Internuclear Ophthalmoplegia: Manifestation of Clinically Isolated Syndrome in Adolescence

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A 13-year-old female with myopia and otherwise healthy was admitted to the emergency department due to diplopia for the previous five days. She had no fever or other symptoms. Trauma or drugs were denied. On physical examination, she presented impairment of adduction of the left eye with abduction horizontal nystagmus of the right eye; convergence was intact (Fig. 1). No other neurological findings were identified. Brain computed tomography and blood studies were negative. Brain magnetic resonance imaging revealed a hyperintense focal lesion in long repetition time (TR) sequences at the floor of the fourth ventricle, near the left medial longitudinal fasciculus, showing slight edema and non-enhancing (Fig. 2). Cerebrospinal fluid showed 13 leukocytes, normal glucose and proteins, and positive oligoclonal bands. Spine magnetic resonance imaging was negative. Findings were consistent with a left internuclear ophthalmoplegia due to a demyelinating lesion of medial longitudinal *fasciculus*. The patient was started on intravenous methylprednisolone for five days with subsequent taper. After 14 months of follow-up, she remains asymptomatic without therapy, with normal neurological examination and no changes in neuroimaging. Internuclear ophthalmoplegia is a specific abnormality in horizontal gaze. It presents with diplopia and results from a lesion (usually demyelinating or ischemic) of the medial longitudinal fasciculus. The medial longitudinal fasciculus is a white matter fiber tract that extends through the brainstem and interconnects the nuclei of cranial nerves III. IV. and VI. providing conjugate eye movements.¹⁻³ In adolescents, the etiology is most often multiple sclerosis² and the presence of oligoclonal bands in cerebrospinal fluid evidences an isolated intrathecal synthesis of immunoglobulins, increasing the likelihood of multiple sclerosis.4

Usually convergence is intact and this is a useful sign that strongly suggests internuclear ophthalmoplegia because it does not occur in oculomotor nerve palsy or in an isolated medial rectus paresis.¹ Due to the lack of clinical or imaging evidence of another demyelinating event, the diagnosis of clinical isolated syndrome was established.⁴⁻⁵ Clinical and imaging surveillance are essential.



Figure 1. Left internuclear ophthalmoplegia. Impaired adduction of the left eye, ipsilateral to the lesion (above). Convergence is intact (below). Abduction nystagmus in the contralateral eye is characteristic.



Figure 2. Hyperintense focal lesion in long TR sequence (blue arrow) in the left paramedian floor of the IV ventricle (topography of the left medial longitudinal *fasciculus*).

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WHAT THIS REPORT ADDS

• Diplopia may arise due to many different etiologies, including neurogenic lesions.

• Lesions in the medial longitudinal *fasciculus* cause internuclear ophthalmoplegia, a specific gaze abnormality characterized by weak adduction of the affected eye on horizontal movements. Abduction nystagmus of the contralateral eye may be present. Often convergence is preserved, and it is a useful sign that strongly suggests internuclear ophthalmoplegia.

• A clinically isolated syndrome is a first episode of neurological symptoms caused by demyelination lesion in the central nervous system, suggestive of multiple sclerosis. The diagnosis of multiple sclerosis is established when the patient fulfills the criteria of the dissemination of demyelinating lesions in space and time.

• Patients with a clinically isolated syndrome need to be monitored for clinical and neuroimaging relapses. Clinically isolated syndrome patients with positive oligoclonal bands were more than twice as likely to convert to multiple sclerosis compared with oligoclonal bands negative patients.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

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Confidentiality of data

The authors declare that they have followed the protocols of their work centre on the publication of patient data.

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