Questions

1. A 78-year-old woman with dementia and rigidity is hospitalized with dehydration. During her hospitalization, she becomes agitated and has prominent visual hallucinations. After a dose of haloperidol, she becomes very rigid and mute. The most likely type of dementia in this patient is:
   a. Alzheimer disease (AD)
   b. Parkinson disease (PD)
   c. Dementia with Lewy bodies
   d. Pick disease
   e. Vascular dementia

2. A 32-year-old woman presents to the ER complaining of blurred vision and pain in the right eye. Your evaluation shows decreased visual acuity in the right eye that does not correct with pinhole testing. There is a relative afferent pupillary defect (RAPD) on the right, and testing of the right visual field shows a small central scotoma. The most likely localization of the lesion is the:
   a. Optic chiasm
   b. Optic nerve
   c. Optic tract
   d. Occipital cortex
   e. Optic radiations

3. In the course of evaluating an infant with developmental regression, a pediatric neurologist notes a cherry-red spot on funduscopic examination. Which of the following diagnoses is consistent with that finding?
   a. Alexander disease
   b. Hurler syndrome
   b. Krabbe disease
   d. Niemann-Pick disease
   e. Canavan disease

4. A 62-year-old woman presents with progressive distal symmetric paresthesias and dysesthesias, with preserved muscle strength. She smokes cigarettes. Your electrodiagnostic study indicates that this is likely a sensory neuronopathy. Which one of the following tests will help to find the possible etiology?
   a. Anti-GM_1 antibodies
   b. Anti-DNA antibodies
   c. Anti-Hu antibodies
   d. MRI of the spine
   e. CT myelogram

5. A 40-year-old woman is evaluated in the ER after a motor vehicle accident resulting in left facial injuries. Examination after she recovers acutely demonstrates that her left seventh and eighth cranial nerves remain dysfunctional. Which of the following skull structures may have been affected by her injury?
   a. Cribriform plate
   b. Optic canal
   c. Superior orbital fissure
   d. Internal auditory meatus
   e. Jugular foramen

6. A 54-year-old woman was seen in the ER complaining of a severe headache. Head CT was normal, and an LP was performed. The opening pressure was 14 cm H_2O, and CSF analysis showed the following: 150 red blood cells, xanthochromic fluid, protein 55 (slightly increased), 15 white blood cells (90% lymphocytes), and normal glucose. Which of the following statements is true?
   a. The xanthochromia may have been caused by a traumatic tap.
   b. The lymphocytic pleocytosis indicates an active infectious process.
   c. Viral meningitis is unlikely because of the normal CSF glucose.
   d. The lymphocytic pleocytosis is likely reactive to the presence of blood within the CSF.
   e. The opening pressure is elevated and reflects pseudotumor cerebri.

7. A 29-year-old woman is brought into the ER in an unresponsive state. Her temperature is 37°C, heart rate 84 per minute, respirations 10, and blood pressure 152/84. On examination, she withdraws to noxious stimulation only. Her right pupil is 10 mm and does not constrict to light. Her left pupil is 5 mm and reacts normally. Which of the following is clinically contraindicated?
a. Raising the head of the bed
b. Intravenous administration of mannitol
c. Hyperventilation
d. LP
e. Neurosurgical consultation

8. A patient presents with gradually worsening weakness of the proximal arm and leg muscles symmetrically over several months. On examination, neck flexors and extensors are found to be weak also. There is no muscle pain or tenderness. What is the most likely site of dysfunction in the nervous system?
a. Peripheral nerve
b. Brachial plexus
c. Spinal nerve root
d. Internal capsule
e. Muscle

9. An 8-year-old boy is brought to a child psychiatrist for evaluation of potential attention deficit–hyperactivity disorder. His mother states that his teachers have been concerned about his attention because they frequently have to repeat instructions to him. At home his brother has noticed that he will stare for several seconds at a time, during which he does not respond to questions. An EEG demonstrates a 3-Hz spike-and-wave pattern. Which of the following is the most appropriate treatment?
a. Methylphenidate (Ritalin)
b. Ethosuximide (Zarontin)
c. Clonidine (Catapres)
d. Fluoxetine (Prozac)
e. Carbamazepine (Tegretol)

10. A 28-year-old woman comes to the ER with a severe unilateral throbbing headache accompanied by photophobia and phonophobia. These headaches started in her teens and she has one every month. Which of the following medications is effective as abortive treatment?
a. Propranolol
b. Sumatriptan
c. Verapamil
d. Amitriptyline
e. Valproic acid

11. A 42-year-old man is brought to the neurologist for evaluation of a few months' history of personality changes. His family indicates that, over the previous year, he has made unusual movements with his hands, and he seems to have some memory difficulties. His father died in his 50s with a similar clinical syndrome, including prominent chorea and dementia. The most likely genetic abnormality will be localized on chromosome:
a. 19
b. 6
c. 4
d. 11
e. 21

12. A 55-year-old woman with a history of ovarian cancer and moderate alcohol consumption is seen in the Neurology ambulatory clinic with a 1-month history of progressive unsteadiness of gait and dysarthria. Examination confirms the presence of both gait and limb ataxia as well as nystagmus. These symptoms were fairly abrupt in onset, progressed over a period of a few weeks, and now appear to have stabilized, but there has been no sign of spontaneous improvement. Which of the following statements is correct?
a. The findings of gait ataxia, dysarthria, and nystagmus indicate diffuse involvement of the cerebellum and suggest that alcohol consumption is the likely cause.
b. The constellation of symptoms and their temporal evolution are most consistent with paraneoplastic cerebellar degeneration, a disorder associated with underlying gynecologic malignancy.
c. The constellation of symptoms and their temporal evolution are most consistent with paraneoplastic cerebellar degeneration (PCD), but ovarian cancer is an unusual cause of this syndrome.
d. The symptoms and signs indicate cerebellar hemispheric dysfunction and are most suggestive of a metastasis from the underlying ovarian cancer.
e. The sudden onset of symptoms and diffuse cerebellar involvement suggest midline cerebellar hemorrhage as the cause.

13. A 45-year-old man with multiple sclerosis (MS) comes to the Neurology clinic complaining of urinary incontinence. He indicates that he experiences increased urgency and frequency of urination. The most likely urodynamic finding in this patient is:
a. An atonic bladder
b. A spastic bladder
c. Stress incontinence
d. Absence of abnormalities
e. Overflow incontinence

14. A 65-year-old man complains of a 3-month history of intermittent urinary incontinence. Urodynamic studies

show an atonic bladder. Which of the following is most likely responsible for his problem?
a. Diabetes
b. Old stroke
c. Multiple sclerosis
d. Right parietal tumor
e. Pineal tumor

15. A 35-year-old man is seen in the Neurology outpatient clinic with the complaint that his fingers occasionally "get stuck" when he tries to open jars. On examination you find subtle weakness of the fingers and toes as well as percussion myotonia. You suspect the diagnosis of myotonic dystrophy. Which of the following statements is true?
a. Myotonic dystrophy is a systemic disorder that may also cause cataracts, diabetes, mental retardation, and cardiac arrhythmias.
b. Myotonic dystrophy is primarily a disorder of skeletal muscle.
c. Myotonic dystrophy should not be considered in the differential diagnosis because it is an inherited disorder that typically manifests itself either at birth or early in life.
d. Myotonic dystrophy is an autosomal recessive disorder caused by a triplet expansion in the DMPK gene.
e. Electromyography is usually normal in myotonic dystrophy and genetic testing is essential to confirm the diagnosis.

16. An 18-year-old woman is brought to the Neurology clinic by her mother, who explains that her daughter has been behaving strangely recently and appears to have paranoid delusions. She has a slight tremor of both hands and the examiner notes that there is a brownish discoloration of the cornea in the vicinity of the limbus. Laboratory studies show a mild transaminitis. Which of the following test results are most likely?
a. Increased serum copper and ceruloplasmin with decreased 24-hour urinary copper
b. Increased serum copper with decreased serum ceruloplasmin and increased 24-hour urinary copper
c. Decreased serum copper and ceruloplasmin and increased 24-hour urinary copper
d. Increased 24-hour urinary copper but decreased copper staining on liver biopsy
e. Increased serum copper and 24-hour urinary copper with decreased serum ceruloplasmin

17. A 40-year-old woman with SLE develops weakness of her right finger and wrist extensors and pain on the right dorsum of her hand several months after being diagnosed with left carpal tunnel syndrome and right sciatic neuropathy. What is the most likely diagnosis?
a. Mononeuropathy multiplex
b. Axonal polyneuropathy
c. Demyelinating polyneuropathy
d. Neuromuscular junction disease
e. Polyradiculopathy

18. A 70-year-old man develops the acute onset of an inability to speak. Examination reveals that he struggles to pronounce a complete word and cannot string words together. He is unable to repeat a sentence but can follow simple and multistep commands. What is the most likely diagnosis?
a. Global aphasia
b. Conduction aphasia
c. Broca aphasia
d. Wernicke aphasia
e. Transcortical motor aphasia

19. A 28-year-old woman is brought to the ER by her husband. In addition to having neck stiffness, she has had a fever for several days, has been somewhat confused, and has not been "acting like herself." LP shows 9 white blood cells with a lymphocytic predominance, 32 red blood cells, protein = 63, and glucose = 65. Gram stain is negative. What is the most likely diagnosis?
a. Bacterial meningitis
b. Viral meningitis
c. Fungal meningitis
d. Meningitis from tuberculosis
e. Subarachnoid hemorrhage

20. A healthy 32-year-old man is brought to the ER after he stopped speaking suddenly, fell to the ground, lost consciousness, and shook for 2 minutes. After the event, he was noted to have a tongue laceration and urinary incontinence. He has no history of similar events. His physical exam shows a mild right hemiparesis. Routine labs and a head CT are normal. An EEG performed the next day is normal. The normal EEG suggests that this man:
a. Had a pseudoseizure and does not require anticonvulsants.
b. Needs admission for long-term video-EEG monitoring.
c. Probably had a seizure, and the normal EEG result is not surprising.
d. Requires hyperventilation to elicit an absence seizure on EEG.
e. Has actually had an ischemic stroke rather than a seizure.

21. Two days after coronary artery bypass surgery, a 62-year-old man with hypertension complains that "there is another man's arm in bed" with him. When asked to hold up

his arms, the patient raises his right arm only. When asked about his left arm, he claims it is the examiner's or another patient's. What is the most likely diagnosis?
a. Right hemispheric stroke with neglect
b. Left hemispheric stroke with neglect
c. Conversion disorder
d. Adjustment disorder
e. Alien-limb phenomenon

22. A 35-year-old man with no known history of seizures is brought in by paramedics in status epilepticus. Which of the following medications is used as initial therapy for this condition?
a. Benzodiazepines
b. Barbiturates
c. Propofol
d. Carbamazepine
e. Lamotrigine

23. Subfalcine herniation most often results in which of the following?
a. Ipsilateral third nerve palsy
b. Contralateral third nerve palsy
c. Ipsilateral hemiparesis
d. Contralateral hemiparesis
e. Bilateral leg weakness

24. A 23-year-old woman presents with loss of vision in the right eye accompanied by slight pain in that eye over a period of 3 days. She has 20/200 acuity, red desaturation, and an afferent pupillary defect in the right eye. The remainder of her examination and MRI are normal. A diagnosis of optic neuritis is made. Which of the following is true about treatment?
a. Interferon beta-1b will hasten recovery from this episode.
b. Mitoxantrone is the most effective treatment.
c. Medical treatment is not likely to help optic neuritis.
d. Oral corticosteroids are preferred for the treatment of optic neuritis.
e. Corticosteroids may delay the development of multiple sclerosis.

