

## Answers and Feedback

### Question 1

Answer E, Radial nerve lesion at the spiral groove

#### **Explanation and Feedback**

Wrist drop may occur from lesions at any of these sites.

A posterior interosseus nerve lesion is unlikely because:

- There was involvement of muscles outside posterior interosseous nerve territory (brachioradialis, extensor carpi radialis longus).
- There was no radial deviation on wrist extension.

A C7 radiculopathy is unlikely because:

- The triceps was not involved.
- Brachioradialis (C5, 6) was involved.

A posterior brachial cord lesion can give a radial palsy but is unlikely because:

- triceps was spared (supplied by the proximal radial nerve, branching from the posterior cord in the lower axilla)
- deltoid was spared (supplied by the axillary nerve, the other branch of the posterior cord).

A demyelinating lesion of the radial nerve at the spiral groove is the most likely answer. This causes conduction block and hence weakness in radial-innervated muscles distal to the site of the lesion. The sensory loss reflects conduction block in the radial nerve. This neuropraxic injury should recover more quickly than axonotmesis or neurotmesis (1,2).

#### **References**

1. Seddon HJ. Three types of nerve injury. *Brain* 1943; 66: 237-288.
2. Fuller G. Focal peripheral neuropathies. *J Neurol Neurosurg Psychiatry* 2003; 74 (suppl 11): ii20-ii24.

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### Question 2

Answer E, vasovagal syncope

#### **Explanation and feedback**

The prodrome of nausea/light-headedness and association with postural change are highly suggestive of syncope as is the rapid recovery. The witness description of the patient being pale and “dead” before moaning and convulsing confirms that this is convulsive syncope and not an epileptic seizure (1)

An ECG should be done in all patients with loss of consciousness (2) to detect underlying abnormalities which can predispose to arrhythmias (such as the long QT syndrome) and is known to be normal in this case. An EEG should

not be done in patients in whom the diagnosis is syncope since this may yield false positive results and may therefore lead to an erroneous diagnosis of epilepsy (2). The history is highly suggestive of severe cardioinhibitory syncope (young patient, rapid loss of consciousness, injury and significant period of loss of consciousness even when supine) (3). The 24-hour ECG is likely to be normal. Imaging of the head will not be useful in patients with syncope. An implantable loop recorder is expensive and invasive. The diagnosis can usually be confirmed by tilt table studies (4).

### **References**

1. Lempert T, Bauer M, Schmidt D. Syncope a videometric analysis of 56 episodes of transient cerebral hypoxia *Ann Neurol* 1994;36:233-7.
2. Diagnosis and management of epilepsy in adults 2003. [www.sign.ac.uk](http://www.sign.ac.uk)
3. Sutton R, Petersen M, Brignole M et al. Proposed classification for vasovagal syncope. *Eur J Cardiac Pacing Electrophysiol* 1992;3:180-3
4. Kurbaan AS, Bowker TJ, Wijesekera N et al Age and hemodynamic responses to tilt testing in those with syncope of unknown origin. *J Am Coll Cardiol* 2003;41:1004-7

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### **Question 3**

Answer B, intra-arterial (formal) cerebral angiogram

### **Explanation and Feedback**

This patient presented with a sudden severe headache and had a normal CT brain scan. The CSF was examined after 3 hours and was uniformly blood stained. This is too early to reliably have developed xanthochromia so the state of the supernatant is irrelevant. This finding would be consistent with (indeed supports) a diagnosis of subarachnoid haemorrhage and in a CSF taken 3 hours post ictus a traumatic tap and a subarachnoid cannot be distinguished even with spectroscopy. Thus the next investigation is an intra-arterial angiogram.

CT or MR angiograms are not as sensitive as formal angiography and, when there is a high pre-test probability, this is important as negative results will not exclude an aneurysm. Repeat CSF analysis would find blood products in CSF following a traumatic tap or a bleed and thus has nothing to add. This scenario indicates the need for delay eg 12 hours in undertaking an LP to allow a traumatic tap to be distinguished from a subarachnoid haemorrhage.

