A severe overlap case of Miller Fisher and the pharingeal-cervical-brachial variant of Guillain Barré Syndrome.

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BACKGROUND

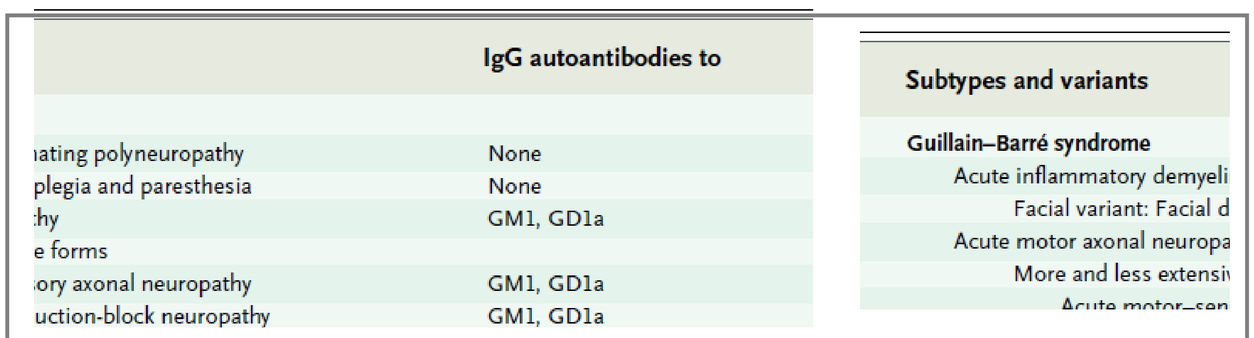
Overlap of pharyngeal–cervical–brachial weakness and Fisher syndrome (PCB/FS) are uncommon (1). The diagnosis is difficult when clinical onset is abrupt and lead to hospitalisation to intensive care unit (ICU). Indeed, neurological examination is limited and diagnosis often relies on paraclinical tests. Nevertheless, diagnosis of Fisher Syndrome (FS) is clinical, EMG/ENG and CSF being normal in the early stage of disease (2-4). Pharyngeal–cervical–brachial weakness (PCB) represents a variant of Guillain–Barré syndrome, characterised by axonal rather than demyelinating neuropathy (5).

CASE REPORT

We herewith report on a 69-year old men, referred to our Unit for an acute diplopia associated with gait disturbance appearing 1 week after an upper respiratory tract infection. Medical history of the patient included a chronic alcohol abuse, type II diabetes mellitus and a prostatic carcinoma, treated with surgery and chemo-radiotherapy resulting in an erectile dysfunction.

Neurological examination at admission showed a bilateral ptosis with reactive pupils, bilateral ophthalmoplegia with complete gaze palsy, bilateral facial nerve paralysis, a generalized apallesthesia with normal osteotendinous reflex, except for bilaterally abolished Achillean reflex. Strength and superficial sensibilities were normal.

A prostigmine test was performed in the hypothesis of Myastenia Gravis and resulted negative. Brain MRI and CSF were strictly normal. Intravenous immunoglobulin therapy was started. The day after, the patient was transferred to the ICU due to a sudden aggravation of the facial paralysis and onset of dysphagia, leading to inhalation and respiratory insufficiency. After starting mechanical ventilation, a refractory hypertension appeared. Spirometry did not disclose a neuromuscular deficit.



	IgG autoantibodies to	Subtypes and variants
		Guillain–Barré syndrome
ıating polyneuropathy	None	Acute inflammatory demyeli
plegia and paresthesia	None	Facial variant: Facial d
hy	GM1, GD1a	Acute motor axonal neuropa
e forms		More and less extensi
ory axonal neuropathy	GM1, GD1a	Acute motor–sen
uction-block neuropathy	GM1, GD1a	

Guillain–Barré syndrome

- Paraparetic variant*
- Bifacial weakness with paraesthesias*
- Acute motor axonal neuropathy (GM1,GD1a)
- Acute motor-sensory axonal neuropathy (GM1,GD1a)
- Acute motor conduction block neuropathy (GM1,GD1a)
- Pharyngeal–cervical–brachial weakness* – Acute pharyngeal weakness*† (GT1a> GQ1b>>GD1a)

Miller Fisher syndrome (GQ1b, GT1a, GD1a)

- Acute ataxic neuropathy† (GQ1b, GT1a > GD1b)
- Acute ophthalmoparesis† (GQ1b, GT1a)
- Acute mydriasis† (GQ1b, GT1a)
- Acute oropharyngeal palsy (GQ1b, GT1a)
- Bickerstaff's brainstem encephalitis‡ – Acute ataxic hypersomnolence‡† (GQ1b, GT1a > GD1b)

Fig 1: Guillain–Barré and Miller Fisher syndromes and their subtypes and associated antibodies. *Localised forms. †Incomplete forms. ‡Central nervous system form. In brackets, autoantibodies associated with the variant. Modified by B R Wakerley and N Yuki, Pract Neurol, 2015.

EMG/ENG did not reveal any conduction block and found an axonal bilateral motor involvement of V, VII and XII cranial nerves.

Routinary blood tests, including HIV test, thyroid exams, B vitamins dosage, a large serological infectious screening were negative. An extensive serum autoantibody screening (AchR, MuSK, rheumatoid factor, ANCA, antinuclear antibodies, anticardiolipines) and complement system protein concentrations were unremarkable. We screened all the antiganglioside antibodies and we found a positive anti GQ1b and GT1a (IgG) and positive anti-GD1a, GD1b, GM1(IgM).

During hospitalisation in the ICU, neurological examination progressed with strenght deficit of the neck flexor muscles, upper limbs symetrical areflexia, bilateral deficit of the hypoglossal nerve. Severe dysautonomic syndrome with refractory hypertension was also disclosed.

CONCLUSIONS

The patient presented clinical aspect of both FS and PCB syndromes, revealing an overlap diagnosis without lower limb weakness and an uncommon rapid onset an progression.

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