to as double effect. The type of medication, the dose, and the intent to relieve suffering, rather than cause death, distinguishes this process from active euthanasia.

References:

McQuillen MP. Ethical issues in neurology. Continuum: lifelong learning in neurol 2003;9(4).

Question 151: Contemporary Issues - Driving

Discussion:
In a survey of over 213 respondents with epilepsy, 19% indicated that in order to drive, they were not completely honest about their seizure frequency. Indeed, 26% reported having had a car accident because of a seizure. It is important to recognize that the loss of a drivers license can profoundly impact an individuals life. In an effort to explore ways to encourage patient compliance, voluntarily exploring other options for driving is imperative if a physician suspects poor compliance. Neurologists are not obligated to serve as policing agents if a patient is driving and is not well-controlled on medication; however, if an individual knowingly drives and injures another, that individual may be liable for both civil and even criminal damages. Adding a second anti-convulsant will not increase compliance for the first drug. Transferring her care to another neurologist is not recommended as a first step, particularly since the treating physician may have better success in at least initially exploring why she is not compliant. Finally, while it may be germane to report a negligent parent for driving her minor children while she is inadequately controlled for her seizures because of her own poor compliance, it is not the ideal first step.

References:


Question 176: Contemporary Issues - Ethics

Discussion:

The standard of best interest states that the medical team is obligated to act on behalf of the child, in whose case, removal of life support is in the childs best interest. However, when conflict exists and cannot be reasonably resolved over the course of several days to weeks, it may be necessary to involve the ethics team. The use of the ethics team can be very helpful in determining how to best resolve issues; however, the patient dialogue between medical team and family is considered the ideal setting to navigate the many difficult decisions that removing life support from a child requires. The hope of this question is to demonstrate that on occasion the medical team has the ethical obligation to act in the best interest of the child when the family may be too distraught to think through their grief. Ideally, the team is patient, calm, and comforting to the family, but also repeatedly clear in their understanding about the lack of meaningful recovery, and the need to act in the child's best interest, rather than the family's interest which may be based in unrealistic expectations.

References:

Tran JM, McCartney JJ. In the best interest of the patient: applying this standard to healthcare decision-making must be done in a community context. Health Prog 1993;74(3):50-56.


Question 179: Contemporary Issues - Patient Safety

Discussion:

The AAN endorses adequate treatment of seizures during pregnancy and recommends discontinuation of AEDs only in situations where it is unlikely that seizures will recur. JME typically requires lifelong therapy. During pregnancy 50% of patients have no change in seizure frequency, 15% to 32% deteriorate, and 25% improve. If seizures are well controlled prior to pregnancy, it is highly likely that they will remain so. There is a 1% to 2% risk of neural tube defects with valproic acid (VPA) and an elevated risk of major congenital malformations.

References:


Question 183: Contemporary Issues - Business

Discussion:

The best CPT code is 99244. A consult is billed in the 99241 through 99245 series. The 99244 is used for a moderate complex problem, or if using the time element, 60 minutes; the 99245 is used when a consult lasts 80 or more minutes, and is complex; finally, the 99255 code is used for an initial inpatient consultation of high complexity, and/or 110 minutes in time. The 99201 through 99205 is used for a new patient evaluation but not for a consult or referral from another medical care provider.

References:


Question 187: Contemporary Issues - End-of-Life/Palliative Care

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 190: Contemporary Issues - Patient Safety

Discussion:

Appropriate health care requires optimum communication. In order for the physician to determine the medical history and accurately obtain a physical examination, interpreters should be used where applicable. The Joint Commission views the issue of the provision of culturally and linguistically appropriate health care services as an important quality and safety issue and a key element in individual-centered care. It is well recognized that the individual's involvement in care decisions is not only an identified right, but is a necessary source for accurate assessment and treatment information.

References:


Question 202: Contemporary Issues - HIPAA

Discussion:
The Health Insurance Portability and Accountability Act (HIPAA) of 1996 was created to establish national standards for health care providers, plans and clearing houses for storage and transmission of personal information as it relates to provision of and payment for health care services. Within the legislation are standards, implementation timelines and penalties for violation. Providers are required to have national identifiers. HIPAA legislation does not have anything to do specifically with clinical research, but mandates secure protection of identifying information in databases. Pre-existing condition clauses are still used by many insurance companies.

References:


Question 339: Contemporary Issues - Core Competencies

Discussion:

A medical expert should strive to provide testimony that is accurate, impartial, and relevant with respect to disputed medical facts or issues. The expert should carefully and thoroughly review relevant medical and scientific data before offering an opinion. Where standard of care is at issue, the expert should become familiar with the relevant standards of practice at the time and practice setting of the occurrence and be prepared to provide detailed, informed, and accurate testimony as to whether a defendant has violated these standards.

Where medical causation is at issue, the expert witness should be prepared to testify as to mechanisms of injury and whether the opinion is based on personal clinical experience, published information, or prevailing expert opinion. It is unethical for a medical expert to tie the level of compensation in a particular case to the outcome of that case.

References:


Question 362: Contemporary Issues - Driving

Discussion:

Deficits in executive function and visuospatial tasks predict performance on driving measures in patients diagnosed with dementia. The majority of patients with mild dementia and all patients found to have impaired executive function should have driving safety assessed. Neurologists should recommend that these patients not drive and report as required by state legislature.

References:


Question 405: Contemporary Issues - Core Competencies

Discussion:

There are very rare and selected situations involving emergency, potentially life-saving treatments in which institutional review board (IRB) approval for an experimental treatment may be obtained. If the treatment involves minimal risk and delay in initiating treatment negates the potential benefit of treatment, such IRB approval is more likely to be obtained. In almost all circumstances, an informed consent process is required, despite the beliefs of the clinician and family that the experimental treatment should be administered.

References:
Question 408: Contemporary Issues - Business

Discussion:

The new Centers for Medicare and Medicaid Services (CMS) criteria for reimbursement of brain FDG-PET are for the evaluation of frontotemporal dementia versus Alzheimer disease in a patient with a dementia syndrome of at least 6 month's duration. The Centers for Medicare and Medicaid Services (CMS) has announced it will expand coverage of positron emission tomography (PET) to improve the care of Medicare beneficiaries with thyroid cancer and those with potential cardiac diseases. The decision expands Medicare coverage of PET scans first made in December 2000. It is also covered for myocardial viability and pre-surgery evaluation of refractory seizures. CMS also announced today that it is designing a demonstration to evaluate the potential role of PET for patients with suspected dementia, as well as a multi-disciplinary expert meeting to fully explore the value of PET for Alzheimer disease.

References:


Neuroimaging

Question 95: Neuroimaging - Critical Care/Trauma

Discussion:

Extracellular methemoglobin is hyperintense on both T1- and T2-weighted images. This is usually the predominating signal in subacute primary intraparenchymal hematomas. Extracellular methemoglobin is also hyperintense on proton density and fluid attenuated inversion recovery images.

References:


Question 159: Neuroimaging - Stroke

Discussion:

The increased signal in the DWI echoplanar images is due to a restriction in the normal diffusion of protons in the ischemic tissue. It occurs during the first week and then tends to disappear. It is not true that the increased signal is caused by T1 shortening or that T2 effects play a larger role than diffusion. No "light bulb" sign heralds reperfusion.

References:


Question 224: Neuroimaging - Technical

Discussion:

The internal cerebral vein is labeled G

References:
Question 225: Neuroimaging - Critical Care/Trauma

Discussion:

The T1 coronal sequence demonstrates the close proximity of the encephalomalacia to the skull surface. FLAIR sequences show encephalomalacia near the cortical surface. T2-weighted view shows a displaced chronic skull fracture, further suggesting that the encephalomalacia is posttraumatic. The patient did have a history of remote head trauma but no history of brain surgery.

References:


Question 226: Neuroimaging - Stroke

Discussion:

There is a cystic structure along the lateral wall of the left lateral ventricle, with signal intensities similar to CSF. Acute infarction has a signal intensity closer to brain tissue. Location is poor for traumatic injury. Absent here are fusion of midline structures and agenesis of the corpus callosum, both associated with holoprosencephaly. Schizencephaly is a cleft which in some cases extends to the ventricular surface, but typically arises from the cortical surface extending inward. Therefore, the findings are best explained by a congenital infarction giving rise to an asymptomatic porencephalic cyst, probably unrelated to the recent episode of right-sided weakness.

References:


Question 227: Neuroimaging - Developmental/Neurogenetic Disorders

Discussion:

This patient has neurofibromatosis, with multiple neurofibromata in most of the roots, larger on the right side (A). Many of the intervertebral foramina are enlarged by the rounded tumors. She also has fibromata on the skin of the lumbosacral region. The other possible answers have different morphology on MRI.

References:


Question 228: Neuroimaging - Multiple Sclerosis

Discussion:

The image shows characteristic demyelinating lesions which are oriented perpendicularly to the ependymal lining (Dawson's fingers). Lyme disease, ischemic demyelination and vasculitis can also result in white matter hyperintensities on FLAIR images but generally do not follow this imaging pattern.

References:


Question 229: Neuroimaging - Technical
Discussion:

A normal spectrum is shown. The tallest peak is N-Acetyl-Aspartate (NAA) and represents neuronal activity (normal). To the left of the NAA peak are the (normal) creatinine and (normal, smaller) choline peaks.

References:


Question 230: Neuroimaging - Epilepsy

Discussion:

Cortical dysplasia can be seen at the posterior extent of the left sylvian fissure, as a thick cortical ribbon. Near the lesion is the angular branch of the middle cerebral artery, of a normal appearance. There is no evidence of an AVM. Cerebral infarction would cause cortical thinning, not thickening. The nodular appearance of an oligodendroglioma and the cystic appearance of porencephaly are not present in this image.

References:


Question 231: Neuroimaging - Brain Tumors

Discussion:

The image shows gadolinium enhancement of the leptomeninges around the cerebellum and in many cortical sulci, most prominently in the calcarine sulcus. Of the primary tumors listed, heavy meningeal infiltration of this kind is most frequent with breast carcinoma.

References:


Question 232: Neuroimaging - Spine

Discussion:

The axis (C2) is easy to identify on sagittal images. From it, one can count the vertebral bodies downwards. C4-C5 is the narrowest portion of the cervical canal in this image.

References:


Question 233: Neuroimaging - Infection

Discussion:
Toxoplasmosis is caused by an obligate intracellular parasite *Toxoplasma gondii*. Patients who develop AIDS are at particularly high risk of disseminated toxoplasmosis which most frequently involves the CNS. Toxoplasmosis is the most common focal mass lesion in AIDS and it most commonly involves the basal ganglia.

References:


**Question 237: Neuroimaging - Dementia**

**Discussion:**

The susceptibility-weighted images show two small hemorrhages in the subcortical region of the right frontal and occipital lobes. There are other punctate hemorrhages in these images, easily appreciable because of the heavy iron content of hemosiderine deposits. Note that they are much more difficult to see on conventional T2-weighted images. In this age group, such hemorrhages are most likely related to amyloid angiopathy. Cavernous angiomas are seen on conventional T2 images. Cysticercosis is not accompanied by heavy iron deposition and the shape is different. Hemorrhagic infarction from emboli may be accompanied by iron deposition, but it tends to affect the cortical ribbon, which is mostly spared in these images. Malignant melanoma may cause metastasis with a high bleeding tendency. However, this type of lesion is much less common than amyloid angiopathy in this age group.

References:

Chao CP, Kotsenas AL, Broderick DF. Cerebral amyloid angiopathy: CT and MR imaging findings. Radiographics 2006;26:1517-1531.


**Question 238: Neuroimaging - Critical Care/Trauma**

**Discussion:**

In this elderly woman with a history of falling, one must consider whether the fall led to the lesion shown or whether a cerebral insult produced the fall. The imaging characteristics are not those of subdural hematoma or lobar intracerebral hematoma. The history of atrial fibrillation and anticoagulant therapy suggest a possible embolic stroke mechanism. The wedge-shaped cortical infarction is characteristic of embolic infarction. Embolic infarctions are more likely to undergo hemorrhagic transformation, as in this case. Thus, an embolic infarction with secondary transformation is the likely cause of these imaging findings. The findings are not suggestive of subarachnoid hemorrhage, and no comment is made about whether this is a contrast-enhanced CT (which it is not). Therefore, tumor and subarachnoid hemorrhage are unlikely in this case.

References:


**Question 239: Neuroimaging - Brain Tumors**

**Discussion:**

The location of this lesion is clearly too cephalad to be a giant aneurysm, and it falls outside of the corpus callosum. There is no surrounding edema, so metastatic disease is unlikely. Lipomas do not enhance. The homogenous enhancement, isointensity on T1 images, and location are characteristic of a falcine meningioma.

References:
Question 240: Neuroimaging - Spine

Discussion:

The images show a swollen cord and the slender vessels of a dural spinal fistula, typically located on the dorsal aspect of the thoracic spinal cord. The vessels are better seen in the higher resolution CISS sequence. Images do not show a herniated disc. The presence of prominent vessels occurs with some ependymomas, but these tumors tend to have sharper margins, whereas here the area of T2 signal increase, which is transitioning gently with the normal cord, suggests more the presence of venous edema typical of dural fistulas. An astrocytoma of this size is unlikely to be as symptomatic and the vessels are atypical, as they are for multiple sclerosis. As indicated in the article by Jellema et al., spinal dural fistulas can present initially with a radicular syndrome, as it did in this patient.