### **References**

UK National external quality assessment scheme for immunochemistry working party, National Guidelines for analysis of cerebrospinal fluid for

bilirubin in suspected subarachnoid haemorrhage, *Ann Clin Biochem* 2003;40:481-88.

**Question setter**

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**Question 4**

Answer B, defer treatment until bladder ultrasound result available

**Explanation and Feedback**

The symptoms are likely to relate to detrusor overactivity and be best treated with antispasmodic / anticholinergic drugs. However, the same symptoms can arise from impaired bladder emptying which would be exacerbated by such medication. Determining the residual volume prior to starting treatment is a simple non-invasive way to distinguish between these two situations.

**References**

Fowler CJ, O'Malley KJ. Management and investigation of neurogenic bladder dysfunction, *J Neurol Neurosurg Psychiatr, Supplement* 2003;74(suppl) iv27-iv31.

**Question setter**

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**Question 5**

Answer A, axonal neuropathy.

**Explanation and feedback**

The clinical presentation is likely to be indicative of a length dependent neuropathy - most likely axonal. In length dependent neuropathies sensory signs in the arms would typically only appear when signs in the legs are above the knee.

B) Chronic inflammatory demyelinating polyradiculopathy. There are a number of factors that make this unlikely: the very slow onset, the length dependent pattern would not be typical for CIDP and the selective loss of the ankle jerks.

C) Dorsal root ganglionitis typically presents with prominent large fibre sensory loss with loss of proprioception and vibration sense and there is not necessarily a length dependent pattern of loss. Reflexes are often lost early.

D) Lumbosacral plexopathy

The signs do not conform to root or plexus lesions.

E) Spinal stenosis of lumbosacral canal

Lumbar canal stenosis is often associated with activity related symptoms, is less often characterised by neurological signs, and when it is these would relate to coexistent radiculopathies, and not signs of a length dependant neuropathy.

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**Question 6**

Answer A arteritic anterior ischaemic optic neuropathy

**Explanation and Feedback**

The presentation of an altitudinal field loss on waking is typical for an ischaemic optic neuropathy. This pattern of field loss is a strong indicator of an ischaemic rather than inflammatory pathology. The changes seen in the optic nerve indicate this is an anterior ischaemic optic neuropathy. A posterior ischaemic optic neuropathy would not be associated with fundal changes. A central retinal artery occlusion produces changes in the retina rather than the disc, and these may be difficult to discern acutely. The preceding malaise and headache make it more likely that this is related to temporal arteritis and is thus an arteritic anterior ischaemic optic neuropathy. A similar presentation with normal disc appearances is seen occasionally in patients with temporal arteritis, accounting for 7% of patients in one series. It is important to consider temporal arteritis in any patient with ischaemic optic nerve disease, even if atypical or without systemic symptoms.

**References**

Hayreh SS, Podhajsky PA, Zimmerman B. Ocular manifestations of cranial arteritis, *Am J Ophthalmology* 1998;125:509-20.

**Question setter**

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**Question 7**

Answer E, variant CJD.

**Explanation and Feedback**

This presentation could be explained by any of several conditions, but the scan, showing the pulvinar sign, is characteristic of variant CJD.

**Clinical features.** The typical clinical presentation is with psychiatric and often persistent painful sensory symptoms at the onset followed by the development of subtle cognitive problems and a movement disorder in a young person. Initially, the movement disorder is often manifested by fidgety movements and later becomes more obvious choreiform movements often progressing to myoclonus as the illness progresses. The mean age at onset is 28, so this lady is a little older than usual, but the range is wide 12 to 74 years. The relatively short history together with the absence of family history and the prominence of sensory symptoms would argue against Huntington's disease or DRPLA. Wilson's disease can present in a very similar way to vCJD at this age, but persistent painful sensory symptoms would again be atypical. Multiple sclerosis would be a possibility but ruled out effectively by the MR scan. Cerebral lupus is a possibility but with normal blood

investigations and no pre-existing diagnosis of systemic lupus would be unlikely.

