



**Association of British Neurologists**  
**Self Assessment Exercise 2006**

**QUESTION 1**

A 40-year-old female gave a 4 month history of daily headache which had started during a bout of "flu". She described a bilateral pressure feeling, occasionally associated with nausea but no photophobia or phonophobia. She would awaken headache free, but the headache would evolve soon after getting up and worsen through the day. She did not usually suffer headaches other than occasional "tension" headaches, and there was no family history of headache. She was a non-smoker and drank 7 units of alcohol per week. She was not taking any regular medication but used ibuprofen 2-3 times per week.

On examination there were no abnormal findings.

The following results were normal:

- CT brain scan
- ESR
- Autoantibody screen.

Which of the following is the most appropriate next step?

**ANSWER OPTIONS**

- A: Trial of indomethacin
- B: Trial of a suitable migraine prophylactic agent
- C: Lumbar puncture
- D: MRI brain with gadolinium
- E: Withdraw ibuprofen

## QUESTION 2

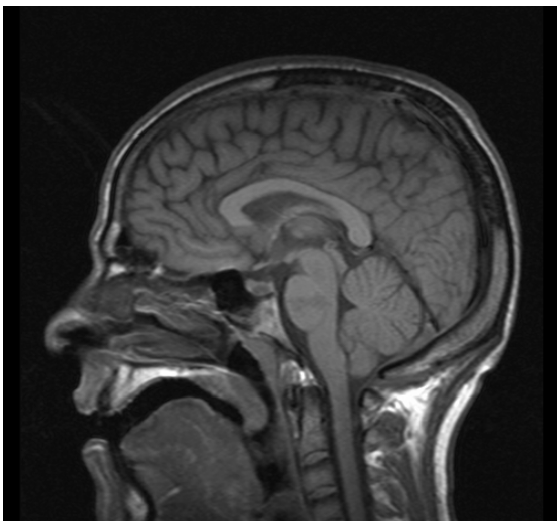
**History:** A 30 year-old man presented with a 5-month history of a constant, dull frontal headache of moderate severity, worst when bending over. It was accompanied by nausea, phonophobia and photophobia. For 2 weeks prior to admission he had lost patches of hair over his scalp, some of his body hair, and his eyelashes. The week prior to presentation he had developed an evolving left hemiparesis, involving the left side of his face. Six months prior to presentation he noted a widespread skin rash with malaise and lymphadenopathy, but had not sought medical attention. He was homosexual, but had no sexual contact for 18 months.

**Examination:** He had generalised alopecia, angular cheilitis, oral aphthous ulceration and ulcerative gingivitis. He had a mild dysarthria, left facial weakness sparing the forehead, and a left hemiparesis (grade 0 in the arm, and grade 2 in the leg). Deep tendon reflexes were brisk throughout and the left plantar response was extensor. The rest of the examination was normal.

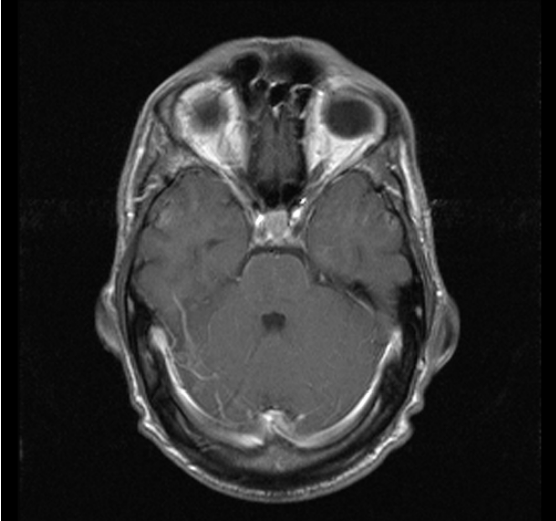
**Investigations:**

- FBC normal other than lymphocytes  $0.95 \times 10^9/l$  (1.5 to 4.0)
- U&E, random glucose, LFT, TFT, B12 normal
- CRP 18
- Hepatitis serology normal.
- Serum EBV and CMV IgG antibodies positive
- Toxoplasma gondii IgG antibody negative.
- CT brain: normal.
- MRI brain: see figure.
- CSF: opening pressure 15cm, protein 2.31g/l, glucose  $<1.1$  mmol/l (paired serum glucose 6.7 mmol/l), WCC 165/mm<sup>3</sup> (polymorphs 90%, lymphocytes 10%), RCC 840/mm<sup>3</sup>, cryptococcal antigen negative, PCR for JC virus, CMV, HSV 1&2, and VZV negative.