25. A 22-year-old right-handed woman develops horizontal diplopia acutely. Your evaluation shows normal right lateral gaze but difficulty with adduction of the right eye while looking to the left and nystagmus in the abducting left eye. What is the most likely anatomic localization?
a. Left paramedian pontine reticular formation (PPRF) producing a right internuclear ophthalmoplegia (INO)
b. Right PPRF producing a right one-and-a-half syndrome
c. Right medial longitudinal fascicle (MLF) producing a right INO
d. Left medial longitudinal fascicle producing a right INO
e. Lateral geniculate nuclei

26. A 55-year-old man with a history of tick bite and erythema chronicum migrans is diagnosed with Lyme disease. He asks his primary care physician about neurologic symptoms he should watch for. Which of the following is an early neurologic manifestation of Lyme disease?
a. Facial nerve palsy
b. Painful polyradiculopathy
c. Spinal cord compression
d. Leukoencephalopathy
e. Generalized epilepsy

27. A 34-year-old man is seen in the Neurology outpatient clinic with symptoms of headache and bilateral lower motor neuron (LMN) facial weakness. There is no history of a skin rash. Neurologic examination discloses a relative afferent papillary defect in the right eye as well as the bilateral facial weakness.
a. Guillain-Barré syndrome (GBS) is the most likely diagnosis, and he should have an LP to help confirm the diagnosis.
b. Lyme disease is the most likely diagnosis, and *Borrelia* serology should be sent to confirm the diagnosis.
c. Sarcoidosis is likely the correct diagnosis, and appropriate investigations include MRI of the brain, LP, and chest x-ray.
d. Multiple sclerosis (MS) is most likely the correct diagnosis, as bilateral facial weakness and optic neuropathy are both common manifestations of this disease.
e. Vasculitis, causing mononeuritis multiplex, is the most likely diagnosis, and so the patient should be referred for rheumatologic evaluation.

28. A 64-year-old man with a history of hypertension presents to the ER with the sudden onset of numbness of his left leg, arm, and face. His motor examination is normal. What is the most likely site of his lesion?
a. Right thalamus
b. Left thalamus
c. Left postcentral gyrus
d. Right precentral gyrus
e. Right corona radiata

29. While playing baseball with some friends, a 15-year-old boy, who was not wearing a helmet, was hit accidentally on the side of the head with a ball. He was knocked unconscious briefly but recovered fully. Two hours later he became increasingly lethargic, so his parents brought him to the ER. When you evaluate the patient, he is barely arousable to your voice. He has mild weakness on the left side of his body, and his right pupil is slightly larger than the left pupil; the right pupil does not appear to react to light. What is the most likely cause of the patient's symptoms and signs?
a. Concussion
b. Epidural hematoma

c. Diffuse axonal injury
d. Ischemic infarct
e. Drug intoxication

30. Which of the following syndromes or diseases could cause bilateral weakness and loss of pain and temperature sensation with preservation of joint position sense in both legs?
a. Amyotrophic lateral sclerosis (ALS)
b. Vitamin B_{12} deficiency
c. Brown-Séquard syndrome
d. Anterior spinal artery syndrome
e. Tabes dorsalis

31. A 2-year-old child presents with new seizures. Her mother tells you that the child is not walking yet. He has a 5-year-old brother with a seizure disorder and mental retardation. On examination, using the Wood's lamp, you find hypomelanotic lesions. The most appropriate next test is:
a. Skeletal surveillance
b. Skin biopsy
c. Head CT or MRI
d. No need for further tests
e. LP

32. A 3-year-old boy is brought to his pediatrician for evaluation of repetitive behaviors, delay of language development, and social isolation. He otherwise has normal motor development. Which of the following is a required feature of autism but not of Asperger syndrome?
a. Abnormal language development
b. Social isolation
c. Onset after age 3
d. Failure to meet milestones for gross motor development
e. Failure to meet milestones for fine motor development

33. A 62-year-old woman with a history of small cell lung carcinoma presents to the Neurology clinic complaining of bilateral paresthesias of the lower extremities. She has no history of diabetes or family history of polyneuropathy. She describes severe pain in the soles of her feet when standing and has difficulty walking. On examination, there is severe pain to light touch over both soles. On your sensory examination description, you will state that this patient has:
a. Hyperesthesia
b. Paresthesia
c. Allodynia
d. Sensory loss
e. Hypesthesia

34. A 38-year-old man presents to the ER complaining of mild headache. He had neck trauma a week earlier. The exam shows anisocoria, with the right pupil being 3 mm and the left 5 mm, both reactive to light. What other findings will help to localize the lesion?
a. Look at the pupils in the dark and check tongue deviation.
b. Look for evidence of ptosis in the left eye and anhidrosis on the left face.
c. Look for evidence of ptosis in the right eye and anhidrosis in the right face.
d. Look for evidence of horizontal diplopia and a cut in the right visual field.
e. Look for evidence of dysarthria and hemiparesis.

35. A 35-year-old woman presents to the ER reporting a few days of progressive ascending muscle weakness. She had a viral infection a few weeks earlier. On examination, you find diffuse weakness and areflexia. The most likely finding in the CSF is:
a. High protein–high cell count
b. High protein–low cell count
c. Low protein–high cell count
d. Low protein–low cell count
e. Normal CSF

36. A 33-year-old man is seen in the ER for difficulty walking. He has paresthesias in his feet and a left footdrop. Initial physical exam shows mild distal weakness in both legs, with absent ankle jerks and reduced reflexes throughout. While the patient is waiting in the ER, his weakness worsens, involving the arms, but he has no difficulty breathing. You want to admit the patient to the intensive care unit. What will be your best argument to convince your ER attending to do so?
a. Absence of upper extremity reflexes
b. Decreased gag reflex
c. A forced vital capacity FVC below 25 mL/kg
d. The patient's weakness is worsening very quickly, and you fear that he may need mechanical ventilation
e. You don't have an argument in this case

37. A 35-year-old man who is HIV-positive presents with radicular pain in the legs and associated bladder distention. The most likely agent responsible for these symptoms is:
a. Cytomegalovirus
b. *Clostridium*
c. *Toxoplasma*
d. *Cryptococcus*
e. *Pneumocystis carinii*

38. A 45-year-old woman presents to the ER with "dizziness," by which she means that she feels a spinning sensation. The sensation is intermittent and seems to be exacerbated by head movement. She has some nausea with the episodes but otherwise has no other symptoms, such as double vision, weakness, hearing loss, tinnitus, or difficulty swallowing. What diagnosis is most likely?
a. Vestibular neuronitis
b. Ménière disease

c. Brainstem infarction
d. Benign positional paroxysmal vertigo (BPPV)
e. Cerebellar infarction

39. A 32-year-old woman is seen in the Neurology outpatient clinic with symptoms of diplopia and ptosis that fluctuate during the course of the day. Examination shows fatigable proximal weakness. You suspect that she has myasthenia gravis (MG). Which of the following statements concerning MG is true?
a. It is an autoimmune disorder caused by antibodies that are directed against presynaptic nicotinic acetylcholine receptors.
b. It is an autoimmune disorder caused by antibodies directed against postsynaptic muscarininc acetylcholine receptors.
c. It is an autoimmune disorder caused by antibodies directed against presynaptic voltage-gated calcium channels.
d. It is an autoimmune disorder caused by antibodies directed against postsynaptic nicotinic acetylcholine receptors.
e. It is an autoimmune disorder caused by antibodies directed against the synaptic enzyme acetylcholinesterase.

40. A patient complains of difficulty chewing. On examination he is found to have decreased strength of his muscles of mastication. Which of the following cranial nerves is responsible for this motor function?
a. Trigeminal
b. Facial
c. Oculomotor
d. Glossopharyngeal
e. Hypoglossal

41. The following patients are being evaluated in a neurologic intensive care unit. For which one would the Glasgow Coma Scale (GCS) be used most commonly to follow his or her clinical status?
a. A 75-year-old man in coma after cardiac arrest
b. A 29-year-old woman with delirium after medication overdose
c. A 69-year-old woman with a thromboembolic stroke and Broca aphasia
d. A 20-year-old man who is unresponsive after head trauma
e. A 59-year-old man with subarachnoid hemorrhage after aneurysm rupture

42. A 68-year-old man taking warfarin falls while in the hospital, is found on the floor, and is difficult to rouse. He has a new right hemiparesis and an intracranial hemorrhage is suspected. What is the most appropriate initial radiologic study?
a. Head CT with contrast
b. Head CT without contrast
c. Skull x-ray
d. Cerebral angiography
e. Brain perfusion scan

43. A 75-year-old man presents to your office with a 1-month history of progressive pain in the left temporal area and pain in his jaw while eating. On laboratory testing, the patient is found to have an elevated ESR of 94. What is the treatment of choice?
a. Sumatriptan
b. Carbamazepine
c. Verapamil
d. Surgical resection of brain tumor
e. Prednisone

44. A 35-year-old man presents with his spouse to your office for difficulty concentrating. Further history also reveals that he has fallen asleep while driving as well as in the middle of important business meetings, despite sleeping at least 8 hours each night. He denies hallucinations or a history of his knees buckling while laughing. His wife reports that he snores loudly while sleeping. His examination is normal with the exception of moderate obesity. Which of the following tests would be most helpful in diagnosing this patient's disorder?
a. Multiple sleep latency test (MSLT)
b. EEG
c. MRI of the brain
d. LP
e. Polysomnography

45. A previously healthy 21-year-old presents to the ER after being involved in a high-speed motor vehicle accident. You note that the patient is unresponsive, makes no spontaneous movement, and has a dilated pupil on the right that is nonreactive to light. What is the best explanation for these signs?
a. Infarction of the left occipital lobe
b. Concussion from the motor vehicle accident
c. Uncal herniation
d. Cervical neck fracture
e. Diffuse axonal injury

46. An 84-year-old man is transferred from another hospital with a reported hypertensive hemorrhage. The films from that hospital are not available, and there are no further details. Which of the following is the most likely location of his hemorrhage?
a. Pons
b. Midbrain
c. Internal capsule

d. Frontal lobe
e. Corpus callosum

47. A 28-year-old man was recently diagnosed with obstructive sleep apnea. Of the following choices, which is the most appropriate treatment?
a. Pemoline
b. Methylphenidate
c. CPAP
d. Benzodiazepine
e. Clomipramine

48. A 45-year-old man with a prior history of migraine headaches with aura presents to the ER complaining of a progressive headache for the last month that is different from his usual migraine. There is no associated nausea or vomiting. His neurologic examination is completely normal. Your next step in management should be:
a. Brain imaging study
b. Abortive migraine treatment
c. Preventive migraine treatment
d. Reassurance and discharge home
e. Administration of pure oxygen

49. Which of the following features is most commonly associated with a pituitary adenoma?
a. Homonymous hemianopia
b. Bitemporal hemianopia
c. Ring enhancement on brain imaging with contrast
d. Seizures
e. Hemiparesis

50. A 24-year-old construction worker falls from a ladder and fractures his cervical spine with resulting signs of upper motor neuron (UMN) dysfunction. Which of the following signs is characteristic of a UMN lesion?
a. Hypotonia
b. Decreased reflexes
c. Flexor plantar response
d. Spasticity
e. Absent reflexes

51. A 67-year-old woman presents to the ER with a new onset of headache, nausea, vomiting, and unsteadiness of gait. Her history is significant for atrial fibrillation, for which she is chronically anticoagulated with warfarin. She also has a pacemaker in place. You are concerned about the possibility of a cerebellar hemorrhage. The imaging modality of choice is:
a. A CT scan, because this is the imaging modality most sensitive to the presence of acute intracranial blood.
b. An MRI, because blood in the posterior fossa will not be visualized on CT.
c. An MRI, because CT is contraindicated by the presence of a pacemaker.
d. A CT scan, because it provides the best images of the contents of the posterior fossa.
e. A SPECT scan to show metabolic activity in the cerebellum.