**Radiology features.** The scan shows the typical appearance of variant CJD, with bilateral pulvinar high signal. It is otherwise normal, although sometimes atrophy is seen in variant CJD. Although the scan appearance is not absolutely diagnostic, in the correct clinical context it is highly sensitive and specific. In patients referred to the National CJD Surveillance Unit, who are subsequently confirmed to have vCJD at post mortem, more than 90% had this appearance. A minor degree of basal ganglia high signal may be seen in sporadic CJD.

### **References**

Knight R. The relationship between new variant CJD and BSE, *Vox Sanguinis* 1999;76:203-208.

Will RG, et al. A new variant of Creutzfeldt-Jakob disease in the UK. *Lancet* 1996;347:921-925.

Will RG, Zeidler M, Stewart G. Diagnosis of New Variant Creutzfeldt-Jakob disease. *Annals of Neurology* 2000;47:575-82.

Lowman A, Knight R, Ironside J. Variant Creutzfeldt-Jakob disease. *Practical Neurology*, 2001;1:2-13.

Lowman A, Ironside J. Diagnosis of variant Creutzfeldt-Jakob disease. *Advances in Clinical Neuroscience and Rehabilitation* 2002;1:6-8.

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### **Question 8**

Answer C, 4 - 10%

### **Explanation and Feedback**

This question aims to address the high risk of stroke in the immediate aftermath of TIA. This is a key issue that explains the need for the urgent clinical assessment of patients presenting with TIA. There have been two recent studies that give surprisingly similar estimates of the risk of stroke at 48 hours (approximately 5%).

### **References**

Johnson SC et al. Short term prognosis after emergency department diagnosis of TIA. *JAMA* 2000;284:2901-2906.

Lovett JK et al. Very early risk of stroke after a first TIA. *Stroke* 2003;34:e138-142.

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### **Question 9**

Answer E, progressive supranuclear palsy

### **Explanation and Feedback**

#### **Diagnosis**

Idiopathic Parkinson's disease (IPD) and corticobasal degeneration (CBD) usually present with an asymmetric akinetic rigid syndrome. In IPD falls are uncommon in the early stages.

Dementia with Lewy bodies could not be fully excluded from the available clinical information, but the lack of fluctuating cognitive impairment and visual hallucinations, with relatively subtle frontal lobe dysfunction would make this diagnosis unlikely. A patient presenting with a symmetric akinetic rigid syndrome could have either multiple system atrophy (MSA) or progressive supranuclear palsy (PSP). Early falls are a feature of PSP due to postural instability. PSP patients may exhibit significant fronto-subcortical cognitive impairment. Early neuropsychiatric features, which can precede clinical features, include personality changes and apathy (1). Significant cognitive impairment is unusual in MSA and is regarded by some as an exclusion to the diagnosis. Thus the early falls, neuropsychiatric features and vertical saccadic eye movement abnormalities are therefore the clinical features that suggest the answer is PSP.

#### **Intermittent sighing**

Intermittent sighing is usually associated with MSA, although was seen in 3 of 62 PSP patients in one recent study (1). Slowed vertical saccadic eye movements and falls within the first year of disease onset fulfil the diagnostic criteria for possible PSP (or evidence of vertical supranuclear gaze palsy) (NINDS-SPSP criteria). CBD and MSA patients usually have normal vertical saccades (2). In the early stages of PSP a supranuclear gaze palsy may not be evident.

#### **Investigations**

Cranial MRI in PSP commonly indicates a reduction in midbrain diameter, as well as signal increase in the midbrain, and globus pallidus (3). The FP-CIT SPECT scan, reflecting dopamine transporter density on the nigrostriatal dopaminergic projections is abnormal in almost all parkinsonian syndromes, except drug induced and vascular disease. It cannot therefore be used to reliably differentiate between typical and atypical parkinsonism. Sphincter EMGs can be abnormal in MSA and PSP and cannot be used to distinguish the two conditions (4).