**Figure A:** T1 sagittal non-contrast MRI



**Figure B:** T1 axial contrast enhanced MRI



**Figure C:** T2 axial MRI



What is the most likely diagnosis?

ANSWER OPTIONS

- A: Progressive multifocal leukoencephalopathy
- B: HIV seroconversion
- C: Cerebral toxoplasmosis
- D: Meningovascular syphilis
- E: Tuberculous meningitis

### QUESTION 3

In which of the following individuals would it be most appropriate to consider chronic subthalamic nucleus stimulation?

#### ANSWER OPTIONS

- A: A 45-year-old man with a 14 year history of Parkinson's disease suffering from frequent 'on-state' freezing episodes each day, in spite of optimal medical treatment.
- B: A 52-year-old woman with a one year history of a rapidly progressive severe bilateral akinetic-rigid form of parkinsonism not responding to levodopa.
- C: A 72-year-old woman with a 18 year history of Parkinson's disease having frequent falls in spite of optimal medical treatment.
- D: A 59-year-old man with a 12 year history of Parkinson's disease experiencing severe frequent dyskinesias whilst in the 'on state' each day.
- E: A 45-year-old man with a history of hypertension having 'lower half' parkinsonism, consisting mainly of gait difficulties.

#### **QUESTION 4**

A 13-year-old boy presented with an 18 month history of failing at school, in part due to problems with bad behaviour and poor concentration. Of late it had also been noted that he had become slower in his movements. He had a normal birth and developmental history, but in the family history his father had died a year ago, at the age of 42, from a neurological disorder that had rendered him bed bound and demented over a 15 year period. His older sister and mother were well with no neurological problems.

On examination he was hard to engage, was bradykinetic with markedly abnormal eye movements, with an almost complete absence of saccades and a very unstable gait. On neuropsychological testing he had clear deficits in a number of domains, but especially with tasks sensitive to the frontal lobe.

His MRI brain was normal as was his routine blood testing including copper studies.

What is the most likely diagnosis?

#### **ANSWER OPTIONS**

- A: New variant CJD (nvCJD)
- B: Juvenile Huntington's disease (JHD)
- C: Metachromatic leukodystrophy (MLD)
- D: Attention deficit hyperactivity disorder (ADHD)
- E: Parkin positive Parkinson's disease

## QUESTION 5

A 63-year-old woman developed transient deafness on the left, followed 24 hours later by severe persistent left-sided deafness, vertigo, loss of balance and left-sided cerebellar signs.

Which of the following disorders best explains this syndrome?

### ANSWER OPTIONS

- A: Anterior inferior cerebellar artery (AICA) infarct
- B: Ménière's disease
- C: Posterior inferior cerebellar artery (PICA) infarct
- D: Ramsay Hunt syndrome (Herpes Zoster oticus)
- E: Vestibulo-cerebellar neuritis

**QUESTION 6**

A 73yr old man presented with sudden-onset headache behind his right eye after cleaning his pond. The severe headache persisted for several days. He was otherwise asymptomatic. He had a CT scan of his head with contrast which showed the abnormality evident in Figure 1. There were no other abnormalities. His headache resolved and he was discharged.

Two weeks later he developed a similar, although milder, headache following sexual intercourse. The following morning he found he could not see all his fingers, missing those in the left visual field. On admission he was found to have a complete left homonymous hemianopia. A further CT scan of the head was performed (Figure 2). CT angiography was normal.

Figure 1: CT scan of the head with contrast (two cuts)

