

## Question 1

### **Scenario**

A 27-year-old woman, who worked as a waitress and was normally fit and well, had woken 3 weeks before with weakness in her left hand. She had noted numbness at the base of the thumb on the dorsum of the hand. Her mother had died in her early 40s with a stroke, but there was no other family history of neurological disease. She was on the oral contraceptive pill. She drank alcohol only occasionally and smoked 15 cigarettes per day.

She had been admitted to a local hospital that day and had a CT brain scan that was normal. Other normal investigations included ECG, FBC, viscosity, glucose, cholesterol, thyroid function, urea and electrolytes, liver function tests, B<sub>12</sub> and folate. When the general medical consultant saw her on the post take ward round he arranged nerve conduction studies and discharged her.

On re-taking the history you established that there had been no unusual circumstances on the night the weakness began and that she had noticed minimal but definite improvement since admission.

Your examination included normal general examination, normal cranial nerves, and normal muscle tone, but with mild weakness of left brachioradialis, and moderate weakness of wrist and finger extension, although with full power in other arm muscles including elbow extension. Reflexes were normal. Sensory examination found a small area of sensory loss over the anatomical snuffbox on the left. Examination of the legs was normal.

### **Question**

What is the single most likely site of the lesion?

- A. C7 nerve root
- B. cerebral cortex
- C. posterior cord of the brachial plexus
- D. posterior interosseus nerve
- E. radial nerve at the spiral groove

## Question 2

### **Scenario**

A 24-year-old man was referred following a "blackout". This had occurred in the early hours of the morning. He recalled waking from sleep, standing up, feeling light-headed and nauseated, lying back on the bed and losing consciousness. He had recovered on the floor. He had not bitten his tongue but he had been incontinent and his head was badly bruised. The next morning he had noted he had a sub-conjunctival haemorrhage. The girlfriend had been awoken by him falling to the ground and described him as being pale and "dead" before giving a loud moan followed by "spasms" of his arms and legs lasting about 30 seconds. He had had a similar episode

4 months previously and reported two "faints" as a teenager associated with a 'flu-like illness.

Examination was normal as was an electrocardiogram taken in the clinic.

**Question**

What is the most likely diagnosis?

- A. cardiac arrhythmia (arrhythmogenic syncope)
- B. epileptic seizure
- C. hypoglycaemia
- D. pseudoseizure
- E. vasovagal syncope

**Question 3**

**Scenario**

A 32-year-old woman presented to the emergency unit with a 1-hour history of headache. She had been awoken at 6.30am with sudden onset of severe generalised headache with vomiting but had remained fully alert.

On examination she was in pain and intolerant of bright lights. There was no neck stiffness and no focal neurological signs.

Investigations included:

CT brain scan normal.

CSF examination (performed at 9.30 am on the day of admission)

- opening pressure 20 cm H<sub>2</sub>O
- three consecutive bottles were blood stained each with a clear supernatant.
- protein 0.6 g.L<sup>-1</sup>
- cell count: erythrocytes 100,000 mm<sup>-3</sup> white blood cells 100 mm<sup>-3</sup>
- glucose 4.6 mmol.L<sup>-1</sup> (matched blood glucose 6.0 mmol.L<sup>-1</sup>)

**Question**

Which of the following would be the next most appropriate investigation?

- A. CT cerebral angiogram
- B. intra-arterial (fornal) cerebral angiogram
- C. MR cerebral angiogram
- D. spectrophotometry of the CSF supernatant
- E. repeat CSF examination after 12 hours

**Question 4**

**Scenario**

A 35-year-old woman with previously mild relapsing-remitting multiple sclerosis reported new symptoms in the previous 6 months of urinary

frequency and urgency, with occasional urge incontinence. A bladder ultrasound scan was requested but had not yet been performed.

**Question**

Which would be the most appropriate next step in her management?

