

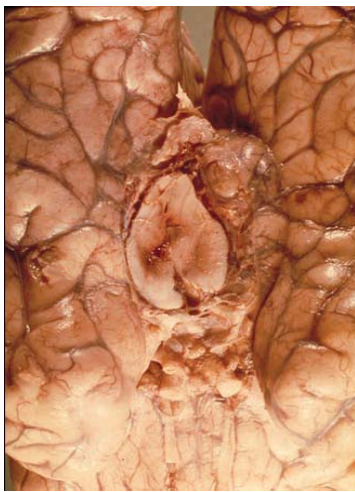
## EANS/UEMS European examination in neurosurgery

### Part I (written)

Variants of questions with answers (compilation - Vyacheslav S. Botev, Department of Neurosurgery, M.Gorky Donetsk National Medical University)

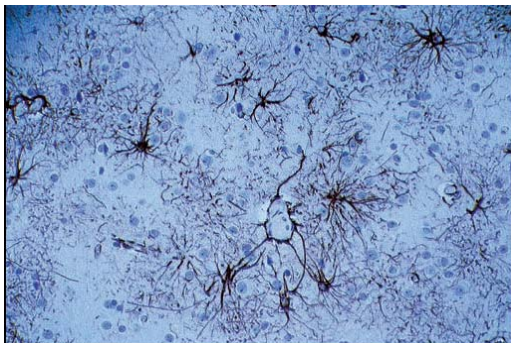
### Neuropathology

**1.** A 54-year-old woman dies 48 hours after suffering severe head injuries in an automobile accident. Just before her death, her left pupil becomes fixed and dilated. An inferior view of the patient's brain at autopsy is shown in the image. Which of the following was the most likely cause of death?



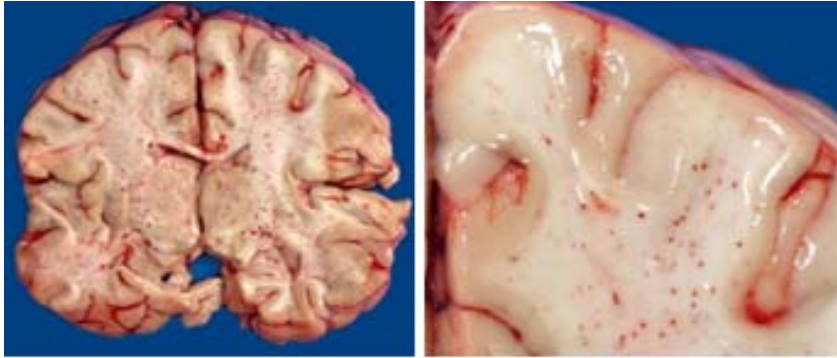
- A. Laminar necrosis
- B. Watershed infarct
- C. Diffuse axonal shearing
- D. Transtentorial herniation
- E. Thrombosis of sagittal sinus

**2.** The patient persists in a vegetative coma for several months and then expires. A section of the temporal lobe shows massive proliferation of cells with a star-shaped appearance (shown in the image). Which of the following best accounts for this cellular response to injury?



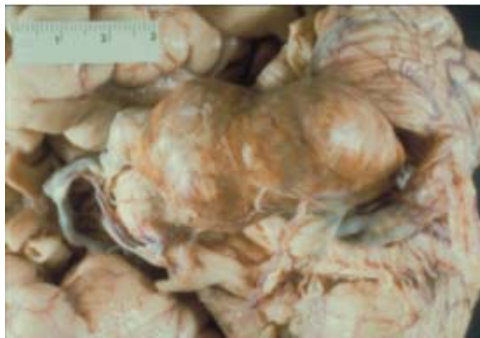
- A. Gliosis
- B. Chromatolysis
- C. Neuronophagia
- D. Leukodystrophy
- E. Axonal regeneration

**3.** A 30-year-old woman suffers massive trauma in an automobile accident and expires 4 days later of respiratory insufficiency. A horizontal section of the patient's brain at autopsy reveals numerous petechiae scattered throughout the white matter. Which of the following is the most likely explanation for this pathologic finding?