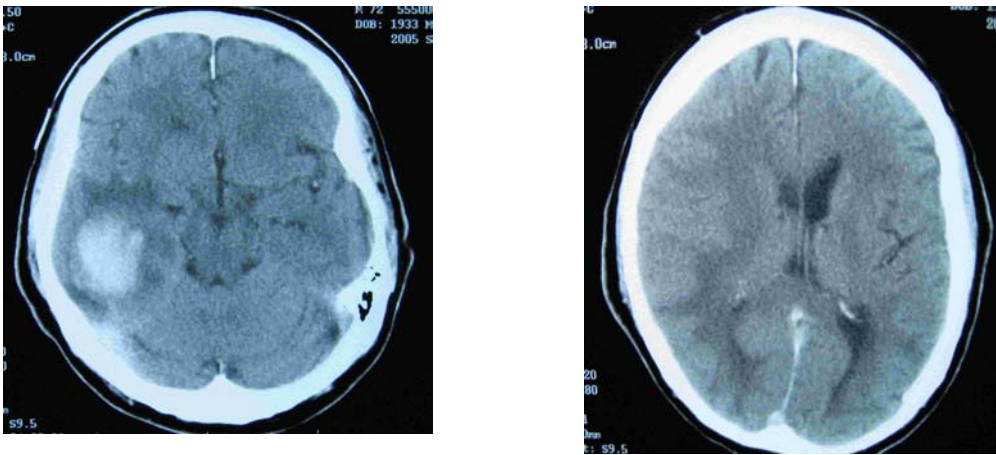
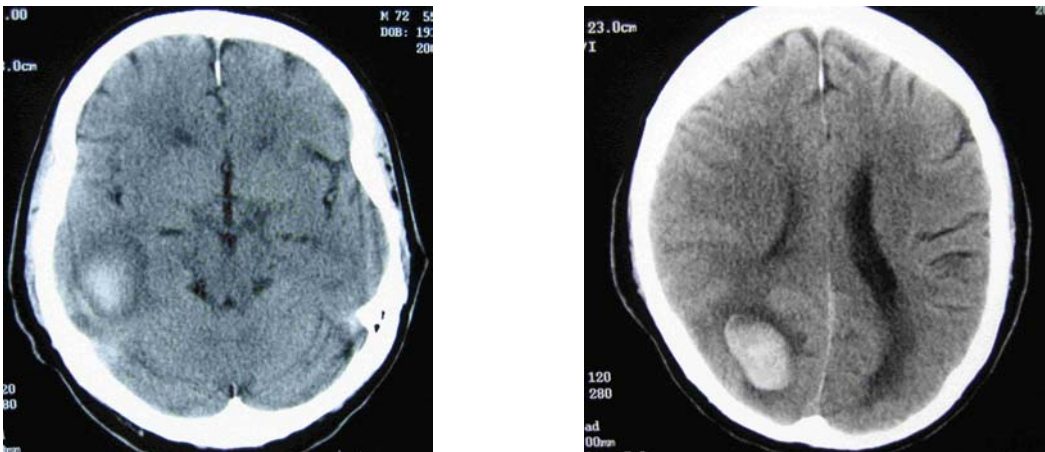


Figure 2: CT Scan of the head (equivalent cuts to original scan)



What is the most likely underlying diagnosis?

ANSWER OPTIONS

- A: Aneurysmal bleed
- B: Arteriovenous malformation
- C: Cerebral amyloid angiopathy
- D: Hypertensive bleed
- E: Metastatic disease

## QUESTION 7

You are asked to see a 56-year-old man on a general surgical ward.

He has Crohn's disease and ankylosing spondylitis. As a young man, he required multiple GI operations, but his inflammatory bowel disease has been relatively quiescent for some years. He had no family history of note and he takes NSAIDs and sulphasalazine.

He was admitted under the surgeons with a six month history of weight loss, abdominal pain and watery diarrhoea, which grew nothing on culture. Eventually, he had a laparotomy at which some adhesions were found, but no evidence of active inflammatory bowel disease. His symptoms settled, more or less, but he failed to mobilise after the operation, because of "dizziness".

You elicit a history of erectile impotence for five years and 10 years of urinary frequency and nocturia for longer, attributed by him to "prostate". He admits to having intermittently felt "dizzy" on standing for at least three years, and to "blacking out" on three occasions in the last year when getting out of bed at night.

On examination, lying down, there are no abnormal signs apart from poor constriction of the pupils to light. Power, sensation and reflexes are normal and there are no signs of parkinsonism. However, he is unable to stand for more than a few seconds, because of postural hypotension: 160/90 lying, 65/undetectable standing.

Blood and urine tests done on the surgical ward, including tests to diagnose diabetes, are normal or negative. An ECG shows sinus rhythm with no R-R variation on respiration.

Which diagnosis best accounts for the clinical picture?

