

CONTINUING MEDICAL EDUCATION

CASE REPORT

A young woman with weakness of the legs

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A previously well 22-year-old woman presented with progressive weakness of her legs and urinary incontinence over 7 days. Clinically she was healthy, with no skin rashes. On neurological examination she had profound bilateral weakness of the lower limbs, hypertonia, hyperreflexia, a positive Babinski sign and a T6 sensory level. Tests for syphilis and HIV and screening for auto-immune conditions were negative.

Magnetic resonance imaging (MRI) of the brain and spinal cord revealed extensive cord swelling between the craniocervical junction and T11 (Fig. 1), a high signal in the right optic nerve and a normal brain. Aquaporin 4 antibodies (neuromyelitis optica immunoglobulin G (NMO-IgG)) were positive with a titre of 1:1 000. These findings confirmed a diagnosis of NMO or Devic's disease.

Discussion

NMO is a chronic, inflammatory, demyelinating disease of the central nervous system characterised by severe attacks of optic neuritis and myelitis that spare the brain in the early stages.^[1] Women are more commonly affected, accounting for 85% of cases.^[2] Sequential or simultaneous optic neuritis and transverse myelitis are typical features. Optic neuritis and myelitis may occur separately during several months or years, but more than half of patients experience optic nerve or spinal cord relapse within one year after the initial clinical event. Optic neuritis usually presents with unilateral or, less often, bilateral ocular pain and vision loss. Myelitis caused by NMO often presents as complete transverse myelitis with tetraplegia or paraplegia, a well-defined sensory level and sphincter dysfunction. Brainstem involvement may cause intractable hiccups and vomiting.^[1]

Spinal cord lesions extending over three or more vertebral segments is the most reliable MRI finding for the diagnosis of NMO. Brain MRI is initially normal in most patients. Cerebrospinal fluid analysis may reveal a pleocytosis of >50 white blood cells/mm³.

The detection of NMO-IgG is more than 75% sensitive and 95% specific for NMO.^[3]

Intravenous high-dose corticosteroid therapy is commonly the initial treatment for acute attacks. Tapering for a period of 2 - 6 months is advised to prevent exacerbations. Rescue therapy with plasma exchange may benefit patients who do not respond to corticosteroid therapy. Maintenance immunosuppressive therapy



Fig. 1. Sagittal T2 weighted MRI of the spine. Extensive cord swelling and cord signal abnormality (arrows) between the cranial cervical junction and T11 are visible.

is a generally accepted strategy for reducing relapses. Azathioprine in combination with oral prednisone reduces the frequency of attacks.^[1]

NMO causes devastating neurological deficits and may cause blindness. A high index of suspicion is needed in patients who present with transverse myelitis and optic neuritis.

References

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