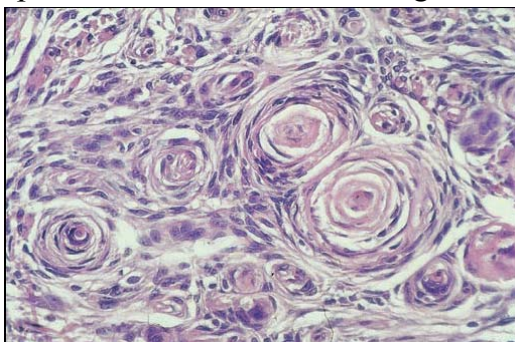
- A. Sepsis
- B. Uremia
- C. Fat embolism
- D. Global ischemia
- E. Occlusion of middle cerebral artery

**4.** A 70-year-old man with a history of senile dementia and a recent myocardial infarct dies of multiple organ system failure following occlusion of the superior mesenteric artery. Examination of the patient's brain at autopsy reveals aneurysmal dilation of the basilar artery (shown in the image). Which of the following is the most common complication of this pathologic finding?



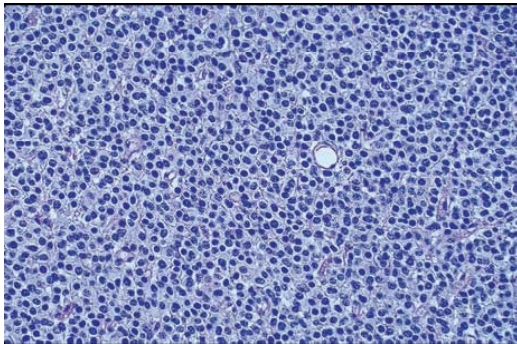
- A. Infection
- B. Dissection
- C. Thrombosis
- D. Hemorrhage
- E. Transformation

**5.** A 45-year-old woman is brought to the emergency room after experiencing a generalized seizure. An X-ray film of the skull reveals a lytic bone mass. A CBC is normal. A portion of the skull and the adherent mass are removed. Microscopic examination of the surgical specimen is shown in the image. What is the appropriate diagnosis?



- A. Meningioma
- B. Medulloblastoma
- C. Hemangioblastoma
- D. Oligodendroglioma
- E. Glioblastoma multiforme

6. A 50-year-old man presents to the emergency room after suffering an epileptic seizure. Vital signs are normal. An X-ray of the patient's head shows a mass in the left cerebral hemisphere with scattered foci of calcification. Histologic examination of a brain biopsy is shown in the image. Which of the following is the appropriate diagnosis?



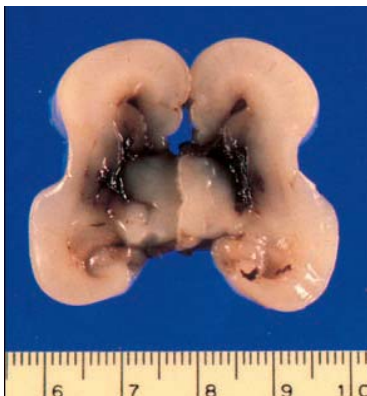
- A. Meningioma
- B. Glioblastoma
- C. Ependymoma
- D. Hemangioblastoma
- E. Oligodendroglioma

7. A 55-year-old woman presents with increasing weight loss and fatigue and subsequently dies of metastatic cancer. The vertebral column at autopsy is shown in the image. What is the diagnosis?



- A. Melanoma
- B. Osteosarcoma
- C. Chondrosarcoma
- D. Multiple myeloma
- E. Rhabdomyosarcoma

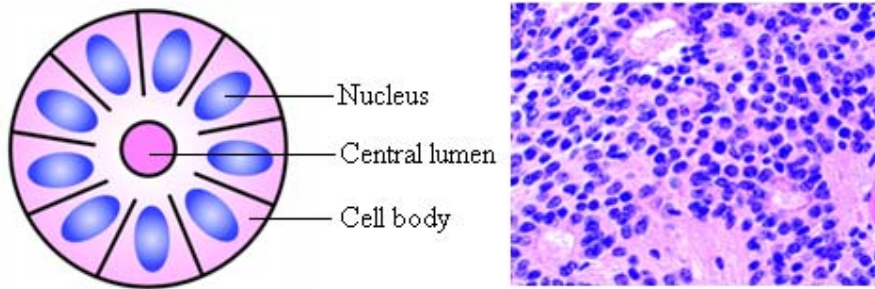
8. A 22-year-old woman delivers a baby at 29 weeks of gestation. Shortly after birth, the neonate becomes short of breath. The neonate is placed on a ventilator, but dies of respiratory insufficiency. The brain at autopsy is shown. Which of the following mechanisms of disease best explains this complication of respiratory distress syndrome (RDS) of the neonate?