### ANSWER OPTIONS

- A: Multiple System Atrophy
- B: Autonomic neuropathy due to primary amyloidosis
- C: Primary autoimmune autonomic neuropathy
- D: Autonomic neuropathy due to amyloidosis secondary to chronic inflammation
- E: Sjogren's syndrome

## QUESTION 8

A 40-year-old man developed progressive weakness and numbness of the legs then arms over 8 weeks. On examination he was bed bound with symmetrical weakness of upper and lower limbs, MRC grade 2-3 proximally and distally. Tendon reflexes were absent. There was glove-and-stocking sensory loss to all modalities. Cranial nerves were normal apart from bilateral papilloedema. His spleen tip was palpable.

Investigations showed:

Full blood count, renal and liver function, calcium all normal

Biochemical evidence of hypothyroidism

IgA lambda paraprotein 2g/L

Immunoglobulin levels otherwise normal

CT brain scan normal

CSF pressure and contents normal

Skeletal survey: solitary sclerotic lesion in right ilium

What is the most likely diagnosis?

### ANSWER OPTIONS

- A: Chronic inflammatory demyelinating polyradiculoneuropathy with monoclonal gammopathy of undetermined significance (CIDP-MGUS)
- B: Guillain Barré syndrome with MGUS
- C: Multiple myeloma
- D: POEMS syndrome
- E: Primary systemic amyloidosis

## QUESTION 9

A 27-year-old Vietnamese man presented to Casualty with rapidly progressive weakness of the limbs which had developed over the previous 3 hours, following a heavy meal. On examination, cranial nerves were normal. There was profound proximal and distal weakness of all 4 limbs. Tendon reflexes were absent. Sensory examination was normal. The admitting medical team managed him for Guillain-Barré syndrome but by the next morning he had returned to normal.

What is the most likely underlying endocrinopathy?

### ANSWER OPTIONS

- A: Hyperaldosteronism
- B: Hyperthyroidism
- C: Hypoaldosteronism
- D: Hypopituitarism
- E: Hypothyroidism

## QUESTION 10

An otherwise healthy man aged 26 was in a psychiatric hospital undergoing an addiction treatment programme for cocaine and alcohol for 10 days. He had been receiving multivitamins and thiamine. On day 11 he went for a fast swim, including a strenuous 25m underwater swim. The following day he passed dark urine on three occasions and his muscles felt a little sore. He had a serum creatine kinase of 30,000 U/L and when this result was discovered two days later he was admitted to the local district general hospital. He recalled having had at least three similar episodes of dark urine after exercise since the age of 20. There was no family history of relevance. General and neurological examination were normal. The serum creatine kinase was now 3,000. Urea and electrolytes and FBC and ESR were normal. He received 2.5 l of saline and was discharged on the following day. Five days later, the serum creatine kinase, electromyography and nerve conduction were normal. An ischaemic exercise test showed normal lactate elevation. A muscle biopsy was planned.

What is the most likely diagnosis at this stage?

### ANSWER OPTIONS

- A: Malignant neuroleptic syndrome
- B: Polymyositis
- C: A form of muscular dystrophy
- D: A disorder of muscle glycogen metabolism (e.g. phosphorylase deficiency)
- E: A disorder of muscle lipid metabolism (e.g. carnitine/carnitine palmitoyl transferase deficiency)

## QUESTION 11

A mildly obese 70-year-old man presented with a one year history of episodes of movements in sleep, occurring most nights, sometimes on several occasions in one night. He himself was not aware of the movements but he was brought to medical attention by his wife, who described stereotyped repetitive flexion/extension movements, apparently confined to the legs, lasting for a few minutes at a time. They sometimes disturbed his sleep and, after bad nights, he felt tired the next day and fell asleep easily. He had a 10 year history of diabetes, treated with dietary restriction and an oral hypoglycaemic agent. Six years earlier, he had had a sudden onset of a left hemiparesis that resolved over two weeks, diagnosed as an ischaemic stroke without investigation. Five years previously, he had had a single generalised tonic-clonic seizure, assumed to be secondary to the stroke, without investigation or treatment. He was reported to snore on some occasions. Neurological examination was generally unremarkable, although the ankle jerks were absent and he had lost vibration sense below the knees.

Which one of the following statements, relating to the episodic movements, is most likely to be true?

