Acute optic neuritis with diffusion restriction on MRI

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DESCRIPTION
A 47-year-old man presented with rapid deterioration in vision and painful eye movements of his left eye. Two years prior he had been diagnosed with frontal left demyelinating lesion by MRI and biopsy after presenting with subacute, though rapidly progressive, right hemiparesis. His neurological deficit worsened despite the treatment with intravenous methylprednisolone and intravenous immunoglobulin, but subsequently responded to intravenous rituximab. He was clinically stable over the 2 years leading up to current presentation, with MRI that showed the residual right frontal lesion, but no other or interval lesions in the brain or spinal cord. He was presumptively diagnosed with acute disseminated encephalomyelitis.

On initial examination after the onset of his visual symptoms, he was only able to perceive light with the left eye, unable to count fingers. He had a positive relative afferent papillary defect on the left, and was clinically diagnosed with acute optic neuritis. MRI (figure 1) performed 5 days after the symptoms onset revealed high T2-weighted signal of the left optic nerve with evidence of diffusion restriction and apparent diffusion coefficient (ADC) reduction in the corresponding location.

The patient’s visual loss remained permanent despite treatment with pulsed methylprednisolone followed by a weaning course of oral prednisolone. Over the next 6 months, he had suffered from four more episodes of acute visual deterioration affecting his right eye. Diffusion restriction and ADC reduction were not demonstrated; however, he did have interval development of high short T1 inversion recovery (STIR) signal associated with contrast-enhancement in the right optic nerve (figure 2). There was persistence of high STIR signal in the left optic nerve (figure 2), this may represent ongoing inflammation or scar tissue formation. This is difficult to differentiate in the absence of new symptoms affecting the left eye in the context of extremely poor visual acuity. The loss of right-sided visual acuity was refractory to...
treatment with methylprednisolone, rituximab and plasma exchange. Subsequently, monthly intravenous cyclophosphamide was started, and as a result he remained disease free for 4 months and had some minimal improvement in right-sided visual acuity. Imaging of patient’s spine demonstrated no cord abnormality.

The results of the tests for anti-aquaporin-4 (AQP4) and anti-myelin oligodendrocyte glycoprotein antibodies were negative. However, the clinical picture was most consistent with neuromyelitis optica (NMO) spectrum disorder. Patients with relapsing disease NMO spectrum disorder tend to have a long interval between the first index event and first relapse. The first relapse is then followed by a cluster of severe relapses usually involving the optic nerve or the spinal cord, which recover poorly despite treatments. This leads to cumulative morbidities in vision and motor function. Bilateral optic neuritis is also more suggestive of NMO spectrum disorder.

Diffusion-weighted imaging is the best method to identify ischaemic lesions in the brain. Uncommonly, acute optic neuritis can also cause diffusion restriction and reduced ADC value in the optic nerve. It is unclear from the literature whether this would be a distinguishing feature between acute optic neuritis secondary to multiple sclerosis or NMO spectrum disorder.

Learning points

- Relapsing neuromyelitis optica (NMO) spectrum disorder is characterised by prolonged interval between the first index event and first relapse, followed by a cluster of severe relapse with poor recovery despite treatment.
- Lesions in the optic nerve with diffusion restriction can be ischaemic or inflammatory.
- NMO spectrum disorder can be seronegative.

REFERENCES