A II-type: Select the most appropriate answer

(  ) 1. 78-year-old woman experienced an intracerebral hemorrhage (ICH) at the left frontal region (Fig a). Four years later she experienced another at the right frontoparietal lobe with secondary subarachnoid hemorrhage (Fig. b).

Post-mortem necropsy had the following finding. Which is the diagnosis?
A. Primary CNS angiitis
B. Cerebral amyloid angiopathy
C. Glioblastoma multiform with recurrent tumor bleeding.
D. Hypertensive intracerebral hemorrhage.
E. Multiple intracerebral aneurysms

Ans: (B)
2. A 36-year-old hypertensive woman recovered from right hemisphere TIAs (left hemiparesis) after right carotid artery stenting. Four months later, she had several episodes of left hemisphere TIA. Her angiography was as follows. What is the most probable diagnosis?

A. Giant cell arteritis.
B. Atherosclerosis
C. Dolichoectasia
D. Fibromuscular dysplasia
E. CADASIL

Ans: (D)
3. A 12-year-old boy suffered from deteriorated performance in school, gait disturbance, and progressive visual loss. Physical examination revealed generalized hyperpigmentation. Brain MRI revealed image below. What’s the most possible diagnosis?
A. Niemann-Pick disease type C
B. GM2-gangliosidoses
C. Prader-Willi syndrome
D. Adrenoleukodystrophy
E. Cerebrotendinous xanthomatosis

Ans: (D)
4. The 10-year-old boy had acute onset of fever, neck stiffness, motor seizure, and altered consciousness. CSF study: 10 cells/mm3 with lymphocyte predominant, sugar: 50mg/dl. Brain MRI showed as followed. Which one is the most likely diagnosis?

A. HSV-1 encephalitis
B. Eastern equine encephalitis
C. St. Louis Encephalitis
D. Subacute sclerosing panencephalitis
E. Japanese encephalitis

Ans: (A)
5. A 19-year-old female patient got seizure disorder with cutaneous disorder as following picture. Which is the most likely diagnosis?

A. Tuberous sclerosis
B. Neurofibromatosis type II
C. Sturge-Weber-Dimitri syndrome
D. Incontinentia pigmenti achromians
E. Von Hippel-Lindau disease

Ans : (A)
6. An 18-year-old woman presents because of two generalized tonic-clonic seizures that have occurred over the past year. Each episode occurred in the morning and she recalls staying up late the night before each seizure. She also described a 2-year history of occasional brief bilateral myoclonic jerks of her arms in the morning. Her maternal aunt had a history of seizure. Neurologic exam and brain MRI showed normal. EEG shows as following. Which of the following is the mostly diagnosis?

A. Juvenile myoclonic epilepsy
B. Unverricht-Lundborg disease
C. Myoclonic epilepsy with ragged red fibers
D. Autosomal dominant nocturnal frontal lobe epilepsy
E. Lafora disease

Ans: (A)
7. A 42 year old male has had increasing difficulty with activities of daily living for the past year, mainly because of choreiform movements. His family has also noted that he has exhibited behavioral changes, though his memory remains intact. His brother is similarly affected. Based upon the appearance of the coronal section of brain shown here, the disease most likely to explain these changes is:

A. Huntington’s disease
B. Parkinson’s disease
C. Alzheimer’s disease
D. Amyotrophic lateral sclerosis
E. Multiple sclerosis

Ans : (A)
8. A 30-year-old girl with severe headache and left hemiparesis. Axial CT without contrast enhancement demonstrates

The most possible diagnosis is
A. Subarachnoid hemorrhage
B. Arteriovenous malformation
C. Subdural hemorrhage
D. Cerebral venous thrombosis
E. Cardioembolic infarction

Ans: (D)
9. A 74-year-old man complained persistent dizziness and unsteadiness. Below is the MRI film. Which is the finding?

A. occlusion of left vertebral artery
B. occlusion of right vertebral artery
C. right cerebellopontine tumor
D. left cerebellar atrophy
E. nasopharyngeal tumor

Ans: (B)
10. A 8-year-old female patient has seizure, right hemiplegia, mental retardation, and glaucoma. The photographic picture above showed the facial skin lesions and enhanced-T1 weight MRI. She is a patient of
A. Sturge-Weber syndrome
B. Neurofibromatosis
C. Tuberous sclerosis
D. Incontinentia pigmenti
E. Von Hippel-Lindau disease

Ans : (A)
11. A patient with optic nerve gliomas and presents with appearance showed like as figures below. What could be the disease most likely?