### ANSWER OPTIONS

- A: The movements are likely to occur in REM-sleep phases.
- B: The most likely diagnosis is epilepsy.
- C: The symptoms may respond to an L-DOPA containing drug.
- D: The problem is likely to be secondary to sleep apnoea.
- E: A cause needs to be sought with cerebral MRI

## QUESTION 12

The general medical team requested a neurological review of a 49-year-old woman who had been admitted a week previously. She had been brought to casualty by her husband following repeated falls at home. Although the patient herself was alert and attempting to be co-operative, she was not able to provide a coherent history. Her husband said that she had, in retrospect, been having increasing difficulty for the previous year. Initially this had manifest with disengagement from her hobbies and friends. She had been treated for depression with fluoxetine by her GP but this had not helped. Things had worsened gradually such that in the week prior to admission she had begun to lose her way around the house, had become incontinent of urine and then started to fall frequently, culminating in her admission.

She scored 10 out of 30 on the Mini-Mental State Examination. Neurological examination was otherwise normal. Blood test results showed normal full blood count, urea and electrolytes, liver function, calcium, thyroid function, B12 / folate and VDRL. ESR was 28.

Plain CT scan performed on admission by the general medical team showed ventricular dilatation but no other changes. Cerebrospinal fluid examination had shown the following: Opening pressure 18 cm of water, 5 white cells (mononuclear), no red cells, protein 0.75 g/l, glucose 3.5 mmol / l (blood 5.2). No sample sent for cytology.

What is the most appropriate next management step?

### ANSWER OPTIONS

- A: EEG
- B: Removal of 30 mls of CSF followed by 10m timed walking test
- C: MR brain imaging with gadolinium
- D: Start treatment with cholinesterase inhibitor
- E: Urgent referral to the neurosurgical team to request ventriculoperitoneal shunt placement

### QUESTION 13

A neurological review was requested for an unconscious 65-year-old man, who had been admitted 4 weeks before with a proven subendocardial myocardial infarction. Three weeks later he had had 2 coronary by-pass grafts, but his postoperative course had been complicated by a period of ventricular fibrillation lasting about 7 minutes. There had been subsequent haemodynamic instability requiring a temporary intra-arterial balloon pump.

On weaning him from the ventilator he had been found to be unresponsive to all painful stimuli. He made inadequate respiratory efforts, and coughed to suction; he had normal pupillary, corneal and doll's head movements. He had not had Propofol for 24 hours, and his urea, electrolytes, glucose and arterial gases were normal.

A representative slice of a CT brain scan is shown:-



What is the most likely cause of the patient's unresponsiveness?

#### ANSWER OPTIONS

- A: Diffuse cortical ischaemia
- B: Brain stem ischaemia
- C: The right frontal infarction
- D: Delayed excretion of anaesthetic drugs
- E: Critical illness neuropathy

**QUESTION 14**

A 48-year-old man was seen in clinic complaining of jerkiness of his arms and stiffness of his neck. He recalled being jerky as a teenager, without ever having had a seizure. He suffered from depression and anxiety in his early 20s, and was prescribed trifluoperazine for 9 months, after which he took diazepam regularly until his late 30s. During this time, his movements were much less prominent. Since stopping medication, his mental state had remained stable, although he admitted to being obsessional. He subsequently noted that his jerks, predominantly affecting the right arm, trunk and face and sparing the legs, had returned. Over the few years prior to referral he had noticed increased stiffness of his neck, which tended to turn backwards and to the right, and that his writing had become jerky. He did not smoke, but drank alcohol regularly. The family history was limited, but his father was said to have had limb jerks, to have been depressed, and to have died of liver failure.

On examination the MMSE was 28/30. There was a jerky torticollis to the right with a degree of retrocollis. The eye movements were normal. Intermittent brief facial jerks were seen, but there was no blepharospasm, or oro-facial dyskinesia, and tongue and palatal movements were normal. In the limbs there were prominent brief jerks of the right hand and arm on movement, which were present to a lesser extent on the left and trunk. Writing was effortful and jerky, but there was no bradykinesia or rest tremor. There were no cerebellar signs. Lower limb examination was entirely normal. The reflexes were all present and the plantar responses were flexor. The gait was normal.

What is the most likely diagnosis?

**ANSWER OPTIONS**

- A: Huntington's disease
- B: Myoclonus-dystonia
- C: Neuroacanthocytosis
- D: Tardive dyskinesia
- E: Tourette's syndrome

## QUESTION 15

A 29-year-old Australian Caucasian female was admitted following a collapse. She lost consciousness for approximately one minute; there was no witnessed seizure activity. When she awoke she had expressive dysphasia but no other neurological symptoms. She had no relevant past medical or family history and the only regular medication was the oral contraceptive pill. She consumed alcohol occasionally, was a regular smoker (10/day) and admitted to smoking cannabis but denied any other recreational drug abuse.