- A. Birth trauma
- B. Hypertension
- C. Anoxic injury
- D. Hemolytic anemia
- E. Chronic passive congestion

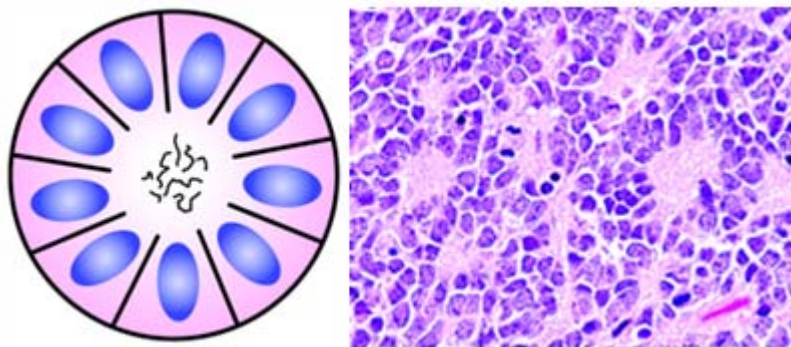


9. Rosettes are groups of tumor cells arranged in a circle around a fibrillary center. Which of the following rosettes is shown below?



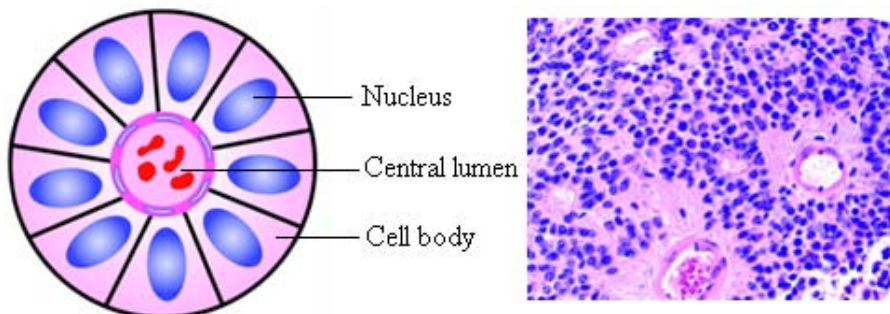
- A. Neurocytic rosette
- B. Homer Wright rosette
- C. True ependymal rosette
- D. Perivascular pseudorosette
- E. Flexner-Wintersteiner rosette

10. Which of the following tumor contains Homer Wright rosette?



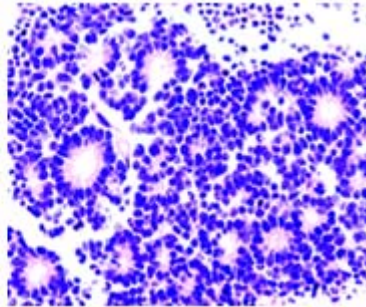
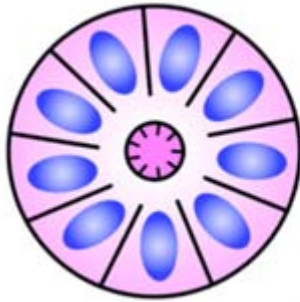
- A. Meningioma
- B. Ependymoma
- C. Medulloblastoma
- D. Pilocytic astrocytoma
- E. Glioblastoma multiforme

11. Which of the following rosettes is shown below?



- A. Neurocytic rosette
- B. Homer Wright rosette
- C. True ependymal rosette
- D. Perivascular pseudorosette
- E. Flexner-Wintersteiner rosette

