Letters to the Editor

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Neuromyelitis optica (Devic’s syndrome) in systemic lupus erythematosus: a case report

Sir. In systemic lupus erythematosus (SLE), neuropsychiatric involvement is one of the main causes of morbidity and mortality. Survival drops significantly after the onset of the first neurological sign, declining to 50% after 5 yr [1]. Neuromyelitis optica is one of the rarest and most serious neurological manifestations of SLE, with about a dozen cases reported in the literature to date [2].

We report the case of a 48-yr-old Irish woman with a 3-yr history of SLE. The diagnosis was based on positivity for antinuclear factor (ANF) and anti-double-stranded DNA antibodies, photosensitivity, malar rash, Raynaud’s phenomenon and non-erosive, symmetrical, inflammatory small-joint arthropathy. She presented to another hospital with acute urinary retention, headache, cold cyanotic fingers with ischaemic changes in the tips of two, and reduced vision in the right eye. Normal admission bloods tests included complete blood count, creatinine, electrolytes, liver function tests, prothrombin time, activated partial prothrombin time and C-reactive protein. The erythrocyte sedimentation rate was raised (64 mm in the first hour) and ANF was positive (1/400). Anti-double-stranded DNA antibody and anti-phospholipid antibody titres were negative, as were anti-phospholipid antibody titres. The patient was catheterized, commenced on azathioprine 75 mg twice daily and given three daily intravenous infusions of Iloprost. A gynaecological review, including transvaginal ultrasound, revealed no cause for the urinary retention. Ophthalmologic review demonstrated right-sided optic neuritis with optic atrophy and normal visual acuity on the left.

A diagnosis of neuromyelitis optica was made and intravenous methylprednisolone 1 g was administered daily for 7 days and plasmapheresis daily for 5 days [5]. Then oral prednisolone 60 mg daily was commenced and azathioprine 75 mg twice daily was continued. Only modest improvement in upper limb power occurred and the patient remained with C5/6 motor paresis. Her head control and cough improved and subsequent chest fluoroscopy demonstrated normal diaphragmatic movement. Management was complicated 1 week after admission by massive rectal bleeding, which required medical management. An area of infarction was present at the tip of her right index finger. MRI of her cervical spine showed an area of high signal extending from the medulla as far as C5/6, with slight expansion of the cord. There was no enhancement after administration of gadolinium contrast (Fig. 1). The appearance was in keeping with longitudinal myelitis.

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resuscitation with 18 units of packed red cells and emergency surgery, at which two large bleeding rectal ulcers were oversewn. As there was no further improvement in neurological status after several weeks despite ongoing azathioprine and corticosteroid therapy, she was transferred to a rehabilitation hospital for further management. Her neurological status has since remained unchanged.

Devic’s neuromyelitis optica is a condition of the central nervous system (CNS) in which visual involvement (retrobulbar optic neuritis or oedematous papillitis) is associated with myelitis of subacute onset. It has recently been distinguished from multiple sclerosis on the basis of several characteristics [progress more severe, no fever at onset, cerebral and cerebellar lesions absent on MRI, no oligoclonal bands shown on electrophoresis of the cerebrospinal fluid (CSF) and no elevation of albumin levels in the CSF] [2]. In fact, CSF analysis is of little benefit in making the diagnosis. Devic’s syndrome is also characterized by longitudinal rather than transverse myelitis.

The underlying pathology is unclear, but in our patient the presence of CNS vasculitis was strongly suggested by the peripheral vasculitic lesion. In one case report of necropsy findings, the optic nerves were strikingly atrophic. The spinal cord was greatly thinned throughout the thoracic region. Myelin staining of the optic nerves showed diffuse pallor with loss of myelin, but normal axons were found with Bodian’s stain. Diffuse demyelination was found in multiple sections of the upper cervical cord. Sections of the mid-thoracic cord showed necrosis as well as demyelination [6].

The best diagnostic tool appears to be MRI. In one study, MRI of 23 patients showed longitudinal, confluent lesions extending across three or more (often six to 10) vertebral segments in 88% of patients. Cord swelling (50%) and gadolinium enhancement (64%) were commonly observed. Twenty-two per cent of patients with follow-up imaging developed focal cord atrophy. In the same study, optic nerve enhancement was seen in five of six patients studied within 2 weeks of an episode of optic neuritis [7].

Diagnostic criteria for neuromyelitis optica have been proposed by Wingerchuk et al. [5]. Staging of the disease needs to take into account visual acuity, motor function, sensory function and sphincter function, and response to treatment can be assessed by measuring these parameters and comparing them with baseline measurements.

The course of neuromyelitis optica is often unfavourable, with poor responses to immunosuppressive regimes and corticosteroids. The development of blindness from optic atrophy is possible, and myelitis may result in permanent tetraparesis and impaired respiratory function. Rapid diagnosis and early initiation of aggressive treatment are essential in most cases if significant recovery is to occur. Currently, Devic’s disease presenting as a manifestation of CNS lupus cannot be separated from idiopathic Devic’s disease. Different authors recommend various treatment regimes, including high-dose intravenous cyclophosphamide and/or pulsed intravenous methylprednisolone [3, 4, 8], a combination of oral prednisolone and azathioprine [9] and/or plasmapheresis [5]. As in our patient, however, the prognosis is often poor, few patients attaining complete recovery [3, 7, 10].

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