On examination she was afebrile, in sinus rhythm with a BP of 120/65 mmHg in both arms. Examination of the cardiovascular and respiratory systems was normal. She had expressive dysphasia but no other neurological signs.

The only abnormal blood results were: ESR 66 mm/hour; total cholesterol 6.5 mmol/L. The following investigations were normal or negative: full blood count, renal, liver and thyroid function, CRP, syphilis serology, coagulation, vitamin B12, red cell folate, glucose, proteins C and S, activated protein C resistance, antithrombin III, autoantibody screen, lupus anticoagulant.

An initial CT demonstrated a left middle cerebral infarct. The patient was commenced on high dose aspirin and admitted for further investigations. The following morning she awoke with a dense right hemiplegia and complete aphasia. MRI confirmed a large left MCA territory infarct with a large clot in the left carotid artery. A transthoracic bubble echocardiogram was normal. A CT angiogram demonstrated a large ring of abnormal soft tissue surrounding the aortic arch, extending cranially along both the left common carotid and, to a lesser extent, the brachiocephalic artery, causing severe narrowing of the left common carotid artery.

What is the most likely diagnosis?

### ANSWER OPTIONS

- A: Cerebral vasculitis
- B: Fibromuscular dysplasia
- C: Patent foramen ovale
- D: Takayasu's arteritis
- E: Todd's paresis

## QUESTION 16

A 30-year-old woman presented with a two-month history of recurrent attacks of generalised shaking associated with loss of consciousness. These were happening up to four times a week. She said that she usually had no warning (although sometimes had had a weird 'aura' before them) and afterwards felt drowsy. She had been incontinent of urine on several occasions and had also sustained carpet burns to her face. Her partner, who had witnessed a number of them, said that she looked quite flushed and upset for several minutes before the attack. The shaking lasted on average around 10 minutes and could include pelvic thrusting. During an attack, her eyes and mouth tended to be shut and afterwards she was often tearful.

She had a history of irritable bowel syndrome and repeated laparoscopy for pelvic pain. She had been under psychiatric care and had received diagnoses of borderline personality disorder, depression and post-traumatic stress disorder. There was a history of recent domestic violence. Subsequent investigation with videotelemetry confirmed the clinical suspicion of non-epileptic attacks.

In hindsight, which of the following aspects of the history was most persuasive for this diagnosis?

### ANSWER OPTIONS

- A: Borderline personality disorder
- B: Pelvic thrusting during the attack
- C: The duration of the attacks
- D: The history of domestic violence
- E: The history of irritable bowel syndrome and pelvic pain

## QUESTION 17

A 54-year-old milkman was referred from the regional chest unit with a query whether he might have motor neurone disease. He had been referred from his local hospital because of daytime somnolence and found to have severe hypercapnia ( $PO_2=9.5$  kPa,  $PCO_2=11$  kPa). An overnight sleep study with video telemetry revealed frequent oxygen desaturation associated with periods of complete apnoea for up to 30 seconds. These episodes were followed by arousals with vocalisations and thrashing around. The patient had no recall for these events.

He was treated with nocturnal non-invasive positive pressure ventilation with complete resolution of daytime symptoms and the nocturnal desaturation. The chest physicians thought they could observe fasciculations in the arms and hands and referred him to the neurology department.

On examination he had a mild degree of cogwheel rigidity and unsteadiness when walking. What had been interpreted as fasciculation was in fact low amplitude myoclonus. A clinical diagnosis was made and the patient was discharged for outpatient follow up. Two weeks later he was found dead by his wife one morning.

Which of the following inclusion bodies was found at autopsy?

### ANSWER OPTIONS

- A: Glial cytoplasmic inclusions
- B: Hirano Bodies
- C: Lewy bodies
- D: Tau positive inclusions
- E: Ubiquitinated inclusions

## QUESTION 18

A 35-year-old woman presented with 3 drop attacks over a period of 6 years. These would occur without warning or provocation. Her legs would give way beneath her without any loss of awareness or consciousness and she would be able to rise immediately after. In addition she complained of feeling a little off balance when walking.

