1. A 35-year-old patient with HIV is assessed for new onset right arm weakness. CT scan of his head shows a temporo-parietal ring enhancing lesion. Which is the most likely cause for this?

A. Cerebral toxoplasmosis
B. Behcet’s disease
C. Primary CNS lymphoma
D. Glioblastoma multiforme
E. Herpes simplex virus (HSV)

2. A 47-year-old man with HIV disease presents to hospital with a tonic-clonic seizure. He had initially presented six months previously with Pneumocystis carinii pneumonia with a CD4 T lymphocyte count of 10 cells/mm³ and had subsequently started on highly active antiretroviral therapy. His most recent CD4 count, taken one month prior to his new presentation, was 50 cells/mm³. On examination he has no focal weakness, but both plantar responses are extensor. Fundoscopy is normal. A CT scan of his brain is shown. The most likely diagnosis is:

A. Cerebral abscess
B. AIDS related dementia
C. Cerebral toxoplasmosis
D. Primary CNS lymphoma
E. Glioblastoma multiforme
3. Which of the following tumors is more likely to be multiple?
A. Lipoma
B. Meningioma
C. Oligodendroglioma
D. Primary CNS lymphoma
E. Glioblastoma multiforme

4. An MRI reveals a large tumor occupying much of the left parietal lobe of a middle aged female patient. An attending resident looks at the image and states that it’s the most common kind of intracranial tumor with, unfortunately, typically a poor prognosis. Such tumors are called...

A. Meningiomas
B. Astrocytomas
C. Schwannomas
D. Ependymomas
E. Oligodendrogliomas

5. A surgeon decides to utilize an infratentorial-supracerebellar corridor to approach a pineal region mass. What blood vessel is frequently cauterized and divided for better exposure of the posterior surface of the tumor during this approach?
A. Vein of Galen
B. Ipsilateral basal vein of Rosenthal
C. Posterior cerebral artery (PCA)
D. Precentral cerebellar vein
E. Superior petrosal sinus

6. During translabyrinthine exposure for acoustic neuroma resection, surgeons find themselves exposing Trautmann’s triangle. All of the following structures delineate this area EXCEPT?
A. A triangular patch of dura on the posterior aspect of the temporal bone facing the cerebellopontine angle.
B. The sigmoid sinus laterally
C. The superior petrosal sinus above
D. The jugular bulb below
E. The foramen magnum medially
7. Which of the following are possible complications of mannitol administration?
   1. Aggravation of vasogenic edema
   2. Development of a hyperosmolar nonketotic state
   3. Acute tubular necrosis
   4. Hypotension

   A. 1, 2, and 3 are correct
   B. 1 and 3 are correct
   C. 2 and 4 are correct
   D. Only 4 is correct
   E. All of the above

8. What is the most sensitive laboratory test for the detection of neurocysticercosis (NCC)?

   A. Peripheral eosinophil count
   B. Complete serum white blood cell count
   C. Stool for ova and parasites
   D. Enzyme-linked immunosorbent assay (ELISA)
   E. Electroimmunotransfer blot (EITB)

9. Questions 1 – 7

   Directions: Match each of the following procedures with the potential complication using each answer once, more than once, or not at all.

   A. Cordotomy
   B. Periaqueductal gray stimulation
   C. Percutaneous trigeminal electrocautery
   D. Sympathectomy
   E. Bilateral thalamotomy
   F. Pallidotomy
   G. Commisural myelotomy

   1. Dysarthria and cognitive decline
   2. Hemiparesis, homonymous hemianopsia
   3. “Ondine’s curse”
   4. Eye movement disorder, pupillary dilation, feeling of fear
   5. Horner’s syndrome
   6. Anesthesia dolorosa
   7. Leg weakness, dysesthesias, bladder dysfunction
Questions 10 – 12

A 45-year-old male undergoes a subtemporal approach for tumor resection with elevation of the dura from the middle fossa floor and petrous bone.

10. Structures visible on the floor of the middle cranial fossa during this exposure may include all of the following EXCEPT?

A. Middle meningeal artery  
B. Trigeminal nerve (V3)  
C. Lesser superficial petrosal nerve  
D. Hypoglossal nerve  
E. Greater superficial petrosal nerve

11. Postoperatively, the patient has decreased lacrimation on the ipsilateral side. What is the most likely etiology of this problem?

