CASE REPORT

A rare case of complete bilateral ophthalmoplegia and ptosis

DANIEL JOHN HALL, TALAL BAZARAA

Medicine, Calderdale and Huddersfield NHS Foundation Trust, Calderdale Royal Hospital Salterhebble, Halifax, West Yorkshire HX30PW, UK

Address correspondence to: D. J. Hall. Tel: 07738913461; Email: daniel-hall@doctors.org.uk

Abstract

We describe the case of an 85-year-old gentleman admitted with bilateral ptosis and complete bilateral ocular paralysis. Initial differential diagnoses included myasthenia gravis, diabetic cranial neuropathy, an ischaemic event and possible occult neoplasm. Investigations did not support any of the differentials and Miller Fisher syndrome (MFS) was considered. Anti-GQ1b IgG antibody was positive, supporting the possibility of anti-ganglioside syndrome. This gentleman was treated with intravenous immunoglobulin (IVIG) and made a full recovery.

Keywords: ophthalmoparesis, Miller Fisher syndrome, Anti-GQ1b antibody, bilateral ptosis, older people

Case report

An 85-year-old gentleman was admitted to the acute medical unit by his general practitioner with a 4-day history of increasing ptosis, ocular paralysis and diplopia. He reported a mild respiratory illness a week prior to visiting his doctor. Past medical history included Type II Diabetes Mellitus and vitamin B12 deficiency. He had never smoked and did not consume alcohol. Clinical examination showed evidence of complete bilateral ptosis and paralysis of all ocular movements. Pupils were normal and fundus examination was unremarkable. He was normoreflexic, without cerebellar signs and no muscle fatigability was present. Neurological and clinical examinations were otherwise unremarkable.

Initial differential diagnoses included myasthenia gravis, diabetic ocular polyneuropathy and a cerebral ischaemic event. Empiric prednisolone and pyridostigmine treatments were begun. Initial investigations including full blood count, electrolytes, liver function tests, thyroid function tests and CRP were within normal ranges. Magnetic resonance imaging of the brain revealed no infarct or mass lesion but age appropriate ventricular dilatation with non-specific white matter changes; indicative of small vessel cerebrovascular disease. Further investigations, including Tensilon test, acetyl-choline receptor antibody, auto-antibody screen, tumour markers, syphilis serology and MuSK-antibodies, were negative. CT chest, abdomen and pelvis revealed no sinister pathology.

Further discussion with neuroradiology led to a second MR brain scan with contrast to investigate the non-specific ischaemic changes previously seen in the brainstem, especially the pre-tectal region. However, this showed no abnormalities, particularly none in the brainstem and tectal area. Neurophysiology showed evidence of symmetrical generalised sensory and motor polyneuropathy, most likely a result of pre-existing diabetes. There was no evidence of any neuromuscular junction disease, either post-synaptic as in myasthenia gravis or pre-synaptic as in Eaton–Lambert syndrome or botulism. A diagnosis of Miller Fisher variant was considered and Anti-GQ 1b antibodies were requested. Lumbar puncture, to check for albuminocytological dissociation was declined by the patient. The steroids and pyridostigmine were gradually withdrawn and he was given intravenous immunoglobulin (IVIG).

To facilitate discharge, he had a sling operation to the right eyelid. He was monitored in the outpatient clinic and 8 weeks later his symptoms resolved completely. Anti GQ 1b antibody was raised at 100 titre (Normal 0–25); thereby confirming the diagnosis (Figures 1 and 2).
Complete bilateral ocular paralysis is a very rare presentation and such patients are difficult to fit into a simple diagnostic criterion. Literature regarding patients presenting with acute ophthalmoparesis and bilateral ptosis is sparse. Common causes of multiple cranial nerve lesions include tumours, vascular disease, trauma, infection and Guillain–Barré syndrome (GBS) [1, 2]. MFS is considered as a variant of GBS [3].

Charles Miller Fisher, in 1956, described three cases characterised by the clinical triad of ophthalmoplegia, ataxia and areflexia [4]. However variants of this triad are known to exist [2, 3]. The condition is usually self-limiting and the outcome is favourable.

Anti GQ1b antibody is positive in over 80% of Miller Fisher syndromes. Positive Anti-GQ1b IgG results are also seen in GBS with ophthalmoplegia, Bickerstaff’s brainstem encephalitis (BBE) and acute ophthalmoparesis [5]. Growing opinion suggests all these conditions exist on a clinically continuous spectrum as part of an anti-GQ1b/anti-ganglioside syndrome and have a common post-infectious autoimmune mechanism in their pathogenesis. The recognition of the aetiological relationship in these conditions is important to support the early use of the same therapeutic options established for GBS (IVIG and plasmaphoresis) as some patients with BBE, MFS and acute ophthalmoparesis have been shown to respond favourably to intervention.

Our patient presented with ophthalmoplegia without ataxia [5] and positive anti-GQ1b antibody, suggesting anti-ganglioside syndrome [6] which can be labelled as atypical Miller Fisher syndrome or acute ophthalmoparesis. He was treated with IVIG and achieved full recovery.

**Key points**

- Bilateral ptosis and complete ophthalmoplegia is a rare occurrence.
- Acute ophthalmoparesis, Miller Fisher syndrome, GBS (with ophthalmoplegia) and BBE are a continuum of a clinical syndrome.
- Anti-GQ1b antibody is a marker for the clinical syndrome and indicates a common aetiology.
- Identification of anti-GQ1b antibody is important to facilitate the use of treatment already established for GBS.

**Acknowledgements**

We are grateful to the neurology team, Dr B. Dafalla, Dr A. Mahmoud and Dr A. Zeidan and consultant ophthalmologist Miss Hollingworth for their help in managing the patient.

**Conflicts of interest**

None declared.

**References**


Received 16 April 2013; accepted in revised form 19 July 2013