- A. advise increased fluid intake
- B. defer treatment until bladder ultrasound result available
- C. prescribe nocturnal desmopressin
- D. prescribe oxybutynin
- E. refer for urodynamic studies

**Question 5**

**Scenario**

A 72-year-old woman presented to outpatients with a 3-year history of progressive numbness in both feet. She described her feet as feeling like lumps of wood but the symptom did not interfere with walking. She had no symptoms in her arms.

She had had mild hypertension for 5 years treated with bendrofluazide 2.5 mg and ramipril 2.5 mg daily. She had not taken other drugs in the recent past. There was no family history of neurological disease. She did not drink or smoke.

General medical examination was normal. Cranial nerves were normal. Tone in the limbs was normal and she had full power in arms and legs. Reflexes were normal in the arms, knee reflexes were normal and the ankle reflexes were absent. Plantar responses were flexor. Vibration sense was lost to the knees, proprioception was normal, and temperature perception was lost to the knee and pinprick to mid calf. Her gait was normal.

The following investigations were normal or negative: routine urine testing; random glucose ( $3.9 \text{ mmol.L}^{-1}$ ); thyroid function tests; serum B<sub>12</sub> and folate; full blood count; ESR, urea and electrolytes; liver function tests; and protein electrophoresis.

**Question**

What is the single most likely diagnosis at this stage?

- A. axonal neuropathy
- B. chronic inflammatory demyelinating polyradiculopathy
- C. dorsal root ganglionitis
- D. lumbosacral plexopathy
- E. spinal stenosis of lumbosacral canal

## Question 6

### **Scenario**

A 72-year-old man awoke with poor vision in his left eye. This was still present when he was seen in the emergency unit 4 hours later. He had been feeling generally unwell for the previous 3 weeks and had reported some mild left sided headache in the previous week.

Examination showed his visual acuity to be 6/6 on the right and 6/24 on the left. He had lost vision in the lower half of the visual field of the left eye (altitudinal defect). The right fundus appeared normal, the left disc was slightly swollen with microhaemorrhages. There were no other physical signs.

### **Question**

What is the likely cause of his visual impairment?

- A. arteritic anterior ischaemic optic neuropathy
- B. central retinal artery occlusion
- C. non-arteritic anterior ischaemic optic neuropathy
- D. optic neuritis
- E. posterior ischaemic optic neuropathy

## Question 7

### **Scenario**

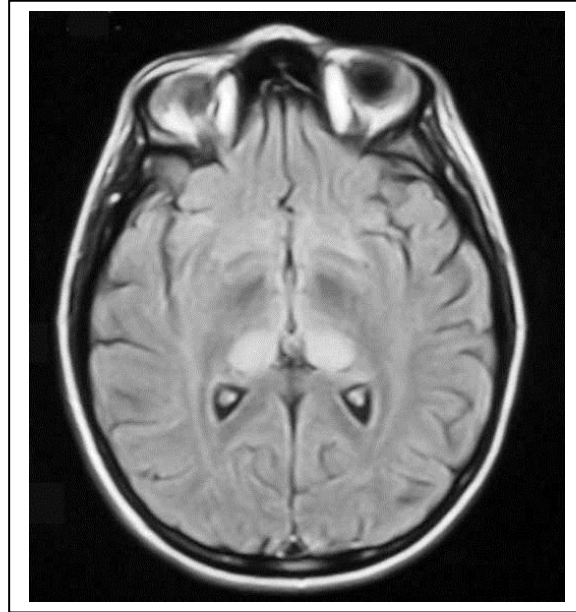
A 40-year-old woman, previously well, presented with a 5-month history of difficulty coping at work. She was being treated for depression. She complained of non-specific and sometimes burning pains in her legs but no definite joint pains or rash. There was no relevant family history.

On examination, she was restless and fidgety. Mini-mental state examination (MMSE) was 25 with errors on attention and concentration and on recalling objects at three minutes. Her gait was unsteady and she showed bilateral intention tremor and also intermittent irregular jerky movements at rest in her limbs and face.

Her routine blood investigations and CT brain scan were normal.

Q7 continued...

Figure 1 shows a section of her MR brain scan (FLAIR sequence).