12. Which of the following rosettes is shown below?



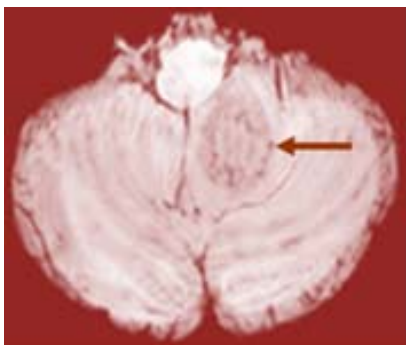
- A. Neurocytic rosette
- B. Homer Wright rosette
- C. True ependymal rosette
- D. Perivascular pseudorosette
- E. Flexner-Wintersteiner rosette

13. What is the pathologic process that the arrow is pointing to?



- A. Dolichoectasia
- B. Berry aneurysm
- C. Radiation change
- D. Acoustic schwannoma
- E. Charcot-Bouchard aneurysm

14. The pathologic specimen below shows the only intracranial lesion found in this patient. This patient would be expected to have exhibited which of the following symptoms?



- A. Seizures
- B. Visual loss
- C. Gait ataxia
- D. Hemiparesis
- E. Hallucinations

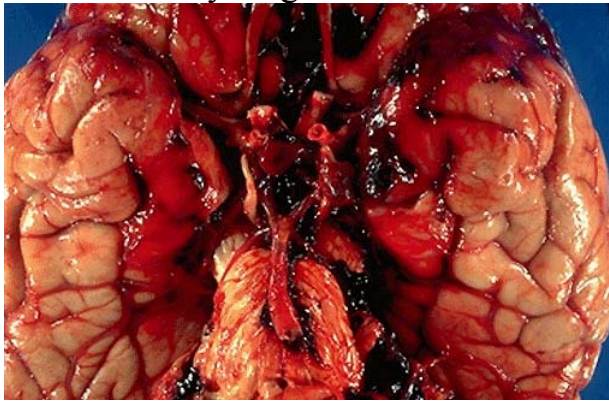
**15.** The structure being illustrated below is removed from the sellar region.



It is most consistent with

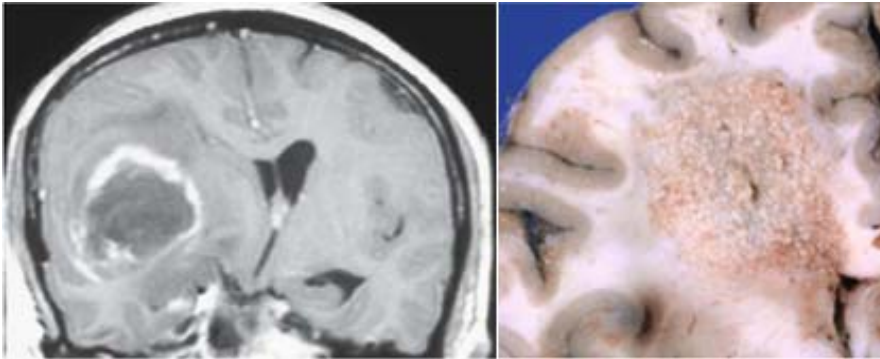
- A. Chordoma
- B. Meningioma
- C. Optic nerve glioma
- D. Craniopharyngioma
- E. Pituitary with a Rathke cleft cyst

**16.** This 39-year-old woman was driving her car when she had the sudden onset of a severe headache. She pulled in to a service station and stopped the car. Then she slumped over the wheel. She was taken to the ED, where she remained comatose and died hours later. This gross appearance is found at autopsy. Which of the following is the most likely diagnosis?



- A. Multiple sclerosis
- B. Huntington disease
- C. Glioblastoma multiforme
- D. Ruptured berry aneurysm
- E. Thromboembolization with cerebral infarction

**17.** The figures shown here are representative of the coronal post contrast MRI and an autopsy specimen from a 45-year-old woman who had a 1-year history of progressive headache and seizures. Which of the following is the most likely diagnosis?