References:


Question 241: Neuroimaging - Brain Tumors

Discussion:

The hyperdense appearance on CT, isointense, nonenhancing appearance on T1 postcontrast, and hyperintense appearance on FLAIR, along with location within the third ventricle adjacent to the foramina of Monro, are typical of colloid cyst.

References:


Question 243: Neuroimaging - Critical Care/Trauma

Discussion:

The middle cerebral artery and its branches are absent. The anterior cerebral artery and the posterior communicating artery and the posterior cerebral artery are present.

References:


Question 244: Neuroimaging - Stroke

Discussion:

The patient has an intraparenchymal frontal hemorrhage due to a ruptured anterior cerebral artery aneurysm which has been coiled. There is extensive subarachnoid hemorrhage, along with small focal infarcts likely due to vasospasm. While these are all dangerous situations, the most concerning is the complete effacement of the sulci supratentorially, as well as the basal cisterns. It will likely progress to herniation and death if not emergently treated.

References:
Question 245: Neuroimaging - Developmental/Neurogenetic Disorders

Discussion:

The most likely diagnosis is autosomal dominant spinocerebellar ataxia type 6 (proved by genetic testing). These patients have pure cerebellar atrophy, affecting predominantly the superior vermis. There is no pontine atrophy, a feature of both olivopontocerebellar atrophy and Machado Joseph disease. There is no downward displacement of the cerebellar tonsils, and the posterior fossa is not smaller than normal, ruling out Chiari type I malformation. Finally, the cerebellar atrophy results in a widened supracerebellar cistern but without any evidence of cerebellar compression, which would be present with a subarachnoid cyst.

References:


Question 247: Neuroimaging - Brain Tumors

Discussion:

The parasagittal, uniformly-enhancing, dural-based mass seen in the MR scans is most consistent with a meningioma. Regarding the other choices, a lipoma would be hyperintense on pre-contrast T1 scans; the sagittal sinus appears patent on the post-contrast coronal image; there is no caudate atrophy to suggest Huntington disease; and toxoplasmosis usually manifests as multiple "ring-enhancing" parenchymal lesions, not present here.

References:


Question 248: Neuroimaging - Infection

Discussion:

There are multiple cystic lesions containing a high intensity/density dot that corresponds to the scolex of tenia solium. The patient had cysticercosis. As often happens in this disorder, intraventricular cysts block the cerebrospinal fluid pathways and hydrocephalus develops. Note that the cyst in the anterior third ventricle also contains a scolex. Hydrocephalus ex vacuo refers to enlargement of cerebrospinal fluid spaces due to atrophy, which is not present in this case. Colloid cysts usually do not contain a central area of focal enhancement. Schistosomiasis usually presents with multiple intraparenchymal edematous lesions with hemorrhage.

References:


Question 254: Neuroimaging - Spine

Discussion:

There is an enhancing nodule within the thecal sac. Most likely considerations would be nerve sheath tumor (schwannoma or neurofibroma) or a drop metastasis. Epidural abscess, chordomas and disc fragments are not in the intradural space. A lipoma would have a much higher signal on unenhanced T1.
Question 256: Neuroimaging - Infection

Discussion:

This patient had a semantic aphasia, often present with lesions of the anterior portion of the left temporal lobe, particularly in older patients. The perisylvian structures, except the insula, are spared. Therefore it is unlikely that this patient had a classical aphasia. Transcortical motor aphasias occur predominantly with frontal lobe lesions, absent here.

References:


Question 259: Neuroimaging - Dementia

Discussion:

There is severe bilateral atrophy in the tip of the temporal lobes, most consistent with a neurodegenerative process such as frontotemporal dementia. Temporal gyri are atrophied, but the gyral crowns are preserved: in trauma they tend to be affected. The symptoms could be related to a prion disorder, but this is not given as an alternative. There is no hydrocephalus. The atrophy is atypical for mesial temporal sclerosis, in that it is very marked and does not predominantly affect the medial temporal region.

References:


Question 260: Neuroimaging - Stroke

Discussion:

The images show decreased cerebral blood flow and decreased cerebral blood volume, along with increased mean transit time in the left middle cerebral artery territory. A CT angiogram demonstrated a thrombus within the M1 segment of the left middle cerebral artery.

References:


Question 262: Neuroimaging - Multiple Sclerosis

Discussion:

Cerebral atrophy is the neuroimaging finding most closely associated with progression of disability in multiple sclerosis patients. Gadolinium-enhancing plaques are the most important factor in the initial diagnosis of multiple sclerosis. T1 hypointensities are correlated with atrophy but may be reversible. T2 lesion areas and the number of T2 lesions are less powerful predictors of disability progression than brain atrophy.

References:

Question 263: Neuroimaging - CSF Circulation Disorders

Discussion:

This patient had symptomatic hydrocephalus. There is no hippocampal atrophy, which would have suggested Alzheimer disease. The frontal lobe is not atrophied, militating against frontotemporal dementia. The sylvian fissures are dilated, but there is no evidence of infarction. Finally, the sulcal dilation present is not relatively homogenous, as in hydrocephalus ex vacuo, but on the contrary, the large sulci are markedly dilated and the small ones (see high parietal convexity) are compressed. CSF is pooling in the large sulci as well as in the ventricles (see reference).

References:


Question 264: Neuroimaging - Stroke

Discussion:

The hemorrhage arose from the right frontal lobe, secondarily rupturing into the lateral body of the lateral ventricle and extending to the temporal horn and, through the foramen of Monro, to the third ventricle. The absence of extraventricular blood in the temporal horn militates against a temporal origin of the hemorrhage. Bleeding from an aneurysm in the circle of Willis is very unlikely because the subarachnoid space at the base of the brain lacks clotted blood.

References:


Question 266: Neuroimaging - Dementia

Discussion:

On biopsy, this woman had a demyelinating disorder of the type known as tumefactive sclerosis, Marburg disease or Schilder disease. The lesions are large, with a cystic appearance on MRI and often have a ring of enhancement that is open: the open-ring sign. The ring of enhancement is open where the lesion touches the gray matter. Lesions circumscribed to the white matter may have a closed ring, as present in the lower left panel. Lesions caused by the other diagnoses listed tend to have more homogenous or closed-ring enhancement.

References:


Question 267: Neuroimaging - Spine

Discussion:

The mass lesion with isointense signal on T1- and bright T2-weighted signal posterior to the dens is most frequently due to pannus. The inflammatory pannus involving the dens is often seen in association with rheumatoid arthritis.

References:

Question 268: Neuroimaging - Spine

Discussion:

Ependymoma is the most likely diagnosis, confirmed here by surgery. The tumor arises from the spinal canal and molds the vertebral bodies. A chordoma would arise from the vertebral bodies, or more likely, from the sacrum, and compress the spinal canal. Although at the sacral level it may be difficult to appreciate the origin of the tumor, it clearly appears intramedullary in higher sections. Similar arguments could be made against the diagnosis of osteochondroma. The appearance of intramedullary coccidiomycosis is different and in general, infections tend to affect the intervertebral discs, relatively spared by this tumor.

References:


Question 269: Neuroimaging - Epilepsy

Discussion:

The images show clefts extending from the extra-axial space to the lateral ventricles bilaterally. These are most characteristic of open-lip schizencephaly. In porencephaly, the cyst is not lined by cortex, as it is here. In hydranencephaly there is little brain tissue around a central cyst. The appearance of encephalomalacia due to trauma is different, with areas of mixed signal intensity in frontal, temporal or occipital regions.

References:


Question 271: Neuroimaging - Critical Care/Trauma

Discussion:

Hypertensive encephalopathy, a cause of reversible posterior leukoencephalopathy syndrome (RPLS), is a rapidly evolving condition that develops in patients with malignant hypertension. If not promptly recognized and aggressively treated it leads to progressive neurological deterioration and death. Renal failure and eclampsia are contributory causes. Similar changes are also seen in toxicity due to immunosuppressive agents. MRI and CT show edema located in the occipital and parietal lobes and other posterior regions. These findings are seen in 85% of patients studied within 72 hours after onset of symptoms. The relative lack of sympathetic innervation in vertebrobasilar vasculature may predispose this region to the development of the cerebral edema.

References:


Question 272: Neuroimaging - Stroke

Discussion:

In the right mesial temporal region there is a region of hyperdensity on noncontrasted head CT, indicating either calcification or bleeding. This corresponds to hypointensity on gradient echo (T2*) sequences, indicating the presence of blood products. On T2-weighted views there is a "popcorn" appearance, most typical of cavernous malformations.

References:

**Question 273: Neuroimaging - Critical Care/Trauma**

**Discussion:**

The DWI sequence shows hyperintensity within the left frontal enhancing lesion. DWI hyperintensity is helpful in differentiating abscesses (bright) from tumors (dark). On the T1-postcontrast views there is a large ring-enhancing lesion in the left frontal lobe, along with enhancement of the meninges on the left. Moderate edema surrounds this abscess on the FLAIR sequences. These findings are most characteristic of an abscess.

**References:**


**Question 274: Neuroimaging - Spine**

**Discussion:**

The axial sequences demonstrate bilateral pars interarticularis defects, resulting in widening of the AP diameter of the spinal canal. The lateral T1 sagittal view demonstrates another view of the pars defect, and the T2 sagittal view shows grade 2 spondylolisthesis. The combination of bilateral pars defects and spondylolisthesis is also known as spondylolysis.

**References:**


**Question 275: Neuroimaging - Stroke**

**Discussion:**

The small dots in the axial section correspond to elongated structures on sagittal sections at the same level, noticeable on the T1 and FLAIR images. These structures are bright on T2, but dark on T1 and FLAIR, suggesting the presence of free water. They are not venules, which would be very noticeable in the susceptibility weighted image. Were they arterioles, the flow void would make them appear as dark dots, even as similar size arteries appear in the subarachnoid space. They are dilated Virchow-Robin or perivascular spaces, around perforating arterioles or venules. Lacunar infarctions are unlikely to be so small and sharply demarcated. They would be bright on FLAIR. Some of the areas that are bright on FLAIR, around the dilated perivascular spaces, could be areas of ischemia or of venous congestion. Hemosiderin deposits would appear dark on T2.

**References:**


**Question 279: Neuroimaging - Critical Care/Trauma**

**Discussion:**

There is a large right frontal parasagittal acute intraparenchymal hemorrhage, along with acute hemorrhage within the interhemispheric fissure and dependent portion of the lateral ventricle. Given the extensive hemorrhage in the fissure and the proximity to the anterior communicating artery (ACOM), an aneurysmal rupture is the most likely etiology. Formal angiography confirmed the presence of a 13mm ACOM aneurysm.

**References:**

**Question 280: Neuroimaging - CSF Circulation Disorders**

**Discussion:**

There is gadolinium enhancement of meningeal structures, suggesting hypotension, carcinomatosis or lymphoma. The orthostatic character of the headache and the absence of other findings favor intracranial hypotension, also favored by the imaging finding of descended tonsils present in the sagittal image.

**References:**


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**Question 281: Neuroimaging - Metabolic**

**Discussion:**

Hyperintensity is seen on DWI images in the bilateral medial thalami. This finding can be seen in deep venous thrombosis, but this is excluded with the normal MRV. The pattern is atypical for a basilar artery ischemic episode and a basilar artery thrombosis is excluded by the normal MRA. The finding most likely represents thiamine deficiency leading to Wernicke encephalopathy.

**References:**


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**Question 282: Neuroimaging - CSF Circulation Disorders**

**Discussion:**

The patient has markedly enlarged lateral and third ventricles on axial views, with a small (or absent) cerebral aqueduct on sagittal views. These findings are most consistent with aqueductal stenosis. In colpocephaly there is dilation of the atrium of the lateral ventricles, as in this patient, but not of the temporal horn, present here. In Dandy-Walker syndrome there is partial absence of the vermis of the corpus callosum, not present here. The Miller-Dieker syndrome includes lissencephaly, not present here. In this age group normal pressure hydrocephalus is not as frequent as aqueductal stenosis. In addition, a communicating hydrocephalus of this kind would be accompanied by some dilation of the fourth ventricle.

**References:**


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**Question 284: Neuroimaging - Brain Tumors**

**Discussion:**

The findings are consistent with glioblastoma. Glioblastomas have characteristic imaging features on postcontrast studies, which show intense, inhomogeneous, nodular, ringlike enhancement that encloses a central isointense necrotic core and delineates the gross tumor margin. Anaplastic astrocytoma, fibrillary astrocytoma, meningioma, and oligodendroglioma tumors typically do not show this bizarre ringlike enhancement.

**References:**

Question 285: Neuroimaging - Epilepsy

Discussion:

A complex formation of flow voids are seen in the left posterior temporal lobe. No significant surrounding edema is seen. These are most characteristic of arteriovenous malformations.

References:


Question 289: Neuroimaging - Brain Tumors

Discussion:

The lesion predominantly affects the white matter, consistent with a vasogenic edema pattern due to high-grade primary or metastatic neoplasm. There is no hyperdensity to suggest bleeding. The relative sparing of the overlying cerebral cortex would be unusual for an operculofrontal middle cerebral artery infarction. A contrast-enhanced MRI is indicated for further clarification.

