How good at neurology are you?

Paul Goldsmith and Graham Lennox
Department of Neurology, Addenbrooke’s Hospital, Cambridge, UK. Email: pg255@hermes.cam.ac.uk; drslennox@aol.com

Questions

1. Please consider the following:
A 50 year-old man presented with a two year history of behavioural changes and slowness. His MRI is shown below. His mother had died from ‘paralysis’.
What is the diagnosis?

Image courtesy of Dr John Thorpe

2. Please read the following passage.
You receive a telephone call regarding a 30 year-old man about to undergo incision and drainage of a pilonidal abscess under general anaesthetic. His father had died unexpectedly following an appendicectomy. This led the anaesthetist to ask him some questions about his muscles. He said he had never been great at sport and he was the one who always dropped the ball in his family. The anaesthetist asks you whether this is relevant:
(a) What would be the best way to confirm the diagnosis?
   (i) EMG
   (ii) Muscle biopsy
   (iii) Genetic test
   (iv) Plasma creatine kinase level
   (v) Other
(b) What would you advise the anaesthetist?
   (i) Reassure
   (ii) Proceed with operation, but be prepared for prolonged post-operative recovery
   (iii) Proceed with operation but do not administer curare-like agents
   (iv) Proceed with operation but do not administer non-depolarizing blockers
   (v) Cancel operation

© 2003 Blackwell Publishing Ltd
3. Please study this plain skull x-ray.
This 84 year-old man presented with deafness. What is the diagnosis?

4. Please consider the following case:
A 66 year old man presented to his family doctor with back pain. His doctor thought the neurological examination was normal and arranged some Xrays. He sends the films through to you and asks whether any further investigation is necessary. If you could only choose one, which of the following would you recommend?
(a) measure blood calcium level
(b) measure parathormone level
(c) reassure
(d) blood cultures
(e) bone scan

5. Please read the following:
You are asked, by his new family doctor, to review a 45-year-old man of low intelligence at a local institution on account of immobility and incontinence. It is unclear whether this is long-standing but, on the basis of your clinical findings, you arrange an MR scan. On the basis of this scan, which of the following would you arrange?
(a) CSF analysis, including oligoclonal bands
(b) Very long chain fatty acids
(c) Organic and aminoacids
(d) White cell enzymes assay
(e) None of the above