

Several of these multiple-choice questions were originally published in *Continuum: Lifelong Learning in Neurology—Neuro-oncology, Volume 21, Issue 2, April 2015* based on the content in the issue developed by the following faculty:

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22. A 45-year-old right-handed, otherwise healthy man is evaluated for progressive headache, behavioral changes, and left hand numbness over the past 5 weeks. Examination shows papilledema, reduced attention, and minimal left upper extremity paresis. MRI of the head shows a 7-cm infiltrating lesion in the right frontal lobe with contrast enhancement and mass effect. Assuming that the histopathologic analysis of the lesion confirms a high-grade glioma, what is the most appropriate approach?

- A. biopsy only, followed by bevacizumab to reduce edema
- B. biopsy only, followed by focused radiation therapy
- C. biopsy only, followed by temozolomide
- D. maximal safe surgical resection, followed by CCNU (lomustine) therapy
- E. maximal safe surgical resection, followed by focused radiation therapy, with concurrent and adjuvant temozolomide

23. A 66-year-old man is evaluated for progressive behavioral changes over the past 6 weeks. Neurologic examination shows reduced attention span and impaired word retrieval. He has papilledema but no focal motor deficits. MRI shows a large lesion involving the right frontal lobe, with contrast enhancement and areas of necrosis. After subtotal resection, neuropathologic findings confirm the presence of a glioblastoma multiforme. Molecular studies show methylation of the O-6-methylguanine-DNA methyltransferase (MGMT) gene. What is the significance of this molecular marker?

- A. it indicates low likelihood of response to CCNU (lomustine)
- B. it indicates that the glioblastoma originated de novo
- C. it predicts additional benefit of bevacizumab in preventing tumor progression
- D. it predicts beneficial effect of temozolomide as an adjuvant of radiation therapy
- E. it predicts high risk of tumor recurrence following resection and radiation therapy

24. A 52-year-old left-handed, otherwise healthy woman is evaluated in the emergency department for a single generalized tonic-clonic seizure that occurred 12 hours ago; she also described a progressive headache over the past 4 months. Examination shows reduced attention but no focal deficits. MRI of the head shows an infiltrating lesion in the right temporal lobe with an area of contrast enhancement and moderate mass effect. The patient undergoes a gross total resection of the tumor, and the histology reveals an anaplastic oligodendroglioma. The presence of which of the following tumor markers would predict a good response to adjuvant procarbazine, CCNU (lomustine), vincristine (PCV) chemotherapy in this patient?

- A. alpha thalassemia/mental retardation syndrome X-linked gene (ATRX) mutation
- B. isocitrate dehydrogenase 1 gene (IDH1) mutation
- C. O-6-methylguanine-DNA-methyltransferase gene (MGMT) acetylation
- D. 1p19q codeletion
- E. 13q14 deletion

25. A 72-year-old man is evaluated for progressive cognitive impairment and gait disturbance over the past 3 months. He has lost 9 kg (20 lbs) since the onset of his symptoms. Examination shows reduced attention span, impaired learning and recall, word finding difficulties, and a spastic gait. MRI shows a large lesion in the left frontal lobe extending across the corpus callosum. Laboratory studies show elevated lactate dehydrogenase level. A stereotactic biopsy shows a diffuse large B-cell lymphoma. According to current evidence, which of the following would be the most appropriate initial treatment for this patient?

- A. autologous stem cell transplant
- B. gross total resection
- C. high-dose methotrexate
- D. temozolomide
- E. whole-brain radiation therapy

26. A 72-year-old woman presents to the emergency department after a generalized tonic-clonic seizure. Over the past 3 weeks she has had progressive headache, memory loss, and behavioral changes. Examination shows papilledema, severely impaired learning and memory, and a superior right quadrantanopia. She is started on phenytoin. MRI shows a large, contrast-enhancing and partially necrotic lesion with a focus of hemorrhage in the left temporal lobe. The patient is started on dexamethasone and undergoes a partial subtotal resection of the mass, leading to the diagnosis of glioblastoma. Despite use of elastic graded compression stockings, she develops a deep venous thrombosis in the right calf on the fourth postoperative day. According to current evidence, which of the following will be the most appropriate approach for this complication?