References:


Question 291: Neuroimaging - Spine

Discussion:

At the L4-5 interspace there is spondylolysis without spondylolisthesis. The images show bilateral lysis of the pars interarticularis. The majority of these cases are the result of stress fractures.

References:


Question 294: Neuroimaging - Dementia

Discussion:

The images show marked atrophy of the frontal pole and of the anterior portion of the temporal lobe, particularly pronounced in the middle and inferior temporal gyri. The posterior portion of the superior temporal gyrus is spared. Some of the gyri are so atrophic that they seem razor thin. All these features are characteristic of lobar atrophy or Pick disease. The topographic distribution of atrophy differs from Alzheimer disease.

References:


Question 295: Neuroimaging - Movement Disorders

Discussion:
The sagittal T1-weighted image through the pons shows the pons and cerebellar vermis to be reduced in size. A glioma would not cause atrophy. Alcoholism would not cause atrophy of the pons. The hallmark of olivopontocerebellar atrophy is loss of the belly of pons, which is also clearly affected. Friedreich ataxia is usually primarily seen with spinal rather than pontine atrophy. Cerebellar, but not pontine atrophy, is present with spinocerebellar ataxia type 6.

References:


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Question 296: Neuroimaging - Critical Care/Trauma

Discussion:

The MRI shows increased signal bilaterally in the basal ganglia and the cerebellum which are most susceptible to anoxic injury. This is not MS as there is no evidence of demyelinating plaques. In superficial siderosis there will be paramagnetic T1 and T2 hypointensity secondary to iron deposition. Head injury would not produce this pattern of diffuse involvement. Multiple acute infarctions are also unlikely as shown above given the symmetrical changes.

References:


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Question 299: Neuroimaging - Infection

Discussion:

The patient has soft tissue swelling involving the right periorbital region, as well as an enhancing soft tissue mass within the right lateral orbit. These findings are consistent with cellulitis. Additionally, on postcontrasted images of the brain, there is enhancement of the meninges, consistent with meningeal spread of the infectious process.

References:


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Question 306: Neuroimaging - Brain Tumors

Discussion:

The images show a large, briskly-enhancing sellar mass extending into the suprasellar region and which causes compression of the optic chiasm. There is no apparent extension from the internal carotid artery to suggest aneurysm. Metastasis could appear similarly, but would be much less common than pituitary macroadenoma in this location. Epidermoid and Rathke cleft cysts do not typically enhance so briskly.

References:


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Question 307: Neuroimaging - Developmental/Neurogenetic Disorders

Discussion:

On the patients MRI there is thinning of the corpus callosum. This finding most often accompanies a familial spastic paraparesis, often caused by a mutation in the SPG11 gene on chromosome 15.
References:


Question 308: Neuroimaging - Stroke

Discussion:

On diffusion-weighted and gradient-echo (T2*) sequences there is a right parietal parasagittal intraparenchymal hemorrhage. On T1 pre-contrasted views there is hyperintensity in the superior sagittal sinus, corresponding to thrombosis of this structure. These findings taken together support the diagnosis of a parietal infarct caused by increased venous pressure. Venous infarctions are often hemorrhagic.

References:


Question 311: Neuroimaging - Spine

Discussion:

The patient presented here has multiple neurofibromas of spinal nerves, and a plexiform neurofibroma infiltrating the nerves forming the left brachial plexus. Although multiple spinal tumors can be seen in both NF-1 (pathology is neurofibroma) and in NF-2 (pathology is schwannoma), plexiform neurofibromas are not seen in NF-2. The remaining diagnoses are not consistent with the images. Metastatic disease may infiltrate through a neuroforaminen but is unlikely to enlarge it. Ependynomas are intramedullary tumors. Nerve sheath diverticulum are hypointense on T1-weighted studies and do not enhance.

References:


Question 312: Neuroimaging - Autoimmune Disorders (Non-MS)

Discussion:

Combining the history and the image, a Tolosa-Hunt syndrome is the most likely diagnosis. A meningioma "en plaque" would not have caused a similar syndrome 2 years previously in the contralateral eye leaving no trace. Myasthenia and the Miller-Fisher syndrome do not show on MRI the enhancing lesion in right cavernous sinus and meninges present in this case. A pseudotumor of the orbit sometime extends into the retro-orbital meninges, but in this case there is no mass effect in the intraconal fat, and the process clearly extends much beyond the orbit.

References:


Question 313: Neuroimaging - Brain Tumors

Discussion:
Epidermoid cysts, as the lesion shown, characteristically are hyperintense on DWI and T2 images. Arachnoid cysts are hypointense on DWI. Dermoids have increased T1 signal due to high fat content. Glioblastomas have mixed signal which enhances inhomogenously and epidermoids generally do not enhance. The lesion is extraparenchymal, therefore, not an infarct.

References:


Question 314: Neuroimaging - Critical Care/Trauma

Discussion:

The image shows a subacute subdural hematoma (SDH). SDHs conform to the shape of convexity and tend to have a "banana" shape. They freely cross cranial sutures but do not cross the midline insertion of the falx or tentorium. SDHs evolve on MRI at a slower rate compared to intraparenchymal hematomas (IPHs). This occurs because the subdural space lacks a blood-brain barrier and the adjacent dura matter remains well vascularized; thus, the oxygen concentration within an SDH remains higher than that expected for an IPH of the same age. This prolongs the evolution of the hemoglobin degradation process by days to weeks at each stage.

References:


Question 315: Neuroimaging - Spine

Discussion:

This patient had documented Enterococcus faecalis osteomyelitis affecting the L2 and L3 vertebrae. There is a ring-enhancing lesion, affecting both of these vertebrae, rather than simply the disc. Thus, there is more pathology than can be explained simply by discitis or expanding L3 neoplasm. There is no evidence of an anterior or posterior spinal approach to suggest surgery. The expanding quality and ring enhancement are not expected with hematoma and compression fracture.

References:


Question 316: Neuroimaging - Stroke

Discussion:

The angiogram shows the distal common carotid artery and its branches. The proximal internal carotid artery has a rounded stump with no distal flow that is consistent with occlusion. A dissection usually has a tapered stump, not a rounded stump. Vessel occlusion in moyamoya disease is usually intracranial.

References:


Question 317: Neuroimaging - Dementia

Discussion:
The images demonstrate marked atrophy of the striatum and relative sparing of the cortical grey matter. These are hallmark imaging features of both Huntington's chorea and neuroacanthocytosis and not seen in association with the other choices. Both neuroacanthocytosis and Huntington disease are distinct genetic disorders with similar clinical and neuroimaging manifestations.

References:


Pathology

Questions 220 - 223: Pathology - Toxic/Metabolic Disease

Discussion:

Bilateral putamenal necrosis is classic for methanol intoxication. Patients may manifest parkinsonism due to this selective basal ganglia damage, as well as blindness secondary to retinal ganglion cell degeneration.

Ingestion of ethylene glycol can either be purposeful as a result of a suicide attempt or accidental when containers that once were used to haul antifreeze are not properly cleaned before they are used for carrying water. Ethylene glycol ingestion may be fatal and at autopsy the brain shows generalized edema, petechial hemorrhages, and oxalate crystal deposition.

Inorganic arsenic causes a peripheral sensory motor neuropathy with axonal degeneration and demyelination. Muscle biopsy is devoid of vacuolar myopathy, unlike the findings in colchicine poisoning.

Urban or Native American youth are groups of individuals who are particularly prone to glue sniffing as a form of recreational drug use. Toluene exposure over time causes cognitive deficits and white matter abnormalities in cerebral white matter.

References:


Question 10: Pathology - Neuromuscular Disease

Discussion:

Trinucleotide repeats underlie myotonic dystrophy, Huntington disease, and fragile X mental retardation, among other conditions. A small number of trinucleotide repeats are present in normal individuals, increased numbers of repeats lead to these autosomal dominant disorders, and transgenerational increase in the number of trinucleotide repeats are associated with earlier onset of disease.

References:


Question 13: 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 25: Pathology - Neurodegenerative Disease

Discussion:

The clinical scenario suggests dementia with Lewy bodies (diffuse Lewy body dementia). 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 53: Pathology - Neuromuscular Disease

Discussion:

Zidovudine (AZT), used for treatment of HIV infection, is a thymidine analog that inhibits reverse transcriptase and mitochondrial DNA polymerase, leading to depletion of mitochondrial DNA. Myalgia, weakness and elevated CK levels occur after 6 to 11 months of exposure with doses of 800 to 1200 mg/day. The diagnosis requires muscle biopsy, which shows ragged red fibers, reflecting mitochondrial proliferation. Recovery occurs over several months after discontinuation of AZT.

References:


Question 130: Pathology - Cerebrovascular Disease

Discussion:

The most common lesion attributed to sickle cell anemia is cerebral infarction. This is often due to occlusion of large arteries. The second most common complication is intracerebral hemorrhage. Subarachnoid hemorrhage is less common and attributed to rupture of aneurysms. Sagittal sinus thrombosis is rare.

References:


Question 138: Pathology - Prion Disease

Discussion:

The most remarkable feature of variant Creutzfeldt-Jakob disease (CJD) is the massive amount of protease-resistant prion protein (PrP) in the form of innumerable mature amyloid plaques. Those particularly in the cerebral cortex are distinctive, being located
in the center of vacuoles and forming "florid plaques"; otherwise known as "daisy" plaques." The other choices may all be seen in prion-related spongiform encephalopathies but do not carry the equivalent specificity as do florid plaques.

References:


Question 144: 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.

References:


Question 177: Pathology - Developmental

Discussion:

Chiari type I malformation consists of elongated cerebellar tonsils and may be an asymptomatic incidental finding on MR imaging or a cause of late-onset hydrocephalus in adults. Approximately 50% of Chiari I patients have associated syringomyelia. Craniolacunia, meningomyelocele, s-shaped kinking of the cervicomедullary junction, and beaking of the midbrain tectum are features associated with the Chiari type II (Arnold-Chiari) malformation.

References:


Question 196: 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 197: Pathology - Epilepsy
**Discussion:**

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**References:**


**Questions 220 - 223: Pathology - Toxic/Metabolic Disease**

**Discussion:**

Bilateral putamenal necrosis is classic for methanol intoxication. Patients may manifest parkinsonism due to this selective basal ganglia damage, as well as blindness secondary to retinal ganglion cell degeneration.

Ingestion of ethylene glycol can either be purposeful as a result of a suicide attempt or accidental when containers that once were used to haul antifreeze are not properly cleaned before they are used for carrying water. Ethylene glycol ingestion may be fatal and at autopsy the brain shows generalized edema, petechial hemorrhages, and oxalate crystal deposition.

Inorganic arsenic causes a peripheral sensory motor neuropathy with axonal degeneration and demyelination. Muscle biopsy is devoid of vacuolar myopathy, unlike the findings in colchicine poisoning.

Urban or Native American youth are groups of individuals who are particularly prone to glue sniffing as a form of recreational drug use. Toluene exposure over time causes cognitive deficits and white matter abnormalities in cerebral white matter.

**References:**


**Question 234: Pathology - Critical Care/Trauma**

**Discussion:**

The photograph shows remote bilateral frontal and left temporal contusions with a slight orange tinge secondary to the presence of residual hemosiderin-laden macrophages. The olfactory nerves are disrupted, which undoubtedly would have led to anosmia.

**References:**


**Question 235: Pathology - Neurodegenerative Disease**

**Discussion:**
Considering the clinical setting, the most likely diagnosis is diffuse Lewy body dementia. The image shows a section of cerebral cortex that was immunostained for alpha-synuclein to demonstrate cortical Lewy bodies.

References:


Question 242: Pathology - Infectious Disease

Discussion:

In this picture, the collections of multiple bubbly cysts in the white matter, caudate and putamen are typical of parenchymal involvement by cryptococcal meningitis. Aspergillosis produces gray irregular necrotic masses. Tuberculoma and metastases tend to be discrete solid firm masses.

References:


Question 246: Pathology - Cerebrovascular Disease

Discussion:

The photographs show a cavernous malformation, not the hemangioblastoma of von Hippel-Lindau disease or the superficial angiomatosis of Sturge-Weber disease. CREST syndrome (calcinosis, Raynaud, esophageal motility disorders, sclerodactyly, telangiectasia) does not have CNS manifestations. Familial amyloidosis usually affects peripheral nerves; most cerebral amyloid angiopathy is sporadic and not familial with the exception of Dutch, Icelandic, and British variants.

Patients with autosomal dominant cerebral cavernous malformation syndrome may have mutations of the KRIT1 gene on chromosome 7q. Other familial cases have been linked to genes on chromosomes 7p and 3q.

References:


Question 249: Pathology - Critical Care/Trauma

Discussion:

The hematoxylin and eosin stained sections demonstrate numerous enlarged, round axonal swellings which stain positively with the silver stain. This is the histologic picture of axonal spheroids, the finding in diffuse axonal injury. Also known as "shear injury," patients with this condition are rendered immediately unconscious with trauma, and subsequently have a course of chronic "closed head injury." Axonal spheroids may be seen at the edge of a cerebral infarction but not in this clinical context and diffuse pattern. Multiple sclerosis may also eventually result in axonal injury with spheroids but with more extensive demyelination than is evident in the figure showing intact myelin.