52. A 22-year-old woman presents with acute bilateral facial nerve palsy and intermittent peripheral nerve symptoms for over 3 weeks. You find elevated Lyme titers in serum and CSF. What treatment would you choose first?
a. Oral doxycycline
b. Intravenous ceftriaxone
c. Oral amoxicillin
d. Oral amoxicillin and doxycycline
e. Fluconazole

53. A 58-year-old man is seen in the Neurology ambulatory clinic with a 3-month history of right-sided resting tremor. On examination, he is noted to have mild masking of facial expression and there is diminished swing of the right arm when he walks. You suspect that he may have early idiopathic Parkinson disease. Which of the following statements concerning this disorder is true?
a. Most cases are familial with mutations in the α-synuclein or parkin genes.
b. It is characterized by the death of dopaminergic neurons in the substantia nigra pars reticulata.
c. The four cardinal features of this disorder are tremor, rigidity, bradykinesia, and postural instability.
d. Impairment of vertical gaze is a common manifestation of this disorder.
e. Early falls are a common problem in this disorder.

54. A 5-year-old boy is seen in the Pediatric Neurology clinic. His motor milestones have been delayed, and examination discloses proximal muscle weakness with difficulty arising from the floor. There is pseudohypertrophy of his calf muscles. He has an older brother with Duchenne muscular dystrophy (DMD) who is confined to a wheelchair. Which of the following statements concerning DMD is true?
a. It is an autosomal recessive disorder caused by mutation in the dystrophin gene.
b. It is an autosomal dominant disorder caused by mutation in the dystrophin gene.
c. DMD and limb-girdle muscular dystrophy are allelic disorders, both being due to mutations in the dystrophin gene.
d. It is a disorder caused by mutation in the dystrophin gene, which is located on the X chromosome.
e. DMD and Becker muscular dystrophy are allelic disorders, due to mutations in the dystrophin gene on chromosome 4.

55. A 9-year-old boy presents with difficulty walking. Neurologic examination demonstrates, among other things, that he performs rapid alternating movements poorly, with a lack of proper rhythm and coordination. This finding, called dysdiadochokinesis, is most typically associated with dysfunction of which of the following brain structures?
a. Basal ganglia
b. Medulla
c. Cerebellum
d. Parietal lobe
e. Thalamus

56. An ischemic stroke involving the right side of the pons could lead to which of the following patterns of weakness?
a. Left facial weakness and right body weakness
b. Right facial weakness and left body weakness
c. Right facial weakness and right body weakness
d. Left arm weakness and right leg weakness
e. Right arm weakness and left leg weakness

57. A 27-year-old woman with complex partial seizures is well controlled on carbamazepine. Which of the following is a characteristic side effect of this medication?
a. Thrombocytopenia
b. Agitation
c. Diabetes insipidus
d. Nephrolithiasis
e. Hyponatremia

58. A 53-year-old construction worker is brought to the ER with a severe, sudden-onset headache accompanied by vomiting. A CT scan of his head demonstrates a subarachnoid hemorrhage. Which of the following is a common cause of subarachnoid hemorrhage?
a. Tearing of bridging veins
b. Laceration of the middle meningeal artery
c. Aneurysmal rupture
d. Amyloid angiopathy
e. Arteriovenous malformation rupture

59. A 45-year-old woman has an MRI scan of the brain for evaluation of progressive headaches. The MRI scan shows a lesion that enhances in a homogeneous manner with contrast administration. Which of the following lesions is most likely to account for the appearance of the MRI scan?
a. Glioblastoma multiforme
b. Meningioma
c. Brain abscess
d. Toxoplasmosis
e. Granuloma

60. A 75-year-old man is brought to the ER after having lost consciousness briefly in his bathroom. By the time he arrives he is feeling fine and is able to give a clear account of what happened. He recalls walking to the bathroom to urinate. Shortly thereafter he became light-headed and felt as if his vision were graying out. These symptoms lasted for about 30 seconds. The next thing he recalls is awakening on his bathroom floor. His wife notes that he was unconscious only briefly. Which of the following descriptions pertinent to this clinical scenario is correct?
a. The symptoms of light-headedness and graying out of vision are atypical symptoms described by patients with syncope.
b. He has micturition syncope.
c. Orthostatic hypotension is the likely explanation for his syncopal episode.
d. Vasovagal syncope is the likely explanation for his syncopal episode.
e. Vestibular neuronitis is the likely explanation for his symptoms.

61. A 36-year-old man comes to the ER with a 4-day history of fever and a generalized unwitnessed seizure 2 hours earlier. MRI of the brain with gadolinium shows contrast enhancement of both temporal lobes and, in a nontraumatic tap, 10,000 RBCs and 15 white blood cells. What is the most likely organism responsible for this clinical picture?
a. Enterovirus
b. *Streptococcus* species
c. *Cryptococcus neoformans*
d. Herpes simplex virus (HSV-1)
e. *Meningococcus*

62. A 55-year-old woman comes to the Neurology clinic complaining of numbness in the last two fingers of her right hand; it tends to worsen at night. On examination, you find a positive Tinel sign at the right elbow (percussion of the ulnar nerve at the right elbow produces a tingling sensation in the last two fingers of the right hand). You are convinced that this is an ulnar neuropathy at the right elbow and perform electrodiagnostic studies. Why do you think that this is a peripheral nerve problem?
a. The acuteness of presentation
b. The physical examination findings
c. The symptoms described by the patient
d. You don't think this is a peripheral nerve problem.
e. There is no CNS complaint.

63. A 25-year-old man is now comatose after suffering blunt-force trauma to the head. On the basis of the clinical history, neurologic exam, and head CT scan, he is diagnosed with an epidural hematoma. Of the following choices, which is the best treatment option?
a. Neurosurgical decompression
b. Hyperventilation
c. Administration of mannitol

d. Conservative management with close monitoring of vital signs and neurologic status
e. Administration of tissue plasminogen activator

64. A 19-year-old man is admitted to a Neurology service with an episode of transverse myelitis. Workup includes an MRI of his head and LP. Which of the following distinguishes acute disseminated encephalomyelitis (ADEM) from multiple sclerosis (MS)?
a. Presence of oligoclonal bands in the CSF
b. Pleocytosis with neutrophilic predominance
c. Monophasic course
d. Multiple lesions on MRI
e. A positive family history of ADEM

65. A 55-year-old man with type 2 diabetes presents with a 5-week history of pain in his right knee, followed by weakness and atrophy of his right quadriceps. Exam shows weakness of the right quadriceps and iliopsoas muscles and an absent right knee jerk. This presentation is most characteristic of what?
a. Diabetic distal symmetric polyneuropathy
b. Proximal diabetic neuropathy or diabetic amyotrophy
c. Mononeuropathy multiplex
d. Stroke
e. These conditions are not seen in diabetics.

66. A 19-year-old man is accidentally hit on the left side of the head with a baseball bat while playing a game with some friends. He loses consciousness and is taken to an emergency room. A head CT scan showed a lenticular shaped hyperdensity in the epidural space over the left temporal region that is exerting some mild mass effect on the brain. Which of the following mechanisms best explains the patient's head CT scan results?
a. Tearing of bridging veins
b. Laceration of the middle meningeal artery
c. Impact of the brain over bony prominences of the skull
d. Rotational acceleration and deceleration of the head
e. Rupture of a cerebral aneurysm

67. A 56-year-old woman is referred to the Neurology clinic by her optometrist, who noted that she had limited movement of her eyes. The patient herself notes only that she has fallen a few times in recent months. Examination confirms that there is marked limitation of vertical eye movements (both up and down gaze). There is mild rigidity in both arms and legs but no tremor. Her postural reflexes are poor. Which of the following is the most likely diagnosis?
a. Parkinson disease (PD)
b. Progressive supranuclear palsy (PSP)
c. Corticobasal ganglionic degeneration
d. Miller-Fisher syndrome (MFS)
e. Chronic progressive external ophthalmoplegia (PEO)

68. A 65-year-old obese woman is referred to the Neurology clinic with complaints of burning pain in both feet, which has been present for several months. You suspect that she may have a small fiber peripheral neuropathy. The most likely findings on examination are:
a. Symmetric weakness and atrophy of intrinsic muscles of the feet with loss of ankle reflexes
b. Symmetric stocking pattern diminution of pinprick and temperature sensation
c. Symmetric stocking pattern diminution of vibration and joint position sense with absent ankle reflexes
d. Symmetric stocking pattern diminution of all sensory modalities with absent DTRs in the arms and legs
e. Symmetric stocking pattern diminution of vibration and joint position sense with retained ankle reflexes

69. A 45-year-old man presents with a several-month history of weakness in his lower and upper extremities. On examination, in addition to weakness in multiple muscle groups, he demonstrates atrophy, hyperreflexia, spasticity of the legs, and bilateral Babinski signs. Fasciculations in multiple muscles are also noted. His sensation to pain, temperature, and joint position sense appear intact. What is his most likely diagnosis?
a. Amyotrophic lateral sclerosis (ALS)
b. Vitamin B_{12} deficiency
c. Anterior spinal artery syndrome
d. Central cord syndrome
e. Brown-Séquard syndrome

70. A 25-year-old man presents to your office with excessive daytime sleepiness, visual hallucinations while falling asleep, and a history of transiently losing tone in his extremities and falling to the ground when he is angry or laughing. Of the following choices, what would be his single best treatment option?
a. Pemoline
b. Venlafaxine
c. Clomipramine
d. CPAP
e. Methylphenidate

