

SHORT REPORT

Late central demyelination after Fisher's syndrome: MRI studies

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Abstract

The case of a patient who presented with clinical, electrophysiological, and MRI evidence of central demyelination is described. The patient had been admitted to hospital for Fisher's syndrome a few years previously. The association of these two events suggests that central and peripheral myelinopathy may be related in Fisher's syndrome.

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Since the initial report,¹ the site of neural injury in Fisher's syndrome has been debated, although most authors favour a peripheral origin and consider it to be a variant of the Guillain-Barré syndrome.² Results from MRI in Fisher's syndrome are usually normal³ even when clinical and electrophysiological studies have shown central dysfunction.⁴ Only once have MRI abnormalities been shown.⁵ We report a patient with delayed central demyelination after the typical ophthalmoplegia-ataxia-areflexia syndrome.

Case report

In October 1979 a 45 year old man had a flu-like illness followed a few days later by diplopia and unsteadiness in walking. Neurological examination showed bilateral ptosis, complete palsy of abduction of rightward and leftward gaze, dysarthria, bilateral facial weakness that was more pronounced on the right side, marked ataxia with mildly impaired joint and vibration sensations, areflexia of the legs, and hyporeflexia of the arms. There was no limb weakness. Protein concentration in cerebrospinal fluid was 600 mg/l, with 15% γ globulin but no oligoclonal banding, and there was no pleiocytosis. Electromyography of limb muscles exhibited a slightly reduced interference pattern without spontaneous activity at rest. Motor nerve conduction velocities and distal latency were normal, but sensory nerve action potentials were absent on median and sural nerves. Fisher's syndrome was diagnosed. By the end of the first week he began to improve and a month later only areflexia persisted.

In May 1986 he began to have intermittent

vertigo and diplopia and a few days later right sided clumsiness. On neurological examination he was drowsy with headache. There was right sided weakness with Babinski's sign, nystagmus, mild bilateral limb ataxia, and areflexia of the legs. Protein concentration in cerebrospinal fluid was 1200 mg/l, the γ globulin ratio was 15% without oligoclonal banding, and there was no pleiocytosis. Head CT with and without contrast gave normal results. MRI studies showed areas of increased signal in the white matter of the occipital region predominantly on the left side. Brainstem auditory evoked potentials were normal, but visual evoked potentials had abnormal responses (P100 latency: 118 ms in left eye, 120 ms in the right). Motor and sensory nerve conduction velocities and sensory nerve action potential amplitude were normal. With corticotrophin treatment he recovered fully within three weeks.

Over the following years the patient suffered from brief bouts of vertigo and diplopia without interictal neurological signs. MRI showed persistent abnormalities with a slight reduction in the size of the supratentorial lesions, and a new infratentorial lesion developed in white matter. Visual evoked potentials showed slowing of visual conduction (P100 latency 140 ms in the right eye in May 1990) without reduction in visual acuity.

Discussion

The clinical features of our patient's illness in 1979 were characteristic of the ophthalmoplegia-ataxia-areflexia syndrome reported by Fisher. As is common, electrophysiological studies confirmed peripheral changes, with abnormal results on electromyography of limbs and normal motor nerve conduction velocities but reduced amplitude of sensory nerve action potential. Neurological signs that appeared in 1986 showed that the central nervous system was affected (drowsiness and hemiparesia with Babinski's sign), and an increase in the latencies of visual evoked potentials suggested central demyelination. Moreover, areas of increased signal intensity in white matter on MRI confirmed demyelination as this 52 year old man fulfilled criteria of Fazekas *et al* for demyelinating disease⁶ by having a supratentorial lesion greater than 6 mm and subsequently developing an

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infratentorial lesion, in the white matter.

In Fisher's syndrome the pathological site of ophthalmoplegia and ataxia remains unclear, but peripheral demyelination seems to be electrophysiologically⁷ and pathological⁸ proved. Central demyelination has never been convincingly proved anatomically,⁸ and we know of only one report of central demyelination shown by MRI.⁵ In most cases MRI results are normal³ even when central signs are associated.⁴ To our knowledge, delayed central demyelination has never been reported, although recurrences of the illness are known.⁹ However, the combination of central and peripheral myelinopathy has been reported in acute¹⁰ and chronic¹¹⁻¹³ inflammatory polyneuropathy and in multiple sclerosis.¹⁰ Both lesions can be simultaneous¹³ or delayed.^{10,11}

In our patient we cannot exclude a mere coincidence of Fisher's syndrome and multiple sclerosis. However, aspects of his central demyelinating disease are rather unusual in multiple sclerosis; age above 50 years for a relapsing-remitting form, protein concentration in cerebrospinal fluid greater than 1000 mg/l; and absence of oligoclonal banding. We believe that the same demyelinating disease occurred first in the peripheral nervous system and then in the central nervous system,

as occurs in other inflammatory polyneuropathies.

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