**Question**

Which of the following is the most likely diagnosis?

- A. cerebral lupus
- B. dentato-rubro-pallido-Luysian atrophy
- C. Huntington's disease
- D. multiple sclerosis
- E. variant Creutzfeldt-Jakob disease

**Question 8**

**Scenario**

A 61-year-old man telephoned his GP to report a transient episode of right arm weakness. The episode occurred in the mid morning whilst he was sitting reading the newspaper. His right arm was so weak that he was unable to hold up the paper. The weakness settled after approximately 2 minutes. He was not aware of any other symptoms. His medical history was otherwise unremarkable. He was a longstanding smoker of 20 cigarettes per day.

**Question**

What is your estimate of his risk of stroke (without intervention) in the 48 hours following this event?

- A. 0.1 – 1%
- B. >1 – 3%

- C. 4 - 10%
- D. 11 - 20%
- E.  $\geq 21\%$

### Question 9

#### **Scenario**

A 67-year-old man presented with a 6-month history of poor mobility and falls. In the preceding year his wife had noticed a mild personality change with apathy and behavioural abnormalities.

On examination there was symmetrical rigidity and bradykinesia but no tremor. He had postural instability and intermittent sighing. His eye movements full but his vertical saccadic movements were slow.

Investigations included:

- MR brain scan: generalised atrophy more pronounced in the midbrain
- FP-CIT SPECT scan: symmetrically reduced uptake in the putamen and caudate
- Serum ferritin:  $235 \text{ mg.L}^{-1}$  (15 –  $300 \text{ mg.L}^{-1}$ )
- Anal sphincter EMG: polyphasic potentials compatible with denervation

#### **Question**

What is the most likely diagnosis?

- A. corticobasal degeneration
- B. dementia with Lewy bodies
- C. multiple system atrophy
- D. Parkinson's disease
- E. progressive supranuclear palsy

### Question 10

#### **Scenario**

A 70-year-old man with ischaemic heart disease and hypertension complained of tingling and numbness in both feet which had been worsening over 6 months.

Examination showed him to have altered pinprick perception over both feet and absent ankle reflexes.

#### **Question**

Which of his medications listed below is most likely to have caused his peripheral neuropathy?

- A. bendrofluazide
- B. clopidogrel

- C. ramipril
- D. simvastatin
- E. spironolactone

### Question 11

#### **Scenario**

A 58-year-old man was referred as an emergency by his GP with left-sided foot drop. The foot drop had come on 14 days earlier after a yoga class, something he had started a few weeks before for fitness. He had noted slight tingling on the dorsum of his foot. He had tripped a few times and on one occasion turned his ankle which had become slightly swollen. The foot drop had substantially improved over the last 10 days. He had retired early because of stresses at work and was taking citalopram for 'his nerves'. He smoked 10 cigarettes per day and drank on average of 3 units of alcohol per day.

General examination and examination of his cranial nerves and arms were normal. He had mild weakness of his left ankle dorsiflexors and evertors, and moderate weakness of extensor hallucis and extensor digitorum; other movements, including ankle inversion and hip abduction, were normal. Reflexes were normal. There was sensory loss on the dorsum of the foot from the base of the hallux to the ankle.

The following investigations were normal or negative:

Urine dipstick testing; random glucose ( $4.5 \text{ mmolL}^{-1}$ ); thyroid function tests, serum B<sub>12</sub> and folate; full blood count; ESR; urea and electrolytes; liver function tests; gamma glutamyl transferase, protein electrophoresis; serum ANCA; rheumatoid factor; and autoimmune profile.

#### **Question**

Which of the following courses of action or investigation is most likely to be helpful in management?

- A. genetic studies
- B. MR scan of the brain
- C. MR scan of the lumbosacral spine
- D. nerve conduction studies/EMG
- E. review in one month

## Question 12

### Scenario

A 19-year-old man presented with a 5-month history of progressive painless visual failure. There was no family history of visual failure and no history of drug abuse or toxin exposure.