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

**18.** What is the pathologic process that the arrow is pointing to?



- A. Cerebral abscess
- B. Oligodendroglioma
- C. Anaplastic astrocytoma
- D. Primary CNS lymphoma
- E. Sphenoid ridge meningioma



## Answers

### 1. D. Transtentorial herniation.

Head trauma can cause extensive intracranial hemorrhage and cerebral edema. After compensatory mechanisms have been exhausted, the brain is shifted laterally away from the side of the lesion. The medial temporal lobe on the side of the hematoma is compressed against the midbrain to displace it downward through the opening created by the tentorium, a fatal event known as transtentorial herniation. Thus, the oculomotor nerve may be compressed against the edge of the tentorium, causing third nerve palsy. The pupil, generally on the side of the lesion, becomes fixed and dilated. The herniated uncus also compresses the vasculature of the midbrain, especially the mesencephalic veins. Venous stagnation in the midbrain causes further hypoxia and impairs neuronal function. Choices A and B are related to global anoxia. Diffuse axonal shearing (choice C) is a microscopic diagnosis.

Diagnosis: Transtentorial herniation. (Lippincott's Illustrated Q&A Review, p 310)

### 2. A. Gliosis.

Astrocytes are star-shaped glial cells that far outnumber neurons throughout the CNS. Astrocytes proliferate locally in response to injuries (e.g., trauma, abscess, tumors, infarcts, and hemorrhages). This process, referred to as astrogliosis or gliosis, is readily demonstrated by immunostaining for glial fibrillary acidic protein (shown in photomicrograph). Astrogliosis evolves in hours to days and persists to an extent that is usually commensurate with the severity of the initiating injury. The result is a "glial scar" composed of reactive astrocytes and their processes. Neuronophagia (choice C) is a function of microglia.

Diagnosis: Astrogliosis. (Lippincott's, p 310)

### 3. C. Fat embolism.

Small emboli, notably those composed of fat or air, occlude capillaries. Fat emboli originating from bone fractures are carried downstream through the cerebral vessels until the caliber of the emboli exceeds that of the blood vessels, at which point they lodge and block blood flow. The distal capillary endothelium becomes hypoxic and permeable, and petechiae develop, most commonly in the white matter. Although sepsis (choice A) sometimes leads to brain petechiae, the patient's condition does not support this conclusion. None of the other choices are characterized by petechiae in the brain.

Diagnosis: Fat embolism. (Lippincott's, p 313)



#### 4. C. Thrombosis.

Aneurysms caused by atherosclerosis are localized mainly in major cerebral arteries (vertebral, basilar, and internal carotid), which are favored sites of atherosclerosis. Fibrous replacement of the media and destruction of the internal elastic membrane weakens the arterial wall and causes aneurysmal dilation. Although dissection (choice B) and hemorrhage (choice D) may occur, the major complication of an atherosclerotic aneurysm is thrombosis.

Diagnosis: Atherosclerotic aneurysm. (Lippincott's, p 314)

#### 5. A. Meningioma.

Meningiomas are benign intracranial tumors that arise from the arachnoid villi and produce symptoms by compressing adjacent brain tissue. They account for almost 20% of all primary intracranial neoplasms. Meningiomas occur at almost any intracranial site but are most common in parasagittal regions of the cerebral hemispheres, the olfactory groove, and the lateral sphenoid wing. On gross examination, most meningiomas appear as well-circumscribed, firm, bosselated masses of variable size. The histologic hallmark of meningiomas is a whorled pattern of "meningothelial" cells (see photomicrograph). The indolent growth of meningiomas enables them to enlarge slowly for years before becoming symptomatic, during which time they displace the brain but do not infiltrate it. Although benign, meningiomas have a propensity to erode contiguous bone.

Choice C (hemangioblastoma) is characterized by vascular proliferation.

Diagnosis: Meningioma. (Lippincott's, p 316)

#### 6. E. Oligodendroglioma.

Oligodendrogliomas arise in the white matter and grow slowly. They occur predominantly in the white matter of the cerebral hemispheres of adults.

Histologically, the tumors have small rounded nuclei similar to normal oligodendrocytes, but they also exhibit increased cell density and cellular pleomorphism. Calcospherites, which may be visualized radiographically, are occasionally scattered randomly throughout the lesion. The other choices do not feature uniform monomorphic cells.

Diagnosis: Oligodendroglioma. (Lippincott's, p 317)

#### 7. A. Melanoma.

The photograph shows pigmented cells in the vertebral bodies of a person who died of malignant melanoma. This autopsy finding illustrates the point that accurate tumor identification depends on morphologic resemblance to normal

tissue. Tumor emboli in this case probably reached bone after surviving passage through the pulmonary microcirculation. None of the other tumors show pigmentation.