A. Lesser petrosal nerve injury  
B. Greater petrosal nerve injury  
C. Geniculate ganglion injury  
D. Chorda tympani injury  
E. Injury of Jacobson’s nerve

12. During surgery, additional exposure is needed to access the upper petroclival region for tumor resection. Which maneuver may assist the surgeon in accomplishing this task?

A. Further drilling of Glasscock’s triangle  
B. Additional exposure through Kawase’s quadrilateral  
C. Further drilling of the arcuate eminence  
D. Identifying Trautmann’s triangle and exposing medially to this landmark.  
E. Modifying the approach by utilizing a presigmoid corridor

13. A 65-year-old woman is investigated for enophthalmos and headache. She is cachetic, anemic and you suspect a metastatic process. CT head demonstrates an infiltrative retrobulbar mass. What is the most likely site of primary disease?

A. Lung  
B. Renal  
C. Breast  
D. Ovarian  
E. Melanoma
14. Which of the following are associated with yolk sac tumors?

1. AFP positivity
2. β-HCG positivity
3. Schiller-Duval bodies
4. Placental alkaline phosphatase positivity

A. 1, 2, and 3 are correct
B. 1 and 3 are correct
C. 2 and 4 are correct
D. Only 4 is correct
E. All of the above

15.

Directions: Match the visual field cut with the most likely lesion site.

Questions 1–4

1. Temporal lobe
2. Lateral posterior choroidal artery
3. Anterior chiasm
4. Anterior choroidal artery

16. A 57-year-old male presents to your office with a 4-month history of unilateral epistaxis and nasal discharge. His MRI is depicted below. Histopathologic analysis of the tumor revealed uniform small cells with round nuclei, scant cytoplasm, a prominent reticular core, and scattered Homer-Wright rosettes. Immunohistochemistry was positive for neuron-specific enolase and S-100 but was cytokeratin, CD20, and CD79a negative. What is the most likely diagnosis?
17. You are called to the OR to do a frozen section of a suprasellar mass in a 5-year-old boy. Which of the following would be most likely?
A. Pituitary adenoma
B. Craniopharyngioma
C. Pilocytic astrocytoma
D. Primary CNS lymphoma
E. Endodermal sinus tumor

18. A 50-year-old woman who works as a paralegal in a law firm comes to her local doctor because of problems with sleep. The patient says that over the past several weeks, she hasn't slept well, feels tired, and has had headaches. Extraocular movements reveal normal conjugate, oblique, and downward movement, but she is unable to look upwards. Which of the following is the most likely diagnosis?
A. Pinealoma
B. Acoustic neuroma
C. Craniopharyngioma
D. Parasagittal meningioma
E. Astrocytoma in the cerebellum

19. A 46-year-old woman with melanoma resected from her chest wall 3 years ago presented to the ED with a week of a severe right parietal headache. Her neurologic examination showed only left upper extremity drift. The radiology resident reported that the CT scan showed a right subdural hematoma. What do you think is the cause of her subdural?
A. Bleeding from a dural metastasis
B. A fender-bender accident last month
C. Excessive use of aspirin for her migraine headache
D. Trauma from bumping her head playing tennis a week ago
E. A misreading of a meningioma on CT scan by an inexperienced resident

20. A 50-year-old previously healthy woman presents to her physician with a 3-week history of progressive gait and limb ataxia, dysarthria, and blurred vision. Examination reveals nystagmus, dysarthria, and severe gait and limb ataxia. The remainder of the neurologic examination is normal. MRI of the head with contrast is normal. A blood test reveals positive serology for anti-Yo antibodies (anti-Purkinje cell antibody). Which of the following malignancies is most likely present in this patient?
   A. Melanoma
   B. Breast cancer
   C. Ovarian cancer
   D. Small cell lung cancer
   E. Non-Hodgkin’s lymphoma

21. Which of the following are the most common organisms found in brain abscesses?
   A. Proteus spp.
   B. Streptococcal
   C. Staphylococcal
   D. Bacteroides spp.
   E. Pseudomonas spp.

22. Which of the following is common the tumor type in the brain of patients with AIDS, but otherwise extremely rare?
   A. Lymphosarcoma
   B. Kaposi’s sarcoma
   C. Metastatic lymphoma
   D. Lymphocytic leukemia
   E. Primary CNS lymphoma

23. A 55-year-old man is referred to the hospital for further assessment having noticed a growth of papules from the nail folds. A doctor confirms that they are periungual fibromas. There is also a history of seizures and renal impairment. What is the likely diagnosis?
24. A 25-year-old woman with no past medical problems visited rural Asia a year ago, now she presents with new onset seizures. As part of her evaluation a CT is obtained and shown below. What is the most likely cause of this patient's symptoms?

