CASE STUDY

Imaging of Neuromyelitis Optica (Devic's Syndrome/Disease)

Introduction

A forty year old female patient developed a sudden onset of weakness in both lower limbs and an inability to walk. Her upper limbs were not affected. She also noticed sensory abnormalities in her lower limbs, abdomen and lower chest. She also had acute urinary retention and constipation.

Within a few days she developed worsening visual function. She was referred to our institution at this stage for further evaluation.

Both pupils were dilated. On fundoscopy she had bilateral optic atrophy and a sluggish direct light reflex on the right and an absent direct light reflex on the left. She had poor visual acuity bilaterally. She did not have any ophthalmoplegia.

She was unable to sit. She had a sensory level at T2-T3 for light touch, pain, temperature and proprioception anteriorly and posteriorly.

A clinical diagnosis of Devic's disease was made with a possible differential diagnosis of Acute Demyelinating Encephalomyelopathy or Guillian Barre Syndrome. Radiological imaging was requested for further evaluation of the patient.

Imaging

Magnetic Resonance Imaging (MRI) of the spinal cord demonstrated an intramedullary lesion of the spinal cord extending from C2 to T4 which was low signal on the T1 sequence (Fig 1a and 1b) and high signal on the T2 (Fig 2a and 2b) and STIR sequences (Fig 3). There was associated expansion of the cord. This was consistent with oedema of the spinal cord.

The MRI of the brain was normal. There was no optic neuritis or atrophy noted.

Discussion

Neuromyelitis optica (NMO) is also known as Devic's disease. It is a demyelinating disease affecting the spinal cord and the optic nerves resulting in paraplegia and blindness with or without recovery. The disease is more common in non-white race groups.[1]

The disease is characterized by acute, severe transverse myelitis followed by simultaneous or sequential bilateral optic neuropathy. In monophasic disease the index events follow each other rapidly whilst with the multiphasic course there is a median interval of 166 days between the index events. Patients in this group experience intermittent relapses, develop worsening disability and have a poor prognosis. [1]

The two most important differential diagnoses to consider are multiple sclerosis (MS) and acute demyelinating encephalomyelopathy (ADEM). In all of the three disease entities, MRI would be the investigation of choice and many studies have identified distinguishing features of these diseases on MRI.

A study by Fillipi et al showed that all the patients in the study with NMO had a spinal cord lesion which was longer than two vertebral bodies in length whilst with MS the lesions were less than two vertebral bodies in length and tended to be multiple. Over 50% of the patients with NMO demonstrated low signal in the lesions on the T1 weighted sequence whilst none of the patients with spinal cord lesions secondary to MS demonstrated low signal on T1 weighted sequences.

The initial brain MRI of patients with NMO is normal and NMO does not include brain pathology as part of the diagnostic criteria.

Studies have found brain pathology in the relapsing type of NMO (R-NMO) and in these patients it is difficult to distinguish from the remitting, relapsing type of MS (RRMS). Cabrera – Gomez et al suggested that the diagnostic criteria be revised to include brain involvement. They found that 65% of patients with R-NMO had one or more lesion in the brain. These lesions were deep, subcortical lesions in 84.2% and were high signal on T2 weighted and FLAIR sequences and were not low signal on the T1 sequences. They were not juxtacortical in location and were <3mm in size. Periventricular lesions were found in 68.4% of patients which were not oval, ovoid or perpendicular orientated. Only 31.5% of patients demonstrated optic neuritis with contrast enhancement. These features help distinguish the disease from MS in the brain. [3]

NMO is rare in children who present with diencephalic, brainstem or cerebral hemisphere involvement, not characteristic of MS.

ADEM may present with supratentorial, assymetrical white matter high signal changes on T2 weighted sequences. Lesions are usually at the grey – white interface. 30-40% of patients may demonstrate involvement of the optic nerves. Lesions may enhance with gadolinium and if enhancement is present it is homogenous and not intense. ADEM in the spine may also present with T2 high signal lesions extending over multiple vertebral body levels. [4]

Conclusion

Neuromyelitis optica is a rare but important demyelinating disease which should be considered in a patient with acute onset of myelopathy and visual disturbances. It is difficult to come to the final diagnosis radiologically but differences from ADEM and MS should be borne in mind as these are the two most important differential diagnoses.

