Total internal and external ophthalmoplegia as presenting symptoms of Miller Fisher syndrome

Bik Ling Man

Department of Medicine and Geriatrics, Tuen Mun Hospital, Hong Kong, Hong Kong

Correspondence to
Dr Bik Ling Man,
beli_man@yahoo.com

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DESCRIPTION

A 46-year-old woman, who enjoyed good past health, presented with progressive diplopia and unsteady gait after a recent upper respiratory tract infection. She had no fever and denied use of any cycloplegic drugs. Physical examination showed bilateral ptosis ([figure 1](#)), and total external ([video 1](#)) and internal ophthalmoplegia ([video 2](#)). She had bilateral facial weakness and ataxia of limbs. Her limb power was full and there was generalised hyporeflexia. Cerebrospinal fluid examinations showed white cell count $<1/\text{mm}^3$ (<5), protein 0.28 g/L (0.15–0.4), glucose 4.3 mmol/L (2.2–3.9). Serum anti-GQ1b IgM and IgG were strongly positive. MRI of the brain and whole spine were normal. NCT and EEG were normal. She was diagnosed to have Miller Fisher syndrome and was treated with a course of intravenous immunoglobulin. Her ophthalmoplegia improved markedly after treatment.

The GQ1b ganglioside is highly enriched in oculomotor, trochlear and abducens nerves, and the ciliary ganglia.¹ Postganglionic involvement of the parasympathetic nerve is associated with internal ophthalmoplegia in anti-GQ1b antibody syndrome.² Internal and external ophthalmoplegia is common in anti-GQ1b antibody syndrome³ and it should be considered in the differential diagnosis of acute or subacute ophthalmoplegia to allow prompt diagnosis and treatment.

Video 1 Total external ophthalmoplegia.

Video 2 Total internal ophthalmoplegia.

Figure 1 Bilateral ptosis.

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Learning points

- ▶ The GQ1b ganglioside is highly enriched in oculomotor, trochlear and abducens nerves and the ciliary ganglia.
- ▶ Postganglionic involvement of the parasympathetic nerve is associated with internal ophthalmoplegia in anti-GQ1b antibody syndrome.
- ▶ Internal and external ophthalmoplegia is common in anti-GQ1b antibody syndrome and it should be considered in the differential diagnosis of acute or subacute ophthalmoplegia.

Competing interests None.

Patient consent Obtained.

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