QUESTION 21

A 42yo man presents with a 2 year history of increasing right facial numbness. He has a history of intermittent unsteadiness, mild hearing loss and vertigo but has otherwise been well. Cranial MRI (T1 weighted following gadolinium contrast) is shown below.

![MRI image]

The most likely diagnosis is:

A. Multiple sclerosis
B. Neurofibromatosis type 2
C. Cerebellar haemangioblastoma
D. Meningioma
E. Pontine glioma

Multiple Sclerosis:

- Multiple hyperintense lesions on T2 weighted images
- Clearly not the answer – the MRI above shows 2 large-ish lesions on T1 – not the appearance of MS
- Also clinical history really doesn’t fit
Meningioma

- Usually benign
- Attached to dura
- May invade the skull but infrequently invade the brain
- Most common across the sagittal sinus, over cerebral convexities, in the cerebellar-pontine angle and along the dorsum of the spinal cord
- Peaks in middle age
- Often found incidentally
- Can present with:
  - Focal or generalised seizures
  - Raised ICP
  - Gradually worsening neurology
- CT: well-defined extra-axial mass; smooth contour, adjacent to dural structures; isointense but uniformly bright enhancement
- MRI: isointense or hypointense on T1 and isointense to hyperintense on T2; Strong homogeneous enhancement with gadolinium; most show “dural tail” of contrast

From Harrisons
http://www.accessmedicine.com/content.aspx?aID=106695
- Location of lesions in this question do not suggest a meningioma
- 2 discrete lesions also makes it unlikely and there is no dural tail

Glioma

- Account for 50 to 60% of brain tumours
- Include astrocytomas, oligodendrogliomas and mixed gliomas
- Astrocytomas are most common primary intracranial neoplasm
- Low-grade astrocytomas are more common in children
- High-grade astrocytomas are more common in adults and are usually supratentorial
- Common presentations include headache, seizures, focal neurological signs
- Usually hypointense on T1 and show heterogeneous enhancement with gadolinium
- On T2 images the oedema is hyperintense and cannot be distinguished from tumour

From Harrison’s

- Oligodendrogliomas are more benign than astrocytomas
- Typically supratentorial
- The MRI in the question shows very homogeneous lesions – not at all typical of gliomas

HAEMANGIOBLASTOMA

- Uncommon
- Occur mainly in cerebellum and spinal cord
- Associated with Von Hippel-Lindau disease – an AD disorder in which patients develop haemangioblastomas, pancreatic cysts and carcinomas, renal masses and phaeochromocytomas
- Can also occur sporadically
- Usually seen in children and young adults
- Multiple tumours suggest VHL disease
- Present due to direct compression or tumour-associated haemorrhage
- Paraneoplastic erythrocytosis can occur due to elevated EPO produced by tumour
- MRI: enhancing nodule associated with a cyst located in the cerebellum or a homogeneously enhancing lesion on the surface of the spinal cord

- Treatment = surgery +/- XRT
- Although possible as the answer, the MRI in the question does not show the cyst within the lesion
- I think VHL is also less common than NF2 (but don’t quote me on that)

NEUROFIBROMATOSIS

- Neurofibromatosis type 1 = cutaneous neurofibromas, cafe au lait spots, freckling of non-exposed areas, hamartomas of the iris and pseudoarthrosis of the tibia
- Neurofibromas are benign peripheral nerve tumours composed of Schwann cells and fibroblasts, generally asymptomatic
- NF1 associated with increased risk of nervous system tumours
- NF2 = bilateral vestibular schwannomas in >90% and a characteristic type of cataract
- Cafe au lait spots and peripheral neurofibromas are rare
- Schwannomas usually present with progressive unilateral deafness in 3rd decade
From Harrisons

- Treatment = surgery in attempt to maintain hearing for as long as possible
- Further schwannomas likely to occur with NF2

Answer: A