71. A 54-year-old man is seen in the Neurology clinic with complaints of resting tremor of the left hand and a general feeling of slowing down. As an example, he explains that it takes him at least 20 minutes to get dressed in the morning. You suspect that he has idiopathic Parkinson disease (PD). If you are correct, examination would be most likely to show which of the following combinations of physical signs?
a. Asymmetric rest tremor, asymmetric rigidity, and poor postural reflexes
b. Symmetric rest tremor, asymmetric rigidity, and poor postural reflexes
c. Asymmetric rest tremor, symmetric rigidity, and poor postural reflexes

d. Symmetric rest tremor and rigidity and poor postural reflexes
e. Asymmetric rest tremor, symmetric rigidity, and impairment of vertical gaze

72. A 55-year-old man with a history of hypertension is seen in the ER with complaints of clumsiness and incoordination; these began 2 days earlier and have increased in severity. He also reports double vision on lateral gaze, which resolves when one eye is covered. He is awake, alert, and oriented. Examination shows restricted eye movements in all directions, with eye abduction in both directions most limited. DTRs are absent, and there is impaired joint position sense. The most likely diagnosis is:
a. Brainstem stroke
b. Cerebellar infarction with compression of the brainstem
c. Miller-Fisher syndrome (MFS)
d. Myasthenia gravis (MG)
e. Alcoholic cerebellar degeneration

73. A 44-year-old woman presents to the ER complaining of urinary incontinence and lower back pain. What will be the most useful next diagnostic procedure to try in the effort to find the etiology of her problem?
a. Urodynamic studies
b. Blood testing including glucose level
c. MRI of the spine
d. Post-void residual
e. LP

74. A 48-year-old woman reports recurrent episodes of stabbing unilateral pain associated with tearing and conjunctival injection. Which of the following is characteristic of chronic paroxysmal hemicrania as opposed to cluster headache?
a. Unilateral pain
b. Conjunctival injection
c. Male predominance
d. Indomethacin responsivity
e. Headache duration of hours

75. A 75-year-old right-handed man with hypertension, diabetes, and hypercholesterolemia is seen in the ER. His family explains that he has had difficulty doing things around the house for the last few days. The patient himself admits that he has found it difficult to get dressed and to prepare his breakfast, but he feels healthy otherwise. On examination his speech is fluent, and he is able to name objects and repeat short phrases without difficulty. He is, however, unable to mimic certain activities described by the examiner, although he seems to have no difficulty understanding what it is that he is supposed to do.
a. He likely has a form of Wernicke aphasia due to a lesion in the left superior temporal lobe.
b. He likely has a form of apraxia due to a lesion in the right frontal lobe.
c. He likely has a form of Wernicke aphasia due to a lesion in the left inferior frontal lobe.
d. He likely has a form of apraxia due to a lesion in the right parietal lobe.
e. He likely has a form of apraxia due to a lesion in the left parietal lobe.

76. The most reliable method for distinguishing between a "traumatic" spinal tap (lumbar puncture) and a subarachnoid hemorrhage is the presence of:
a. Increased opening pressure
b. Increased red cell count
c. Increased white cell count
d. Xanthochromia
e. Pain upon needle insertion

77. The magnetic resonance imaging sequence that is most sensitive for the presence of blood breakdown products (e.g., after a hemorrhage) is:
a. T1
b. T2
c. Contrast-enhanced T1
d. FLAIR
e. Susceptibility

78. Which of the following disorders is most closely associated with REM sleep behavior disorder?
a. Alzheimer disease
b. Multiple sclerosis
c. Multiple system atrophy
d. Myasthenia gravis
e. Primary lateral sclerosis

79. Which of the following vascular malformations is the most likely to result in an intracranial hemorrhage?
a. Arteriovenous malformation
b. Capillary telangiectasia
c. Cavernous hemangioma
d. Developmental venous anomaly
e. Vein of Galen

80. An 81-year-old right-handed man with hypertension and hypercholesterolemia presents with the sudden onset of a dense right hemiplegia. His language is normal, and he has normal eye movements and pupillary reactions. He has no sensory deficits. What is the most likely localization of his stroke?
a. Left motor cortex
b. Left internal capsule
c. Left thalamus
d. Left midbrain
e. Left lateral medulla

81. In which of the following disorders would the highest elevation of creatine kinase (CK) be expected?
a. Becker muscular dystrophy
b. Duchenne muscular dystrophy
c. Lambert-Eaton myasthenic syndrome
d. Limb-girdle muscular dystrophy
e. Myotonic dystrophy

82. A 68-year-old man with no major medical problems attends his annual visit with his primary care physician. The doctor wishes to perform a brief screening neurologic examination. Which of the following parts of the neurologic exam would be the most sensitive for the detection of potential abnormalities in multiple different parts of the nervous system?
a. Deep tendon (muscle stretch) reflexes
b. Gait evaluation
c. Visual field examination
d. Joint position sense testing
e. Total weight-lifting capacity

83. A 78-year-old woman with a history of coronary artery disease and hypercholesterolemia develops the sudden onset of paralysis of all four extremities. On examination her eyes are open, she appears alert, and she can consistently respond to complex questions and commands by blinking her eyes but otherwise has minimal facial movement and no movement of the extremities. This condition is best described as:
a. Locked-in syndrome
b. Persistent vegetative state
c. Brain death
d. Coma
e. Stupor

84. A 30-year-old man is found to have increased intracranial pressure after a head injury. Which of the following treatments can serve to lower intracranial pressure?
a. Lowering the head of the bed
b. Intravenous fluid load
c. Depression of respiratory rate
d. Mannitol
e. Basilar artery stent

85. An 80-year-old man has developed gradually worsening memory over the past several years. His wife also reports that he appears to have vivid visual hallucinations at times, and his alertness has been fluctuating on a day-to-day basis. Examination demonstrates bradykinesia and rigidity in the extremities, without any dyskinesias. What is the most likely diagnosis?
a. Alzheimer disease
b. Dementia with Lewy bodies
c. Vascular dementia
d. Huntington disease
e. Progressive supranuclear palsy

86. A 71-year-old with a clinical diagnosis of Alzheimer disease comes to autopsy after a fatal motor vehicle accident. Which of the following is a neuropathological hallmark of Alzheimer disease?
a. Lewy bodies in the substantia nigra
b. Lewy bodies in cortical neurons
c. Prominent atrophy of caudate
d. Spongiform changes in cortex
e. Neurofibrillary tangles

87. A 34-year-old man comes for neurologic consultation because of paroxysmal episodes of speech difficulty that have occurred recently. The best way to distinguish whether these are seizures or other types of events is:
a. History
b. Neurologic examination
c. Brain MRI
d. Routine EEG
e. Empiric anticonvulsant trial

88. A 16-year-old college student is seen in the emergency room for fever, confusion, and headache. Lumbar puncture is performed. Which of the following cerebrospinal fluid profiles is most consistent with acute bacterial meningitis?
a. Normal white blood cell count, high protein, low glucose
b. Normal white blood cell count, high protein, high glucose
c. Elevated white blood cell count, high protein, low glucose
d. Elevated white blood cell count, high protein, high glucose
e. Decreased white blood cell count, high protein, high glucose

89. A 55-year-old man presents with headache and right hand weakness. On examination you find bilateral papilledema and upper motor neuron weakness in the right arm and leg. MRI with contrast shows an enhancing mass in the left frontal region, crossing the corpus callosum in a "butterfly" pattern with surrounding edema. Prior to requesting a biopsy, you discuss with your resident that this is most likely a:
a. Meningioma
b. Astrocytoma
c. Glioblastoma
d. Ependymoma
e. Schwannoma

90. You evaluate a 22-year-old woman complaining of visual problems. Your examination shows bitemporal visual field defects. Where is the lesion?
a. Right optic nerve
b. Right occipital lobe
c. Left optic radiation
d. Optic chiasm

e. This visual field defect is nonphysiologic, suggesting a psychiatric explanation.

91. You are asked to evaluate a 33-year-old construction worker who is complaining of paresthesias in the first and second digits of his right hand. Your physical examination shows no weakness but a mild decrease in light touch over the thumb. You request a nerve conduction study to rule out carpal tunnel syndrome, and it turns out to be normal. On repeated history, the patient indicates that on occasion, he gets a sharp, "electric" pain travelling from his neck to the right hand. What are you missing?
a. A median neuropathy at the wrist
b. A neuromuscular junction disorder affecting distal hand muscles
c. A C8–T1 radiculopathy
d. A lower trunk brachial plexopathy
e. A C6-7 radiculopathy

92. A 75-year-old man underwent surgery to correct a large abdominal aortic aneurysm. The procedure appeared to go well, but you are called a few hours later to evaluate the patient who states that he cannot move or feel his legs. On the way to the ICU, you consider the possible causes of his symptoms and plan your physical examination. What is the most important test to help localize the lesion?
a. MRI of the spine
b. Sensory level
c. Reflexes in lower extremities
d. Plantar flexion reflex
e. Toe position sense

93. A 19-year-old man presents to the emergency department with 2 days of ascending weakness. He had diarrhea 3 weeks earlier. He looks comfortable. On examination, you find moderate weakness in all limbs, with normal strength in facial muscles. You suspect Guillain-Barré syndrome and recommend admission to Neurology. What should be done first?
a. Spinal tap to look for albuminocytologic dissociation
b. Call for an emergency EMG to verify diagnosis
c. Send the patient for a whole spine MRI to rule out cord compression
d. Obtain pulmonary function tests, including FVC
e. Start high-dose steroids and then move to the floor

94. A 55-year-old woman is seen in the emergency room with the complaint that when she rolled over in bed in the morning she felt acutely vertiginous for about 30 seconds, with associated nausea and vomiting. Over the course of the day, the vertigo has recurred, each time precipitated by turning her head. She has no other symptoms and feels well in between the episodes of vertigo. She is known to have poorly controlled diabetes and hypertension. When examined in the emergency room, she is found to have rotatory and downbeating nystagmus during the Dix-Hallpike maneuver with the head tilted one way but not the other. The neurologic examination is otherwise entirely normal. Which of the following is the most likely diagnosis?
a. Meniere disease
b. Cerebellar stroke
c. Benign positional paroxysmal vertigo (BPPV)
d. Perilymph fistula
e. Vestibular neuronitis

95. A 77-year-old woman with a history of migraine in her 20s and 30s is seen by her primary care physician with the complaint that she has experienced headaches again for the first time in many years. Upon further enquiry she reports a sense of generalized fatigue and notes that there is discomfort over her right temple when she brushes her hair. The neurologic examination is normal. Which of the following would be the most appropriate clinical course?
a. Reassurance that her headaches likely represent recurrence of her old migraines. No further investigations are needed
b. MRI of the brain to rule out an intracranial mass lesion, as new-onset headaches in the elderly are commonly caused by raised intracranial pressure
c. Lumbar puncture to rule out high or low pressure headaches
d. ESR, CRP, and temporal artery biopsy, as giant cell arteritis is the most likely diagnosis
e. Explanation that she likely has trigeminal neuralgia, given the distribution of her symptoms over the right temporal region

96. A 63-year-old man with poorly controlled hypertension is brought to the emergency room after being found by his wife in a confused state. He had been fine when she left to go shopping, but when she returned several hours later, she found that he wasn't making any sense when talking. When you examine him you find that his naming is impaired and his verbal fluency is reduced, but other language functions are intact. You note that his speech is soft. Which of the following conclusions is most accurate?
a. The reduced verbal fluency, together with an anomia, is most suggestive of a posterior (Broca) aphasia.
b. The hypophonia, together with the evidence for a language impairment, is most suggestive of a subcortical aphasia.
c. He does not have an aphasia given that his language functions other than verbal fluency and naming are intact.
d. The impaired ability to name objects (anomia) is most suggestive of a conduction aphasia.
e. The soft speech, together with the anomia and reduced verbal fluency, suggests that he has a transcortical motor aphasia.

