

1) A 52-year-old man has had headaches and difficulty concentrating for the past 2 months. He then begins to exhibit odd behavior, including shooting his rifle in his back yard, which the neighbors find disconcerting. He then suffers a generalized tonic-clonic motor ('grand mal') seizure and is admitted to the hospital. MR imaging of his brain reveals a large mass with extensive necrosis in the left cerebral hemisphere extending across corpus callosum into the right hemisphere. Which of the following neoplasms is he most likely to have?

- A. Meningioma
- B Glioblastoma
- C Medulloblastoma
- D Metastatic adenocarcinoma
- E Pilocytic astrocytoma

Ans: (B) CORRECT. A glioblastoma is the highest grade glioma and the most malignant of all primary brain tumors, with rapid and extensive growth. Gliomas in adults usually arise above the tentorium.

2) A 74-year-old man has exhibited memory problems for the past 7 months, and he is noted by his immediate family to confabulate. He dies as a consequence of a hepatocellular carcinoma. At autopsy, his brain demonstrates bilaterally small mamillary bodies that show brown discoloration. Microscopically, there is gliosis and vascular proliferation and hemosiderin deposition in the mamillary bodies and periaqueductal gray matter. Which of the following is the most likely diagnosis?

- A Multiple sclerosis
- B Parkinson disease
- C Amyotrophic lateral sclerosis
- D Wernicke-Korsakoff syndrome
- E Huntington disease

Ans: (D) CORRECT. Wernicke disease can also lead to hemorrhage and/or loss of periaqueductal grey matter. The Wernicke-Korsakoff syndrome is seen with chronic alcoholism. The mechanism may have to do with thiamine deficiency.

(Micronodular cirrhosis with alcoholism is a risk for hepatocellular carcinoma.)

3) A previously healthy 42-year-old former major league baseball player develops progressive, symmetric muscular weakness of his upper extremities along with fasciculations over the course of 3 years. Then he develops difficulty speaking and swallowing. He does not have myalgias or arthralgias. He remains afebrile. His mental function has not become diminished. Which of the following is his most likely diagnosis?

- A Amyotrophic lateral sclerosis
- B von Recklinghausen disease
- C Multiple sclerosis
- D Werdnig-Hoffman disease
- E Guillain-Barré syndrome

Ans: (A) CORRECT. The course of ALS is progressive, with motor impairment due to either upper or lower motor neuron loss, or both. Bulbar involvement can lead to problems speaking and eating, with risk for aspiration. This disease often goes by the eponym of another baseball player, Lou Gehrig, who could not play first base anymore once the disease became advanced.

4) A 50-year-old man is noted by his wife to have undergone personality changes over the last year. In the past, he was noted to be obsessive-compulsive, but he became slovenly and now does not appear to take an interest in his work. He has become more forgetful. On physical examination he has frontal release signs and memory loss. He appears unconcerned about his illness. MR imaging of the brain is performed and shows a 3 cm diameter left frontal lobe mass with areas of calcification. Which of the following diagnoses is most likely to be made on microscopic examination of this mass?

- A Vascular malformation
- B Oligodendroglioma
- C Meningioma
- D Schwannoma
- E Organizing abscess
- F Remote infarct

Ans: (B) CORRECT. The location and radiographic changes suggest a glial neoplasm, most likely an oligodendroglioma.

5) A 9-year-old boy has had the new onset of headaches for the past 4 months. The headaches are associated with dull pain and seem diffuse, but they are becoming more frequent and prolonged. On physical examination he has difficulty abducting his right eye. MR imaging reveals enlargement of the lateral ventricles. There is a 4 cm homogenous, well-circumscribed mass within the fourth ventricle. Which of the following is the most likely diagnosis?

- A Astrocytoma
- B Choroid plexus papilloma
- C Ependymoma
- D Meningioma
- E Metastatic bronchogenic carcinoma
- F Schwannoma

Ans: (C) CORRECT. Ependymomas can arise from ependymal lining cells in ventricles. The mass effect can produce an obstructive hydrocephalus, as well as impinge upon adjacent structures, such as the abducens nerve (CN VI) in this case. The most common location in children is the fourth ventricle, and in adults they are most often found in the spinal cord.

6) An 86-year-old man has become progressively unable to live independently for the past 10 years, and he now requires assistance with bathing, dressing, toileting, feeding, and transfers in and out of chairs and bed. On physical examination, he has no motor or sensory deficits. He cannot give the current date or state where he is. Six months later, he suddenly becomes comatose and dies. At autopsy, there is a large superficial left parietal lobe hemorrhage. Histologic examination of the brain shows numerous neocortical neuritic plaques and neurofibrillary tangles. The peripheral cerebral arteries and the core of each plaque stain positively with Congo red. Which of the following mechanisms is most likely responsible for his disease?

- A Aggregation of A β peptide
- B Conformational change in the prion protein (PrP)
- C Dopamine deficiency
- D Expansion of polyglutamine repeats
- E Mutations in the tau gene

Ans: A

7) A 63-year-old man had increasing irritability over 3 years. He wandered about his neighborhood, complaining to the neighbors about everything. He had no memory loss and was always able to find his way home. The neighbors were pleased when he developed aphasia. On physical examination, there were no motor or sensory deficits and no gait disturbances or tremor. MRI of the brain showed bilateral marked temporal and frontal lobe gyral atrophy. He died of pneumonia 1 year later. At autopsy, the frontal cortex microscopically shows extensive neuronal loss, and some remaining neurons show intracytoplasmic, faintly eosinophilic, rounded inclusions that stain immunohistochemically for tau protein. What is the most likely diagnosis?