Diagnosis: Melanoma. (Lippincott's, p 44)

### **8. C. Anoxic injury.**

The pathogenesis of RDS of the newborn is intimately linked to a deficiency of surfactant. This material lowers the surface tension of the alveoli at low lung volumes and thereby prevents collapse (atelectasis) of the alveoli during expiration. Atelectasis secondary to surfactant deficiency results in perfused but not ventilated alveoli, a situation that leads to hypoxia and acidosis.

Intraventricular cerebral hemorrhage is a major complication of RDS. The periventricular germinal matrix in the newborn brain is particularly vulnerable to hemorrhage because the dilated, thin-walled veins in this area rupture easily (see photograph). The pathogenesis of this complication is believed to reflect anoxic injury to the periventricular capillaries, venous sludging and thrombosis, and impaired vascular autoregulation. Despite advances in neonatal intensive care, the overall mortality of RDS is about 15%, and one third of infants born before 30 weeks of gestational age die of this disorder. Although the other choices are associated with bleeding, they are unlikely causes of periventricular hemorrhage in a baby with RDS.

Diagnosis: Respiratory distress syndrome of the neonate. (Lippincott's, p 68)

### **9. C. True ependymal rosette.**

This rosette (seen in ependymoma) consists of tumor cells surrounding an empty lumen. It is thought that these structures represent attempts by the tumor cells to recreate little ventricles with ependymal lining. One thing to note: although these guys are characteristic of ependymoma, they're not seen in every case. In fact, it's fairly uncommon to find them at all (they're only present in a small percentage of well-differentiated ependymoma). Diagram of true ependymal rosette. A halo of cells surrounds an empty lumen. Photomicrograph from an ependymoma showing several true ependymal rosettes. The halo-like cluster of cells in each rosette surrounds an empty central lumen (H&E; original magnification 400×).

### **10. C. Medulloblastoma.**

This rosette, named for James Homer Wright, the first director of the Massachusetts General Hospital, is typically seen in neuroblastomas, medulloblastomas, and primitive neuroectodermal tumors (PNETs). It consists of a halo of tumor cells surrounding a central region containing neuropil (hence its association with tumors of neuronal origin).

Diagram of Homer Wright rosette. A halo of cells surrounds a central hub that contains a meshwork of fibers.

Photomicrograph from a PNET demonstrating multiple Homer Wright rosettes. The halo-like cluster of cells in each rosette surrounds a central area of fiber-rich neuropil (H&E; original magnification 400×).

Rosettes are little round groupings of cells found in tumors. They usually consist of cells in a spoke-wheel or halo arrangement surrounding a central, acellular region. Rosettes are so named for their resemblance to the rose windows found in gothic cathedrals (check out the beautiful rose window in the Cathedral of Notre-Dame in Strasbourg).

There are a bunch of different kinds of rosettes, each with different types of cells and different names. Most of them are found in tumors of the nervous system. It's useful to be able to recognize these, because they help with the diagnoses of difficult tumors.



Rosette patterns and associated tumors

Rosette type	Associated tumors
Homer Wright	Neuroblastoma, medulloblastoma, PNET
Flexner-Wintersteiner	Retinoblastoma, pineoblastoma
True ependymal	Ependymoma
Perivascular pseudorosette	Ependymoma, medulloblastoma, central neurocytoma, glioblastoma,
Pineocytomatous	Pineocytoma
Neurocytic	Central neurocytoma

#### 11. D. Perivascular pseudorosette.

This rosette consists of tumor cells collected around a blood vessel. It's called a pseudorosette because the central structure isn't part of the tumor. In the past, the term pseudorosette meant any rosette that didn't have a truly empty lumen...but it seems that designation is kind of outdated. These rosettes are common in ependymomas, but you also see them in medulloblastoma, PNET, central neurocytomas, and glioblastomas.

Diagram of perivascular pseudorosette. A halo of cells surrounds a blood vessel. Photomicrograph from an ependymoma showing 2 prominent perivascular pseudorosettes. The halo-like cluster of cells in each rosette surrounds a blood vessel. Note the several smaller true ependymal rosettes (H&E; original magnification 200×).