97. A 27-year-old African-American woman with diabetes presents to the outpatient Neurology clinic with subacute onset of bilateral facial weakness. She explains that the symptoms have developed over the course of the last few days. She also reports having a slightly raised and tender rash over the anterior aspects of both shins. On examination you find bilateral lower motor neuron facial palsy as well as tender erythematous nodules over both shins. Which of the following is the most likely diagnosis?
a. Guillain-Barré syndrome
b. Lyme disease
c. Neurosarcoidosis
d. Diabetes
e. Tuberculosis

98. Ataxia may be a manifestation of which vitamin deficiency?
a. Vitamin A
b. Vitamin B
c. Vitamin C
d. Vitamin D
e. Vitamin E

99. Which of the following are the cardinal features of idiopathic Parkinson disease?
a. Tremor, bradykinesia, rigidity, and postural instability
b. Bradykinesia, dementia, tremor, and rigidity
c. Rigidity, hallucinations, tremor, and postural instability
d. Tremor, rigidity, bradykinesia, and gaze palsy
e. Tremor, autonomic dysfunction, bradykinesia, and rigidity

100. Which of the following statements regarding higher cortical function is true?
a. Aphasia is characterized by a problem with articulation of words.
b. Apraxia is defined by the inability to carry out an automatic or unlearned motor task.
c. Agnosia is an inability to recognize objects through a sensory modality even when the primary sensory modality is unimpaired.
d. Neglect is a form of apraxia in which there is insufficient attention paid to one hemispace.
e. Agraphia is a disorder in which affected individuals are unable to draw pictures.

A

Answers

1. c (Chapter 12)

The presence of visual hallucinations is an early symptom of dementia with Lewy bodies (DLB). Other characteristics include cognitive decline, fluctuations of alertness, extrapyramidal symptoms, and an extraordinary sensitivity to neuroleptics. Visual hallucinations and sensitivity to neuroleptics are not early signs of AD, PD, Pick disease, or vascular dementia.

2. b (Chapter 4)

Decreased visual acuity that does not correct with pinhole testing, an RAPD, and a central scotoma is characteristic of optic nerve disease. A lesion affecting the optic chiasm will produce a bitemporal heteronymous hemianopia. If a lesion affects the optic tract, the optic radiations (in both temporal and parietal areas) or the occipital cortex (unilaterally), it will produce a homonymous hemianopia; that in the occipital cortex may be "macular sparing."

3. d (Chapter 25)

Niemann-Pick disease, Gaucher disease, and Tay-Sachs disease are all associated with cherry-red spots in the macula. Niemann-Pick disease is an autosomal recessive disorder caused by sphingomyelinase deficiency. Alexander disease and Canavan disease are dysmyelinating disorders with prominent macrocephaly but not cherry-red spots. The classic ophthalmologic finding of Hurler syndrome is clouding of the cornea rather than a cherry-red spot in the macula. Krabbe disease is an autosomal recessive disorder caused by galactosylceramide §-galactosidase deficiency. It does not produce a cherry-red spot in the macula.

4. c (Chapter 23)

One possible etiology of sensory neuronopathies is a paraneoplastic disorder, in particular small cell lung cancer. This is generally associated with positive anti-Hu antibodies (antineuronal antibodies) and can also be associated with paraneoplastic encephalomyelitis, ataxia, and autonomic neuropathy. Anti-GM_1 has been associated with multifocal motor neuropathy with conduction block. MRI of the spine would not help at this stage. Other causes of sensory neuronopathy include Sjögren syndrome, pyridoxine intoxication, and chemotherapy (cisplatin). Chest CT to search for occult malignancy is also recommended.

5. d (Chapter 1)

Each cranial nerve courses through a particular foramen, or opening, in the skull. Skull base fractures and other such injuries can result in damage to these structures and injury to the associated cranial nerves. The seventh (facial) and eighth (vestibulocochlear) nerves both course through the internal auditory meatus, which may have been damaged in this woman's case.

6. d (Chapter 2)

This woman has had a subarachnoid hemorrhage. Bleeding into the subarachnoid (CSF) space typically initiates an inflammatory response, one manifestation of which is a lymphocytic pleocytosis. Xanthochromia is the result of breakdown of blood within the subarachnoid space. Its presence in a bloody CSF sample helps to distinguish intrathecal hemorrhage from a traumatic tap. The CSF glucose is typically normal in both subarachnoid hemorrhage and viral meningitis. It is frequently low in bacterial, mycobacterial, and carcinomatous meningitis. An opening pressure of 14 cm H_2O is within the normal range of 6 to 15 cm H_2O.

7. d (Chapter 3)

This patient's clinical presentation suggests increased intracranial pressure (ICP) from a right hemispheric lesion.

The "blown" right pupil suggests that herniation of the right hemisphere has compressed the right oculomotor nerve. Choices A through C are all measures that acutely decrease ICP, while neurosurgery may be needed as a more definitive intervention. Performing an LP in this situation could be dangerous and could actually precipitate worsening herniation.

8. e (Chapter 5)

Symmetric proximal weakness usually suggests a primary muscle problem, as does weakness of neck flexors and extensors. The absence of muscle pain and tenderness does not argue against a primary muscle pathology. The other listed choices would not usually result in this pattern of weakness.

9. b (Chapter 15)

This child likely has absence seizures, which are frequently diagnosed after a teacher or parent notices inattention, "daydreaming," or staring episodes. Absence seizures last a few seconds each, can occur many times a day, and have a classic EEG appearance. Ethosuximide and valproic acid are typical drugs of choice.

10. b (Chapter 10)

This woman is suffering from a migraine headache. Sumatriptan is effective as abortive treatment. The other medications are effective in decreasing the severity and frequency of attacks and are used as preventive therapy.

11. c (Chapter 12)

This case represents an early onset of dementia with associated personality changes and movement disorder (chorea)—the classic triad of HD. HD is linked to chromosome 4p16.3, also known as the HD gene, encoding for a protein called huntingtin. The mutation produces an unstable CAG repeat sequence with more than 40 repeats. HD is not linked to the other chromosomes listed.

12. b (Chapter 8)

PCD is typically a pancerebellar syndrome with clinical manifestations including ataxia, dysarthria, and nystagmus. The underlying malignancy is typically a gynecologic one or breast cancer. The temporal evolution is typically that of acute or subacute onset with fairly rapid progression over weeks to months, followed by stabilization. Metastatic cerebellar disease would more likely affect a cerebellar hemisphere and produce lateralized cerebellar dysfunction. Alcoholic cerebellar degeneration typically affects the vermis, and the characteristic manifestation is that of a gait ataxia. Stroke (ischemic or hemorrhagic), although abrupt in onset, would not be expected to progress over a period of weeks to months.

13. b (Chapter 9)

MS characteristically produces an upper motor neuron bladder or spastic bladder with increased frequency and urgency. Stress incontinence is an involuntary loss of urine during coughing, sneezing, laughing, or other physical activities that increase intra-abdominal pressure. An atonic bladder is characterized by overflow incontinence and increased capacity and compliance.

14. a (Chapter 9)

Atonic bladder implies an LMN lesion at the level of the conus medullaris, cauda equina, sacral plexus, or peripheral nerves. It is characterized by overflow incontinence and increased capacity and compliance. Diabetes is the only one in the group able to produce that type of lesion.

15. a (Chapter 24)

Myotonic dystrophy is a multisystem disorder that may also cause frontal balding, diabetes, and gastrointestinal symptoms. It is the most common adult-onset muscular dystrophy. It is inherited in an autosomal dominant fashion and is caused by a triplet repeat expansion in the DMPK gene. EMG typically shows myotonic discharges.

16. b (Chapter 16)

She likely has Wilson disease, an autosomal dominant disorder of copper metabolism that presents with neuropsychiatric symptoms as well as a movement disorder. The pigment changes in the cornea are Kayser-Fleischer rings and are characteristic of Wilson disease.

17. a (Chapter 5)

The patient's current symptoms are suggestive of a right radial neuropathy. Multiple sequential mononeuropathies, each affecting a single peripheral nerve, are known as mononeuropathy multiplex. Pain is a typical feature. Patients with rheumatologic conditions are susceptible; vasculitis may be involved.

18. c (Chapter 11)

Broca aphasia is characterized by effortful nonfluent speech and an inability to repeat, with relatively preserved comprehension. Transcortical motor aphasia is similar but features preserved repetition.

19. b (Chapter 21)

Along with the clinical picture, a CSF profile of lymphocytic pleocytosis, elevated protein, normal glucose, and a negative Gram stain point to a viral or aseptic meningitis. The clinical presentations of problems A through D could appear very similar, but the CSF analysis is crucial in identifying the responsible organism. Bacterial meningitis tends to produce a granulocytic

pleocytosis. Fungal meningitis is usually associated with hypoglycorrhacia (defined as a CSF-serum glucose ratio below 0.4). Subarachnoid hemorrhage characteristically produces a large number of RBCs (thousands).

20. c (Chapter 2)

About 50% of patients with epilepsy have normal routine EEGs. A seizure is a clinical diagnosis, and this patient's convincing story supersedes the negative EEG. Long-term video-EEG monitoring is not required to prove the diagnosis of seizure. While hyperventilation can help to elicit absence seizure activity on EEG, his history and age make an absence seizure unlikely. Although an ischemic stroke can precipitate a seizure, this man's history is most suggestive of seizure. The right hemiparesis is more likely a Todd paralysis rather than an ischemic stroke.

21. a (Chapter 11)

This patient exhibits a form of neglect, in which he does not recognize his left arm as his. Right frontal or parietal lesions are the most common etiology. In the alien-limb phenomenon, patients retain awareness of the limb but feel that it is not under their control.

22. a (Chapter 15)

Benzodiazepines are the first agents used in the treatment algorithm for status epilepticus. Typically, phenytoin and then phenobarbital are used subsequently. Propofol is used if status epilepticus becomes refractory, while carbamazepine and lamotrigine are anti-epileptic drugs that are not available in parenteral form.