#### **References**

1. Nath U, Ben-Shlomo Y, Thompson RG, et al. *Clinical features and natural history of progressive supranuclear palsy. A clinical cohort study.* Neurology 2003; 60:910 – 916

2. Hamilton SR. *Neuro-ophthalmology of movement disorders*. Current opinion in Ophthalmology 2000; 11:403-407
3. Burn DJ, Lees A. *Progressive supranuclear palsy: where are we now?* Lancet Neurology 2002;1: 359-69
4. Kashmere J, Camicioli R and Martin W. *Parkinsonian syndromes and differential diagnosis*. Curr Opin Neurol 2002; 15:461-466

### **Learning Points**

- In a parkinsonian syndrome early falls and abnormal vertical saccadic eye movements and are typical of PSP
- Cognitive impairment is uncommon in MSA
- Sighing is not specific to MSA
- Radiological and neurophysiological investigations cannot reliably distinguish between parkinsonian syndromes

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### **Question 10**

Answer D, simvastatin

### **Explanation and Feedback**

Statins are associated with peripheral neuropathy. In a case controlled study, patients who had been on statins for 2 years or more were 26 times more likely to have a neuropathy than matched controls, a substantial increased risk.

The other drugs listed are not associated with neuropathy.

### **References**

Gaist D, Jeppesen U, Anderson M et al. Statins and risk of polyneuropathy: a case controlled study, Neurology 2002;58:1333-7.

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### **Question 11**

Answer E review in one month, (2<sup>nd</sup> choice D Nerve conduction studies)

### **Explanation and Feedback**

This man has a left sided common peroneal nerve lesion that occurred after undertaking yoga, an activity he was unaccustomed to which is likely to have included external compression of the common peroneal nerve at the fibular head. Preserved ankle inversion and hip abduction indicates this is probably not an L5 radiculopathy. The degree of weakness and the beginnings of recovery indicates this is an incomplete lesion probably a neuropraxia, and

likely to improve over the next weeks and months. This being the case, management is going to be conservative with avoidance of further compression. Nerve conduction studies would not change this management, though might uncover an associated subclinical generalised neuropathy. However the presence of a clear trigger and negative screening bloods make this relatively unlikely. Imaging the lumbar spine would not change management even in the unlikely event this were to be related to an L5 disc. Thus the most helpful test is to review after 1 month, with an expectation that this will be substantially improved. If however it is not, the other investigations may then prove helpful.

### **References**

Willison HJ, Winer JB. Clinical evaluation and investigation of neuropathy. *J Neurol Neurosurg Psychiat* 2003;74(Supplii):ii3-ii8.

Fuller GN. Focal Peripheral neuropathies. *J Neurol Neurosurg Psychiat* 2003;74(Supplii):ii20-ii24.

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### **Question 12**

Answer E, MR brain scan with gadolinium contrast.

His MR brain scan following gadolinium contrast showed extensive abnormal signal in the suprasellar region and pineal gland (Figure 1). This was diagnosed as a germinoma, and treated with radiotherapy.

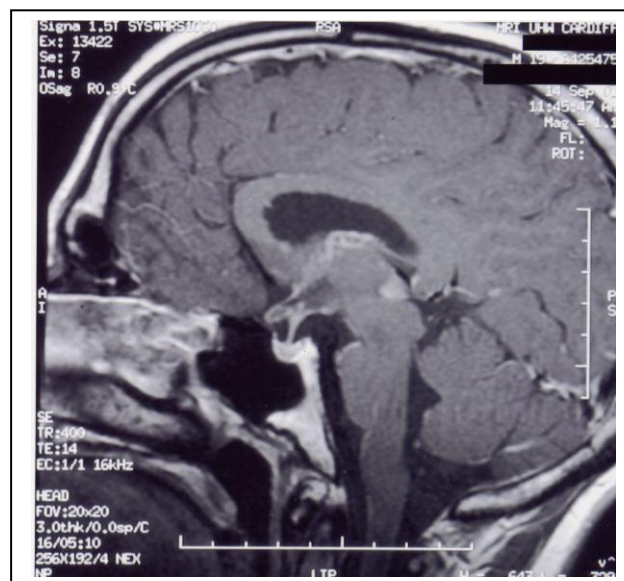


Figure 1. MR brain scan following gadolinium contrast (sagittal T1 weighted image)