References:


Question 250: Pathology - Cerebrovascular Disease
Discussion:

Acute cerebral infarcts are characterized by swelling, effacement of the gray-white junction, and cracking at the interface between the intact and infarcted brain tissue. The photo illustrates these findings as well as small cortical hemorrhages in this large fresh cerebral infarct in the middle cerebral artery territory.

References:


**Question 251: Pathology - Neuromuscular Disease**

Discussion:

This muscle biopsy demonstrates degenerating fibers undergoing phagocytosis, hypercontracted fibers, excessive fibrosis and variation in fiber size. The proliferation of connective tissue, especially, is most consistent with a dystrophic process, such as Duchenne muscular dystrophy. Kugelberg-Welander syndrome is a spinal motor neuron atrophy which gives a neurogenic appearance in muscle biopsies. Nemaline rod myopathy contains classic eosinophilic inclusions in the muscle fibers. Congenital muscular dystrophy would have an earlier age of onset, while Pompe disease is manifested by glycogen storage in the myofibers.

References:


**Question 253: Pathology - Infectious Disease**

Discussion:

The correct answer was HHV-6 encephalitis, a well known complication of bone marrow transplantation. Histologic features that were confined to the mesial temporal lobe are those of viral encephalitis. Images show perivascular mononuclear cell inflammatory infiltrates, coupled with microglial clusters. Toxic conditions would not show encephalitis-like features. Progressive multifocal leukoencephalopathy shows viral inclusions and demyelination, not an encephalitic-like picture and has no predilection for the mesial temporal lobe.

References:


**Question 255: Pathology - Tumors**

Discussion:

The tissue section from the resected intraventricular mass showed large cells with abundant eosinophilic cytoplasm, as well as many multiple nuclei. Although these features closely resemble those of a number of primary central nervous system tumors, including gemistocytic astrocytoma and ganglion cell tumor, the intraventricular location eliminates these entities, which are intraparenchymal tumors, and points to subependymal giant cell astrocytoma as the correct diagnosis. Of the remaining choices listed, the differential diagnosis of an intraventricular tumor would include subependymoma and choroid plexus papilloma; however, the tumor in the present case does not display the papillary architecture of choroid plexus papilloma or the multilobulation and abundant fibrillar matrix of subependymoma. Subependymal giant cell astrocytomas are commonly associated with tuberous sclerosis and may be the presenting clinical feature, as in the present case.
References:


Question 257: Pathology - Developmental

Discussion:

The image shows numerous small gyri over the surface of the brain. The correct diagnostic term is polymicrogyria.

References:


Question 261: Pathology - Demyelinating Disease

Discussion:

Intravascular lymphoma, also known as angiotropic lymphoma, has a predilection to affect brain and skin and a significant number of patients come to clinical attention due to neurological symptoms. The photomicrograph shows intravascular cytologically atypical cells. No granulomas are present, as would be expected in neurosarcoidosis and the blood vessel wall was undamaged.

References:


Question 270: Pathology - Neurodegenerative Disease

Discussion:

The picture shows marked caudate atrophy diagnostic of Huntington disease, which results from a trinucleotide repeat amplification mutation in the huntingtin gene residing on chromosome 4.

References:


Question 277: Pathology - Critical Care/Trauma

Discussion:

The graphic shows a circumscribed collection of purulent material (empyema) on the surface of the brain (i.e., subdural as opposed to epidural or subarachnoid). Subdural empyema may complicate head injury or sinus infections. Organizing subdural hematoma would usually be densely adherent to the overlying dura and not display this color, being tan or rusty in coloration.

References:

**Question 278: Pathology - Neurodegenerative Disease**

**Discussion:**

The features show small ubiquitinated inclusions in the cytoplasm of neurons of the dentate gyrus of the mesial temporal lobe. With the additional information that no tau immunoreactive inclusions were seen, Alzheimer disease, Pick disease, and FTDP-17 can be eliminated since all are tauopathies. FTDP-17 usually presents with more extrapyramidal symptoms and is an autosomal dominant condition. FTLD-U is a sporadic condition that accounts for approximately 50% of all cases of frontotemporal lobar degeneration. TDP43 immunoreactive neuronal inclusions are characteristic of FTLD-MND, which was the best answer. Motor signs may be subtle as in this case, being limited to fasiculations of the upper extremities.

**References:**


**Question 283: Pathology - Developmental**

**Discussion:**

Agyria (lissencephaly) is a developmental lesion of the cerebral cortex in which there is no formation of cortical gyri. Lissencephaly is regarded as a neuronal migration abnormality.

**References:**


**Question 286: Pathology - Neurodegenerative Disease**

**Discussion:**

Progressive supranuclear palsy presents with falls, postural instability, retrocolis, bradykinesia, pseudobulbar palsy and dementia. At autopsy, globoid neurofibrillary tangles are characteristically found in substantia nigra, brainstem tegmentum, putamen, and select cortical areas. These are immunoreactive for tau. Also seen are tau-positive astrocytic inclusions, so called tufted astrocytes. Tufted astrocytes are almost exclusively seen in progressive supranuclear palsy (PSP) and can be found in striatum, thalamus, subthalamic nucleus and precentral gyrus and are rare to absent in a similar disorder, corticobasal degeneration.

**References:**


**Question 287: Pathology - Neurodegenerative Disease**

**Discussion:**

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**References:**

Question 288: Pathology - Neurodegenerative Disease

Discussion:

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References:


Question 290: Pathology - Cerebrovascular Disease

Discussion:

The low (left) and higher (right) power images show a mass of closely juxtaposed, large caliber blood vessels. Most of these are "arterialized" veins with hyalinization and damage to their walls, since arteriovenous malformations fail to have capillaries interposed between the veins and arteries to dissipate the high arterial pressures.

References:


Question 292: Pathology - Cerebrovascular Disease

Discussion:

The process shown is cerebral amyloid angiopathy (CAA), which occurs most often sporadically. It is highly associated with clinical dementia (although it can occur in its absence) and Alzheimer-like neurodegenerative changes. CAA accounts for about 5% to 15% of nontraumatic brain hemorrhage in adults. Rare familial forms of the disease are well recognized. Patients may present with large lobar hemorrhages that are superficially located and distributed around the cerebral cortex. Basal ganglia and thalamic hemorrhages typically occur in patients with chronic hypertension, not CAA. The incidence of CAA increases with age.

References:


Question 293: Pathology - Demyelinating Disease

Discussion:

Acute demyelinating encephalomyelitis (ADEM) is the only choice from the list that would produce this distinctive pattern of perivenous demyelination. The disease often occurs as a postinfectious phenomenon.

References:


Question 298: Pathology - Hypothalamus/Pituitary
Discussion:

A 55-year-old male with an endocrinologically inactive sellar mass could potentially have a craniopharyngioma, meningioma, null cell pituitary adenoma, or Rathke cleft cyst; germinoma in this age group would be less likely. The images show a bland, benign epithelial tumor without whorls, calcification, or ghost cells; diagnosis is null cell pituitary adenoma.

References:


Question 301: Pathology - Tumors

Discussion:

The images show epithelial cords or strings of vacuolated "physaliphorous" cells within a mucoid matrix chordoma is the most common neural crest-derived tumor of the sacrum and almost always arises within bone. Chordoma is derived from notochordal remnants and occurs predominantly in the sacrum or the skull base near the clivus. Myxopapillary ependymomas arise in the filum terminale, and neurofibromas and schwannomas arise in the nerve roots. While these types of tumors can also attain sufficient size to erode the sacrum, it is uncommon. Teratomas can also erode the sacrum but do so less frequently than chordomas. Ewing sarcoma is a tumor type that is rare in this location.

References:


Question 303: Pathology - Infectious Disease

Discussion:

The photograph shows the larval form of the pork tapeworm *Taenia solium*, the causative agent in cysticercosis. (The larval form is called *Cysticercus*).

Neurocysticercosis is more likely to affect the ventricular system than amebiasis, coccidioidomycosis, tuberculosis or nocardiosis. However, the diagnosis is made firm by the finding of a large, complex infectious agent.

References:


Question 305: Pathology - Tumors

Discussion:

The photomicrographs show classic perivascular pseudorosettes and ependymal canal-like structures characteristic of ependymoma.

References:
Question 309: Pathology - Hypothalamus/Pituitary

Discussion:

The histologic figure shows a distinctive form of keratin composed of clusters of plump keratinocytes that is referred to as "wet" keratin. Among tumors of the central nervous system, this type of keratin is unique to the adamantinomatous craniopharyngioma. Wet keratin nodules frequently undergo dystrophic calcification. In contrast, the keratin seen in epidermoid and dermoid cysts consists of layers of very thin, flat, flaky anucleate squamous cells. Papillary craniopharyngioma arises primarily in adults rather than children and is composed of squamous epithelium that, in contrast to that of the adamantinomatous variant, does not form significant amounts of keratin. The lining of Rathke cleft cysts consists of pseudostratified ciliated columnar epithelium with goblet cells; although focal squamous metaplasia can occur, the distinctive nodules of wet keratin characteristic of adamantinomatous craniopharyngioma are not seen.

References:


Question 310: Pathology - Demyelinating Disease

Discussion:

Central pontine myelinolysis (CPM) has been most closely associated with the too-rapid correction of profound hyponatremia. It has been described in a variety of patients including alcoholics with Wernicke-Korsakoff syndrome, but many non-alcoholic patient groups have also been reported. The latter include liver transplant recipients and patients with severe burns, malnutrition, severe electrolyte disorders, and anorexia. Hypophosphatemia, but not thyroid disorders or hypoglycemia, has also been postulated to be related to CPM.

References:


Question 318: Pathology - Demyelinating Disease

Discussion:

Multiple periventricular areas of demyelination are characteristically seen in multiple sclerosis. These plaques have a grey hue because of the loss of myelin that normally confers a glistening white appearance to the white matter.

References:


Question 322: Pathology - Basic Reactions

Discussion:
Alzheimer type II astrocytes are devoid of visible cytoplasm and possess large watery nuclei with marginated chromatin. They are characteristically seen in hepatic encephalopathy such as occurs in alcoholic cirrhosis, Wilson's disease, and other conditions that severely compromise liver function and elevate the blood ammonia level.

References:


Question 349: Pathology - Cerebrovascular Disease

Discussion:

Of the choices provided, an acute hypertensive hemorrhage of the basis pontis is most likely. Autopsy also revealed hyalinization of basal ganglia arteries, left ventricular cardiac hypertrophy, and arteriolonephrosclerosis, which are all related to chronic hypertension. There is no history of a preceding event, which may have led to brainstem herniation and secondary (Duret) hemorrhage. Cerebral amyloid angiopathy does not typically involve the brainstem. Neoplasms and vascular malformations are uncommon causes of acute pontine hemorrhage.

References:


Question 381: Pathology - Demyelinating Disease

Discussion:

Shadow plaques in multiple sclerosis were formerly thought to represent areas of partial demyelination but are now realized to be areas of partial remyelination. Why the myelin is not fully restored in these plaques is unknown but may be important to understanding the multiple sclerosis disease process.

References:


Question 390: Pathology - Tumors

Discussion:

The potential for invasion of mesenchymal tissues, including the dura (and superior sagittal sinus), cranial bones, muscles and scalp, is an intrinsic property that is frequently seen to a greater or lesser extent in many ordinary (WHO grade I) meningiomas. Invasion of these mesenchymal tissues may complicate surgical resection but does not constitute a criterion for upgrading to atypical (WHO grade II) or anaplastic (WHO grade III) meningioma. Brain invasion, in contrast, is much less commonly seen and its presence in the absence of prior surgical procedures warrants a diagnosis of atypical (WHO grade II) meningioma in the new 2007 WHO classification. Highly vascular meningiomas are sometimes referred to as angiomatic and have no association with aggressive behavior. The older term "angioblastic meningioma" encompassed a heterogeneous group of vascular dural-based tumors that included angiomatic meningioma, hemangiopericytoma and hemangioblastoma; it is an imprecise and obsolete term that should be avoided. Four histologic subtypes merit upgrading based on their potential for early recurrence and/or aggressive clinical behavior: clear cell (WHO grade II), chordoid (WHO grade II), rhabdoid (WHO grade III), and papillary (WHO grade III). In contrast, the remaining nine WHO-recognized meningioma subtypes are classified as low grade (WHO grade I): meningothelial, fibrous, transitional, psammomatous, angiomatic, microcystic, secretory, lymphoplasmacyte-rich, and metaplastic.

References:

Question 394: Pathology - Neuromuscular Disease

Discussion:

Vacuole formation in muscle fibers is the most common change in hypokalemic periodic paralysis. The vacuoles are most prominent during the attack and are much less prominent between attacks. Group atrophy is indicative of neurogenic atrophy. Increased numbers of mitochondria may be seen in mitochondrial myopathies, which may also exhibit abnormally shaped mitochondria with paracrystalline inclusions. Lymphorrhages are small collections of lymphocytes seen at the motor end plate in myasthenia gravis. Abnormalities of the Z band are seen in nemaline myopathy.

References:


Question 404: Pathology - Neuromuscular Disease

Discussion:

Peripheral myelin protein 22 (PMP22) and P0 protein are peripheral nerve myelin proteins that are abnormal in the hereditary motor and sensory neuropathies. Overexpression of PMP22 occurs because of duplication of the PMP22 gene in Charcot-Marie-Tooth disease type 1A (CMT1A) while mutations of the gene encoding P0 protein occur in CMT1B.