General clinical examination was unremarkable. His corrected visual acuity was counting fingers on the right and 6/24 on the left. Colour vision was impaired bilaterally. Fundus examination (pupils dilated) revealed bilateral pale optic discs, but the macula and retina were normal. There were no other abnormal neurological signs.

Investigations included:

- Visual evoked potentials were reduced in amplitude, but not delayed.
- Electro-retinogram (ERG) was normal.
- FBC, U+Es, LFTs, TFTs, ACE were normal and autoantibody screen was negative.
- Chest radiograph was normal.
- MR brain scan (T1 weighted) before contrast was reported as normal (Figure 1)
- MR scan of the whole spine before and after contrast was normal.
- Cerebrospinal fluid was acellular, with normal pressure, protein and glucose.

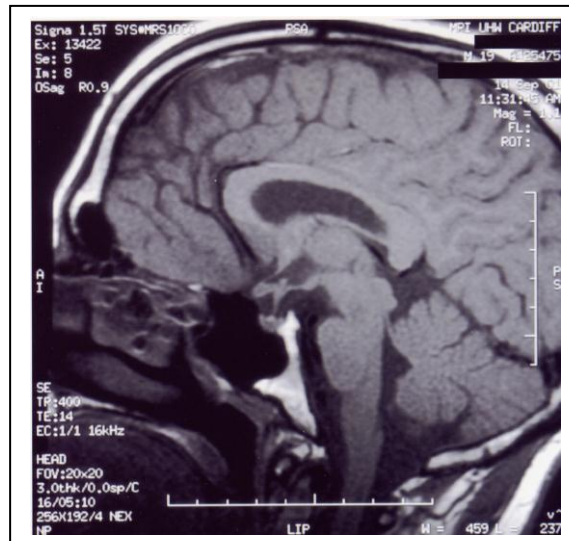


Figure 1. Unenhanced MR brain scan (sagittal T1 weighted image)

### Question

What would be single most helpful next investigation?

- A. blood for VDRL
- B. bone marrow aspiration and biopsy
- C. cerebrospinal fluid for oligoclonal bands
- D. Leber's gene testing
- E. MR brain scan with gadolinium contrast

### Question 13

#### **Scenario**

A 27-year-old male injecting heroin addict presented with rapidly progressive flaccid dysarthria, dysphagia, dry mouth and a mild generalised flaccid limb weakness. Twelve hours after admission he suddenly deteriorated resulting in a respiratory arrest following which he required artificial ventilation on Intensive Care. After resuscitation he was alert and orientated and over subsequent days he developed bilateral ptosis, dilated unreactive pupils, diplopia secondary to lateral rectus weakness, profound bilateral facial weakness and a severe flaccid quadriplegia with depressed reflexes. Sensation remained intact. Medication on admission included oral antibiotics, prescribed by his GP, for an infected injection site in his groin.

#### **Question**

What is the most likely diagnosis?

- A. botulism
- B. Guillain-Barré syndrome
- C. Lambert-Eaton myasthenic syndrome
- D. myasthenia gravis
- E. poliomyelitis

### Question 14

#### **Scenario**

A 36-year-old man presented with a 1-day history of sudden onset nausea, vomiting, unsteadiness, difficulty speaking and swallowing, and left sided neck pain. There was no history of trauma.

On examination he was lying in bed and vomiting intermittently. He was reluctant to sit up. Neurological examination of the head and neck revealed a left ptosis and miosis, deviation of the soft palate and post pharyngeal wall to the right on phonation, and left facial and intra-oral sensory loss. He was unable to swallow 10 mls of water without coughing. He was dysphonic and had an abnormal cough. He had left sided cerebellar signs and was unable to stand.

Investigations revealed a normal CT head and CSF and routine bloods were unremarkable.

**Question**

What is most likely clinical diagnosis?