### **Explanation and Feedback**

- Germinomas arise through neoplastic change in germ cells during embryogenesis. Most arise in the midline axis, somewhere between the supra-sellar region and the pineal gland.
- They usually affect young patients (less than 20 years), and especially men (male: female 4:1). Visual failure is a common presentation.
- Germinoma is usually too small to see on unenhanced MRI (as in this case), but it brightens prominently and homogeneously with contrast.
- A relatively homogeneous, well circumscribed, extra-axial, enhancing pineal region mass in a young male is highly characteristic of a germinoma (see answer 1).
- Surgery for germinoma, even biopsy for tissue diagnosis, is potentially hazardous. Surgical resection exposes the patient to the operative risks and to the possibility of regional tumour dissemination.
- Germinomas are exquisitely sensitive to radiotherapy, and this would be justified, perhaps even without a tissue diagnosis.
- Current survival rate with radiotherapy is 90% after a median follow-up of 5 years.

### **References**

Horowitz MB, Hall WA. Central Nervous System Germinomas. *Arch Neurol* 1991; 48: 653-657.

Jakacki R. Central nervous system germ-cell tumor. *Curr Treat Options Neurol* 2002; 4: 139-145.

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### **Question 13**

Answer A, Botulism

### **Explanation and Feedback**

This is a real case which turned out to be “wound botulism” in an injecting heroin addict. The main differential diagnosis is a variant of GBS which can produce a similar clinical picture of cranial nerve palsies and autonomic dysfunction.

Although only recently reported in the UK<sup>1</sup>, the association of wound botulism and injecting drug addiction has been described in the USA<sup>2,3,4</sup>. Wound botulism should be considered in any injecting drug user who presents with descending motor and autonomic signs. Early diagnosis and treatment is vital. Nerve conduction studies were normal other than for low amplitude compound motor action potentials. Repetitive stimulation at low frequency (3Hz) produced no decrement suggestive of myasthenia gravis. However, tetanic stimulation (20Hz) produced a prominent incremental response (38%) consistent with a pre-synaptic neuromuscular transmission defect.

Management is primarily supportive, however where the condition is recognised early anti-toxin shortens hospital stay and duration of ventilation (5). Wound debridement and benzypenicillin or metronidazole should be given.

### **References**

1. Mulleague L., S.M. Bonner, A. Samuel, P. Nichols, M. Khan, S. Shaw, T. Gruning. Wound botulism in drug addicts in the United Kingdom. *Anaesthesia* 2001;56:120-123.
2. Centre for Disease Control. Botulism in United States 1899-1996, handbook for epidemiologists, clinicians, and lab workers. US dept of health 1998.
3. Merson MH, Dowell VR. Epidemiologic, clinical, and laboratory aspects of wound botulism. *NEJM* 1973; 289: 1005-10.
4. CDC Morbidity and Mortality Weekly Report. Wound botulism-California, 1995. *Journal of the American Medical Association* 1996; 275: 95-6.
- 5 Tacket CO, Shandera WX, Mann JY, Hargrett NT, Blake PA, Equine antitoxin use and other factors that predict the outcome in foodbourne botulism. *Am J Med* 1984;76:794-8.

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### **Question 14**

Answer B, lateral medullary infarction.

### **Explanation and Feedback**

The constellation of symptoms and signs is entirely consistent with lateral medullary infarction and the age of the patient and the prominent pain makes a vertebral dissection the most likely pathology.

- A left sided carotid artery dissection would explain the Horner's syndrome but not the left sided signs.
- A posterior fossa lesion would present with progressive rather than sudden onset symptoms.
- A subarachnoid haemorrhage might present with sudden onset symptoms with focal signs but not with Horner's syndrome.
- Migraine is sometimes associated with Horner's syndrome but would not explain the pattern of sensory deficit or lower cranial nerve deficit.
- Venous sinus thrombosis might cause lower cranial nerve palsies but does not explain the Horner's syndrome and is not usually of sudden onset.