References:


Question 411: Pathology - Critical Care/Trauma

Discussion:

Kinetic energy as defined by classical mechanics is equal to one-half the product of the mass of the projectile multiplied by the square of its velocity. Kinetic energy increases linearly with projectile mass, but increases as the square of the velocity; therefore, velocity is the single most important determinant of tissue injury during missile passage. Tumble and fragmentation can determine the rate at which kinetic energy is transferred and can alter the amount of damage done by a bullet possessed of a given kinetic energy.

References:


Question 420: Pathology - Tumors

Discussion:

Loss of heterozygosity for chromosomes 1p and 19q in anaplastic oligodendrogliomas is associated with increased responsiveness to chemotherapy, particularly PCV therapy (procarbazine, CCNU, vincristine).

References:

Pharmacology/Chemistry

Question 3: 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 27: Pharmacology/Chemistry - Movement Disorders

Discussion:

Beta blockers, calcium channel blockers, lamotrigine, and dixogin have not been implicated in drug-induced tremors. Neurologic side effects were reported in 20% to 40% of patients treated with amiodarone (Hilleman, et al 1998), at times associated with tremor, ataxia, peripheral neuropathy, malaise or fatigue, sleep disturbances, dizziness, and headaches.

References:


Question 34: Pharmacology/Chemistry - Headache

Discussion:

The efficacy, safety and tolerability of topiramate has been demonstrated in three large multicenter, randomized, double-blind, placebo-controlled trials. Overall, AEs led to treatment discontinuation in 24.9% of patients receiving topiramate 100 mg/day and 11.0% receiving placebo (P < 0.001). AEs leading to discontinuation during the double-blind phase in > or =2% of patients included paresthesia (8.0% discontinued), any cognitive symptoms (7.3% discontinued), fatigue (4.7% discontinued), insomnia (3.4% discontinued), nausea (2.3% discontinued), loss of appetite, anxiety, and dizziness (2.1% discontinued because of each AE). Most AEs began during the titration period.

References:


Question 43: Pharmacology/Chemistry - Neurogenetics

Discussion:

Spinocerebellar ataxia type 6 (SCA6) is a dominantly inherited degenerative disorder of the cerebellum characterized by nearly selective and progressive death of Purkinje cells. The underlying mutation in SCA6 consists of an expansion of a trinucleotide
CAG repeat in the 3’ region of the gene, CACNA1A, encoding the 1A subunit of the neuronal P/Q-type voltage-gated calcium channel. Affected individuals have 20 to 33 CAG repeats.

It is characterized by adult-onset, slowly progressive cerebellar ataxia, dysarthria, and nystagmus. Mean age of onset is 43 to 52 years. Initial symptoms are gait unsteadiness, stumbling, and imbalance (in ~90%) and dysarthria (in ~10%). Eventually all persons have gait ataxia, upper-limb incoordination, intention tremor, and dysarthria. Dysphagia and choking are common. Visual disturbances may result from diplopia, difficulty fixating on moving objects, horizontal gaze-evoked nystagmus, and vertical nystagmus. Hyperreflexia and extensor plantar responses occur in up to 40% to 50%. Basal ganglia signs, including dystonia and blepharospasm, occur in up to 25%. Mentation is generally preserved.

Episodic ataxia type 2 is also due to point mutations in the CACNA1A gene coding for the alpha 1A voltage-dependent calcium channel subunit. However, small repeat expansions of the CAG motif have been found in family members presenting with either spinocerebellar ataxia or episodic ataxia.

References:


Question 44: Pharmacology/Chemistry - Neuromuscular Disorders

Discussion:

Nifedipine and other calcium channel blockers can cause increased weakness in myasthenic patients. A variety of drugs induce or worsen myasthenic symptoms through actions on pre- or postsynaptic mechanisms. Certain antibiotics impair transmitter release by interfering with calcium flux. Other drugs affect metabolism of acetylcholine or cholinergic receptor function. Bromocriptine, chloramphenicol, fluoxetine, and gabapentin do not adversely affect the myasthenic patient.

References:


Question 48: Pharmacology/Chemistry - Movement Disorders

Discussion:

Paraneoplastic syndromes refers to symptoms or signs due to damage to organs or tissues that are remote from site of a malignancy. Most are rare, affecting 0.01 percent of patients with cancer, with the exception of Lambert-Eaton myasthenic syndrome, myasthenia gravis, and polyneuropathy in association with a osteosclerotic plasmacytoma. Antibodies directed at neural nuclear or cytoplasmic epitopes can be measured in spinal fluid and serum. Indentification of these antibodies and their target antigens has advanced the ability to make an early diagnosis if present in a symptomatic patient.

The presence of antigen specific cytotoxic T cells has been documented in patients with acute and subacute pan cerebellar degeneration and circulating anti-Yo antibodies. These antibodies are directed against the cytoplasm of cerebellar purkinje cells. Other antibodies have also been implicated in paraneoplastic subacute cerebellar degeneration, including anti-Tr and anti-Hu.

References:


Question 61: 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 63: Pharmacology/Chemistry - Dementia

Discussion:

Memantine (Namenda) is a moderate-affinity, uncompetitive, voltage-dependent, N-methyl-D-aspartate (NMDA) -receptor antagonist with fast on/off kinetics that inhibits excessive calcium influx induced by chronic overstimulation of the NMDA receptor.

References:


Question 74: Pharmacology/Chemistry - Headache

Discussion:

According to the *International Classification of Headache Disorders*, 2nd edition, the definition of medication overuse headache (MOH) continues to evolve over time. The recently published new appendix criteria for a broader concept of chronic migraine of the International Headache Society define MOH as 1) headache present > 15 days/month; 2) regular overuse for 3 months of one or more acute/symptomatic treatment drugs, defined as: ergotamine, triptans, opioids, or combination analgesic medication on 10 days/month on a regular basis for >3 months; or simple analgesics or any combination of ergotamine, triptans, analgesics, opioids on 15 days/month on a regular basis for >3 months without overuse of any single class alone.

References:


Question 83: Pharmacology/Chemistry - Epilepsy

Discussion:

Much has been said about the dangers of the liver toxicity of valproic acid. It seems that patients with specific inborn errors of metabolism such as mitochondrial disease or urea cycle defect are particularly susceptible to this complication.

References:


Question 85: Pharmacology/Chemistry - Movement Disorders

Discussion:
Botulinum neurotoxin (BoNT) is a microbial protein that exists in seven serotypes, designated A through G. Although the individual serotypes are immunologically distinct, all members of the group possess similar subunit structures, act on the same target organs, and produce similar functional outcomes. Each molecule is typically released from bacteria as part of a noncovalent complex with other proteins.

BoNT is an enzyme that acts in the cytosol of nerve endings to cleave three polypeptides that govern exocytosis. Serotypes A and E cleave synaptosomal-associated protein (SNAP)-25, serotypes B, D, F, and G cleave vesicle-associated membrane protein (VAMP), and serotype C cleaves both syntaxin and SNAP-25. 3,4 The ability of BoNT to block acetylcholine release at neuromuscular junctions accounts for its therapeutic action to relieve dystonia, spasticity, and related disorders.

The toxin has additional therapeutic benefits, not necessarily related to neuromuscular transmission, including: 1) blockade of acetylcholine release at autonomic nerve endings and 2) blockade of transmitter release at peripheral nerve endings that use mediators other than acetylcholine.

References:


Question 86: Pharmacology/Chemistry - Epilepsy

Discussion:

According to the current AAN guidelines and the US Food and Drug Administration, the main indication for lamotrigine in children is new onset of absence seizures. The main advantage of the newer antiepileptic medication compared to the older classic ones is a lower incidence of side effects. Carbamazepine and phenytoin are ineffective or can worsen absence seizures. Levetiracetam and topiramate may be useful as adjunct therapies for other generalized types of epilepsy.

References:


Question 88: 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:


Question 89: Pharmacology/Chemistry - Other Pain Syndromes

Discussion:

Thalamic pain syndromes can result in a minority of patients with ischemic lesions in this area and involves the contralateral side including face, arm, leg. The risk of pain or dyesthesias appears to be higher with lesions involving the inferolateral regions of
the thalamus, specifically the ventrocaudalis and ventro-oralis nuclei of the thalamus. Time lag between stroke onset and pain complaint can be between 2 to 15 days.

Parietal strokes, including the operculum, and posterior insula can produce a variety of sensory syndromes which are primarily negative signs or loss of sensory modalities, not pain.

References:


Question 92: Pharmacology/Chemistry - Neuromuscular Disorders

Discussion:

The neurologic manifestations of acquired copper deficiency in humans has been recognized, the most common being a myelopathy presenting with a spastic gait and prominent sensory ataxia. The known causes of acquired copper deficiency include prior gastric surgery, excessive zinc ingestion, and malabsorption; however, often the cause is unclear.

Hyperzinccemia may be present even in the absence of exogenous zinc ingestion. The clinical features and neuroimaging findings are similar to the subacute combined degeneration seen in patients with vitamin B₁₂ deficiency. Copper and vitamin B₁₂ deficiency may coexist. The neurologic syndrome may be present without the hematologic manifestations.

B₁₂ deficiency, which can present similarly, is often associated with elevated serum methylmalonic acid and homocystine levels. Iron supplementation has been associated with copper deficiency as well due to less than clear mechanisms.

References:


Question 99: Pharmacology/Chemistry - Neurogenetics

Discussion:

Ataxia-telangiectasia (A-T) is a rare autosomal recessive genetic disorder characterized by progressive neurodegeneration, a high risk of cancer and immunodeficiency. These patients are also hypersensitive to radiotherapy. The gene product defective in this syndrome, ATM (ataxia-telangiectasia mutated), normally recognizes DNA damage and signal to the DNA repair machinery and the cell cycle checkpoints to minimize the risk of genetic damage. No curative strategy for this disease exists. Treatment has focused on slowing the progress of the neurodegeneration; devising approaches for the treatment of tumors while minimizing side effects and treatment with immunoglobulin for the immunodeficiency.

References:


Question 119: Pharmacology/Chemistry - Headache

Discussion:

This patient has typical cluster headache; oxygen, at a typical dosing of 100% given via a nonrebreather face mask at 7 L/min to 10 L/min for 20 minutes. In some patients, oxygen is completely effective at aborting an attack if taken when the pain is at maximal intensity, while in others, the attack is only delayed for minutes to hours rather than completely alleviated. Sumatriptan and DHE are effective but carry risk for patients with coronary artery disease. Verapamil is effective for prophylaxis, but not aborting an acute cluster headache. Propranolol (beta adrenergic receptor blockers) have demonstrated efficacy for prophylaxis of migraine, but not cluster headaches.
Question 135: Pharmacology/Chemistry - Epilepsy

Discussion:

Pyridoxine-dependent seizures are believed to result from diminished activity of glutamic acid decarboxylase (GAD), which is responsive to pharmacologic doses of its cofactor, pyridoxine (vitamin B₆). Deficient action of GAD would be expected to produce elevated levels of the excitatory neurotransmitter glutamic acid, with corresponding low levels of gamma aminobutyric acid, the major inhibitory neurotransmitter in the brain. Such findings have been reported in patients with pyridoxine-dependent seizures, both prior to and when off therapy. The cerebrospinal fluid glucose concentration is normal in this condition.

References:


Question 143: Pharmacology/Chemistry - Movement Disorders

Discussion:

Based on American Academy of Neurology criteria, the highest quality literature available for the respective indications is as follows: blepharospasm (two Class II studies); hemifacial spasm (one Class II and one Class III study); cervical dystonia (seven Class I studies); focal upper extremity dystonia (one Class I and three Class II studies); focal lower extremity dystonia (one Class II study); laryngeal dystonia (one Class I study); motor tics (one Class II study); and upper extremity essential tremor (two Class II studies).

Therefore, botulinum neurotoxin should be offered as a treatment option for the treatment of cervical dystonia (Level A), may be offered for blepharospasm, focal upper extremity dystonia, adductor laryngeal dystonia, and upper extremity essential tremor (Level B), and may be considered for hemifacial spasm, focal lower limb dystonia, and motor tics (Level C). Spastic dysphonia is due to upper motor neuron lesions. Though case series have been reported using botulinum toxin for other movement disorders (such as myoclonus and tics), there is not enough evidence, or clinical consensus, to recommend it as first-line therapy.

References:


Question 146: Pharmacology/Chemistry - Cerebrovascular Disease

Discussion:

Clopidogrel is a potent oral antiplatelet agent often used in the treatment of coronary artery disease, peripheral vascular disease, and cerebrovascular disease. The mechanism of action of clopidogrel is an irreversible blockade of the adenosine diphosphate (ADP) receptor on platelet cell membranes. This receptor is named P2Y12 and is important in platelet aggregation, the cross-linking of platelets by fibrin. The blockade of this receptor inhibits platelet aggregation by blocking activation of the glycoprotein IIb/IIIa pathway.