She walked at 11 months, was never good at sports at school but learned to ride a bicycle. There was no family history of neurological disease.

General examination was normal. Cranial nerves were normal. Muscle bulk and tone were normal in all 4 limbs. There was mild symmetrical weakness of hip flexion/extension, and knee flexion (MRC 4+). Upper limb reflexes were normal, knee jerks brisk, ankle jerks just present without reinforcement, plantars mute. There was mild 4 limb ataxia and mild gait ataxia. Romberg's test was negative. Sensory examination was normal.

The following investigations were normal:

FBC, ESR, U&Es, LFTs, Calcium, TFTs.

MRI brain and CSF analysis (protein, glucose, cell count)

Muscle biopsy (including oxidative stains)

Abnormal results:

Serum lactate: 3.00 mmol/l (0.5-2.00).

EEG: markedly abnormal, with florid, generalised, short lived polyspike and wave complexes on a slow background.

What is the most likely diagnosis?

### ANSWER OPTIONS

A: Baltic myoclonus

B: MELAS

C: MERRF

D: SCA 2

E: Sialidosis

## QUESTION 19

A 45-year-old man presented with a history of evolving sensory disturbance in the lower limbs and a depressive illness, unresponsive to treatment. He subsequently developed memory problems and was seen in a neurology clinic by a specialist registrar, who included variant Creutzfeldt-Jakob disease in his differential diagnosis.

Regarding investigations, which of the following is the least compatible with variant CJD?

### ANSWER OPTIONS

- A: Generalised periodic EEG discharges
- B: Methionine homozygosity at codon 129 of the prion protein gene
- C: Negative CSF 14-3-3 protein
- D: Raised CSF protein
- E: Symmetrical posterior thalamic hyperintensity on MRI

## QUESTION 20

A 42-year-old scientist, presented to her General Practitioner with blurring of the vision in the infero-nasal quadrant of her right eye. This progressed over about a day, but never became a complete scotoma – there was just marked blurring. She had previously been well, there was no pain, and there were no other neurological symptoms. She had never travelled outside the country other than to attend conferences. An optometrist was unable to find any abnormality on automated perimetry and sent her to an ophthalmologist. On examination, the ophthalmologist found her visual acuities to be 6/6 on the right and 6/5 on the left. She was able to see 15/17 Ishihara plates with the right eye, and 17/17 with the left. No abnormality was found to confrontational visual fields, and fundoscopy was normal. There was no relative afferent pupillary defect. The patient was reassured and discharged.

The visual disturbance settled after two weeks, but after another two weeks the lady returned to her General Practitioner, stating that the area of visual loss was now somewhat larger. A CT scan was organised by the General Practitioner. This showed some reduced attenuation in the left frontal lobe which was felt most likely to be due to demyelination. The patient was therefore referred as an emergency to the local neurologist.

Like the ophthalmologist, the neurologist could find nothing on examination, but performed visual evoked potentials. In the right eye, the latency of the P100 was 123 msec, and that of the left eye was 106 msec (normal range 95 – 110 msec). Blood tests were normal, as was a lumbar puncture; in particular, no oligoclonal bands were found. A brain MRI showed the appearances in the figures below. An MRI scan of the spinal cord was normal. The visual disturbance subsequently resolved over the next few months and the patient felt her vision had returned back to normal.

Figure 1: Axial T2 and Gd-enhanced T1 weighted MRI scans:

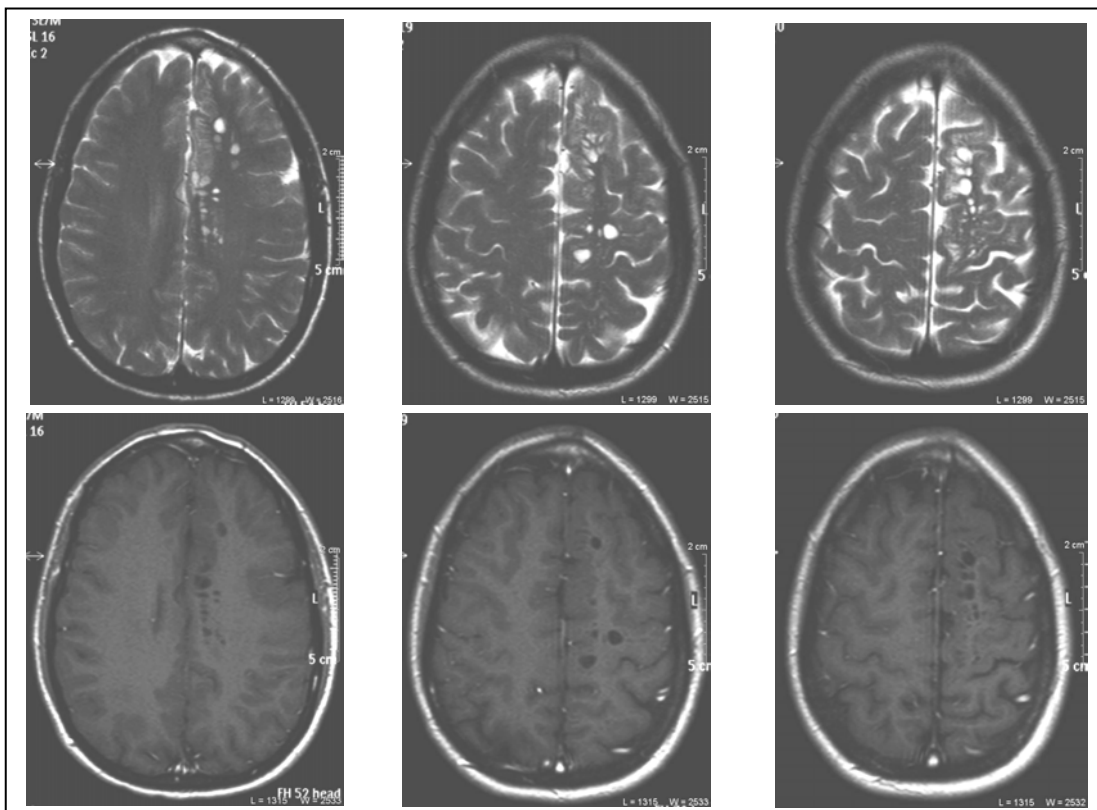
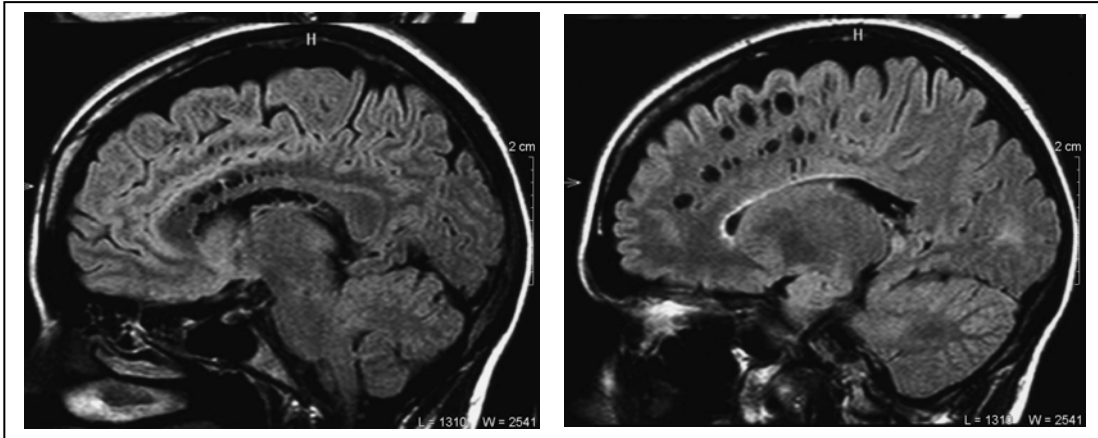


Figure 2: Unenhanced sagittal T1 images:



Which of the following statements is correct about the diagnosis and management of this individual?

ANSWER OPTIONS

- A: The patient has multiple sclerosis according to the McDonald criteria, and should ideally be started on a disease-modifying agent (beta interferon or glatiramer acetate).
- B: The patient has multiple sclerosis according to the McDonald criteria, but should not be started on a disease-modifying agent (beta interferon or glatiramer acetate).
- C: The patient has multiple sclerosis according to the Poser criteria, and should ideally be started on a disease-modifying agent (beta interferon or glatiramer acetate).
- D: The patient has probably had a clinically isolated syndrome, and should be started on a disease-modifying agent (beta interferon or glatiramer acetate).
- E: The patient has probably had a clinically isolated syndrome but should not be started on a disease-modifying agent (beta interferon or glatiramer acetate).