23. e (Chapter 17)

Subfalcine herniation may result in compression of the anterior cerebral artery with leg weakness as the result. Ipsilateral third nerve palsy is the first sign of uncal (not subfalcine) herniation. Continued uncal herniation may result in compression of the contralateral cerebral peduncle against the free edge of the tentorium, leading to ipsilateral hemiparesis (the "Kernohan's notch" phenomenon). Contralateral third nerve palsy and contralateral hemiparesis are not features of subfalcine herniation.

24. e (Chapter 20)

Intravenous corticosteroids may delay but not prevent the development of MS in a patient with optic neuritis. They are preferred to oral corticosteroids. Inteferon beta-1b and mitoxantrone are used for MS but not in the treatment of isolated optic neuritis.

25. c (Chapter 4)

Lesions of the MLF produce an INO. The clinical characteristics of a right INO include inability to adduct the right eye in left lateral gaze plus nystagmus of the abducting left eye. Adduction during convergence is maintained because this action does not depend on the MLF. "One-and-a-half syndrome" occurs as a consequence of a lesion involving the PPRF or sixth-nerve nucleus and the adjacent ipsilateral MLF. This produces an ipsilateral gaze palsy and INO on the contralateral side; the only eye movement present in the lateral plane is abduction of the contralateral eye.

26. a (Chapter 21)

Aseptic meningitis and cranial nerve palsies (such as facial nerve palsy) are among the early manifestations when the nervous system becomes involved in Lyme disease. Painful polyradiculopathy or leukoencephalopathy are two (typically later) nervous system complications of Lyme. Spinal cord compression and generalized epilepsy would not be expected to occur as a direct consequence of this disorder.

27. c (Chapter 18)

Sarcoidosis is one of the most common causes of bilateral LMN facial weakness. It is also an important cause of a lymphocytic meningitis (hence the headache) and may cause a variety of other cranial neuropathies, including optic neuropathy (hence the relative afferent papillary defect). MS is another important cause of optic neuritis, but bilateral facial weakness would be unusual. GBS may cause bilateral facial weakness but typically in the context of areflexia and generalized weakness; the relative afferent papillary defect would be unusual. Lyme disease may cause bilateral facial weakness similarly (although unilateral facial weakness would be more common); the afferent papillary defect would not be expected. Vasculitis is an extremely unusual cause of bilateral facial weakness.

28. a (Chapter 14)

Because of the sudden onset of symptoms along with the patient's stroke risk factors, he most probably has had a pure sensory stroke. The most likely lesion is in the contralateral thalamus, because the sensory pathways cross prior to synapsing in the thalamus. The left postcentral gyrus is on the wrong side to explain the patient's deficit. Also, it is unusual to have sensory loss of the face, arm, and leg equally from a stroke affecting the postcentral gyrus. This is because the middle cerebral artery provides blood to the face and arm regions of the cortex, while the anterior cerebral artery supplies blood to the leg region. The precentral gyrus is predominantly involved in motor pathways and not the sensory system. A lesion of the right corona radiata would be expected to cause a left hemiparesis rather than left hemisensory loss.

29. b (Chapter 17)

The middle meningeal artery travels between the skull and the dura. When this vessel is damaged (typically due to trauma resulting in a skull fracture that lacerates that artery),

blood accumulates in the epidural space, resulting in an epidural hematoma. Patients often have a brief episode of loss of consciousness at the time of trauma, followed by a lucid interval and then clinical deterioration as the bleeding continues. Diagnosis and treatment constitute an emergency, because the blood will continue to collect and may cause brain herniation if untreated.

Diffuse axonal injury or an ischemic infarct would be expected to have a sudden onset without a progressive decline in function. Likewise, a concussion should not cause progressive neurologic decline and, like drug intoxication, would not result in the physical signs seen in this case.

30. d (Chapter 22)

ALS is a motor neuron disease with involvement of the lower motor neurons and corticospinal tracts. Weakness, muscle atrophy, and muscle fasciculations are prominent features. Sensory findings are not typical of ALS. Vitamin B_{12} deficiency classically results in degeneration of the dorsal columns and corticospinal tracts. Therefore joint position sense loss and weakness are typical features, whereas pain and temperature are spared. Brown-Séquard syndrome results from hemisection of the spinal cord. The classic features are ipsilateral weakness and loss of joint position sense with contralateral loss of pain and temperature sensation below the lesion. Tabes dorsalis is a late complication of neurosyphilis and is characterized by isolated dorsal column dysfunction resulting in loss of joint position sense. Anterior spinal artery syndrome usually results from infarction of the anterior spinal artery, causing ischemia to the anterior two-thirds of the spinal cord. Therefore dorsal columns are spared but weakness and loss of pain and temperature sensation result because of involvement of the ventral horns and spinothalamic tracts.

31. c (Chapter 25)

This patient meets the diagnostic criteria for tuberous sclerosis complex (TSC). A head CT or MRI may identify cortical tubers, subependymal giant cell astrocytomas, or other lesions. The other tests do not help in the evaluation of TSC.

32. a (Chapter 25)

A diagnosis of autism requires a combination of social, behavioral, and language abnormalities with onset before age three. Asperger syndrome shares social isolation and eccentric behavior with autism. Language is normal in Asperger syndrome. Neither gross nor fine motor delay is a required feature of either condition.

33. c (Chapter 6)

Allodynia is pain provoked by normally innocuous stimuli; hyperesthesia is increased sensitivity to sensory stimuli, and paresthesias are abnormal spontaneous sensations. "Hypesthesia" refers to decreased sensation.

34. c (Chapter 4)

This patient appears to have a Horner syndrome on the right, likely produced by a carotid dissection as a consequence of neck trauma. Horner syndrome is characterized by unilateral miosis, ptosis, and (sometimes) ipsilateral facial anhidrosis as a result of impaired sympathetic innervation. Examine the pupils in the dark (turn the lights off and look at the pupils during the first 5 to 10 seconds). A dilation lag in the small pupil and anisocoria greater in darkness means a sympathetic defect in the smaller pupil and will help with the diagnosis.

35. b (Chapter 23)

This patient appears to have a Guillain-Barré syndrome. Albuminocytologic dissociation means high protein with almost no cells in the CSF, which is characteristic of this syndrome. Immediately after the onset of weakness, however (the first 3 to 4 days), the CSF could be completely normal. Additional studies to corroborate the diagnosis include nerve conduction studies and EMG to demonstrate slowing of conduction velocities, prolongation of F-wave latency, and possible conduction block.

36. d (Chapter 23)

This patient appears to have acute ascending weakness with loss of reflexes characteristic of Guillain-Barré syndrome or acute inflammatory demyelinating polyradiculoneuropathy. His exam worsens while in the ER, and that should be an indication that he is deteriorating quickly and needs to be admitted to the ICU for close observation. An FVC below 15 mL/kg is an indication for intubation and mechanical ventilation.

37. a (Chapter 23)

Cytomegalovirus (CMV) infection is the most common cause of polyradiculitis or cauda equina syndrome in an immunocompromised individual. The other agents do not affect the nerve roots or cauda equina primarily. Cytomegalovirus polyradiculitis occurs in about 2% of AIDS cases and is characterized by the subacute onset of a flaccid paraparesis, sacral pain, paresthesias, and sphincter dysfunction. PCR evaluation of the CSF for CMV can provide a definitive diagnosis. Treatment is with ganciclovir or foscarnet or, in severe cases, both drugs.

38. d (Chapter 7)

Her symptoms consist of a feeling of movement—which is vertigo. The intermittent nature of her vertigo, the exacerbation with head movement, and the absence of brainstem signs are consistent with BPPV. In order to confirm the diagnosis, one can perform the Dix-Hallpike maneuver at the bedside. Brainstem and cerebellar infarctions rarely present with isolated vertigo, and Meniere disease is characterized by hearing loss and tinnitus along with episodic vertigo.

39. d (Chapter 24)

The primary antigenic target in autoimmune MG is the postsynaptic acetylcholine receptor. Presynaptic voltage-gated calcium channels are the target of the Lambert-Eaton myasthenic syndrome.

40. a (Chapter 1)

The trigeminal nerve is responsible for the muscles of mastication. The facial nerve innervates the muscles of facial expression, the oculomotor nerve subserves eye movements, the glossopharyngeal nerve innervates some pharyngeal muscles, and the hypoglossal nerve moves the tongue.

41. d (Chapter 3)

The GCS—which provides a composite assessment of unresponsive patients based on their eye movements, motor function, and language ability—is typically used for patients after head trauma. It has prognostic value for head-injured patients and is easy for nonphysicians to use.

42. b (Chapter 3)

A noncontrast head CT is the imaging study of choice in suspected intracranial hemorrhage. This allows for the easiest delineation of acute blood, which should appear hyperdense (bright) on this study. Head CT with contrast, skull x-ray, and brain perfusion scan do not help to identify acute blood. Cerebral angiography would be indicated only if a ruptured aneurysm or other vascular anomaly were suspected as the cause of an intracranial hemorrhage.

43. e (Chapter 10)

The patient's clinical presentation is typical for temporal arteritis: age over 50, pain over the temporal arteries, jaw claudication, and an elevated ESR. Definitive diagnosis is made by temporal artery biopsy. Treatment with prednisone for several months must be initiated early, because involvement of the ophthalmic artery can lead to blindness if diagnosis and treatment are delayed.

44. e (Chapter 13)

The patient's history and obesity are most consistent with a diagnosis of obstructive sleep apnea. While narcolepsy is also associated with excessive daytime sleepiness, patients typically have associated hypnagogic hallucinations or cataplexy, which are absent in this patient. Therefore, polysomnography is the test that would be most helpful in confirming the diagnosis. The MSLT is useful for diagnosing narcolepsy while MRI of the brain, LP, and EEG would be of no diagnostic value in this patient.

45. c (Chapter 17)

Uncal herniation results from mass lesions of the middle cranial fossa. This patient most likely has a hemorrhage in the middle cranial fossa from head trauma. If large enough, the mass lesion causes displacement of the medial portion of the temporal lobe (uncus) downward over the tentorium cerebelli. This typically results in compression of the brainstem and entrapment of the third cranial nerve. This compression can cause coma due to disruption of the ascending arousal system from the brainstem. It causes an ipsilateral dilated pupil due to compression of the parasympathetic nerve fibers (traveling with the third cranial nerve) that normally cause pupillary constriction. Diffuse axonal injury can result in coma but would not be expected to be responsible for a unilateral dilated pupil that is nonreactive to light.

46. a (Chapter 14)

Intracerebral hemorrhages caused by hypertension are most often found in the basal ganglia, thalamus, pons, and cerebellum, in order of decreasing frequency.

47. c (Chapter 13)

Pemoline and methylphenidate are stimulants used for the treatment of narcolepsy. Clomipramine is a tricyclic antidepressant used for the treatment of cataplexy. Because obstructive sleep apnea is characterized by repetitive episodes of upper airway obstruction during sleep, treatment is often with CPAP, which helps maintain airway patency during sleep. Alcohol and sedating drugs such as benzodiazepines can decrease upper airway tone, resulting in worsened symptoms. Last, obesity is a risk factor for obstructive sleep apnea, so weight loss may prove beneficial in obese patients.