- A. carotid artery dissection
- B. lateral medullary infarction
- C. posterior fossa space occupying lesion
- D. subarachnoid haemorrhage
- E. venous sinus thrombosis

**Question 15**

**Scenario**

A previously fit 45-year-old man presented with a 3-month history of weakness in his left hand. He had some difficulty opening bottles and opening doors with a key. He had some cramps in his hand, but no sensory symptoms. He had noted some wasting over the last month.

On examination he was well. Cranial nerves were normal. He had significant wasting of the left interossei and abductor digiti minimi with fasciculations visible in the first dorsal interosseous. He had weakness of finger abduction and adduction. Abductor pollicis brevis, the long flexors of the little and ring fingers and all other muscles in the arm were normal. Reflexes were normal. Sensation was normal. The legs were normal.

**Question**

What is the most likely diagnosis?

- A. deep ulnar nerve lesion in the hand
- B. motor neurone disease
- C. syringomyelia
- D. T1 root lesion
- E. ulnar nerve lesion at the elbow

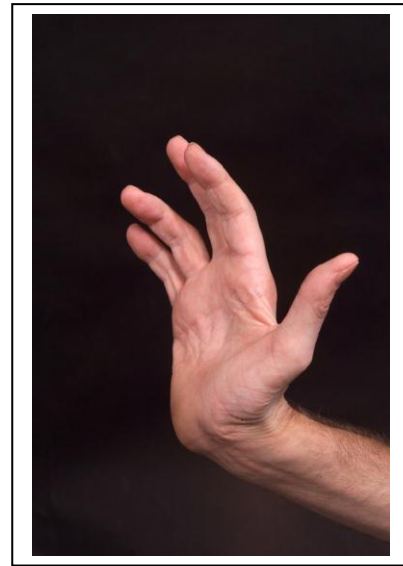
**Question 16**

**Scenario**

A 43-year-old man was referred for neurophysiological examination following unsuccessful right ulnar nerve decompression at the elbow. He had an aching pain in his right hand and forearm, which failed to respond to the surgery, and which had now begun in his left hand and forearm. His most bitter complaint, however, was the progressive loss of strength in his arms and hands and the worsening stiffness in both his legs. There were no bladder symptoms.

His arms and hands were wasted (Figures 1 and 2) with some fasciculations. His upper limb reflexes were absent and there was definite sensory loss to pinprick in his hands and forearms. Position sense in the

hands was normal. The lower limb reflexes were pathologically brisk and the plantars were equivocal.



His nerve conduction studies were as follows

Right	Distal motor latency (ms)	Amplitude	Velocity (m.s <sup>-1</sup> ) (normal >45)	F wave (ms)
<b>Motor</b>		(mV)		
Median: wrist	4.3 (<4.2)	1.9 (>5)		33.3
elbow		1.8 (>5)	56.5	
Ulnar: wrist	4.1 (<3.5)	0.2 (>4)		absent
below elbow		0.19 (>4)	61.8	
above elbow		0.19 (>4)	55.0	
<b>Sensory</b>		(μV)		
Radial		20 (>10)	60	
Median: digit 1		10 (>8)	51	
Ulnar: digit 5		15 (>6)	60	
Sural		15 (>6)	60	

EMG demonstrated positive sharp waves, fibrillations at rest and large polyphasic neurogenic motor units in right deltoid, triceps, biceps, flexor carpi ulnaris, extensor digitorum communis, first dorsal interosseus, abductor pollicis brevis, cervical paraspinals and mid thoracic paraspinals. EMG findings were normal in all lower limb muscles sampled, as well as tongue and masseters.

**Question**

What is the most likely diagnosis?

- A. axonal peripheral neuropathy
- B. cervical spondylotic radiculomyelopathy
- C. Kennedy's syndrome
- D. mononeuritis multiplex
- E. syringomyelia

### Question 17

#### **Scenario**

A 40 year old woman was referred because of unsteadiness, slurred speech and personality changes. She had become forgetful, withdrawn and had made errors with money. Her relationship with her husband had deteriorated due to irritability. She was employed as a school secretary and her performance at work had deteriorated with some administrative mistakes and omissions. She admitted to feeling depressed. There was no other relevant past medical history: in particular she had never taken neuroleptic drugs. Her father had committed suicide at the age of 55: he had been affected by a neurological illness diagnosed as multiple sclerosis: the features were unsteadiness, rigidity and depression. The patient had had little contact with him but her mother supplied these details in the clinic.

