Internuclear ophthalmoplegia: a case study

BY MIGUEL KURC

A 44-year-old patient presents with binocular, mostly horizontal diplopia when fixating on moving objects. This has been ongoing for approximately five years. He is known to have multiple sclerosis and had an episode of optic neuritis six years prior in the right eye which was treated with intravenous methylprednisolone infusions. The patient has begun to fixate with his left eye only to alleviate his symptoms. He is also noticing that his right eye has become more divergent than before.

During his examination, the visual acuity was 0.54logMAR (uncorrected) in the right eye and 0.52 (uncorrected) in the left eye and did not improve with pinhole. His intraocular pressures were 10 and 15 in the right and left eye respectively. There was no colour vision deficit but there was a right relative afferent pupillary defect (RAPD). Anterior and posterior segment abnormalities were largely unremarkable but the Heidelberg macula optical coherence tomography showed minimal reduction in the retinal nerve fibre layer of the right eye when compared with the left. Ocular motility testing showed a minimal elevation deficit (appears to be superior rectus weakness) as well as bilateral adduction deficit (right more than left). There was also a small end-point nystagmus in the abducting eye noticed on both sides.

The initial diagnosis for this patient was a binocular internuclear ophthalmoplegia and urgent MRI head was arranged with referral to neurology for their input.

What is internuclear ophthalmoplegia?

Internuclear ophthalmoplegia (INO) is an eye movement disorder affecting the conjugate horizontal movement of the eyes. This is usually caused by damage of the interneurons between the nuclei of the third and sixth cranial nerves located in the dorsal midbrain and pons [1]. This interneuron is called the medial longitudinal fasciculus (MLF). The horizontal eye movements are mediated by the paramedian pontine reticular formation (PPRF) and the sixth cranial nerve nucleus, which controls contraction of the ipsilateral lateral rectus muscle, and they transmit their signal along the MLF to the contralateral third cranial nerve nucleus, which controls the contraction of the medial rectus muscle [1]. This simultaneous and equal contraction of the ipsilateral lateral rectus and contralateral medial rectus allows for horizontal eye movement and follows Hering's law of equal innervation. The laterality of the INO is defined by the side with adduction deficit, which is ipsilateral to the MLF affected.

Symptoms and signs

Patients with INO may have symptoms of varying severity, ranging from asymptomatic to diplopia or even oscillopsia. If diplopia is present it tends to be on gazes contralateral to the INO. Some of the more common presenting complaints of presents is reading fatigue, visual confusion, headaches / vertigo and loss of depth-perception among others [2].

The main finding on clinical examination will be an ipsilateral adduction deficit of the affected eye and a contralateral horizontal nystagmus with fast phase towards position of gaze, this may be unilateral or bilateral. In some cases, there will also be a skew deviation (a vertical eye movement with an excyclotorsion element) and depending on the location of the lesion, convergence may be spared.

Causes

Internuclear ophthalmoplegia can be caused by any lesion or insult affecting the dorsal midbrain or pons. The most common causes are cerebrovascular disease and multiple sclerosis (as is the case with the above case). Other less common causes include trauma, brainstem tumours, infections, hydrocephalus, Arnold-Chiari malformations and lupus erythematosus [3]. An important condition to be aware of that can mimic INO is myasthenia gravis. An important way to differentiate these is by their history (fatigability, onset, duration) as well as the potential for convergence (which may be preserved with INO) [2].

Other associations with INO

Apart from an isolated INO, a patient may present with few other syndromes that involve INO. One and a half syndrome, where a lesion affects the MLF and the PPRF / sixth nerve nucleus resulting in the inability for the ipsilateral eye to move horizontally and the contralateral eye may abduct. There is also eight and a half syndrome, where the lesions affect the MLF, PPRF / sixth nucleus and the adjacent facial fascicular nerve (cranial nerve seven) resulting in the above as well as an ipsilateral facial weakness or droop.

In conclusion, INO is a well-known condition with ranging symptomology and aetiology. Its presentation warrants thorough examination and investigations as its common causes may have severe health implications if left undiagnosed. Collaboration and discussion with a multi-disciplinary approach is warranted to help optimise management for these patients.

References

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