DEVIC’S DISEASE OR MULTIPLE SCLEROSIS?

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CASE HISTORY

A previously healthy 38 year old woman of Bangladeshi origin presented to A&E with a three day history of flu-like symptoms, left sided headache and blurred vision.

She was apyrexial but looked tired. Examination was normal. Visual acuity was not formally assessed. She was discharged from A&E with a suspected diagnosis of viral labyrinthitis, and advised to rest and continue taking regular fluids and paracetamol.

Three days later she re-presented to A&E with worsening visual acuity and continued localised left sided headache, despite analgesics. Further questioning revealed that three months earlier she had suffered an episode of ataxia and lower limb numbness, resolving spontaneously after three weeks. MRI lumbar spine had shown only left-sided scoliosis at L2 with minor degenerative changes.

There was no history, or signs, of meningism or examthematous disease, and the patient reported no recent vaccinations or new medications. There was no similar past or family history. She remained apyrexial. Examination of visual acuity was extremely poor (grossly finger counting). Fundoscopy appeared normal, with pupils equal and reactive to light. Neurological examination revealed an unsteady gait (with a tendency to fall forwards), which was attributed to the patient’s poor vision. Tone, power, sensation and co-ordination were normal in all four limbs, as were reflexes and proprioception.

Ophthalmology review confirmed extremely poor vision, with the patient unable to see fine detail at all, and revealed slight right eye relative afferent pupillary defect (suggestive of optic neuritis) and bitemporal pallor on fundoscopy.

INVESTIGATIONS

Baseline bloods were normal (full blood count, clotting, renal function, thyroid function, liver function, glucose, calcium/phosphate, CRP, ESR, B12, folate).
CT and MRI (including with gadolinium) scans of the brain showed no conclusive abnormality. MRI spine showed two to three small high T2 signal lesions in the lower part of the thoracic cord.

**Figure 1 MRI Spine.** The most typical appearance is that a section of the cord (typically more than 3 vertebral segments) is swollen and “hyper-intense” (white on a T2 MRI). The arrows point to areas of demyelination. [3]

Lumbar puncture revealed clear and colourless CSF, with opening pressure of 24cmH₂O. Analysis revealed glucose 3.2mmol/l, protein 0.85g/l (moderately raised), lymphocytes <2, polymorphs <1, erythrocytes 90 (cells/mm³/10⁶ cells/l). No growth (including a culture for AAFB). Oligoclonal banding, syphilis serology and Purkinje cell antibodies were negative.

Blood cultures showed no growth.

Electrodiagnostic testing of the optic tract revealed no response to pattern visual evoked response (VER) and a delayed response to flash VER.

**OUTCOME**

The pattern of events suggested a clinical diagnosis of acute demyelination with deteriorating vision. The patient was treated with IV methylprednisolone followed by oral steroids, resulting in complete remission of symptoms.

Devic’s disease, also known as neuromyelitis optica (NMO), is defined as optic neuritis with myelitis [1]. The main clue to diagnosis here was the onset of bilateral reduction in visual acuity. Here is a useful aide for differential diagnosis in rapid onset blindness [1]:

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Rapid onset of blindness

- Atypical Causes of Optic Neuritis
  - CO poisoning
  - Meningitis
  - Neoplasm
  - SLE

- Ischaemic Optic Atrophy
  - Alcohol / Tobacco
  - Heavy Metals
  - Leber’s OA

- Causes of Optic Atrophy
  - Optic Pathway Lesions
  - Optic Tract
  - Occipital Lobe

- Higher visual processing
  - Amblyopia

- Ischaemic Optic Atrophy
- Causes of Optic Atrophy
- Optic Pathway Lesions
- Higher visual processing

* Leber’s Optic Atrophy, and macular dystrophies
‡ E.g. Meningiomas

The pairing of optic neuritis with myelitis also occurs in patients with multiple sclerosis (MS), acute disseminated encephalomyelitis (ADEM), systemic lupus erythematosis (SLE) and Sjogren’s syndrome. It may also be associated with viral and bacterial infection, however, often no underlying cause is found. The table below highlights some key differences between Devic’s disease and MS.

DIFFERENCES BETWEEN DEVIC’S DISEASE & MULTIPLE SCLEROSIS [2]:

**DEVIC’S**
- Asian communities
- Male:Female ratio equal
- Negative brain MRI at onset plaques/demyelination
- Oligoclonal bands positive 30% 80%
- Unilateral or bilateral optic neuritis neuritis
- MRI abnormality > 3 vertebral segments

**MS**
- Western communities
- Female preponderance
- MRI brain:
  - Oligoclonal bands positive
  - Usually unilateral optic

When faced with a presentation of sudden-onset, rapidly worsening vision in an Asian patient with normal MRI brain, the possibility of Devic’s disease should be borne in mind.
REFERENCES