Dipyridamole inhibits the uptake of adenosine into platelets, endothelial cells and erythrocytes in vitro and in vivo; the inhibition occurs in a dose-dependent manner at therapeutic concentrations (0.5-1.9 g/mL). This inhibition results in an increase in local concentrations of adenosine which acts on the platelet A2-receptor thereby stimulating platelet adenylate cyclase and increasing platelet cyclic-3',5'-adenosine monophosphate (cAMP) levels. Via this mechanism, platelet aggregation is inhibited in response to various stimuli such as platelet activating factor (PAF), collagen and adenosine diphosphate (ADP).
Aspirin's ability to suppress the production of prostaglandins and thromboxanes is due to its irreversible inactivation of the cyclooxygenase (COX) enzyme. Cyclooxygenase is required for prostaglandin and thromboxane synthesis. Low-dose, long-term aspirin use irreversibly blocks the formation of thromboxane A2 in platelets, producing an inhibitory effect on platelet aggregation.

Tissue-type plasminogen activator (tPA) is one of the two mammalian serine proteases that activates plasminogen into plasmin, the primary plasmatic function being fibrinolysis.

References:


Question 147: Pharmacology/Chemistry - Epilepsy

Discussion:

Unlike other antiepileptic drugs, levetiracetam is not primarily metabolized in the liver and therefore is less likely to interfere with the metabolism of porphyrins or other drugs. The safety and efficacy of levetiracetam in treatment of seizures in patients with acute intermittent porphyria (AIP) have been reported.

References:


Question 153: Pharmacology/Chemistry - Sleep Disorders

Discussion:

Modafinil (Provigil) is a medication used to promote wakefulness. Originally marketed in 1998, modafinil, a Schedule C-IV drug, was approved by the Food and Drug Administration (FDA) for the treatment of excessive sleepiness (ES) associated with narcolepsy. In 2003, the FDA approved modafinil for the treatment of ES associated with obstructive sleep apnea-hypopnea syndrome (only as an adjunct to standard treatment) and shift work sleep disorder.

References:


Question 156: Pharmacology/Chemistry - Other Pain Syndromes

Discussion:

Reflex sympathetic dystrophy (RSD) is composed of five major features: pain, swelling, autonomic dysregulation, movement disorders, and atrophy and dystrophy. RSD is caused by an injury to a specific nerve or the C- and A-delta fibers that innervate the involved tissue and alterations of sensory, autonomic, and possibly motor processing centrally. It is a progressive illness that spreads with time and may encompass the entire body. There is no psychological disposition to the problem, but all patients are severely depressed because of the constant pain, lack of sleep, and complete disruption of their lifestyle. Atrophy and dystrophy are partly due to loss of nutritive blood supply to the affected tissues. The movement disorder is partly due to deficiency of GABAergic mechanisms; the tremor is an exaggeration of the normal physiologic tremor.

Treatment consists of decreasing the afferent pain, maintaining barrage from the underlying defect, and blocking the sympathetic component of the process. Physical therapy is important as well. New developments include the use of neurotrophic factors to reverse the phenotypic changes that occur in the dorsal horn and the use of pharmacologic agents to block the activity-dependent
NMDA channels that appear to be instrumental in maintaining central sensitization. Late diagnosis is often associated with irreversible damage.

The use of magnetoencephalography has documented cortical reorganization of regional representation in the somatosensory cortex that correlates with pain severity.

References:


Question 161: Pharmacology/Chemistry - Dementia

Discussion:

This patient has a Parkinson-dementia syndrome, likely either dementia with Lewy bodies (DLB) or Parkinson disease dementia. Rivastigmine inhibits acetylcholinesterase and thus augments cholinergic activity and can lessen fluctuations in sensorium and hallucinations in DLB that is a cholinergic-deficient state. Patients with DLB are often pathologically sensitive to standard neuroleptics which can cause severe parkinsonism and neuroleptic malignant syndrome. This is an absolute contraindication. Over-the-counter antihistamine and prescription anticholinergic agents should be avoided. Memantine has not been shown to be beneficial for this indication.

References:


Question 172: Pharmacology/Chemistry - Neurogenetics

Discussion:

Black box warning for all carbamazepine formulations indicates that studies in patients of Chinese ancestry have found a strong association between the risk of developing Stevens-Johnson syndrome (SJS/TEN) and the presence of HLA-B*1502, an inherited allelic variant of the HLA-B gene. HLA-B*1502 is found almost exclusively in patients with ancestry across broad areas of Asia. Patients with ancestry in genetically at risk populations should be screened for the presence of HLA-B*1502 prior to initiating treatment with all carbamazepine containing medications.

References:


Question 178: Pharmacology/Chemistry - Aging, Degenerative Diseases

Discussion:

Diffuse Lewy body dementia (DLB) is commonly considered the second most common form of dementia, although some experts believe vascular dementia to be the second most common form. DLB is often under-diagnosed and misdiagnosed as Alzheimer disease or Parkinson related dementia. The core features of dementia with Lewy bodies are cognitive decline plus at least one of the following: fluctuations in cognition, visual hallucinations, and parkinsonism. Other supportive features include: neuroleptic sensitivity, repeated falls, syncope, transient loss of consciousness, REM sleep disturbances, depression, delusions, and nonvisual
hallucinations. Increased prudence with the use of neuroleptic agents is essential in DLBs because the use of these agents is associated with physical and cognitive decline and increased mortality. In most cases the symptoms of the parkinsonian syndrome (akinesia, rigidity, tremor) can be improved by levodopa and 300 mg/d is the maximal dose because of psychiatric side effects.

While neuroleptic sensitivity has been reported with the use of both typical and atypical antipsychotic medications, visual hallucinations and delusions can be improved by therapy with atypical neuroleptics (clozapin, risperidon, quetiapin). Clozapin seems to be the drug of first choice. Doses of 3 x 12.5 up to 3 x 50 mg/d can be used. Rarely atypical neuroleptics worsen parkinsonian symptoms and state of consciousness. Decreases in neuroleptic sensitivity can often be achieved by dose reductions, although neuroleptic discontinuation is sometimes necessary. Haloperidol is a typical antipsychotic with a higher risk of neurologic worsening. Cholinesterase inhibitors may be especially useful in the treatment of DLB. Cholinergic deficits are associated with visual hallucinations, and cholinesterase inhibitors often result in resolution of hallucinations, improved cognition, and decreased behavioral disturbances.

References:

Henriksen AL, St Dennis C, Setter SM, Tran JT. Dementia with lewy bodies: therapeutic opportunities and pitfalls. Consult Pharm 2006;21(7):563-575.

Benecke R. Diffuse Lewy body disease - a clinical syndrome or a disease entity? J Neurol 2003;250[Suppl 1]:139-142.

Question 184: Pharmacology/Chemistry - Epilepsy

Discussion:

Topiramate (TPM) is a neuromodulatory agent that was initially approved as an antiepileptic drug and is increasingly used in the treatment of a number of neurological and metabolic disorders. Among its various pharmacological actions, TPM has been shown to inhibit the activity of specific carbonic anhydrase enzymes in the kidney. This action is associated with the development of metabolic acidosis, hypocitraturia, hypercalciuria and elevated urine pH, leading to an increased risk of kidney stone disease.

References:


Question 185: Pharmacology/Chemistry - Neuromuscular Disorders

Discussion:

Malignant hyperthermia is an autosomal-dominant inherited disorder of the skeletal muscle cell, characterized by a hypermetabolic response to all commonly used inhalational anaesthetics and depolarizing muscle relaxants. The clinical syndrome includes muscle rigidity, hypercapnia, tachycardia and myoglobinuria as result of increased carbon dioxide production, oxygen consumption and muscle membrane breakdown.

References:


Question 188: Pharmacology/Chemistry - Toxic/Metabolic Disease

Discussion:

Bariatric surgical procedures are increasingly common and there are neurologic complications that can result. The most common presentations are peripheral neuropathy (62%) and encephalopathy (31%). Of those with peripheral neuropathy, 67% have polynephropathy and 30% have mononeuropathies, which include meralgia paresthetica and foot drop. Neurologic emergencies,
including Wernicke syndrome, rhabdomyolysis, and Guillain-Barré syndrome, have also been reported. There is evidence to suggest a role for inflammation or an immunologic mechanism in neuropathy after gastric bypass. Micronutrient deficiencies following gastric bypass are also prevalent, with up to 25% patients with B12 deficiency and 1% with thiamine deficiency.

Although copper deficiency has been reported in bariatric surgery patients, the neurologic disorder associated with it is a subacute combined degeneration very similar to that seen with B12 deficiency.

References:

Question 189: Pharmacology/Chemistry - Epilepsy

Discussion:
Of the anticonvulsant medications listed, only lamotrigine is not an inducer of vitamin D metabolism.

References:

Question 198: Pharmacology/Chemistry - Movement Disorders

Discussion:
Restless legs syndrome (RLS) is clinically defined as an urge to move the legs with or without paresthesia, worsening of symptoms with rest and transient improvement with activity, and worsening of symptoms in the evening and night. Endstage renal disease, increasing age, female gender, pregnancy, frequent blood donations, iron deficiency and neuropathy are considered to be risk factors for this disorder. The association to RLS is less definitely established for other conditions, such as Parkinson disease (PD) or diabetes.

References:

Question 327: 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:
Discussion:

Some patients with autonomic failure exhibit a degree of hypoproliferative anemia. Administration of recombinant human erythropoietin-alpha, 25-75 U/kg subcutaneously two to three times a week for 6 weeks, rapidly corrects the anemia and improves orthostatic tolerance.

References:


Question 340: Pharmacology/Chemistry - Headache

Discussion:

Migraine is a very common disorder. An estimated 18% of women and 6% of men experience migraine, but many go undiagnosed and undertreated. Use migraine-specific agents (triptans, dihydroergotamine [DHE]) in patients with moderate or severe migraine or whose mild-to-moderate headaches respond poorly to nonsteroidal anti-inflammatory drugs (NSAIDs) or combinations such as aspirin plus acetaminophen plus caffeine. Failure to use an effective treatment promptly may increase pain, disability, and the impact of the headache.

Serotonin (5-HT) receptors are part of the G protein-coupled and ligand-gated ion channel families. 5-HT exerts its diverse actions by binding to cell surface receptors which can be classified into seven distinct families (5-HT1 to 5-HT7) according to their structural diversity and mode of action. Some of the 5-HT families are comprised of multiple receptors which share similar structural and mechanistic properties but display very different operational profiles. Evidence continues to mount in support of the important roles of the 5-HT receptors in various neuropsychiatric disorders such as anxiety, depression, schizophrenia, migraine and drug addiction. The 5-HT receptors may also play an important role in obesity, aggression, sexual behavior, mood, learning, gastrointestinal motility, and cardiovascular disorders. A number of selective/non-selective 5-HT agonist and antagonist ligands (drugs) have been developed to challenge many of these disease states.

The triptans are a class of drug specifically designed and developed for the acute treatment of migraine. All triptans are small, synthetic molecules that selectively activate 2 closely related subtypes of receptor (5-HT1B and 5-HT1D) for 5-hydroxytryptamine (serotonin). 5-HT1B receptors are localized in cranial blood vessels and also trigeminal nerve terminals. 5-HT1D receptor subtypes has very similar pharmacology to that of 5-HT1B receptors and is similarly located in neurons but absent from cranial vascular smooth muscle. In migraines, benefit is likely derived from vasoconstriction of cranial blood vessels and modulation of neuropeptide release, such as CGRP and substance P.

References:


Question 344: Pharmacology/Chemistry - Other Pain Syndromes

Discussion:

Serotonin syndrome (SS) is a potentially life-threatening adverse drug reaction caused by excessive serotonergic agonism in central and peripheral nervous system serotonergic receptors. Symptoms are characterized by a triad of neuron-excitatory features, which include (a) neuromuscular hyperactivity -- tremor, clonus, myoclonus, hyperreflexia and, in advanced stages, pyramidal rigidity; (b) autonomic hyperactivity -- diaphoresis, fever, tachycardia and tachypnea; (c) altered mental status -- agitation, excitement and, in advanced stages, confusion. Monoamine oxidase inhibitors and opioid analgesics can cause serotonin toxicity. It arises when pharmacological agents increase serotonin neurotransmission at postsynaptic 5-hydroxytryptamine 1A and 5-hydroxytryptamine 2A receptors through increased serotonin synthesis, decreased serotonin metabolism, increased serotonin release, inhibition of serotonin reuptake or direct agonism of the serotonin receptors.
Multiple drugs and supplements have been implicated in this syndrome, including serotonin reuptake inhibitors (SSRIs), serotonin and norepinephrine reuptake inhibitors (SNRIs), bupropion, monoamine oxidase inhibitors, triptans, opioid analgesics, lithium, illicit drugs such as LSD, cocaine, and amphetamines, herbal supplements (e.g., St. John's wort and ginseng), dextromethorphan, ritalin, linezolid, and some antinausea medications (e.g., metoclopramide, ondansetron, and granisetron). The etiology is often the result of therapeutic drug use, intentional overdosing of serotonergic agents or complex interactions between drugs that directly or indirectly modulate the serotonin system. Serotonin syndrome in this case resulted from coadministration of tramadol, venlafaxine, and mirtazapine. It is likely that the activation of 5-HT(1A) receptors by mirtazapine, the combined serotonin reuptake inhibition by venlafaxine and tramadol, as well as possible serotonin release by tramadol, contributed to the development of SS in this case.

