Bilateral third nerve palsy with pupil involvement as the first manifestation of multiple sclerosis

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Isolated ocular cranial nerve palsy is a rare presentation of multiple sclerosis (MS). In one study of 1489 patients with MS, 24 had isolated nerve palsies in the course of the disease (1.6%) and only 14 presented with isolated cranial nerve palsies as the first sign of the disease; among these, only one had a third nerve palsy [1]. Isolated cranial nerve palsies were observed in 35 patients out of 483 patients with a CIS/MS series, and only 2 had a third cranial nerve affection [2].

A 34-year-old woman presented with progressive binocular double vision, right upper eyelid ptosis and headache for two weeks. On neurological examination she had bilateral third nerve palsy, with pupil involvement on the right side; the remaining neurological examination was normal. A brain MRI scan showed multiple focal white matter high signal intensity foci on T_2-weighted sequences consistent with demyelinating lesions; one of these lesions involved the midline of the midbrain tegmentum, extending anteriorly to the interpeduncular fossa and posteriorly in front of the aqueduct (Figure). This lesion, which showed contrast-uptake after intravenous injection of gadolinium, was likely responsible of the clinical symptoms, as involved the theoretical location of both oculomotor nerve nucleus. The MRI lesions fulfilled criteria for dissemination in space according to the Barkhof criteria. The analytical work-up, including infectious serologies, thyroid function and immunologic studies, was normal. Oligoclonal bands were present in CSF but not in serum. Visual evoked potentials were normal. Other reasons for a bilateral third nerve lesion were reasonably discarded. The diagnosis of brainstem clinically isolated syndrome (CIS) was made, and the patient received treatment with intravenous methilprednisolone pulses. There was a satisfactory recovery of the third nerve deficits. Two months later she developed a mild sensory myelitis, confirming clinically the diagnosis of multiple sclerosis (MS). Disease modifying treatment was proposed and the patient started treatment with interferon β-1a.

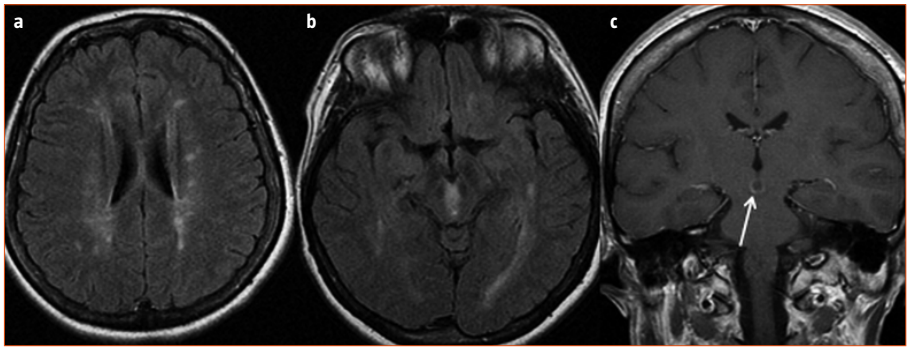

Figure. Brain MR imaging: a, b) Transverse FLAIR T_2-weighted images showing multiple focal high intensity lesions involving the hemispheric white matter and the midline of the midbrain tegmentum; c) Observe the ring-enhancement of the midbrain lesion on the coronal contrast-enhanced T_1-weighted image (arrow).

To the best of our knowledge, only seven other patients in whom the initial sign of multiple sclerosis was an isolated third nerve palsy, have been reported in English-language literature [3-9]. Clinical presentation varied in terms of associated eye pain or headache, presence of ptosis, degree of ophtalmoparesis, pupil involvement and unilaterality/bilaterality. Apart from the case presented here, only another one of bilateral third nerve palsy was previously described [10].

Isolated third nerve palsy is a rare presentation of MS; yet, this diagnosis must be considered, especially in young patients, but remains an exclusionary diagnosis. MR imaging demonstration of lesions of the kind of seen in MS, including the anatomical location of the oculomotor nerve nucleus in the midbrain, would support this clinical diagnosis.

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