48. a (Chapter 19)

A headache that is either different from the normal pattern or progressive deserves to be investigated further with a brain imaging study. Slowly progressive brain tumors can be associated with a normal neurologic exam or minor abnormalities. Nausea and vomiting need not be present, especially in the early stages of a tumor. Administration of pure oxygen is an effective treatment for cluster headaches, but the patient's description is not consistent with this diagnosis.

49. b (Chapter 19)

Seizures or hemiparesis are not usual features of pituitary adenoma. Varying degrees of bitemporal hemianopia (a visual field deficit in the temporal visual fields bilaterally) may be caused by compression of the optic chiasm. A homonymous hemianopia results from dysfunction of the optic radiations or visual cortex posterior to the chiasm. On brain imaging with contrast, pituitary adenomas usually enhance in a homogeneous manner and do not typically exhibit ring enhancement.

50. d (Chapter 22)

Signs of UMN or corticospinal tract dysfunction include hypertonia, spasticity, increased reflexes, and an extensor plantar

response (Babinski sign). Signs of lower motor neuron (LMN) dysfunction include hypotonia, decreased or absent reflexes, and a flexor plantar response (downgoing toe). Weakness may be present with either UMN or LMN dysfunction.

51. a (Chapter 2)

CT is the imaging modality of choice for demonstrating acute intracranial bleeding. While it is true that MRI provides better visualization of the contents of the posterior fossa, a cerebellar hemorrhage usually will be visible on CT. Patients with pacemakers and other implanted metal objects cannot undergo MRI. Although MRI with diffusion-weighted imaging is the most sensitive modality for ischemic stroke, a susceptibility-weighted MRI sequence is preferred for detecting intracranial blood.

52. b (Chapter 21)

In the presence of severe Lyme disease with CNS involvement, as in this case, intravenous antibiotics followed by oral therapy comprise the first choice. Here, intravenous ceftriaxone is the first choice. The combination of oral amoxicillin and doxycycline is the most common treatment for uncomplicated Lyme disease. Fluconazole is an antifungal and has no value in the treatment of *Borrelia burgdorferi* infection.

53. c (Chapter 12)

Pathologically, PD is characterized by progressive death of dopaminergic neurons of the substantia nigra pars compacta. Most cases of PD are sporadic, but there are reports of familial cases in which mutations in the parkin and α-synuclein genes have been described. Impairment of vertical gaze is a common feature of progressive supranuclear palsy (PSP), a neurodegenerative disorder that is also characterized by parkinsonian features. Despite the gait manifestations of PD, early falls are actually uncommon (but are common in PSP).

54. d (Chapter 24)

DMD and BMD are allelic disorders due to mutations in the dystrophin gene, located on the X chromosome. The inheritance pattern is X-linked. The limb-girdle muscular dystrophies are a heterogeneous group of disorders, some with autosomal dominant and some with autosomal recessive inheritance. Mutations in a wide variety of genes have been reported in patients with limb-girdle muscular dystrophy, including the sarcoglycan genes.

55. c (Chapter 1)

The cerebellum is the primary brain structure involved in coordination, although other components of the motor pathways are involved as well. Testing for rapid alternating movements is part of the coordination exam. The other choices listed have less, or no, primary role in coordination.

56. b (Chapter 5)

"Crossed signs" can occur with unilateral lesions in the pons if descending motor fibers heading for the ipsilateral facial nucleus are affected, with the descending fibers heading for the contralateral spinal cord. With right pontine lesions, the right face and left body could be weak.

57. e (Chapter 15)

Characteristic side effects of carbamazepine include hyponatremia, agranulocytosis, and the risk for Stevens-Johnson syndrome. Except for the hyponatremia, these side effects are rare.

58. c (Chapters 14 and 17)

Tearing of bridging veins produces a subdural hematoma. Laceration of the middle meningeal artery causes an epidural hematoma. Amyloid angiopathy is a cause of lobar hemorrhage in the elderly. Although arteriovenous malformation (AVM) rupture is a cause of subarachnoid hemorrhage, aneurysmal rupture is a more common cause.

59. b (Chapter 19)

Meningiomas enhance in a bright and mainly homogeneous manner. Certain tumors (particularly glioblastoma multiforme and metastatic lesions), brain abscesses, toxoplasmosis, granulomas, and active demyelinating lesions typically show ring enhancement after contrast administration. While lymphomas can enhance in a homogeneous manner, they can also be ring-enhancing.

60. b (Chapter 7)

Micturition syncope is a form of reflex or neurogenic syncope that involves the triggering of cardioinhibitory and/or vasodepressor responses. The symptoms of light-headedness and graying of vision are typically reported by patients with syncope. Other symptoms might include a heavy feeling at the base of the neck, buckling at the knees, and tinnitus. Although orthostatic hypotension is a common cause of syncope, the occurrence of syncope after micturition, rather than upon standing, suggests that this is not the cause in this case. Vasovagal syncope is another common cause of syncope but typically occurs in the setting of acute pain or with a strong emotional response. Vestibular neuronitis is characterized by vertigo, and there is no associated loss of consciousness.

61. d (Chapter 21)

In HSV infection, the MRI often shows contrast enhancement and edema of the temporal lobes. An EEG can also be helpful and may show sharp-wave discharges in the temporal lobes. Treatment for viral meningitis is mainly supportive, because there are no specific treatments for most viral infections. If

HSV infection is suspected, however, treatment should begin promptly with intravenous acyclovir even while tests are pending, because mortality is close to 70% in untreated cases.

62. b (Chapter 23)

Physical examination is the most important information to define symptoms as belonging to the peripheral nervous system (PNS). Sensory symptoms can have a central or peripheral origin. The acuteness of the presentation does not help localization in this case. Paresthesias may be seen in both PNS and CNS dysfunction.

63. a (Chapter 17)

Neurosurgical decompression is the treatment of choice for an epidural hematoma that has resulted in uncal herniation. This is a neurosurgical emergency, so conservative management would only result in further neurologic decline. While hyperventilation and administration of mannitol may help to decrease intracranial pressure, these are temporizing measures; neurosurgical decompression is necessary to remove the accumulating blood. Because the patient has a hemorrhage, tissue plasminogen activator, which is used in acute ischemic strokes, would be contraindicated.

64. c (Chapter 20)

ADEM is a monophasic demyelinating illness. MS is characterized by multiple white matter lesions separated in space and time and is therefore not monophasic. Oligoclonal bands in the CSF are more common in MS than in ADEM. The pleocytosis of ADEM is lymphocytic. Both MS and ADEM can produce multiple lesions on MRI. ADEM is acquired and commonly occurs after viral infections or vaccinations. A positive family history is more likely to be relevant for a patient with MS.

65. b (Chapter 23)

This is a common presentation of proximal diabetic neuropathy, also known as diabetic amyotrophy. It represents a form of polyradiculoneuropathy that has a predilection for the lumbosacral plexus and in general tends to recover spontaneously over months to years. The etiology is likely different from the more common distal symmetric polyneuropathy seen in diabetes.

66. b (Chapter 17)

The patient's symptoms and head CT findings are consistent with an epidural hematoma, which results from laceration of the middle meningeal artery. The classic head CT finding of an epidural hematoma is a hyperdense region with a biconvex or lenticular shape. Tearing of bridging veins results in a subdural hematoma. Impact of the brain over the bony prominences of the skull results in cerebral contusions. On head CT, these areas appear as hyperdensities within the brain parenchyma and not in the epidural or subdural spaces. Diffuse axonal injury results from rotational acceleration and deceleration of the head and can be associated with either a normal head CT scan or hemorrhages within the deep white matter of the brain. Lastly, rupture of a cerebral aneurysm results in subarachnoid hemorrhage and not an epidural hematoma.

67. b (Chapter 16)

Progressive supranuclear palsy is a disorder characterized by parkinsonism, supranuclear impairment of eye movements (vertical gaze typically affected more prominently than horizontal gaze), and impaired postural reflexes. Corticobasal ganglionic degeneration and PD may also cause rigidity and poor postural reflexes, but are not typically associated with eye movement abnormalities. The MFS and chronic PEO are both associated with eye movement abnormalities, but these disorders affect the external ocular muscles rather than the supranuclear gaze centers and are not associated with extrapyramidal features.

68. b (Chapter 18)

Small-fiber neuropathy typically produces symptoms of neuropathic pain, and examination discloses impaired temperature and pinprick sensation. Other sensory modalities are mediated by large fibers. Weakness and atrophy reflect involvement of motor fibers rather than small-fiber sensory function.

69. a (Chapter 22)

The patient exhibits both upper motor neuron signs (hyperreflexia, spasticity, and Babinski signs) and lower motor neuron signs (atrophy and fasciculations), which are the hallmark of ALS. Weakness can occur with either lower motor neuron (LMN) or upper motor neuron (UMN) dysfunction. None of the other options listed would cause widespread findings in both. Vitamin B_{12} deficiency classically results in degeneration of the dorsal columns (loss of joint position sense) and corticospinal tracts (UMN signs). Anterior spinal artery syndrome usually results from infarction of the anterior spinal artery, causing ischemia to the anterior two-thirds of the spinal cord. Therefore dorsal columns are spared, but weakness and loss of pain and temperature sensation result because of involvement of the ventral horns and spinothalamic tracts. Central cord syndrome is most common in the cervical cord and typically results in loss of pain and temperature sensation in a cape-like distribution. Brown-Séquard syndrome results from hemisection of the spinal cord. The classic features are ipsilateral weakness and loss of joint position sense with contralateral loss of pain and temperature sensation below the lesion.

70. e (Chapter 13)

The patient's history of excessive daytime sleepiness, visual hallucinations while falling asleep (hypnagogic hallucinations),

and transient loss of tone triggered by emotional states (cataplexy) is characteristic of narcolepsy. Of the choices listed, methylphenidate is the best option, as it will treat the excessive daytime sleepiness and cataplexy. Clomipramine and venlafaxine are primarily effective for treating the cataplexy but will not improve the patient's daytime sleepiness. CPAP is a treatment for obstructive sleep apnea. Pemoline is an effective medication, but due to possible hepatic toxicity, it is usually reserved for use when other medications have failed.

71. a (Chapter 16)

The extrapyramidal features of idiopathic PD are typically asymmetric. Postural reflexes may be impaired in a number of extrapyramidal disorders including idiopathic PD. Impaired vertical gaze is more typical of progressive supranuclear palsy than of idiopathic PD.

72. c (Chapter 8)

MFS is a disorder characterized by ataxia, ophthalmoplegia, and areflexia. It is considered a variant of the Guillain-Barré syndrome and is associated with the finding of anti-GQ1b antibodies in the serum. Stroke (involving either the brainstem or cerebellum) should be sudden in onset and typically would not be expected to progress over a period of several days. Ophthalmoplegia may be seen in both MG and the MFS, but areflexia is not a feature of MG. Alcoholic cerebellar degeneration may be associated with a peripheral neuropathy and loss of joint position sense and deep tendon reflexes but should not produce ophthalmoplegia (unless associated with Wernicke encephalopathy, in which case confusion should also be present).