Neuroleptic malignant syndrome (NMS) is caused almost exclusively by antipsychotics, including all types of neuroleptic medicines along with newer antipsychotic drugs. The higher the dosage, the more common the occurrence. Rapid and large increases in dosage can also trigger the development of NMS. Other drugs, environmental or psychological factors, hereditary conditions, and specific demographics may cause greater risk, but to date no conclusive evidence has been found to support this. The disorder typically develops within 2 weeks of the initial treatment with the drug, but may develop at any time the drug is being taken.

References:


Question 347: Pharmacology/Chemistry - Toxic/Metabolic Disease

Discussion:

Paralytic shellfish poisoning is caused by consumption of bivalve mollusks (mussels, clams, scallops, oysters) contaminated with the dinoflagellate-produced toxin called saxitoxin, which blocks sodium channels and, thus, interferes with impulse conduction in peripheral nerves and muscles. Respiratory failure can result. A number of other marine toxins can produce very similar symptoms. Ciguatera toxin is another dinoflagellate-derived toxin that concentrates in certain large fish. Neurotoxic shellfish poisoning is due to consumption of oysters and clams contaminated with brevotoxin B, yet another dinoflagellate toxin. Domoic acid poisoning occurs following consumption of mussels contaminated with the marine diatom Nitzchia pungens. Neurologic features include confusion and altered states of arousal. Seizures may also occur. Domoic acid is a glutamate agonist. Tetrodotoxin, unlike the previously mentioned toxins, is produced by the puffer fish itself. Saxitoxin poisoning occurs in the Northeast and Northwest USA, ciguatoxin poisoning in Florida and Hawaii, brevotoxin B poisoning in the Gulf of Mexico and Caribbean Sea, domoic acid poisoning in eastern Canada and tetrodotoxin poisoning wherever puffer fish are eaten, primarily in Japan.

References:


Question 354: Pharmacology/Chemistry - Neuromuscular Disorders

Discussion:

Periodic paralysis describes a clinical syndrome of episodic partial or general weakness often associated with abnormal ion channel conductance. Hyperkalemic periodic paralysis (hyperPP) is an autosomal dominant disorder which manifests as episodic weakness usually during the first decade. Attacks usually last 1 to 4 hours, are generalized, and triggered by rest after exercise, stress, fatigue, and are potassium sensitive. Electrical myotonia is found in up to 75% of affected individuals. It is typically associated with elevated ictal serum K levels, however many individuals may be normokalemic during an attack. Most cases of hyperPP, as well as the allelic disorders paramyotonia congenita and potassium-aggravated myotonia are caused by mutations in the sodium channel, SCN4A. Management of the periodic paralyses is symptomatic and behavioral. Affected individuals learn to avoid precipitating triggers. Hyperkalemic periodic paralysis patients stay away from K rich foods, medications that increase serum K levels, and fasting. First line prophylactic treatment is a thiazide diuretic to reduce frequency of attacks. Carbonic anhydrase inhibitors anecdotally have shown benefit in preventing attacks of periodic paralysis (PP), but require further adequate study. Inhaled beta agonist have been shown to ameliorate attacks. During an attack, a patient can be give IV glucose or insulin, inhaled beta agonist, or oral carbohydrates in the ER setting. In contrast, hypokalemic periodic paralysis is associated commonly
with decreased serum potassium levels. It is the most frequent of the PP's and is also inherited in an autosomal dominant pattern. The majority of cases are due to mutations in the calcium channel, CACNA1S. Onset of onset is usually in the first to third decade. Attacks are usually longer than those of hyperPP and last between hours to days. Although symptoms can be associated with rest after exercise, these patients are also sensitive to things that lower serum potassium, including exercise, large carbohydrate meals, ethanol, cold, and certain medications (steroids, beta agonists, or insulin). Although there can be significant overlap in presentation and associated triggers between hyperPP and hypoPP, association with fasting and presence of interictal lid lag and eyelid myotonia is specific for hyperPP.

References:


Question 364: Pharmacology/Chemistry - Demyelinating Disorders

Discussion:

Central pontine myelinolysis (CPM) consists of demyelination in the base of the pons, with a relative sparing of the axons and the nerve cells. Myelinolysis in the pons is frequently associated with demyelination in other areas of the brain where the gray and the white matter are closely admixed. These extrapontine lesions are thought to coexist with CPM in 10% of the cases. The pathogenesis of CPM is unknown. It is hypothesized that in the chronic hyponatremic state, rapid treatment with hypertonic saline solution creates an osmotic stress on the brain. This leads to an osmotic injury to the vascular endothelial cells resulting in the release of myelinotoxic factors or vasogenic edema. The brain dehydration may lead to the separation of the axons from its myelin sheath with resultant injury of oligodendrocytes. Neurological deterioration may often be preceded by a transient improvement paralleling the correction of electrolyte disturbance. Fluctuating levels of consciousness, convulsions, hypoventilation, or hypotension may herald the onset of this syndrome. Eventually pseudobulbar palsy and quadriplegia develops. Swallowing dysfunction and inability to speak may be dominant features. In severe cases, the patient may develop a "locked-in syndrome". Predisposing factors include severe underlying medical illness or nutritional deficiency. The disease may develop in patients undergoing liver transplantation, long term alcohol abuse and liver cirrhosis, due to uremia or as a complication of dialysis, and in patients with severe malnutrition.

References:


Question 369: Pharmacology/Chemistry - Cerebrovascular Disease

Discussion:

The current guidelines recommend treating ischemic stroke patients who present within 3 hours of symptom onset with recombinant tissue plasminogen activator (r-tPA) at a dose of 0.9 mg/kg (maximum dose 90 mg) within 3 hours of symptom onset with 10% of the dose given as bolus followed by infusion lasting 60 minutes. Exclusion criteria include prior intracranial hemorrhage; history of intracranial neoplasm, aneurysm or anteriovenous malformation; stroke or head trauma within the previous 3 months; major surgery or biopsy of a parenchymal organ within preceding 14 days; gastrointestinal or urinary bleeding within the previous 21 days; recent myocardial infarction; seizure at onset of stroke; history of known hereditary or acquired abnormal hemostasis; current use of oral anticoagulants with prothrombin time>15 seconds, use of heparin in previous 48 hours with prolonged partial thromboplastin time; platelet count <100,000/mm3, and evidence on CT of major hypodensity or sulcal effacement (>1/3 of middle cerebral artery territory), and systolic blood pressure>185 mmHg or diastolic blood pressure >110mmHg, and blood glucose <50 mg/dl or >400 mg/dl.

References:

Question 388: Pharmacology/Chemistry - Other Pain Syndromes

Discussion:

Post-herpetic neuralgia (PHN) is the term used for the condition that exists if the pain persists after a herpes zoster reactivation rash (shingles) has resolved. Advanced age and compromised cell-mediated immunity are significant risk factors for reactivation of herpes zoster and the subsequent development of PHN. Though the pathophysiology of PHN is unclear, studies suggest peripheral and central demyelination as well as neuronal destruction are involved. Both the varicella vaccine, and the newly released zoster vaccine, against herpes zoster may lead to substantial reductions in morbidity from herpes zoster and PHN. In addition, current evidence suggests that multiple medications are effective in reducing the pain associated with PHN; these include tricyclic antidepressants, antiepileptics, opioids, N-methyl-D-aspartate (NMDA) receptor antagonists, as well as topical lidocaine (lignocaine) and capsaicin. Reasonable evidence supports the use of intrathecal corticosteroids, but the potential for neurologic sequelae should prompt caution with their application. Epidural corticosteroids have not been shown to provide effective analgesia for PHN. Sympathetic blockade may assist in treating the pain of herpes zoster or PHN. For intractable PHN pain, practitioners have performed delicate surgeries and attempted novel therapies. Although such therapies may help reduce pain, they have been associated with disappointing results, with up to 50% of patients failing to receive acceptable pain relief.

References:


Question 397: Pharmacology/Chemistry - Epilepsy

Discussion:

Grapefruit juice leads to inhibition of CYP3A4, predisposing to increased plasma levels and toxicity of carbamazepine. There is no interaction between dietary proteins or carbohydrates nor is there an effect of carbamazepine on iron metabolism. Carbamazepine does not increase the risk of nephrolithiasis.

References:


Question 413: 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 417: Pharmacology/Chemistry - Headache

Discussion:
Calcitonin gene related peptide (CGRP) is derived, with calcitonin, from the CT/CGRP gene located on chromosome 11. It is primarily produced in nervous tissue, however, its receptors are expressed throughout the body. It is found in every location described in migraine genesis and processing, including meninges, trigeminal ganglion, trigeminocervical complex, brainstem nuclei, and cortex. It is released in animal models following stimulation of the CNS similar to that seen in migraine, and triptans inhibit this release. Injection of CGRP into migraineurs results in delayed headache similar to migraine. Elevation of CGRP occurs during migraine, resolving following migraine-specific treatment. Finally, and most importantly, CGRP receptor antagonists terminate migraine with efficacy similar to triptans.

References:


Question 438: Pharmacology/Chemistry - Movement Disorders

Discussion:

Frucht and Fahn (2000) reviewed more than 100 cases of Landes-Adams syndrome (LAS) and found that clonazepam, sodium valproate, and piracetam were significantly effective in approximately 50% of patients. Clonazepam, sodium valproate, piracetam, and levetiracetam may be recommended as first-line agents to treat patients with post-anoxic action myoclonus (PHAM). Piracetam is not available in the US.

References:


Question 442: Pharmacology/Chemistry - Movement Disorders

Discussion:

Restless leg syndrome (RLS) is characterized by an unpleasant sensation in the extremities that occurs before sleep and is associated with a strong urge to move limbs. This results in temporary relief of symptoms, yet symptoms typically recur when movement stops. Many patients also experience periodic limb movements during sleep. The cause is unknown but is common with iron deficiency, pregnancy, and metabolic dysfunction such as renal failure. Medications reported to be beneficial include dopaminergics (eg, levodopa, bromocriptine, pramipexole, and pergolide), and opiates (eg, codeine, propanolol), and benzodiazepines (clonazepam, diazepam, triazolam). All serotonin uptake inhibitors except bupropion have been reported to worsen RLS.

References:


Physiology

Question 4: Physiology - EEG

Discussion:

Polymorphic delta activity is produced by processes involving subcortical white matter.

References:
Question 22: Physiology - Sleep

Discussion:

In restless leg syndrome, an associated clinical feature is serum ferritin <50 micrograms/L. Overnight polysomnogram would show periodic limb movements in sleep with episodes of at least 4 consecutive movements, 0.5 to 5 seconds long, and between 5 and 90 seconds apart.

References:

Walters AS. Restless Legs Syndrome and PLMS. Continuum 2007; 13(3):115-138

Question 24: Physiology - EEG

Discussion:

This EEG shows slow spike waves in generalized distribution. It is characteristic of Lennox- Gastaut syndrome. This often follows infantile spasms which is associated with hypsarrythmia in EEG.

References:


Question 26: Physiology - EEG

Discussion:

Rapid deterioration of mental function with an EEG that shows generalized sharp and slow wave complexes is most consistent with nonconvulsive status epilepticus, which can present clinically with an abrupt deterioration in mental function with confusion or stupor. Hepatic coma is associated with triphasic waves, and drug toxicity is usually associated with generalized slow wave or excessive beta activity. Herpes simplex encephalitis, which can also present in a subacute fashion, is associated with periodic lateral epileptiform discharges (PLEDs).

References:


Question 33: Physiology - EMG

Discussion:

Ischemia due to arterial compression can involve all nerves distal to the site of the injury. This can result in severe denervation and pain with weakness, worse in distal muscles.

References:


Question 36: Physiology - Evoked Potentials
Discussion:

Brainstem auditory evoked potentials (BAEPs) are normal in patients with cortical deafness as the responses are recorded from the brainstem and not the cortex.

References:


Question 39: Physiology - EEG

Discussion:

Wicket waves, or wicket spikes, constitute a benign pattern of uncertain clinical significance, occurring predominantly in the EEG of older adults during light sleep.

References:


Question 40: Physiology - EMG

Discussion:

Sedation especially when deep can cause loss of late responses like F waves and H waves. In this patient since the motor and sensory nerve conduction studies are normal, the absence of F waves is secondary to sedation and the depression of the central excitatory state.

References:


Question 42: Physiology - EEG

Discussion:

Hypsarrhythmia is characteristic EEG pattern seen in children with infantile spasms. The pattern is characterized by high voltage diffuse slowing with multifocal spikes and sharps in a chaotic fashion.

References:


Question 45: Physiology - EMG

Discussion:

Symptomatic infants with myotonic muscular dystrophy in neonatal period show myopathic motor units and low amplitude motor CMAP. Myotonia develops after child is older.

References:
Foot drop may present secondary to a lesion at various levels. The tibialis anterior is supplied by deep peroneal nerve, peroneus longus is supplied by the superficial peroneal nerve, and the tibialis posterior is supplied by the tibial nerve. Because the tibialis posterior is involved, the lesion is at the sciatic level or higher. If the gluteus medius were involved, the lesion would then be proximal to the sciatic nerve.

References:

On repetitive nerve stimulation at 3 Hz, a greater than 10% decrement is abnormal in patients with MG.

References:

Low temperature would result in increased latency, increased amplitude and increased duration of the response.

