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An unusual case of Miller Fisher syndrome presenting with proptosis and chemosis

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Abstract

Miller Fisher syndrome (MFS), a rare variant of Guillain-Barré syndrome, is characterized by ophthalmoplegia, ataxia, and areflexia. In addition to this classic triad, symptoms may include bulbar palsy, weakness, and sensory loss. The anti-GQ1b IgG antibody is a sensitive and specific marker for MFS; it is found in more than 90% of affected patients. We describe an unusual case of MFS that presented with dramatic bilateral proptosis and chemosis.

Keywords

Miller Fisher syndrome; Guillain Barré syndrome; proptosis; GQ1b antibody; ophthalmoplegia

Case report

A 62-year-old African-American woman was transferred to our hospital with proptosis, diplopia, and difficulty walking. Her symptoms began two weeks earlier, shortly after an episode of sore throat and productive cough. She also experienced malaise, dysarthria, dysphagia, and paresthesias in her hands and feet. She had no pain. Medical history included hypertension and hyperlipidemia. She had no history of ophthalmologic disorders.

Physical examination demonstrated prominent bilateral proptosis and ptosis (greater on the right), lagophthalmos, right exotropia and left esotropia. She exhibited nearly complete ophthalmoplegia, 2-3+ conjunctival injection, chemosis, and lid edema (Figure, A). Her pupils were dilated and minimally reactive. Visual acuity was decreased (20/100 OD, 20/50 OS) compared to her baseline (20/20 OD, 20/25 OS). Disc margins were sharp on fundus examination. Visual fields were full bilaterally to 3-stage confrontation. Intraocular pressures were elevated (28-31 mmHg, normal range 10-21).

Speech was nasal. Strength was mildly decreased symmetrically in all extremities. Sensation was intact. Reflexes were absent throughout. Coordination testing demonstrated mild dysmetria and ataxia.

Laboratory studies showed normal thyroid function and negative HIV, rapid plasma reagin (RPR), and acetylcholine receptor antibody serologies. CSF examination demonstrated elevated protein concentration (75 mg/dL, normal range 40-70) and normal white blood cell count (3 cells/mm³, normal range 0-5). The CSF glucose level was normal. Bacterial, viral, and fungal testing were negative. Testing for anti-GQ1b IgG antibody, a sensitive and

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specific marker for Miller Fisher syndrome (MFS) found in more than 90% of affected patients (1), was positive at a titer of 1:200.

Noncontrast CT of the head and CT angiography showed no acute intracranial vascular or parenchymal pathology. The only abnormality of the orbits was protrusion of the globes and thinner than normal extraocular muscles (Figure, C). MRI of the brain with and without gadolinium contrast demonstrated only nonspecific periventricular white matter changes.

Nerve conduction studies demonstrated absent sensory nerve action potentials in the median, ulnar and sural nerves, with preserved compound muscle action potentials. Baseline testing and repetitive stimulation testing of the right facial nerve at 3 Hz were normal.

Following admission, her ophthalmologic examination progressed to complete bilateral ophthalmoplegia, with worsened bilateral chemosis and subconjunctival hemorrhages. Swallowing also worsened. Cinematic barium esophagogram indicated severe flaccid, ataxic oropharyngeal dysphagia. Empiric treatment was initiated with intravenous immunoglobulin (IVIG) (0.4 g/kg for 5 days). In addition, she received timolol-dorzolamide eye drops to decrease intraocular pressure and aggressive corneal lubrication to treat exposure keratopathy. Enteral nutrition was provided through a Dobhoff tube. Over the following week, her eye swelling began to resolve, speech improved, and deep tendon reflexes returned. Five days after she completed treatment with IVIG, she could tolerate a soft mechanical diet without aspiration. Her ataxia resolved gradually with physical therapy. Intraocular pressure normalized. Ophthalmoplegia improved slowly, beginning with incremental recovery of vertical eye movements. She started to regain horizontal eye movements one month after hospital admission. At the time of discharge from inpatient rehabilitation, one month after IVIG therapy, her proptosis had resolved (Figure, B), and the GQ1b antibody level was undetectable.

Discussion

Ophthalmoplegia is a characteristic feature of MFS. Other reported eye findings include pupillary abnormalities and ptosis (2). Less common symptoms include acute angle-closure glaucoma (3) and optic neuritis (4). We report an unusual case of serologically-confirmed MFS that presented with dramatic proptosis and chemosis, apparently due to the disease itself.

We are aware of only two other reports of proptosis in MFS. In one, this symptom was due to concurrent Graves disease (5). In the second, the diagnosis of MFS is questionable (6). The patient in that report was a 44-year-old woman with a history of irregular treatment for thyroid dysfunction who presented with diplopia and proptosis. There is no mention of ataxia or areflexia, and GQ1b antibodies were not tested. The authors base the diagnosis of MFS on the patient's diplopia and incomplete ophthalmoplegia, albuminocytologic dissociation with a CSF albumin level of 27.83 mg/dl (which is within the reference range for adults), and the recovery of normal vision within two months in association with treatment with intravenous and oral steroids. The authors assert that myasthenia gravis was excluded as a cause of diplopia, because repetitive nerve stimulation testing was normal; no mention is made of assessment of serum acetylcholine receptor antibodies, which is a more sensitive test for that disorder. Moreover, proptosis caused by thyroid dysfunction may not improve even after suitable thyroid treatment.

The ganglioside GQ1b is found abundantly in cranial nerves and spinal roots, and anti-GQ1b antibodies are thought to be important in the pathogenesis of MFS (1,7). We suggest that thin extraocular muscles may have been an idiopathic finding that predisposed our patient to laxity in globe position after cranial nerve impairment and led to development of

proptosis. Consistent with this hypothesis, two cases of passive proptosis occurring after oculomotor nerve injury have been reported (8). Ophthalmoplegia and weakness of eye closure may then have propagated a cycle of increasing desiccation, conjunctival inflammation, and chemosis.

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Abbreviations

CSF	cerebrospinal fluid
CT	computed tomography
HIV	human immunodeficiency virus
IVIG	intravenous immunoglobulin
MFS	Miller Fisher syndrome
MRI	magnetic resonance imaging
OD	right eye (oculus dexter)
OS	left eye (oculus sinister)
RPR	rapid plasma reagin

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Figure.
 Photographs and CT scan of patient
 (A) Photograph illustrates proptosis, chemosis, and lagophthalmos. (B) One month later, ocular symptoms resolved. (C) CT scan of the orbits demonstrates proptosis, with protrusion of globes beyond the orbital rims, as well as thin extraocular muscles, with no indication of intraorbital pathology.