73. c (Chapter 9)

Acute urinary incontinence is an emergency. MRI of the spine will help to determine whether an acute lesion is responsible for the incontinence (cauda equina or conus medullaris syndrome, spinal cord compression, etc.). Determination of the PVR would not help in this situation, and urodynamic studies are not indicated in the acute setting. Lumbar puncture is not indicated in this situation.

74. d (Chapter 10)

Both chronic paroxysmal hemicrania and cluster headache are unilateral and can produce conjunctival injection. Chronic paroxysmal headache is more common in women, whereas cluster headache is more common in men. Response to indomethacin is seen in chronic paroxysmal hemicrania but not in cluster headache. Episodes of chronic paroxysmal hemicrania typically last for 20 minutes rather than hours.

75. e (Chapter 11)

Although the patient's symptoms are somewhat nonspecific, examination shows that he has normal language function but with inability to perform certain actions described by the examiner. "Apraxia" refers to the inability to perform a learned motor task and it is typically caused by lesions in either the frontal or parietal lobe of the dominant hemisphere.

76. d (Chapter 2)

Xanthochromia is the yellow discoloration of the supernatant (if blood has been present for a few hours) in a spun sample of CSF that characterizes the presence of blood due to a subarachnoid hemorrhage. In a traumatic tap, the red cells precipitate, and the supernatant is colorless. Increased opening pressure, increased white cell count, and pain upon needle insertion may or may not be present in either condition. Increased red cell count is seen in both subarachnoid hemorrhage and a traumatic tap.

77. e (Chapter 2)

While T1 and T2 images may detect blood breakdown products, susceptibility imaging or gradient-echo imaging is the most sensitive technique for determining the presence of intracranial hemorrhage. Contrast-enhanced T1 is more useful for detecting the presence of a brain tumor. FLAIR imaging is the single best MRI technique for screening for most types of intracranial lesions.

78. c (Chapter 13)

Synucleinopathies are associated with REM sleep behavior disorder. Examples of synucleinopathies are Parkinson disease, Lewy body dementia, and multiple system atrophy. Alzheimer disease, multiple sclerosis, myasthenia gravis, and primary lateral sclerosis are not synucleinopathies and are not associated with REM sleep behavior disorder.

79. a (Chapter 14)

Of the vascular malformations, arteriovenous malformations are at the greatest risk to bleed, with a rate of bleeding of approximately 2% to 3% per year. Capillary telangiectasias, cavernous hemangiomas, and developmental venous anomalies are vascular malformations which rupture much less frequently, and are thus less likely to result in an intracranial hemorrhage. The vein of Galen is a normal anatomic structure.

80. b (Chapter 14)

The patient has a pure motor stroke involving the right side. Possible localization for this syndrome includes the left corona radiata, left internal capsule, and the left side of the base of the pons. Infarction of the motor cortex capable of producing a right hemiplegia would also likely cause an aphasia. Thalamic strokes produce more prominent sensory than motor deficits. Infarction of the midbrain sufficient to produce a hemiplegia would also be associated with eye movement abnormalities. The lateral medullary (Wallenberg) syndrome is associated

with ipsilateral ataxia, ipsilateral Horner syndrome, and ipsilateral facial sensory loss, with contralateral impairment of pain and temperature in the arm and leg, nystagmus, and vertigo. Weakness is absent in a Wallenberg syndrome because motor fibers travel more anteriorly within the medulla.

81. b (Chapter 24)

Duchenne muscular dystrophy is associated with a marked elevation of the serum CK level. Becker muscular dystrophy and limb-girdle muscular dystrophy are also associated with increased CK levels, but the elevations are less marked than for Duchenne dystrophy. Myotonic dystrophy may be associated with a normal CK or only mild elevation. Lambert-Eaton myasthenic syndrome is a neuromuscular junction disorder which is not typically associated with elevations in CK.

82. c (Chapter 1)

The ability to walk in a steady, coordinated manner requires the concerted functioning of multiple parts of the nervous system, including motor pathways, sensory tracts, and the cerebellum, among other things. Testing for gait abnormalities is thus a sensitive way to detect abnormalities in many different nervous system functions.

83. a (Chapter 3)

Locked-in syndrome, which generally occurs with large lesions in the base of the pons, such as infarcts from cardiac embolism or basilar artery stenosis, is characterized by loss of all significant motor function except eye blinking or perhaps vertical eye movements, with preservation of awareness and cognitive function. A large pontine lesion will typically affect corticobulbar and corticospinal fibers bilaterally, but blinking and vertical eye movements are preserved because of intact midbrain function.

84. d (Chapter 3)

Mannitol is an osmotic diuretic which can be used to lower increased intracranial pressure, although the benefit may be transient. Lowering the head of the bed, loading intravenous fluids, or a decrease in respiratory rate would all have the effect of raising intracranial pressure. Basilar artery stenting is not an appropriate intervention for increased intracranial pressure.

85. b (Chapter 12)

Dementia with Lewy bodies may be the second most common type of dementing illness after Alzheimer disease. It is characterized by a parkinsonian motor syndrome, visual hallucinations, and marked fluctuations in alertness, as well as an exquisite sensitivity to neuroleptic medications.

86. e (Chapter 12)

The two neuropathological hallmarks of Alzheimer disease are amyloid plaques and neurofibrillary tangles. The presence of Lewy bodies in the substantia nigra suggests Parkinson disease, and in cortical neurons suggests dementia with Lewy bodies. Prominent atrophy of the caudate is seen in Huntington disease. Spongiform changes in cortex suggest the possibility of Creutzfeldt-Jakob disease.

87. a (Chapter 15)

The diagnosis of seizures is a clinical one. Except on the rare occasions when a paroxysmal event is directly observed by the physician, the best way to distinguish a seizure from other episodes of neurologic dysfunction such as syncope, migraine, or transient ischemic attack is by detailed characteristics obtained from the history. The other listed choices may contribute to the diagnostic workup but none is as important as the history in making the diagnosis.

88. c (Chapter 21)

A CSF leukocytosis, elevated protein, and depressed glucose level form the characteristic profile in acute bacterial meningitis. Viral meningitides typically do not depress the CSF glucose. Fungal and tuberculous meningitides can share a common profile with bacterial meningitis except that the leukocytosis predominantly involves lymphocytes rather than neutrophils (except initially).

89. c (Chapter 19)

The typical "butterfly" pattern is characteristics of glioblastoma multiforme. There are other tumors that can cross white matter tracts, like lymphoma. The other options are very unlikely to produce this radiologic pattern. Moreover, the usual location of meningiomas, ependymomas, and schwannomas is not those found on this MRI.

90. d (Chapter 4)

Bitemporal visual field defects are seen in conditions affecting the optic chiasm, such as suprasellar tumors. Lesions in the left optic radiation produce a right homonymous hemianopia, and lesions of the right occipital lobe a left homonymous hemianopia. Optic nerve lesions produce unilateral visual field defects.

91. e (Chapter 6)

Sensory symptoms in the thumb can be related to median neuropathies (as in a carpal tunnel syndrome) but also to higher lesions such as those seen in a C6 radiculopathy. Lower trunk brachial plexopathy or C8 or T1 radiculopathies could involve the 4th and 5th digits and intrinsic muscles of the hand (including those innervated by the median nerve). The fact that the EMG did not show median neuropathy at the wrist, plus the history of radicular pain, makes a C6 or C7 radiculopathy the most likely cause. Neuromuscular junction disorders do not present with pain or sensory symptoms.

92. e (Chapter 22)

The patient appears to have an anterior spinal artery syndrome (ASAS), a well-recognized complication of abdominal aortic surgery. It usually includes a dissociated sensory loss as the anterior two-thirds of the spinal cord is perfused by the ASA, while the posterior third (posterior columns) is perfused by posterior spinal arteries. Therefore, the corticospinal and spinothalamic tracts are affected in ASAS, but the posterior columns remain intact, with preservation of joint position sense. A sensory level should be present but may not help to define the location of the lesion (nor will reflexes). MRI of the spine can help corroborate your clinical diagnosis but not show much quickly.

93. d (Chapter 23)

The patient has ascending weakness, and the preceding gastrointestinal syndrome makes Guillain-Barré syndrome more likely. Other tests need to be considered to confirm the diagnosis, but it is important to know the current respiratory status before any other test. If the FVC is less than 15 mL/kg, the patient should be transferred to the ICU and intubated. If the FVC is normal, the patient should have frequent FVC checks early during hospitalization, because a very rapid deterioration can occur. Steroids are not a treatment option for Guillain-Barré, which is treated with IVIg or plasmapheresis.

94. c (Chapter 8)

This is a very characteristic history of BPPV. Typically episodes of vertigo are brief, lasting 10 to 30 seconds, with no symptoms in between attacks. The absence of any other associated symptoms, the normal neurologic examination, and the characteristic nystagmus when the affected ear is closer to the ground during the Dix-Hallpike maneuver, are all highly characteristic. Meniere disease is associated with tinnitus and hearing loss. A cerebellar stroke would not produce recurrent positional symptoms and would likely be associated with other neurologic deficits.

95. d (Chapter 10)

This history is very characteristic of giant cell (temporal) arteritis. Other symptoms might include jaw claudication. Testing the ESR and CRP and proceeding to temporal artery biopsy are critical if the feared complication of blindness from an anterior ischemic optic neuropathy is to be avoided. Migraine is very unlikely; it would be a mistake to assume that a new headache in a 77-year-old woman represents recurrence of an old problem. MRI and LP are of no value in suspected GCA.

96. b (Chapter 11)

The anomia (impaired naming) and reduced verbal fluency indicate the presence of an aphasia (language disorder). The reduced verbal fluency does suggest an anterior location, but the hypophonia is most suggestive of a subcortical (usually thalamic) syndrome.

97. c (Chapter 18)

The rash on her shins likely represents erythema nodosum. Sarcoidosis is one of the most common causes of bilateral lower motor neuron facial palsy, especially in the African-American population.

98. e (Chapter 18)

Vitamin E deficiency may cause ataxia, myelopathy, and polyneuropathy. Vitamin B deficiency may cause subacute combined degeneration of the cord and dementia.

99. a (Chapter 16)

Tremor, bradykinesia, rigidity, and postural instability are the four cardinal features of idiopathic Parkinson disease. Dementia may occur but is late and is not characteristic. Hallucinations may occur as part of diffuse Lewy body disease and are frequently encountered in idiopathic PD, but are not a cardinal feature. Gaze palsy suggests the diagnosis of progressive supranuclear palsy rather than idiopathic PD. Autonomic dysfunction is common in PD, but if prominent should raise the prospect of multiple system atrophy.

100. c (Chapter 11)

Aphasia is a language (not a speech) disorder. Apraxia is the inability to carry out a learned motor task. Neglect is not a form of apraxia. Agraphia describes an inability to write.