A. Brain metastasis
B. Multiple sclerosis
C. Pyogenic abscess
D. Neurocysticercosis
E. Strongyloidiasis

25. Button sequestrum of skull is seen in all except:

A. Epidermoid
B. Tuberculosis
C. Hemangioma
D. Eosinophilic granuloma

26. Which of the following is the most likely cause of drop metastases?
A. Epidermoid
B. Dysgerminoma
C. Medulloblastoma
D. Hemangioblastoma
E. Oligodendroglioma
27. The structures being shown here are:

1. Often seen in neuroblastoma
2. Can be seen in medulloblastoma
3. Homer Wright rosettes
4. Flexner-Wintersteiner rosettes

A. 1, 2, and 3 are true
B. Only 1 and 3 are true
C. Only 2 and 4 are true
D. Only 4 is true
E. None of the above

Answers

1. A. Cerebral toxoplasmosis.
2. C. Cerebral toxoplasmosis.

Primary central nervous system lymphoma tends to be multiple in 50% of cases. Meningiomas are multiple in less than 10% of cases. Glioblastoma multiforme appears multicentric in less than 5% of cases. Lipomas are usually incidental findings and almost never multiple.

4. B. Astrocytomas.

5. D. Precentral cerebellar vein.

Cauterizing and dividing the precentral cerebellar vein will often expose the posterior surface of pineal region tumors. The veins of Galen and Rosenthal should be preserved during this operation, as well as the vermian vein, which often can be spared in this approach. The choroidal arteries may supply feeders to the tumor but rarely need to be cauterized and ligated for adequate tumor resection.
6. E. The foramen magnum medially.

There are two goals of the translabyrinthine approach for acoustic neuroma resection that may help achieve maximal tumor resection. The first is to remove enough bone to identify the nerves lateral to the tumor as they course through the IAC, and the second is to expose the dura of the posterior aspect of the temporal bone that faces the cerebellopontine angle (CPA). This triangular patch of dura facing the CPA is called Trautmann’s triangle and extends from the sigmoid sinus laterally, the superior petrosal sinus above, and the jugular bulb below. The foramen magnum is not included in Trautmann’s triangle.

7. E. All of the above.

8. E. Electroimmunotransfer blot (EITB).

Complete white blood cell count, peripheral eosinophil level, and serum anticysticercal antibody levels should be obtained in all patients suspected of having NCC. Patients requiring ventriculostomy placement should have cerebrospinal fluid (CSF) analyzed for eosinophil and anticysticercal antibody levels. Stool testing for ova and parasites is helpful in patients with simultaneous intestinal tape worm infection but is insensitive and nonspecific for *T. solium* species and is found in less than 33% of cases.

Several laboratory methods have been developed to detect host antibodies against circulating cysticercal antigens. From the many tests performed, current data indicate that enzyme-linked immunosorbent assay (ELISA) and electroimmunotransfer blot (EITB) tests are the most effective. Studies comparing these diagnostic modalities have shown that the EITB assay is more sensitive overall than ELISA, especially when serum is being tested. Both techniques are more sensitive in cases with multiple cysts than in cases with solitary or confined lesions. Additionally, no global difference among cases was found with parasites located in different compartments (ventricles, subarachnoid space, parenchyma) of the CNS.

9.

1. Dysarthria and cognitive decline - E. Bilateral thalamotomy
2. Hemiparesis, homonymous hemianopsia - F. Pallidotomy
3. “Ondine’s curse” - A. Cordotomy
4. Eye movement disorder, pupillary dilation, feeling of fear - B. Periaqueductal gray stimulation
5. Horner’s syndrome - D. Sympathectomy
6. Anesthesia dolorosa - C. Percutaneous trigeminal electrocautery
7. Leg weakness, dysesthesias, bladder dysfunction - G. Commisural myelotomy

10. D. Hypoglossal nerve.
11. B. Greater petrosal nerve injury.
12. B. Additional exposure through Kawase’s quadrilateral.