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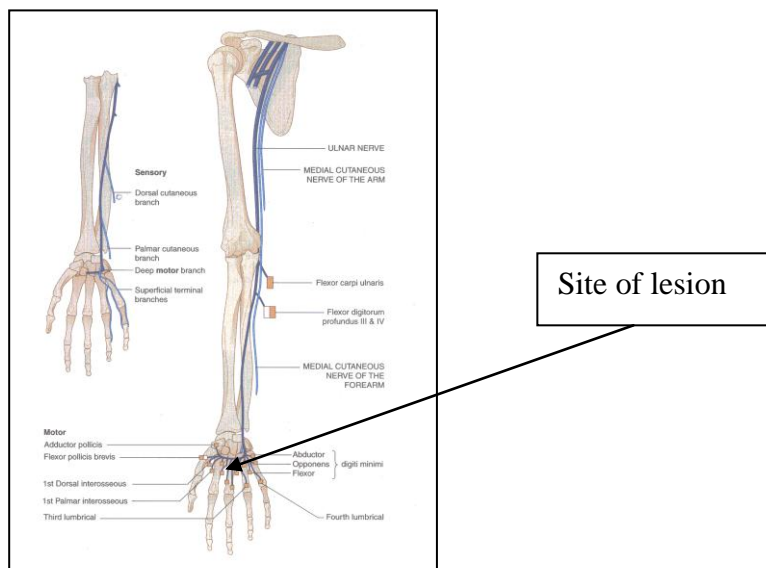
### Question 15

Answer A, deep ulnar nerve lesion in the hand

#### **Explanation and feedback**

A lesion of the deep motor branch of the ulnar nerve (arrow) produces the pattern of weakness described without sensory loss. It is possible this could reflect a fascicular lesion at the elbow, however the severity of the loss distally, with significant recent onset wasting and the lack of other symptoms from the ulnar more proximally is against that.

A T1 lesion would also involve the median innervated muscles and syringomyelia and motor neurone disease would be expected to produce more widespread signs.



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### Question 16

Answer E, syringomyelia

#### **Explanation and Feedback**

- The sensory nerve action potential amplitudes (SNAPs) are normal but the compound muscle action potential (CMAPs) amplitudes are reduced. This together with EMG neurogenic changes implies motor fibre axonal loss. This may occur in anterior horn cell pathologies (including intrinsic spinal cord disease), radiculopathies, myopathies or Lambert-Eaton myasthenic syndrome.

- An axonal neuropathy or plexopathy would show reduction in both SNAPs and CMAPs, because of axonal loss in both sensory and motor axons.
- Sensory loss with normal SNAPs indicates a lesion proximal to the dorsal root ganglion (DRG). If the DRG is intact there is no wallerian degeneration in distal sensory axons and therefore a normal SNAP is recorded. This may be seen in central causes of sensory loss or in radiculopathies.
- Syringomyelia causes both loss of anterior horn cells (and hence neurogenic EMG and small CMAPs) and spinothalamic sensory loss (proximal to the DRG, so no wallerian degeneration and normal SNAPs).
- The definite sensory loss makes motor neurone disease very unlikely. Kennedy's syndrome (X-linked bulbospinal neuronopathy) affects anterior horn cells and interestingly is often associated with low SNAPs, but without prominent sensory symptoms and no upper motor neurone involvement (Olney et al., 1991).

The cervical MR scan confirmed the patient to have a large syrinx.

### **Reference**

Olney RK, Aminoff MJ, So YT. Clinical and electrodiagnostic features of X-linked recessive bulbospinal neuronopathy. *Neurology* 1991; 41: 823-8.

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### **Question 17**

Answer: 2

1. The clinical features are very suggestive<sup>1</sup> of Huntington's disease and this is the most likely clinical diagnosis. It is common for affected relatives in earlier generations not to have been diagnosed accurately and the true nature of the father's illness may have been concealed or not recognised. Suicide is common in Huntington's disease.
2. If Huntington's disease seems likely on clinical grounds, a single blood test may establish the diagnosis without the need for lengthy laboratory tests and scans. This is only reasonable if the diagnosis seems clinically secure; in atypical cases or patients in whom the diagnosis is not a serious possibility this approach is inappropriate as a positive result may be misleading. This is particularly true of patients with psychiatric illness and a vague family history.
3. These tests are unlikely to help and so cannot be regarded as "important" preliminaries to DNA testing unless Huntington's disease is clinically unlikely (eg chorea with absent reflexes). Thyrotoxicosis, SLE, polycythaemia and neuroacanthocytosis are unlikely (the latter should be considered in cases of chorea and areflexia).

4. Most genetic laboratories require consent for DNA testing even in affected individuals; the patient should know what is being tested and the implications of a positive result. Remember that you are testing the family as well and it is sensible to try to involve them in the decision if possible and if the patient is in agreement.
5. It is sensible to encourage the patient to involve family members but this is not essential; for example, the patient may not agree to this.

### **REFERENCES**

1. Fletcher NA. Movement Disorders. In: Donaghy M, editor. *Brain's Diseases of the Nervous System*. 11th ed. Oxford: Oxford University Press, 2001:1015-1096.
2. Davis MB, Bateman D, Quinn NP, et al. Mutation analysis in patients with possible but apparently sporadic Huntington's disease. *Lancet* 1994;344:714-7.
3. Harper PS. The natural history of Huntington's disease. In: Harper PS, editor. *Huntington's disease*. London: W.B. Saunders, 1991:127-140.
4. Harper PS, Newcombe RG. Age at onset and life table risks in genetic counselling for Huntington's disease. *J Med Genet* 1992;29:239-42.

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### **Question 18**

Answer E, verapamil.

### **Explanation and Feedback**

Verapamil was found to be more effective than placebo in a randomised controlled trial in cluster headache (1). Studies of valproate (2) and propranolol (3) have been negative, though these were small and do not rule out an effect. There are no trials for propranolol or prednisolone.

### **References**

- 1) Leone M, D'Amico D, Frediani F, et al. Verapamil in the prophylaxis of episodic cluster headache: a double blind study versus placebo, *Neurology* 2000;54:1382-5.
- 2) El Amrani M, Massiou H, Bousser MG. A negative trial of sodium valproate in cluster headache: methodological issues, *Cephalalgia* 2002;22:205-8.
- 3) Grottemeyer KH, Husstedt IW, Schlake HP. Betablockers vs placebo in vasomotor headache. A double blind crossover study, *Dtsch Med Wochenschr* 1987;112:1740-3.

### **Question setter**

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### Question 19

Answer D, MELAS

#### **Explanation and feedback**

- MELAS (mitochondrial encephalomyopathy with lactic acidosis and stroke like episodes) is the only answer which ties together the clinical (including family history), biochemical and radiographic findings.
- Familial hemiplegic migraine may present in a similar manner but is not associated with seizures. Although subtle changes on MR imaging have been reported those shown here are far too extensive.
- Cerebral venous thrombosis may be clinically indistinguishable from this case, though the family history is not explained. MR findings are usually that of a haemorrhagic infarct not pertaining to a single arterial territory and CSF opening pressures were not elevated in this case.
- A diagnosis of CADASIL would be consistent with recurrent migraine and a dominant family history. Imaging in CADASIL shows deep white matter abnormalities, in contrast to the findings in this patient.
- Cerebral vasculitis might also present in this manner, and whilst one would expect a reactive CSF this is not always the case. The presence of an elevated random glucose in the diabetic range is not explained however and the strong family history of migraine would not favour this diagnosis.

#### **References**

- 1) Schaefer AM, Taylor RW, Turnbull DM. The mitochondrial genome and mitochondrial muscle disorders *Current Opinion in Pharmacology* 2001,1:288-293.
- 2) Chinnery PF, Howell N, Andrews RM, Turnbull DM. Clinical mitochondrial genetics. *J Med Genet* 1999;36:425-436.
- 3) <http://www.emedicine.com/ped/topic1406.htm>

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### Question 20

Answer E = shagreen patch of tuberous sclerosis

#### **Explanation and feedback**

This patch is in a typical site and is a raised and leathery with the consistency of orange-peel. This finding makes the diagnosis of tuberous sclerosis. The word *shagreen*, along with its French and German equivalents, *chagrin*, is derived from the Persian word *saghari*, applied to a leather produced from an ass.

The other dermatological diagnoses are:

- A. Darier's disease
- B. granuloma annulare
- C. local excoriation (non-specific)
- D. morphea (limited scleroderma)

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