### **12. E. Flexner-Wintersteiner rosette.**

This rosette (named for pathologist Simon Flexner and ophthalmologist Hugo Wintersteiner) is characteristic of retinoblastomas. It consists of tumor cells surrounding a central lumen that contains cytoplasmic extensions from the tumor cells. If you look at the tumor cells under electron microscopy, they have features of primitive photoreceptor cells.

Diagram of Flexner-Wintersteiner rosette. A halo of cells surrounds a largely empty central hub. Small cytoplasmic extensions from the cells project into the lumen.

Photomicrograph from a retinoblastoma showing multiple Flexner-Wintersteiner rosettes. The halo-like cluster of cells in each rosette surrounds a nearly empty appearing central lumen containing fine cytoplasmic processes (H&E; original magnification 400×).

### **13. A. Dolichoectasia.**

Location: This is a view taken from the base of the brain and the structure being labeled is the basilar artery. Dolichoectasia: (dolicho- means long). The artery is dilated, with a rigid appearance and have yellowish discoloration. These changes are diagnostic for atherosclerosis and the vessel is also dilated. It is also elongated, widened, and tortuous. These features are that of dolichoectasia and are common in patients with advanced atherosclerosis. The most common sites are the basilar artery and the changes can extend to the vertebral artery. The other common site is the supraclinoid segment of the internal carotid artery with possible extension into the middle cerebral artery. They have also been described in young non-atheromatous patients and some of them are associated with Ehler-Danlo's syndrome type IV, Marfan's syndrome, pseudoxanthoma elasticum, and  $\alpha$ 1-antitrypsin deficiency. Schwannomas occur in this location (the cerebellar-pontine angle). The structure in question is a blood vessels and does not have the slightest suggestion of a tumor.

Berry (saccular) aneurysms, as reflected by its name, appear as small berry-like structures attached to the blood vessels. They are usually thin-walled. Berry aneurysms are most commonly found at or very close to the point of bifurcation of intracranial arteries with the circle of Willis as the most common site.

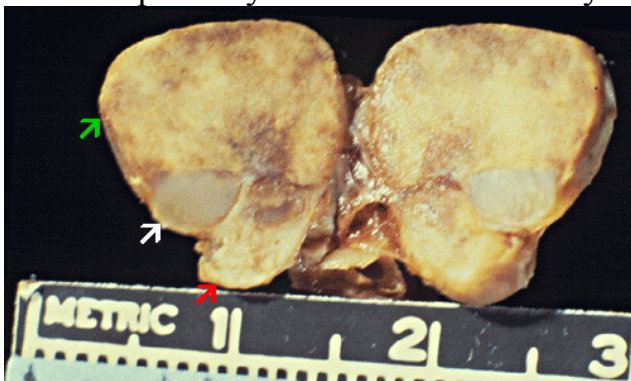
Charcot-Bouchard aneurysms, also known as miliary aneurysms or microaneurysms, occur in small arteries around 100-300  $\mu$ m in diameter. They are found most commonly at the putamen, globus pallidus, and thalamus. They can also be seen in other areas such as the caudate, internal capsule, centrum semiovale, and cortical gray matter. In most cases, they are associated with hypertension.



**14. C. Gait ataxia.**

This specimen is a transverse section through the brainstem and cerebellum. There is a large area of discoloration and disturbed anatomy in the left cerebellar hemisphere that is producing little mass effect. Because this is the only lesion postulated for this patient, there is no reason to suspect seizure activity, because that phenomenon would be unlikely in the absence of a cerebrocortical (or at least cerebral) lesion. The other findings listed would similarly not be expected in a patient with cerebellar damage. (Bradley, pp 1932–1933.)

**15. E. A pituitary with a Rathke cleft cyst.**



The shape of the specimen is consistent with the bisected nodular structure that is consistent with a pituitary gland. The anterior (↗) and posterior (↘) pituitary can be well recognized. A cystic structure containing consolidated gelatinous material (↖) is present and is most consistent with a Rathke cleft cyst.

**16. D. Ruptured berry aneurysm.**

Rupture of a berry aneurysm involving a cerebral artery at the circle of Willis and its branches is a sudden event that produces hemorrhage into the subarachnoid space at the base of the brain, as shown here.

**17. E. Glioblastoma multiforme.**

**18. E. Sphenoid ridge meningioma.**