Structures often visible on the middle cranial fossa floor during subtemporal approach include the middle meningeal artery (often sacrificed by cautery and packing of the foramen spinosum), trigeminal nerve (V3), lesser superficial petrosal nerve, greater superficial petrosal nerve, ICA (if there is small dehiscence in the bone), as well as the arcuate eminence, which overlies the superior semicircular canal. Decreased tearing after surgery most likely resulted from injury of the greater superficial petrosal nerve, which provides parasympathetic supply to the lacrimal and nasal gland. Additional exposure to the posterior fossa during a subtemporal approach may be gained by removing the bone of Kawase’s quadrilateral located in the medial petrous apex, medial to Glasscock’s triangle. Kawase’s quadrilateral is bounded laterally by the greater superficial petrosal nerve, medially by the petrous ridge and V3 of the trigeminal nerve, and at its base by the arcuate eminence. Glasscock’s triangle is bounded laterally by a line from the foramen spinosum to the facial hiatus, medially by the greater superficial petrosal nerve (GSPN), and at its base by the mandibular division of the trigeminal nerve.

13. C. Breast.
Most retrobulbar metastases are extraconal (outside the muscle cone). Neuroblastoma and Ewing’s sarcoma are the most common in children and produce smooth extraconal masses related to the posterior lateral wall of the orbit. In adults, an infiltrative retrobulbar mass and enophthalmos is characteristic of scirrhous carcinoma of the breast (invasive ductal carcinoma). Enophthalmia is also considered to be one of the earliest signs of metastatic breast cancer.

14. B. 1 and 3 are correct.

Yolk sac tumor is a germ cell tumor that exhibits loosely arranged cells with clear cytoplasm and prominent eosinophilic bodies. Yolk sac tumors are positive for AFP, and they often exhibit Schiller-Duval bodies.

15.

1. Temporal lobe - D

Superior homonymous quadrantic defects (“pie-in-the-sky”, D) may result from a lesion along Meyer’s loop (after temporal lobectomy) or along the inferior bank of the calcarine fissure.

2. Lateral posterior choroidal artery – C
The central portion of the lateral geniculate body receives blood flow primarily from the lateral posterior choroidal artery. Interruption of this vessel causes a horizontal homonymous sector defect (wedge-shaped, C).

3. Anterior chiasm – A

The anterior chiasm or junctional syndrome results in a unilateral optic nerve defect of one eye and a superior temporal defect in the other eye (A) due to the loop made by the inferonasal retina of the other eye (Willebrand’s knee).

4. Anterior choroidal artery – B

Occlusion of the anterior choroidal artery causes a homonymous defect in the upper and lower quadrants, with sparing of the horizontal sector (quadruple sectoranopsia, B), which is usually characteristic of a lateral geniculate body infarct that is supplied by the anterior choroidal artery.

16. C. Esthesioneuroblastoma.

The clinical history of this patient and destructive MRI appearance of this lesion are highly suggestive of a malignant neoplasm involving the paranasal sinuses. The histologic and immunohistochemical markers are most consistent with esthesioneuroblastoma.

17. C. Pilocytic astrocytoma.

Pilocytic astrocytoma, followed by a craniopharyngioma, is the most common supracellular tumor in children.

18. A. Pinealoma.

This patient has a pinealoma. Tumors of the pineal gland compress the vertical gaze center in the tectum of the midbrain. The pineal gland manufactures melatonin from its precursor serotonin; an inadequate supply of melatonin results in insomnia. Tumors of the pineal gland will not compress the cerebral cortex or the rest of the brainstem. Frequently, the only physical sign noted is failure of upward gaze. An acoustic neuroma (choice B) is a schwannoma of the eighth cranial nerve. It results in deafness, ataxia, and dysarthria. Nystagmus may be present. The gaze centers are not affected. Craniopharyngiomas (choice C) are usually seen in children. There is failure of growth, headaches, and bitemporal hemianopia. Parasagittal meningiomas (choice D) usually result in headache,
spastic paresis, and urinary incontinence. Astrocytomas of the cerebellum (choice 
E) are usually seen in children. These tumors present with headache, nausea, 
vomiting, papilledema, and cerebellar signs such as ataxia, dysarthria, nystagmus, 
and intention tremor. The gaze centers are not affected.

19. A. Bleeding from a dural metastasis.
This woman has bleeding from a dural metastasis. Dural metastases result from 
direct extension of skull metastases or from hematogenous spread. They are 
found at autopsy in about 10% of patients with advanced systemic cancer and 
may be clinically asymptomatic. When they bleed, these metastases can present 
as subdural hematomas. The dural metastasis may be mistaken for a meningioma 
on imaging. The prognosis in this patient is poor, because her chest radiograph 
showed extensive infiltration with tumor.

Brain metastases account for approximately a third of all intracranial tumors and 
are the most common intracranial neoplasm. They characteristically occur at the 
corticomedullary junction of the brain and have surrounding edema that typically 
exceeds the tumor volume. Multiple lesions are present in approximately two-
thirds of cases and should be searched for with administration of intravenous 
contrast. Most are hypodense on CT unless hemorrhagic or hypercellular, hence 
the lesion in this case is hemorrhagic. This lends itself to a differential of primary 
neoplasms which includes melanoma, renal cell carcinoma, thyroid carcinoma, 
bronchogenic carcinoma and breast carcinoma. The history of back pain also 
suggests bone metastases.

20. C. Ovarian cancer.
This patient has paraneoplastic cerebellar degeneration. Paraneoplastic neurologic 
disorders are rare complications of cancer. In this patient, a structural lesion of the 
posterior fossa, such as a tumor or infarct, has been appropriately excluded by 
imaging. In each of the paraneoplastic syndromes, the systemic tumor cell is 
believed to express an “onconeural antigen” that produces an immune response in 
the patient. This onconeural antigen shares similarities to antigens normally 
expressed by specific neural tissue. The host immune response (cell mediated and 
humoral) then attacks both the tumor and the specific neural tissue that share 
antigenic similarity. This theory is strengthened by the fact that patients’ tumors 
often share antigenic similarities with neural tissue and by the frequent 
observation that patients with paraneoplastic neurologic disease often have 
limited or no metastatic disease and small primary tumors. The latter observation 
suggests that the immune response to tumor may be particularly strong in these 
patients.
The presence in this patient’s serum of the anti-Yo antibodies is very specific for 
paraneoplastic cerebellar degeneration in the setting of ovarian, uterine, fallopian
tube, or breast cancer. Anti-Hu (anti-neuronal antibody type I) is the antibody associated with small cell lung cancer and paraneoplastic neurologic disease. Paraneoplastic neurologic disorders are very rare in non-Hodgkin’s lymphoma or melanoma, and there is no serologic antibody specifically associated with either tumor.

21. B. Streptococcal.
Both aerobic and anaerobic streptococcal bacteria occur in more than half of all brain abscesses. Staphylococcus aureus most often occurs in patients who have had penetrating head wounds or have undergone neurosurgical procedures. Enteric bacteria (e.g., Escherichia coli, Proteus, and Pseudomonas) account for twice as many abscesses as S. aureus.

22. E. Primary CNS lymphoma.
Kaposi’s sarcoma is unusually common in patients with AIDS, but it is rarely metastatic to the brain. Metastatic lymphomas producing meningeal lymphomatosis are not especially rare in the general population, but primary lymphomas (that is, lymphomas apparently arising in the CNS) were rare before the AIDS epidemic. The primary brain lymphoma usually presents as a solitary mass and can occur anywhere in the brain, but it does have a predilection for the periventricular structures.

This is an inherited (autosomal dominant) hamartomatous condition in which there are facial angiomas (adenoma sebaceum), subungual fibromas, angiomyolipomas, cardiac rhabdomyomas, pulmonary lymphatic involvement, skin changes such as shagreen patches and ashleaf macules.

Neuroimaging with CT or MRI is the most useful method of diagnosis. CT scan shows both calcified and uncalcified cysts, as well as distinguishing active and inactive cysts. Cystic lesions can show ring enhancement and focal enhancing lesions. Some cystic lesions, especially the ones in ventricles and subarachnoid space may not be visible on CT scan, since the cyst fluid is isodense with CSF. Thus diagnosis of extraparenchymal cysts usually relies on signs like hydrocephalus or enhanced basilar meninges. In such cases CT scan with intraventricular contrast or MRI can be used. MRI is more sensitive in detection of intraventricular cysts.
Photograph shows multiple *T solium* cysts in vitro.

25. C. Hemangioma.
The button sequestrum sign refers to a lesion that is located in the bone and consists of bone opacity surrounded by a relatively well-defined lucent area.

26. C. Medulloblastoma.
Medulloblastoma is the most common to drop metastasize. Rarely, oligodendroglialoma can have drop mets. Other tumors do not produce drop metastasis.

27. A: 1, 2, and 3 are true.
- Often seen in neuroblastoma
- Can be seen in medulloblastoma
- Homer Wright rosettes.

This photo was taken from a medulloblastoma and the structures being shown here (delimited by the arrows) are Homer Wright rosettes. This type of rosette is often seen in neuroblastomas arising from the adrenal glands and other locations. They are also seen in some cases of medulloblastomas. Although Homer Wright rosettes can occur in retinoblastoma, Flexner-Wintersteiner rosettes are far more common in retinoblastomas.