References:

Frequency refers to the number of times a repetitive wave recurs in 1 second.

References:
Increase in size and duration of motor units result from increase in number of muscle fibers innervated by a single motor unit. This is seen on muscle biopsy on ATPase reaction as grouping of fibers of same type.

References:

Question 76: Physiology - EEG

Discussion:
Three Hertz slow wave activity can be seen as a normal finding during hyperventilation in a child. Simple absence seizures are associated with 3 Hz spike-and-wave discharges during hyperventilation.

References:

Question 79: Physiology - Basic Physiology

Discussion:
Fusimotor fibers transmit impulses to muscle spindles.

References:

Question 81: Physiology - EEG

Discussion:
Triphasic waves are frontally prominent high voltage positive potential preceded and followed by low voltage negative potential. Seen characteristically in patients with hepatic coma.

References:

Question 84: Physiology - EMG

Discussion:
Wrist extension is performed by the Extensor Carpi Ulnaris (ECU) and Extensor Carpi Radialis (ECR). The ECR is supplied by the radial nerve and the ECU is innervated by the posterior interosseous nerve (PIN). Radial deviation on wrist extension suggests weakness of ECU with normal function of ECR.

References:
Discussion:

This infant has clinical phenotype of Krabbe's disease which is associated with central demyelination and predominantly demyelinating sensory motor polyneuropathy.

References:


Question 104: Physiology - EEG

Discussion:

Alpha coma pattern is a generalized invariant monorhythmic pattern in the alpha frequency range that shows little or no reactivity. This can occur after cardiopulmonary arrest, brainstem strokes, or drug overdose.

References:


Question 109: Physiology - EEG

Discussion:

Because the alpha rhythm frequency is variable depending on the behavioral state of the individual, it should only be measured when it is clear the subject is awake and not drowsy.

References:


Question 112: Physiology - EEG

Discussion:

After 2 years of age asynchronous sleep spindles are considered abnormal.

References:


Question 124: Physiology - EEG

Discussion:

Absence seizures can be misdiagnosed as attention deficit disorder. Observance at school may be helpful in gaining an appropriate history. Family history of seizures is also helpful. EEG shows 3 Hz spike and wave.

References:

Question 125: Physiology - EEG

Discussion:

For absence seizures, hyperventilation is still the best activation procedure for bringing out abnormalities on the EEG.

References:


Question 129: Physiology - EEG

Discussion:

Beta frequency coma is usually seen with drug toxicity or anesthesia.

References:


Question 145: Physiology - EEG

Discussion:

Patients with myoclonic epilepsy have interictal pattern of generalized polyspike and wave discharges.

References:


Question 155: Physiology - EEG

Discussion:

A sensitivity of at least 2 V/mm, or a lower sensitivity should be used for at least 30 minutes to make the diagnosis of electrocerebral silence.

References:


Question 158: Physiology - Basic Physiology

Discussion:

Rapid firing at the depolarization at the presynaptic terminal during maximal exercise of the muscle causes a net increase in intracellular calcium at the presynaptic terminal of the neuromuscular junction. This occurs as the time for extrusion of calcium is longer than the time for entry at the presynaptic terminal. At fast rates of depolarization the extrusion of calcium lags behind calcium entry and results in transient net increase in intracellular calcium. This improves the release of acetylcholine from the presynaptic terminal.

References:
Question 162: Physiology - EMG

Discussion:

Carpal tunnel syndrome (median neuropathy at the wrist) is commonly bilateral, worse in the dominant hand, has numbness involving the thumb, index, third, and frequently radial half of the fourth finger with sparing over the thenar eminence, weakness of the abductor pollicis brevis muscle with denervation is seen in more severe cases.

References:


Question 165: Physiology - Sleep

Discussion:

In the diagnosis of narcolepsy, overnight polysomnographic recording is done first to exclude any other primary cause of hypersomnia. A multiple sleep latency test (MSLT) is performed subsequently, which documents severe daytime sleepiness and abnormal occurrence of REM sleep.

References:


Question 168: Physiology - EMG

Discussion:

H reflexes are suppressed by supramaximal stimulation, are optimal with submaximal stimulation, have amplitude 50% to 100% of M wave, have relatively constant latency, have constant morphology, and are recorded from forearm and leg.

F waves are maximal with supramaximal stimulation, are infrequent with submaximal stimulation, have amplitude 10% of M wave, have variable latency, have variable morphology, and are recorded from hand and foot muscles.

References:


Question 171: Physiology - Evoked Potentials

Discussion:

Prolongation of P100 latency on one side results from slowing of conduction in the optic nerves. Acute optic neuritis causes prolongation of P100 latency and is a common cause of monocular vision loss in young adults.

References:


Question 182: Physiology - EMG
Discussion:

The muscle membrane in hypokalemic periodic paralysis is inexcitable during paralysis in patients with periodic paralysis. Myotonic discharges are seen in hyperkalemic periodic paralysis.

References:

Question 186: Physiology - EMG
Discussion:

EMG needle examination evidence of fibrillation potentials and decreased recruitment of motor units in the iliopsoas, vastus medialis, adductor longus, and vastus lateralis muscles without other abnormality localizes the lesion to the lumbar plexus.

References:

Question 191: Physiology - EMG
Discussion:

Fasciculation potentials result from irritation of the alpha motor neuron, and are seen in a variety of neurogenic conditions, but can occur in normal individuals. They are frequently seen in motor neuron disease, but usually in association with other abnormalities of spontaneous activity, motor unit action potential morphology or recruitment.

References:

Question 195: Physiology - EMG
Discussion:

Multiplets (myokymic and/or neuromyotonic discharges) can cause sustained contractions of muscles. Carbamazepine can reduce symptoms by reducing sodium and potassium channel conductance and reduce the resting membrane potential. Phenytoin is also highly effective. Benzodiazepines and anesthesia drugs have no effect.

References:

Question 200: Physiology - EMG
Discussion:

With acute (less than 1 week) nerve root compression, the only EMG abnormality may be a reduced recruitment of motor unit potentials. Fibrillation potentials are usually not seen for 7 - 10 days after onset of injury. Fasciculation potentials and large, polyphasic motor unit action potentials reflect chronicity.

References:
Question 236: Physiology - EEG

Discussion:

Periodic lateralized epileptiform discharges (PLEDs) are often seen with acute or subacute cerebral dysfunction as produced by herpes simplex encephalitis, vascular insults like stroke, abscess, or subdural hematoma. Small sharp spikes are a benign variant seen in adults, usually during drowsiness and light sleep. Triphasic waves are generally seen in metabolic encephalopathies. A 3 Hz spike-and-wave pattern is seen in children between 5 and 15 years of age suffering from absence seizures. Positive occipital sharp transients of sleep (POSTS) are transient sharp waves seen in the occipital region spontaneously during sleep.

References:

Question 252: Physiology - EEG

Discussion:

Breach rhythm is seen on EEG with breach in the cranial vault. It appears as accentuation or increased amplitude of rhythms, especially theta, alpha, and beta, underlying the breach in the skull.

References:

Question 265: Physiology - EMG

Discussion:

The tracings demonstrate prolonged latencies with a markedly reduced conduction velocity of 17 and 11 m/sec. There is no evidence of conduction block and the waveforms are uniform (no temporal dispersion) in nature. The lack of conduction block or temporal dispersion makes this more likely hereditary in nature rather than acquired. HMSN 1 is the demyelinating form, which is what is shown in the recording.

References:

Question 297: Physiology - EMG

Discussion:

The tracing shows a markedly prolonged distal latency with a small, dispersed motor response and decreased velocity. The presence of temporal dispersion and/or conduction block favors an acquired demyelinating polyneuropathy such as chronic inflammatory demyelinating polyneuropathy (CIDP).

References:
Question 302: Physiology - EEG

Discussion:

14- and 6-per-second positive spikes are a benign phenomenon in the EEG and are seen over posterior and temporal head regions during drowsiness and light sleep.

References:

Question 319: Physiology - EEG

Discussion:

Creutzfeldt-Jakob disease (CJD) is a prion disease that causes rapidly progressive dementia, myoclonus, and periodic, short-interval (0.5 to 1.0 second), generalized, bisynchronous discharges on EEG. The most likely EEG finding in a patient with early Alzheimer's disease is normal background or mild slowing of alpha activity.

References:

Question 329: Physiology - EMG

Discussion:

Children who develop infantile botulism typically are normal at birth and develop normally until the second to fifth month of life. Hypotonia then develops, accompanied by constipation. On examination the patient is quite weak and areflexic. Compound muscle action potential recording in response to 50 Hz stimulation produces a diagnostic incrementing response.

References:


Question 337: Physiology - EEG

Discussion:

Rare frontal sharps can be seen on EEG in a normal full-term baby and does not indicate underlying abnormality or epileptiform activity.

References:


Question 338: Physiology - EMG

Discussion:

Fast and slow rates of stimulation in patients with myotonic disorders causes a decline in size of motor unit amplitude.

References:
Question 348: Physiology - EMG

Discussion:

The right femoral nerve innervates quadriceps muscle and mediates the sensory and motor portions of the knee reflex. The obturator nerve shows axonal injury. The site of lesion for involvement of both these nerves is most likely from a lesion in the lumbar plexus.

References:


Question 360: Physiology - EMG

Discussion:

The electrodiagnostic studies in this patient localizes the lesion to the axillary nerve. Axillary nerve provides motor supply to deltoid and teres minor muscles and supplies sensory input to the outer aspect of the shoulder.

References:


Question 363: Physiology - Autonomic Studies

Discussion:

Heart rate changes in response to deep breathing depends on the presence of normal vagal innervation of heart.

References:

Low P.. Autonomic Disorders.

Question 376: Physiology - EMG

Discussion:

The presence of fibrillations suggests the process to be at least 2-3 weeks old. The normal motor unit morphology implies that reinnervation has not taken place and thus the lesion is sub acute in nature. The biceps (lateral cord/musculocutaneous nerve), deltoid (posterior cord/axillary nerve) and infraspinatus (suprascapular nerve) are all innervated by the C5 nerve root.

References:


Question 377: Physiology - EMG

Discussion:
In demyelinating lesions of the nerve, the nerve distal to the site demyelination show normal excitability. This patient demonstrates normal facial CMAP similar to the opposite side with reduced motor unit recruitment of the facial muscles. These features indicate the presence of a demyelinating lesion of the nerve.

References:


**Question 389: Physiology - EMG**

**Discussion:**

Homozygous mutation in SMN gene results in spinal muscular atrophy. These children will show normal sensory conduction studies with reduced motor CMAP and normal conduction velocity for age.

References:


**Question 395: Physiology - Sleep**

**Discussion:**

Sleep spindles occur during stage 2 sleep. Sleep spindles are >0.5 seconds duration.

References:


**Question 399: Physiology - EEG**

**Discussion:**

Centrotemporal spikes are characteristically seen in benign local epilepsy of childhood (Rolandic epilepsy) in which seizures are characterized by focal twitching of the face and/or hand, excessive salivation, and inability to speak. The seizures often become generalized.

References:


**Question 403: Physiology - EEG**

**Discussion:**

Eye blinks are most commonly recorded from frontopolar electrodes, although may spread into temporal and central electrodes.

References:

**Question 407: Physiology - EEG**

**Discussion:**
An EEG with reactivity, variability, and increase in background frequency demonstrates a favorable prognosis after cardiac arrest.

**References:**

**Question 415: Physiology - EMG**

**Discussion:**

Adductor magnus is innervated by L2 and L3 roots through the obturator nerver. Iliopsoas is innervated by L1 and L2 roots through the femoral nerve. Preganglionic involvement spares the sensory nerve action potentials, although degeneration of motor axons leads to muscle atrophy and reduction in amplitude of compound muscle potentials.

**References:**


**Question 416: Physiology - Sleep**

**Discussion:**

During REM sleep, there is normally loss of muscle tone noted with decreased EMG activity. In REM behavior disorder, there is increased tone and limb movements during REM sleep, resulting in movements that may end with patients falling out of bed or hurting bed partner unintentionally.

**References:**

AAN. Clinical Practice Guidelines 2006: Techniques Associated with the Diagnosis and Management of Sleep Disorders.

**Question 421: Physiology - EMG**

**Discussion:**

There is an increase in the amplitude in the proximal segments as compared distally. This is typically seen when there is an accessory peroneal nerve. This is usually a normal variant and can be tested by stimulating under the lateral malleolus and recording the EDB.

**References:**


**Question 429: Physiology - Evoked Potentials**

**Discussion:**

Median SEP with delayed N13 but normal N13 - N20 interpeak latency indicates a lesion prior to the cervical cord. In ALS, SEPs are normal, as this is not a test of the motor system.

**References:**


**Question 430: Physiology - EEG**
Discussion:

Burst suppression pattern in hypoxic ischemic coma carries an extremely poor prognosis.

References:

**Question 433: Physiology - EEG**

Discussion:

A prominent waveform that can only be recorded from one electrode is artifact until proven otherwise.

References:


**Question 434: Physiology - EMG**

Discussion:

The study demonstrates focal slowing across the palmar segment of the ulnar nerve distal to the origin of digital branch of the ulnar nerve. Lesion at the elbow and origin of FCU show focal slowing and or conduction block at those sites. Medial cord lesion reduces the size of thenar response.

References:


**Question 439: 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: