Internuclear ophthalmoplegia as a presenting sign of Lyme disease: a case report

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Abstract

Aim: To describe an unusual case presentation of INO associated with possible Lyme disease.

Methods: This is a case report of a 30 year old man who presented to the emergency department complaining of right orbital pain, double vision and an inability to look to the left. He was seen by the ophthalmologist and neurologist and reported worsening symptoms of a frontal headache and difficulties in walking. He had a history of tick bites and a consequent rash 2 months earlier. There was no vomiting, limb weakness or fever and also no significant history of trauma.

Results: On examination visual acuity was 6/9 in the eye; pupils, fundus and discs were normal. Orthoptic assessment revealed a right/alternating exotropia with diplopia and right INO was confirmed on ocular movement testing. Vertical nystagmus was also noted on upgaze. Convergence was normal. He was diagnosed with a right internuclear ophthalmoplegia with intact convergence. CT scan and MRI were normal and so he was commenced on ceftriaxone 2 mg IV once daily for probable neuroborreliosis which after 3 days was switched to oral doxycycline 100 mg for 2 weeks. Tests for Lyme disease proved inconclusive. Four weeks later the patient was reassessed and his INO had resolved.

Conclusion: Diagnosis of Lyme disease should be considered for sudden onset internuclear ophthalmoplegia. Internuclear ophthalmoplegia (INO) as the first sign of neuroborreliosis is extremely rare. To our knowledge there is only one other documented case of an adult patient with an isolated INO.

Key words: Incomitant strabismus, Internuclear ophthalmoplegia, Nystagmus, Orbital pain, Sudden onset strabismus

Introduction

Internuclear ophthalmoplegia (INO) is a lesion of the medial longitudinal fasciculus resulting in a palsy of the medial rectus and a dissociated gaze-evoked nystagmus in the abducting eye. The commonest cause of unilateral INO in the younger patient is multiple sclerosis (MS).

Other causes such as tumours are rarely purely unilateral. The ophthalmoplegia is generally the presenting feature of the disease. However other causes must be considered.1 Lyme disease is a tick-borne illness caused by spirochete Borrelia burgdorferi, first described in 1975.2-3 The disease is characterized by a broad variety of symptoms ranging from neurological, dermatological, cardiac and rheumatological signs. Often this wide variety can lead to a late diagnosis of the disease. The neurological disorders found in Lyme disease are collectively referred to as neuroborreliosis, which occurs in two forms: the acute or early stage, and the chronic stage with symptoms persisting for more than 6 months. Diagnosis and treatment of the disease is of extreme importance as serious complications such as meningitis and encephalitis can result.

Clinical features suggestive of Lyme disease include a history of tick bite(s) and erythema migrans rash. The diagnosis is confirmed by the presence of specific antibodies in both serum and cerebrospinal fluid (CSF).3 The clinical course of Lyme disease has itself been divided into three stages. Stage one directly follows the infected tick bite which results in a rash. After several weeks or months, spirochetes spread in the body. At this stage neurologic symptoms may develop and the patient may see skin and cardiac manifestations: stage two. Stage three is reported to be similar to MS. However the extent to which patients may be affected can differ as well as the timeframe.4,5 Ocular manifestations are a rare feature. Follicular conjunctivitis has been reported in patients with early Lyme disease and iridocyclitis, pars planitis, vitritis, choroiditis, acute multifocal posterior placoid pigment-epithiopathy (AMPPPE), and retinal vasculitis have been reported within a few months of onset. Neuro-ophthalmic conditions including multiple cranial nerves, optic atrophy and disc edema have also been documented. Lyme disease is also one of the few conditions that may cause bilateral simultaneous facial paresis.4-9 To our knowledge there has been only one reported case of an adult patient presenting with an isolated internuclear ophthalmoplegia as the only clinical symptom of early neuroborreliosis.4

Case report

A 30 year old man presented to the emergency clinic on the 7 October 2012 complaining of right orbital pain, double vision and an inability to look to the left. He also experienced a mild frontal headache. Two months earlier
whilst exercising he had received a significant number of tick bites to both legs and subsequently developed a rash, which had now healed. He was also having problems walking and was noted to have ataxia. There were no other systemic complaints and he was referred to the on call ophthalmologist.

On examination by the ophthalmologist his visual acuity was recorded as 6/9 either eye but he struggled to read the letters, saying they were jumping. Fundus and disc examination was normal. He was unable to look to the left with his right eye and horizontal nystagmus of the left eye was noted on left gaze. Upbeat nystagmus was also seen on elevation. He was admitted and a CT scan, ECG and blood tests were carried out. The CT scan was returned as normal that night and an MRI was requested to be followed by CSF studies to Mo oligoclonal bands.

In view of the history of tick bites, rash and joint pain following the bites a diagnosis of probable neuroborreliosis was made and following discussion with the microbiologist he was commenced on IV ceftriaxone 2 mg OD. Orthoptic assessment was requested.

Orthoptic assessment 8 October 2012

The patient was aware that his right eye would not move to the left and had taken photos himself to show this (Fig. 1).

He had no previous history of any eye problems. On assessment visual acuity was −0.100 either eye with logMAR. Cover test revealed a small right/alternating exotropia with diplopia measuring 20³ base in (BI) for near fixing right eye (FRE) and 12³ BI for distance FRE. Convergence was recorded as to nose with nystagmus. Ocular motility testing revealed a moderate underaction of the right eye with horizontal nystagmus of the left eye on left gaze. Vertical nystagmus was seen on up gaze. Horizontal and vertical saccades were hypermetric. A diagnosis of right internuclear ophthalmoplegia with intact convergence was made. A Hess chart was done which demonstrates the ocular motility problem (Fig. 2).

The following day he was seen by the neurologist who indicated right INO that was possibly now resolving. MRI was normal and a lumbar puncture was requested for the following day. He was switched onto oral doxycycline.

Three days after admission the patient reported a significant improvement with less diplopia and he was able to look to the left. The patient was discharged on 11 October 2012 with a follow-up with the neurologist. Lumbar puncture results were returned as unreliable.

Orthoptic assessment 1 November 2012

The patient was seen for orthoptic follow up 1 month after initial presentation. He reported a big improvement and was discharged on 11 October 2012 with a follow-up with the neurologist. Lumbar puncture results were returned as unreliable.

Fig. 1. Photos taken by the patient showing attempted left gaze.

Fig. 2. Hess chart, 8 October 2012.

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the right eye on laevoversion with a small amount of horizontal nystagmus of the left eye. Slight overaction of the right eye with underaction of the left eye on laevo elevation was also seen (Figs. 3, 4).

Discussion
This case demonstrates the importance of considering the diagnosis of Lyme disease in patients with INO. In addition it illustrates the importance of early intervention. Although the diagnosis of Lyme disease is not confirmed the patient showed a prompt improvement in the INO when started on IV antibiotics. This is usually the case with most other cranial neuropathies that have been reported as rapidly improving once treatment was initiated.3,8

Serologic tests may confirm a clinical diagnosis of Lyme disease; however a negative result does not exclude the diagnosis.7 In this case it was reported by the microbiologist that the antibody response may take several weeks to develop, and may be abrogated by the prompt treatment he received. It has also been reported that according to the test used, Lyme borreliosis may be seronegative in up to 50% of cases. In that instance a cerebrospinal fluid examination is recommended.10

Conclusion
To our knowledge there has been only one reported case of an adult patient presenting with an isolated inter-nuclear ophthalmoplegia as the only clinical symptom of early neuroborreliosis.9 In this case the patient was of similar age but presented with a bilateral INO. Serum titres against Borrelia burgdorferi were negative; however diagnosis was confirmed by lumbar puncture.

In the case of our patient the CSF sample was returned as unreliable. The investigations and history would suggest the possibility that the INO was a result of Lyme disease. As the MRI scan of the brain was normal, with no inflammatory changes seen, it does leave his undoubted ophthalmoplegia otherwise unexplained.

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