Unravelling a case of pseudo one and a half syndrome

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DESCRIPTION
This is a female in her 50s with a medical history of hypertension who presented to her outpatient clinic for evaluation of ‘blurred vision’ for 2 weeks, subsequent to having placed faux lashes with latex glue, the latter to which she claimed to be significantly allergic to. She complained of facial pain and a headache, though denied fever, dysphonia, dysphagia, weakness or other symptoms. On presentation, her examination demonstrated bilateral chemosis of both eyes with left upper eyelid swelling. Her extraocular movements revealed defects with abduction and adduction of the right eye, along with restricted adduction and vertical gaze movements of her left eye, a possible 1½ syndrome or a pseudo-1½ syndrome (see figure 1). Her vision in each eye was normal and her fundi were unremarkable bilaterally.

With concerns of an urgent pathology such as orbital cellulitis, cavernous sinus thrombosis, this patient underwent an emergent CT scan of orbits and sinuses along with MRI and magnetic resonance venography (MRV) of the brain, which were unremarkable. Her labs demonstrated normal thyroid functions but positive acetylcholine receptor-binding antibodies.

On a follow-up 2 weeks later, the chemosis in both eyes and the left eyelid swelling had resolved, although a left eye ptosis in addition to her external ophthalmoplegia was now noticeable.

After receiving two doses of 60 mg of pyridostigmine orally, this patient’s symptoms of ‘binocular diplopia’ had improved significantly, as did her left eye ptosis and extraocular movements in both eyes.

With a diagnosis of ocular myasthenia gravis, this patient was continued on pyridostigmine 60 mg three times a day along with prednisone 5 mg per day. A CT chest was obtained which failed to reveal a thymoma. She continued to demonstrate improvement with her binocular diplopia over the following weeks.

Myasthenia gravis may affect any of the six extraocular muscles, masquerading as any type of ocular motor pathology.1 Multiple studies have examined the pattern of ocular findings in myasthenia gravis but no clear clinical pattern has yet been described.2

One and a half syndrome classically involves the pontine region of the brainstem but the presence of a vertical gaze palsy as demonstrated in our patient above raises the possibility of midbrain or cerebral hemisphere involvement and had to be ruled out. Although pseudo one and a half syndrome has been commonly associated with myasthenia gravis, it can present similarly to Guillain-Barré syndrome (GBS), or more specifically, the Miller-Fisher variant of GBS which is characterised by a triad of ataxia, areflexia and ophthalmoplegia.3 Internuclear ophthalmoplegia (INO) is also commonly seen in...

Figure 1 Ocular examination with patient demonstrating left lateral gaze (A), right lateral gaze (B) and vertical gaze (C).

Learning points
► One and a half syndrome, a central nervous system pathology, typically occurs due to involvement of the paramedian pontine reticular formation or abducens nucleus and the medial longitudinal fasciculus, presenting as a ipsilateral conjugate horizontal gaze palsy and a contralateral internuclear ophthalmoplegia (inability of the contralateral eye to adduct).
► Pseudo one and a half syndrome most often has aetiologies related to the nerves innervating the extraocular muscles (Miller Fisher syndrome) or the neuromuscular junction (ocular myasthenia gravis/myasthenia gravis). These diseases can present similarly to a true one and a half syndrome but with wider possibilities of gaze palsies.
► With our patient’s acute and dramatic presentation with facial/eye pain and headache, a history of severe latex allergy, significant chemosis of both eyes and eyelid oedema, aetiologies such as orbital cellulitis, cavernous sinus thrombosis and stroke had to be ruled out before considering a diagnosis of ocular myasthenia gravis.
multiple sclerosis however demyelination of the medial longitudinal fasciculus is frequently bilateral, leading to bilateral INO.3

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

REFERENCES