Unusual manifestations in Miller-Fisher syndrome

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Introduction

Miller-Fisher syndrome (MFS) is an uncommon, post-infectious acute neuropathy affecting 0.1 per 100 000 population annually worldwide [1]. It is characterised by areflexia, ataxia and ophthalmoplegia. Optic neuritis in MFS is very rare; only 3 cases have been reported in the literature [2]. Development of facial nerve palsy while on plasmapheresis was found in only 2 cases of MFS in an extensive literature survey. In a review of 109 reports describing 243 cases of MFS, internuclear ophthalmoplegia (INO) was seen in only 15 cases [3].

We report 2 cases of MFS, one with optic neuritis, ptosis, papillary abnormalities, dysautonomia and development of facial palsy while on plasmapheresis, and the other with internuclear ophthalmoplegia.

Case reports

Case 1

A previously healthy, 25-year old man was admitted with a 4-day history of diplopia, unsteady gait and numbness of both feet, 3 weeks after an upper respiratory tract infection (URTI). On examination he was drowsy, febrile, ataxic and had ptosis of the right eye. There was bilateral asymmetrical external ophthalmoplegia and INO.

Funduscopy revealed venous congestion on the right. Muscle power was grade 4 on the right side, grade 5 on the left in both upper and lower limbs, with global areflexia. There was no sensory loss. Pulse was 50/min and blood pressure was normal.

Routine biochemical tests were normal. Nerve conduction studies (NCS) showed normal motor conduction velocities, but the amplitude of sensory nerve action potentials was reduced. CSF was acellular with a high protein concentration (170mg/dl) and bacterial culture, acid fast bacilli (AFB), viral studies were negative. Mycoplasma antibodies were negative, and CT and MRI scans of brain were normal.

After 72 hours of starting of plasmapheresis, he showed improvement in ataxia, ptosis, ophthalmoplegia and distal numbness. Reflexes returned to normal on day 4.

Case 2

A 45-year old previously healthy man noticed diplopia, drooping of the right eyelid, severe headache and right eyeball pain, 10 days after an URTI. Next day he developed numbness of both hands and feet with unsteady gait. He had no hypertension.

On admission to hospital on day 3, he was afebrile and had ptosis of the right eye, mild mydriasis, proptosis and bilateral complete external ophthalmoplegia. Funduscopy revealed venous congestion. He had truncal ataxia with global areflexia without limb weakness. There was no sensory loss or facial palsy. Pulse was 60/min and blood pressure fluctuated between 130/100 and 160/110 mmHg.

CT and MRI imaging of the brain were normal. Routine biochemical tests were also normal. NCS revealed normal motor conduction with reduced amplitude of sensory nerve action potentials. CSF was acellular with high protein concentration (320mg/dl). CSF bacterial culture, AFB and viral studies were negative.

On day 5 he developed partial ptosis of the left eye and optic neuritis as evidenced by the presence of bilateral afferent pupillary defects, reduced visual acuity (6/24 on right, 6/9 on left) and peripheral field constriction on perimetry. By day 12, ataxia, ptosis of the left eye and optic neuritis recovered.

On day 16 (day 4 on plasmapheresis) he developed left facial palsy. During his clinic follow up facial palsy, ophthalmoplegia and areflexia improved.

Discussion

MFS is a variant of Guillain-Barre syndrome (GBS) sharing clinical and neurophysiological features. Often the causative agent is not identified, although Campylobacter jejuni and Haemophilus influenzae can induce antibodies that cross-react with neural antigens leading to neuropathy [1].

MFS is closely associated with antibodies to GQ1b gangliosides (Anti GQ1b Ab), which are present in over

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90% of patients with MFS and may contribute to the pathogenesis [1]. Anti GQ1b Ab is also associated with Bickerstaff's brainstem encephalitis (BBE) with similar clinical picture, together with drowsiness and extensor plantar response, overlap of GBS with ophthalmoplegia and acute ophthalmoplegias without ataxia suggesting the clinical spectrum of Anti GQ1b Ab syndrome [4].

GQ1b gangliosides are present in appreciable amounts in 2nd, 3rd, 4th and 6th cranial nerves suggesting its involvement in ophthalmoplegia and optic neuritis [4]. The first patient was drowsy on admission with flexor plantar response and impairment in sensory nerve action potentials suggesting a disease entity in the spectrum of GBS, MFS and BBE. Bilateral INO is rare in MFS, which was seen in case 1. This has been reported before [5,6,7].

Case 2 had optic neuritis. Optic nerve involvement in MFS was reported only in 6 cases in the literature, two with visual pathway dysfunction, one with demyelinating optic neuropathy and 3 with optic neuritis [2].

Facial palsy occurs in about 45% of MFS [7] but case 2 developed facial palsy while on plasmapheresis when other signs were improving, which is rare. GQ1b gangliosides are not present in facial nerves. Different antibodies may be involved in the development of facial palsy or various pathomechanisms are involved in different stages of MFS [8].

Dysautonomia in MFS can manifest as hypertension, bradycardia, postural hypotension or cardiac arrhythmia. The second patient had an asymmetrical complete third nerve palsy which is not common in MFS. He also had unilateral proptosis, which is unusual in MFS. These two reports highlight the importance of early detection of MFS even with unusual manifestations as it is amenable to treatment with immunomodulatory therapies.

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References