On examination, the patient was withdrawn and upset. There was mild dysarthria. Eye movements were slightly slow in all directions. There was an unsteady staggering gait, brisk reflexes and definite generalised chorea. Her minimental score was 29/30: she was assessed as quite competent to consent to investigation of her condition.

#### **Question**

Which of the following statements is most correct?

- A. There is a wide differential diagnosis here and this is not necessarily Huntington's disease. Her father probably had MS as this is a common disorder.
- B. The most appropriate diagnostic approach at this stage is to proceed directly to a DNA test for a Huntingtin gene expansion.
- C. It is important to check the patient's thyroid function, ANF, blood count and a fresh blood film for acanthocytes before considering a DNA test.
- D. It is acceptable to carry out a diagnostic DNA test (in contrast to a predictive test) without a detailed explanation of the genetic implications of a positive result and specific consent to the blood test is not necessary in this situation (in a patient who is already clinically affected).
- E. It is essential to obtain consent from the patient's mother and children before considering a DNA test.

### Question 18

#### Scenario

A 40-year-old man described severe intermittent unilateral orbital pain and tearing, waking him from sleep every night for the previous 7 days. He had previously been diagnosed as having cluster headache.

#### Question

Which of the following has the strongest evidence base to support its use as a prophylactic medication for cluster headache?

- A. pizotifen
- B. prednisolone
- C. propranolol
- D. sodium valproate
- E. verapamil

### Question 19

#### Scenario

A 33-year-old right-handed male was admitted to hospital with a 24-hour history of right-sided headache, nausea and photophobia. The headache had worsened overnight before he suffered a generalised tonic-clonic seizure shortly before admission. He had a past history of migraine. His sister had a history of diabetes mellitus, his mother and a maternal aunt had had strokes in their 50s.

On examination the patient was confused with a GCS of 13. He had a left-sided homonymous hemianopia. Blood pressure was 160/95 mmHg but there were no other abnormalities.

Investigations revealed:

FBC/Electrolytes/ESR – Normal

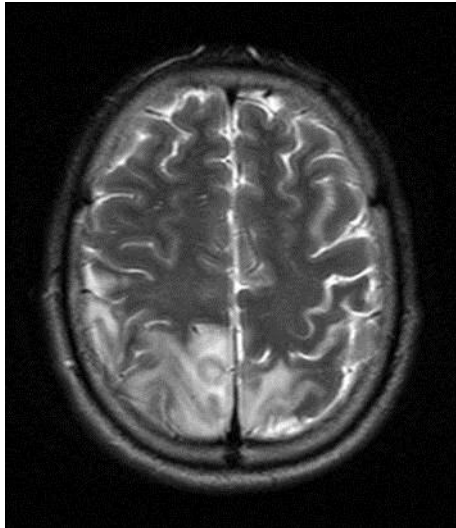
Random blood glucose – 14.2 g.dl<sup>-1</sup>

MRI (see figure below – T2 weighted image)

CSF – opening pressure 12 cm H<sub>2</sub>O.

Protein 0.42 g.L<sup>-1</sup>

Cytology: acellular



**Question**

What is the most likely diagnosis?

- A familial hemiplegic migraine
- B sagittal sinus thrombosis
- C CADASIL (cerebral autosomal dominant arteriopathy with subcortical infarcts and leucoencephalopathy)
- D MELAS (mitochondrial encephalomyopathy with lactic acidosis and stroke-like episodes)
- E cerebral vasculitis

**Question 20**

**Scenario**

A patient with longstanding epilepsy reported a chronic skin lesion at his first consultation with you.

**Question**

Which of the following skin abnormalities would alter your diagnosis if you found them on a patient with epilepsy?

A



B



C



D



E