Figure 1a

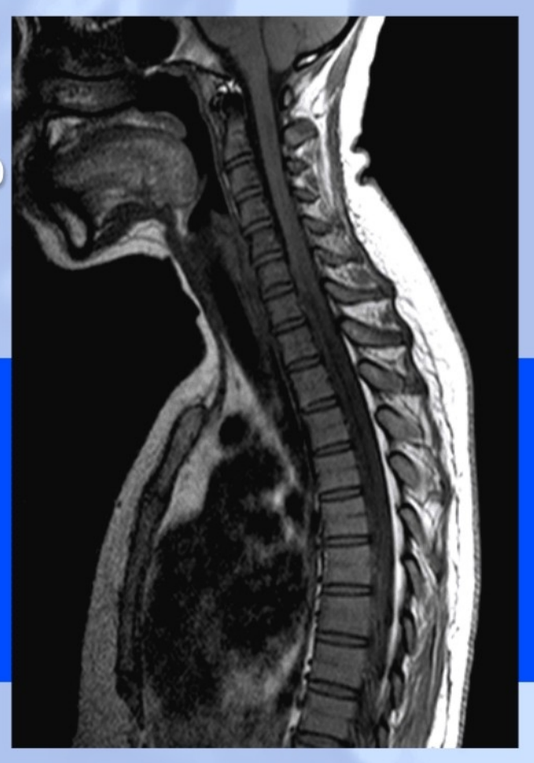
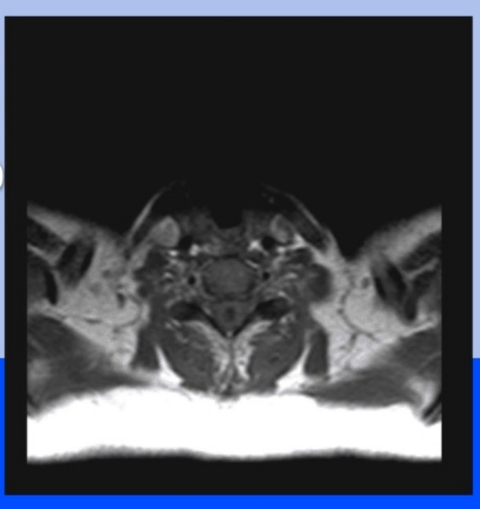


Figure 1b



Sagittal and axial MRI images show irregular, intramedullary low signal of the cord across multiple vertebral bodies i.e. from C2 to T4

Figure 2a

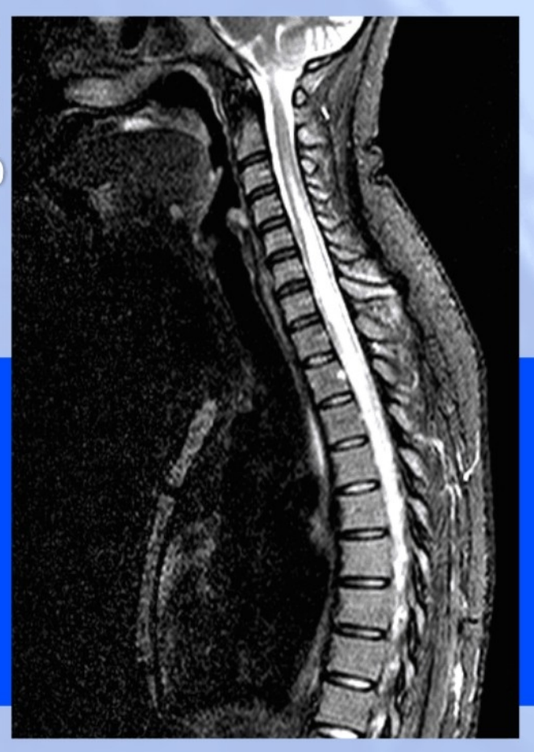
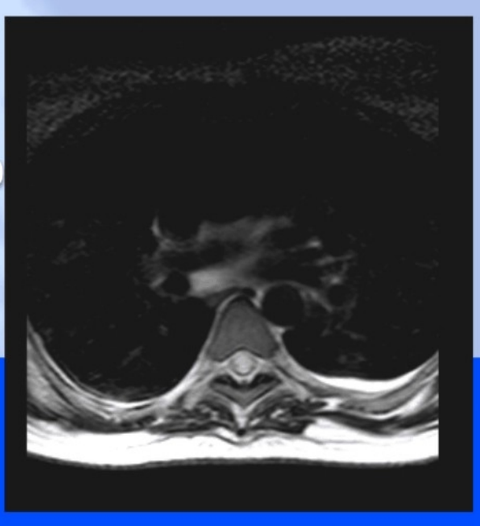
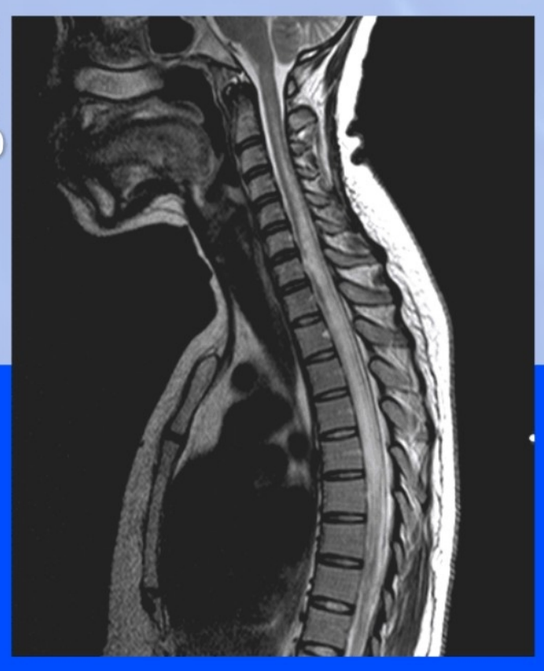


Figure 2b



Sagittal and axial T2 weighted sequence images shows diffuse intramedullary high signal in the spinal cord with expansion of the cord extending across multiple vertebral body levels

Figure 3



Sagittal STIR MRI sequence demonstrating diffuse intramedullary high signal in the spinal cord extending across multiple vertebral body levels

References

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