- A Alzheimer disease
- B Huntington disease
- C Leigh disease
- D Multiple system atrophy
- E Parkinson disease
- F Pick disease
- G Vascular dementia

Ans: F

8) A 60-year-old woman had problems related to movement for 5 years. Physical examination showed cogwheel rigidity of limbs and a festinating gait, which she had difficulty initiating. Her face was expressionless. She was given levodopa/carbidopa, and her condition improved. Two years later, she had difficulty performing activities of daily living and showed marked cognitive decline. She died of aspiration pneumonia. Autopsy findings include mild cerebral atrophy and loss of substantia nigra pigmentation. Microscopically, cortical neurons show spheroidal, intraneuronal, cytoplasmic, and eosinophilic inclusions.

Immunohistochemical staining for which of the following proteins is most likely to be positive in these inclusions?

- A α -Synuclein
- B Amyloid precursor protein
- C Apolipoprotein E
- D Huntingtin \ E Presenilin

Ans: A

9) A 36-year-old man who had been healthy all his life now has progressive, symmetric muscular weakness. A year ago, he noted weakness in the area of the head and neck, which caused difficulty with speech, eye movements, and swallowing. In the past year, the weakness in the upper and lower extremities has increased, and he can no longer stand, walk, or feed himself. His mental function remains intact. Which of the following cells is most likely being destroyed in this man?

A Ependymal cell

B Lower motor neuron

C Microglial cell

D Oligodendrocyte

E Pigmented neuron

F Spiny neuron

Ans: B

10) A 49-year-old man develops an acute psychosis. He has a lengthy history of chronic alcoholism. He has difficulty performing a finger-to-nose test, and there is paralysis of the lateral rectus muscles. A deficiency of which of the following nutrients is most likely to produce these findings?

A Cobalamin

B Folate

C Niacin

D Pyridoxine

E Thiamine

Ans: E

11) A 40-year-old man has been experiencing headaches for the past 6 months. He had a seizure 1 day ago. On physical examination, there are no remarkable findings. MRI of the brain shows a solitary, circumscribed 3-cm mass in the right parietal centrum semiovale. The mass has small cysts and areas of calcification and hemorrhage. Neurosurgery is performed, and the mass is removed.

Microscopically, the mass consists of sheets of cells with round nuclei that have granular chromatin. The cells have a moderate amount of clear cytoplasm, and they mark with GFAP by immunohistochemical staining. The patient receives adjuvant radiation and chemotherapy, and there is no recurrence. Which of the

following molecular markers is most likely to be found in the cells of this mass?

A BRAF mutation

B CD20 expression

C c-MYC amplification

D EGFR amplification

E 1p and 19q co-deletions

Ans: E

12) An 11-year-old girl has had increasing headaches upon awakening for the past month. On examination, papilledema is present bilaterally. An MRI of her brain reveals a 3-cm solid circumscribed mass within the fourth ventricle. There is third and lateral cerebral ventricular dilation. The mass is excised and microscopically shows perivascular pseudo rosettes with round, regular tumor cells arranged around vessels. Which of the following neoplasms is she most likely to have?

A Astrocytoma

B Ependymoma

C Glioblastoma

D Medulloblastoma

E Schwannoma

Ans: B

13) A 45-year-old woman has had unilateral headaches on the right for the past 5 months. Physical examination yields no remarkable findings. The representative gross appearance of the lesion seen on CT scan of the head is shown in the figure. The mass is surgically removed and microscopic examination shows elongated cells with pale, oblong nuclei and pink cytoplasm with occasional psammoma bodies. Cytogenetic analysis shows 22q-. What is the most likely diagnosis?

A Astrocytoma

B Ependymoma

C Meningioma

D Metastasis

E Tuberculoma

Ans: C

14) A 76-year-old man has a single episode of grand mal seizure. On physical examination, he is afebrile and normotensive. Motor strength is intact, and there is no loss of sensation. Cranial nerves are intact. His mental function is not diminished. There is a 1-cm, darkly pigmented skin lesion on the upper back. Brain MRI shows three solid, 1- to 3-cm mass lesions, without ring enhancement or surrounding edema, located at the gray-white junction in the right and left frontal lobes. The cerebral ventricles appear normal in size. What is the most likely diagnosis?

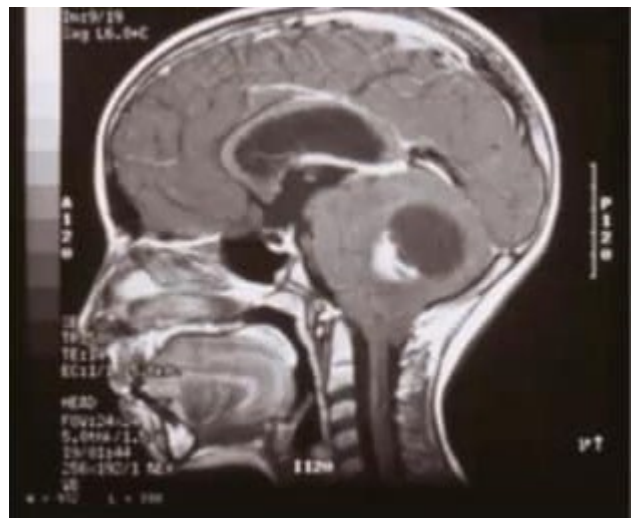
- A Glioblastoma
- B Hemangioblastoma
- C Meningioma
- D Metastatic carcinoma
- E Non-Hodgkin lymphoma
- F Oligodendroglioma

Ans: D

UWORLD) Which of the following is the most likely diagnosis?

- A. Pilocytic astrocytoma
- B. Ependymoma
- C. Oligodendroglioma
- D. Glioblastoma multiforme
- E. Medulloblastoma

Ans: A



Done by: Ahmad AlHurani
Good Luck!