LETTER TO THE EDITOR

Jerky seesaw nystagmus with internuclear ophthalmoplegia as the presenting finding in systemic lupus erythematosus

Le nystagmus à bascule avec ophtalmoplégie internuncléaire comme le symptôme révélateur d’un lupus érythémateux disséminé

Introduction

Internuclear ophthalmoplegia (INO) is characterized by adduction paresis of the ipsilesional eye and dissociated abducting nystagmus of the contralesional eye on attempted gaze to the contralesional side. It is caused by the lesions involving the medial longitudinal fasciculus (MLF) including arterial infarcts, demyelinating diseases, trauma and inflammation [1]. Oculomotor abnormalities such as up-beating of the ipsilesional eye, abnormal head position and rarely seesaw or hemi-seesaw nystagmus may be also associated with INO [2—4].

Seesaw nystagmus is a nystagmus subtype characterized by the combination of cyclic conjugated torsional eye movements and simultaneous vertical eye movements; as alternating elevation and intorsion of one eye and simultaneous depression and extorsion of the other eye. It has been reported in the literature with parasellar lesions, mesodiencephalic pathologies, brainstem infarcts or haemorrhages, severe head trauma, multiple sclerosis (MS), cranial radiotherapy, intrathecal methotrexate and congenital achiasma [5].

INO has been reported rarely in association with systemic lupus erythematosus (SLE). However, there is no case presented with INO and seesaw nystagmus as the first manifestation of SLE, to the best of our knowledge. Herein, we report a case with INO and seesaw nystagmus as the presenting finding of SLE.

Case presentation

A 45-years-old female who presented with sudden-onset dizziness, ataxia and diplopia to the emergency department was referred to our department. There was no history of ocular or systemic disorders. Her best-corrected visual acuity was 10/10 in both eyes. Examination of the anterior segment and fundus was unremarkable. Ocular motility examination revealed the right INO and jerky seesaw nystagmus (Fig. 1): adduction paralysis of the right eye with associated abducting nystagmus in the left eye and jerk wave-form torsional-vertical nystagmus in both eyes with the following characteristics:

• the torsional component was conjugate. The fast phases were anticlockwise;
• the vertical component was disjunctive. The fast phases were up-beating in the intorting right eye, down-beating in the extorting left eye;
• the nystagmus was present in all gaze directions and unaffected by changes in head position or in vergence angle. Convergence was preserved.

An MRI scan of brain showed hyperintense lesions in a linear pattern and perpendicular to corpus callosum in accordance with the course of vascular structures located in periventricular area, in T1-weighted sagittal images (Fig. 2). The lesions did not show enhancement. Besides, a tiny hyperintense lesion that could explain the present clinical picture was found at the pontine level in axial thin-section T2-weighted MRI image (Fig. 3). With suspect of demyelinating disease or vasculitis, treatment with intravenous pulsed methylprednisolone was started immediately. Laboratory test results showed a white blood cell count of 5400/μL, a platelet count of 274,400/μL, hemoglobin of 12.8 g/dL, C-reactive protein of 6.2 mg/L (normal < 5), an erythrocyte sedimentation rate of 70 mm/h (normal < 20), anti-ds-DNA of 50 U/mL (normal < 25), MPO ANCA: 19 U/mL (normal < 5), PR3 ANCA: 20 U/mL (normal < 5) and positive anti-Smith antibodies. Cardiolipin levels were within normal limits. In protein electrophoresis, it was seen that alpha-1, alpha-2 and beta bands were increased and albumin band was decreased. The results of lumbar puncture (LP) were within normal limits and oligoclonal band was determined to be negative.

In consequence of LP and the other laboratory investigations (especially positive anti-Smith antibodies), the patient was diagnosed with SLE presented with CNS involvement as first attack. Intravenous pulsed methylprednisolone was...
Ocular motility examination revealed the right internuclear ophthalmoplegia and jerky seesaw nystagmus: adduction paralysis of the right eye with associated abducting nystagmus in the left eye (a, c) and jerk wave-form torsional-vertical nystagmus in both eyes (b).

given for 5 days at a dose of 1 g/day. The complaints and the eye symptoms started to improve on 2nd day of treatment and returned to normal on the 10th day of admission. After the pulse steroid therapy, methylprednisolone (16 mg), azathiopurine (100 mg), acetyl salicylic acid (100 mg) and colchicum dispert (0.5 mg t.i.d) with daily doses were started by rheumatology department. The patient has been followed up without attacks for 1 year.

Discussion

Internuclear ophthalmoplegia (INO) is a complex oculomotor disorder occurring due to MLF lesion. Seesaw nystagmus is rarely associated with INO and it is considered that concurrent involvement of the descending fibers of contralateral semicircular canals together with MLF injury causes this condition [2]. Although infarctions are accused commonly as etiological factors, vasculitis may also cause INO. In a series including 410 cases with INO, the followings were reported as etiological factors: infarction in 157 patients (38%), multiple sclerosis in 139 patients (34%), the other causes in 114 patients (28%) and vasculitis in only 7 patients [6]. The most common vasculitis causing INO were SLE, Sjögren’s syndrome and temporal arteritis. In the literature, INO as an unusual presentation of SLE was reported by Abel et al. [7]. However,
there is no case presented with INO and seesaw nystagmus as the first manifestation of SLE to the best of our knowledge.

Dehaene et al. [8] reported a case with INO and extorsional nystagmus in the right eye and intorsional-upbeat nystagmus in the left eye due to a small ponto-mesencephalic infarction similar in our case. They suggested that this could be due to the selective damage of the fibers from the contralateral anterior semicircular canal. Choi et al. [3] reported a case with INO and hemi-seesaw nystagmus developing due to a small infarction in the right dorsomedial pontine tegumentum. The authors suggested for this condition that MLF lesion could cause ipsilateral inactivation of the interstitial nucleus of Cajal with sparing the rostral interstitial nucleus of MLF and simultaneous involvement of the pathways from contralateral antero-posterior semicircular canals results in various patterns of dissociated torsional-vertical nystagmus. Our case shows similar clinical and radiological findings of the cases presented by Choi and Dehaene, and we consider that this complex condition occurs due to the involvement of the fibers from contralateral semicircular anterior-posterior canals together with MLF injury.

On the other hand, even though the patient was diagnosed with SLE in our case, it must be considered that SLE can be easily misdiagnosed with MS or both diseases may occur in the same individual [9–11]. Fanouriakis et al. [9] reported nine patients who fulfilled the diagnostic criteria for both SLE and MS. They added that the diagnosis of SLE preceded the development of MS in five patients, with a time lag ≤ 5 years in four of them [9]. Although challenging, some diagnostic tests may help to differentiate between these entities but some points still remain unclear [10]. Consequently, the possible coexistence of these entities should always be kept in mind.

In conclusion, in this article, we aimed to present a SLE patient with neurological involvement presenting with isolated right INO and jerky seesaw nystagmus, and to explain the possible pathophysiological mechanisms of this clinical picture by reviewing previous literature. Although they are rare, vasculitic diseases should always be kept in mind in case of complex ocular motility disorders.

Disclosure of interest

The authors declare that they have no conflict of interest concerning this article.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.jfo.2014.04.016.

References


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