- A. aspirin
- B. inferior vena cava filter
- C. low-dose warfarin
- D. low-molecular-weight heparinoid
- E. rivaroxaban

27. A 66-year-old woman with a history of breast cancer treated with resection and chemotherapy 5 years ago is evaluated for progressive headache over the past week. She has lost 10 kg (22 lbs) over the past 6 months and has been disabled by severe low back pain due to widespread vertebral metastasis. Examination shows reduced attention span, papilledema, and left arm weakness. MRI of the brain shows multiple metastases involving the right frontal lobe, left parietal lobe, left temporal lobe, and right cerebellum. Treatment with steroids and whole-brain radiation therapy is considered. Which of the following drugs may be potentially beneficial in delaying cognitive decline from whole-brain radiation therapy in this patient?

- A. donepezil
- B. memantine
- C. methylphenidate
- D. modafinil
- E. rasagiline

28. A 45-year-old man with recently diagnosed Hodgkin lymphoma is evaluated for progressive gait ataxia over the past 3 months. Neurologic examination shows cerebellar ataxia and nystagmus. Which of the following autoantibodies is most likely to be found in this patient?

- A. anti-CRMP (collapsin response mediator protein)
- B. anti-Hu (antineuronal nuclear antibody type 1)
- C. anti-NMDA (N-methyl-D-aspartate) receptor
- D. anti-Purkinje cell type 1 (PCA-1)
- E. anti-Tr (delta/notchlike epidermal growth factor-related receptor)

29. A 53-year-old woman comes to the emergency department because of severe low back pain and right lower extremity weakness developing over the past 12 hours. She has a history of breast cancer treated with resection and hormone therapy 15 years ago. She feels otherwise well, has continued to be physically active, and was disease free upon a recent evaluation 3 months ago. Examination shows tenderness in the right lumbosacral region and weakness in the right gluteus medius, hamstrings, peronei, and anterior and posterior tibialis muscles. Sensation is reduced in the right lateral leg and dorsum of the foot; knee and ankle reflexes are normal. MRI shows multiple vertebral metastases; there is a pathologic fracture at the L4-L5 interspace with a bone fragment and anterolisthesis with compression of the right L5 root. She is immediately started on dexamethasone therapy. Which of the following would be the most appropriate next step in treatment?

- A. bisphosphonate therapy followed by chemotherapy
- B. bracing followed by chemotherapy
- C. decompressive laminectomy
- D. decompressive resection and stabilization followed by local radiation therapy
- E. whole-spine radiation therapy

30. A 6-year-old boy is evaluated for severe headache, vomiting, and gait difficulties over the past month. Examination shows gait ataxia and papilledema. MRI of the head shows a midline cerebellar tumor consistent with medulloblastoma, with obstructive hydrocephalus. A second cousin was treated for the same neoplasm 2 years ago. Mutations of which of the following genes is most likely to be found in this family?

- A. adenomatous polyposis coli (APC)
- B. isocitrate dehydrogenase (IDH)
- C. neurofibromin 1 (NF1)
- D. tuberous sclerosis 2 (TSC2)
- E. von Hippel-Lindau (VHL)

31. A 28-year-old man is evaluated for progressive gait instability over the past 6 months. Neurologic examination shows gait ataxia, left upper limb dysmetria, diffuse hyperreflexia, and bilateral Babinski signs. Ophthalmologic examination is normal. MRI of the brain and cervical spine shows cystic lesions with a contrast-enhancing nodule in the left cerebellar hemisphere and at the C2-C3 level of the spinal cord. The patient's father died of renal cell carcinoma. This disorder reflects a mutation of a gene encoding a tumor protein that inhibits which of the following targets?

- A. epidermal growth factor receptor
- B. mechanistic target of rapamycin

- C. mitogen activated protein kinase
- D. Ras-GTPase
- E. vascular endothelial growth factor

32. Which of the following can be seen as an early complication of surgical resection in patients with medulloblastoma?

- A . accelerated intracranial atherosclerosis
- B. endocrinopathy
- C. hearing loss
- D. neurocognitive dysfunction
- E . posterior fossa mutism

33. Which of the following best describes the medical management of patients with brain tumors who are transitioning to a hospice approach?

- A. antiepileptic therapy should be continued as long as possible
- B. corticosteroids should be discontinued
- C. low-molecular-weight heparinoids should be started for venous thromboembolism prophylaxis
- D. palliative chemotherapy should be continued
- E. stimulant therapy with modafinil should be started