Pseudointernuclear ophthalmoplegia as a presenting feature of ocular myasthenia gravis

Britta Nijsse,[1] Marijke Wefers Bettink,[2] Rinze F Neuteboom[1]

[1]Department of Neurology, Erasmus Medical Center, Rotterdam, The Netherlands
[2]Department of Ophthalmology, Rotterdam Eye Hospital, Rotterdam, The Netherlands

Correspondence to
Britta Nijsse,
b.nijsse@elisabeth.nl

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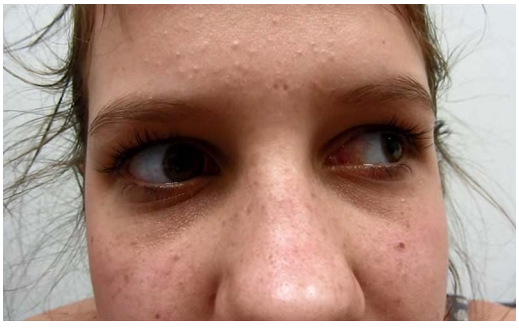
DESCRIPTION

A 16-year-old girl presented with fluctuating diplopia, ptosis of the left eye and an internuclear ophthalmoplegia (INO; video 1), which resolved after administration of neostigmine. Antiacetylcholine receptor antibodies were present in the serum: 0.34 nmol/L (normal range 0–0.25 nmol/L). These findings led to the diagnosis of ocular myasthenia.

INO is mostly seen with lesions of the medial longitudinal fasciculus in the dorsomedial brainstem tegmentum of either the pons or the midbrain, causing adduction weakness of the ipsilateral eye and a contralateral abduction nystagmus. The underlying mechanisms causing abduction nystagmus are not fully understood. Probably, it reflects an adaptive process that helps to overcome the adduction weakness of the opposite eye.[1] This is consistent with Hering's law of equal innervation, which states that increased innervation to a weak muscle in one eye is accompanied by a commensurate increase in innervation to the yoke muscle in the other eye.

In myasthenia gravis (MG), extraocular muscle weakness can cause the same oculomotor pattern as in INO.[2] However, in such cases there is no lesion in the central nervous system and therefore it has been referred to as pseudo-INO. In MG, there is intermittent block of neuromuscular conduction to extraocular muscles, which causes muscle weakness. Although the adduction weakness has a different cause as in INO, the assumed pathogenetic mechanism of the abduction nystagmus in pseudo-INO is the same.[3] Pseudo-INO can also be a clinical manifestation of Guillain-Barré syndrome (GBS) or the Miller-Fisher variant of GBS.[3]

The symptoms in our patient completely resolved rapidly after treatment with low doses of pyridostigmine (30 mg four times a day).



Video 1 Nystagmus of the abducting left eye and impaired adduction of the right eye, representing a pseudo-internuclear ophthalmoplegia (pseudo-INO) in a patient with ocular myasthenia gravis. Also note the slight ptosis of the left eye.

Learning points

- In any patient with an internuclear ophthalmoplegia (INO) which can not be explained by a lesion in the brainstem, consider pseudo-INO.
- In the case of pseudo-INO, it is important to perform additional tests on diagnosing myasthenia gravis.

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Competing interests None.

Patient consent Obtained.

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