A. Wilson disease
B. Neurofibromatosis type 1
C. Leber’s optic Neuropathy
D. Neurosyphilis
E. Tuberous sclerosis

Ans : (B)
A 35-year-old man experienced progressive weakness for ten years. His father and two elderly sisters also have the same problems. When asked to elevate his arm laterally, he has much difficulty. (see the following pictures). Muscle biopsy showed the following findings. Eye fundi examination disclosed retinal vasculopathies. EKG was normal. Which would be the probable diagnosis?

A. Becker muscular dystrophy.
B. Facioscapulohumeral muscular dystrophy
C. Myotonic dystrophy
D. Inflammatory myopathy.
E. Spinomuscular atrophy type 3.

(Gomri-Trichrome) (ATPase 4.3)

Ans: (B)
13. A 42-year-old man experienced progressive memory decline and emotional liability for several years. He used to be a migraineur since age 25. No other systemic diseases such as hypertension and diabetics had been documented. His father and grandfather both died at around age 60. Physical examinations disclosed generalized increased DTRs and presence of the left Babinski’s sign. His brain MRI is as follows. Which of the following diagnosis is most likely?

A. Binswanger’s disease
B. Multiple sclerosis
C. Progressive multifocal leukoencephalopathy
D. CADASIL
E. Moya-Moya disease

Ans: (D)
14. Abnormal nerve conduction study depicted below could commonly be seen in the following conditions, except?
A. Hereditary neuropathy with liability to pressure palsy (HNPP)
B. Charcot-Marie-Tooth 1A (CMT1A)
C. Multiple motor neuropathy
D. Acute axonal injury for 1 day
E. Guillain-Barre syndrome

Ans: (B)
15. A 35-year-old lady with history of migraine for 2 years suffered from an episode of acute cerebral infarct with good recovery. Brain MRI showed picture depicted below. What is the most possible diagnosis?

A. MELAS (mitochondrial encephalomyopathy, lactic acidosis and stroke-like episode)
B. CADASIL (cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathies)
C. Binswanger’s diseases
D. Syndrome X
E. Progressive multifocal leukoencephalopathy

Ans: (B)
16. A 22-year-old female present with episodes of dizziness, left side numbness and mild left hand weakness. Brain images are shown below.

Which of the following diagnosis is most likely?

A. Multiple lacuna stroke
B. Multiple sclerosis
C. Acute disseminated encephalomyelitis
D. Tuberculoma of central nervous system
E. Autoimmune disease

Ans: (B)
17. Which description is correct about the marked waveform?
   A. can be augmented by the movement of contralateral limb
   B. can be augmented by the movement of ipsilateral limb
   C. can be suppressed by pure imaginary movement
   D. more prevalent in old ages
   E. is pathognomonic for Rolandic seizure

Answer: (C)
18. A 65 year-old female patient complained of numbness over the left shoulder and arm for two years. The results of SSEP study with stimuli at the left median nerve are as followed.

What is the most likely diagnosis?
A. Cervical myelopathy
B. Left brachial plexopathy
C. Right thalamus lesion
D. Left C5-6 radiculopathy
E. Right parietal lobe lesion

Ans: (B)
19. Severe and daily headache for 2 years in a 65-year-old man without increased intracranial pressure (IICP) (T1 MRI with contrast). What is the most likely diagnosis?

A. spontaneous intracranial hypotension
B. TB meningitis
C. idiopathic hypertrophic pachymeningitis
D. idiopathic intracranial hypertension (pseudotumor cerebri)
E. Meningeal carcinomatosis

Ans: (C)
A 16-year-old girl developed nystagmus, dizziness and gait disturbance for 2 weeks. The patient died in 4 days after admission (A: MRI with contrast, B: gross and C histology of neuropathology). Which of the following is correct for her?

A. The enhanced region (arrow) of the MRI is pineal body
B. The gross pathology showed infarction
C. Microscopy revealed perivascular hemorrhage, which is typical for viral infection
D. Treatment should include thiamine
E. The disease is not reversible

(A) T1 with contrast, (B) gross anatomy, and (C) histopathology

Ans: (D)