Arq Neuropsiquiatr 2008;66(2-A):254-255

ISOLATED OCULOMOTOR NERVE PARESIS AS THE PRESENTING SIGN OF MULTIPLE SCLEROSIS

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In multiple sclerosis (MS), ocular motor disturbances such as nystagmus or internuclear ophthalmoplegia are frequent and their pathophysiological processes are relatively well known. On the contrary, other rare and not so well studied manifestations such as isolated ocular motor nerve palsy may be observed and can represent a diagnostic challenge for the clinician as in the case we report.

CASE

A 35-year-old woman was admitted in the emergency department complaining of double vision on right gaze of sudden onset lasting for three days. Additionally from the beginning of clinical onset she referred constant right periocular pain not related with ocular movements, which spontaneously reverted within 24 hours. She had no recent infectious or traumatic events and her personal and familiar medical history was otherwise unremarkable. On neurological examination showed a left isolated incomplete III cranial nerve (CN) palsy characterized by eyelid ptosis, limitation on adduction and supraversion of left eye, paresis of the four extraocular muscles confirmed by Hess screen and no pupillary dysfunction, either on light or accommodation. The remaining neurological examination, namely visual acuity, fundoscopy, other cranial nerves and long tracts systems were normal. Also, orbital murmurs, proptosis, quemosis or conjunctival hyperemia were not found. She was apyretic and normotensive. All the analytic work-up was within normal limits. It included hemogram, C-reactive protein, sedimentation rate, glycemia, lipid profile, thyroid function, infectious serologies (lyme disease, syphilis), immunologic study (antinuclear antibodies, anticardiolipin antibodies, anti-neutrophilic cytoplasmic antibodies, anti-double stranded antibodies, angiotensin-converting enzyme) and CSF cytochemistry studies. The MRI and the conventional angiography performed at 24 hours and at the fifth day after admission respectively were normal. A complete and spontaneous reversion of the III CN palsy occurred within three weeks.

Regarding the clinical follow-up, one year and half afterwards she presented another deficit episode, now consisting of left eye blurred vision which had partially recovered without medication. Further, one month later she had numbness of the left lower limb. At that time, examination revealed low visual acuity in the left eye (1/10) with normal fundoscopy, bilateral pyramidal syndrome without motor deficits and a left D10 hyposthesic level. The MRI showed multiple small T2 and Flair hyperintense lesions located in the subcortical white matter on the high frontal convexity, interhemispheric cortex and left middle cerebellar peduncle, without disruption of the bloodbrain barrier (Fig 1). Left optic nerve was found to be thick although did not show signal abnormalities or gadolinium enhancement. The spinal cord MRI disclosed multiple intraspinal cord lesions in the transitions of C1-C2, C5-C6 and D9-D10 showing T2 hyperintensity and gadolinium enhancement (Fig 2). Oligoclonal bands were present in the CSF although not in the serum.

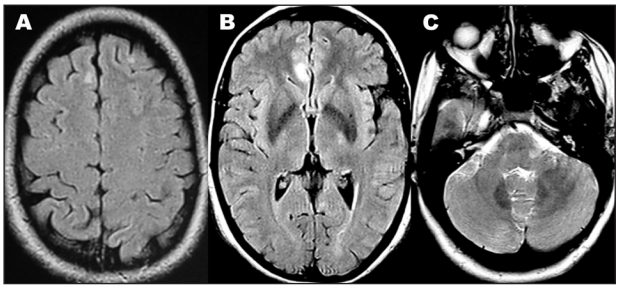

Fig 1. Brain MRI: (A) and (B), axial fluid-attenuated inversion recovery (FLAIR), show right subcortical frontal (A) and anterior ipsilateral periventricular (B) hyperintense lesions. (C) axial T2-turbo spin-echo (T2-TSE), reveal an hyperintense lesion localized in the left midle cerebellar peduncle.

Thus, the patient had clinical criteria of time and space dissemination, corresponding to the optic nevritis and sensitive attack, even not considering the cranial nerve episode. The diagnosis of MS was additionally corroborated by the presence of Barkhof criteria for space dissemination[1] and the finding of oligoclonal bands only in the CSF. Thus, she was provided beta-1b interferon and untill the present time had no other attack (12 months of follow-up) although keeps a sequelar visual deficit (EDSS=2).

DISCUSSION

We describe a case of a 35-year-old woman with an unremarkable medical history who presented an iso-

PARESIA ISOLADA DO NERVO OCULOMOTOR COMUM: UMA FORMA DE APRESENTAÇÃO INVULGAR DE ESCLEROSE MÚLTIPLA

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Received 26 September 2007, received in final form 13 December 2007. Accepted 14 February 2008.

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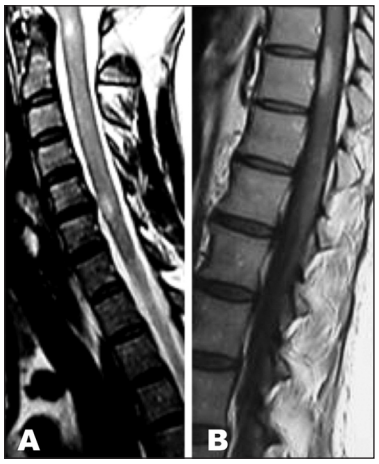

Fig 2. Spinal cord MRI: (A) sagittal T2-TSE show hypersignal lesions in C1-C2 and C5-C6. (B) sagittal T1 after gadolinium, displays contrast enhancement in the lesion localized in D9-D10.

lated external III CN ophthalmoparesis associated with ipsilateral periocular pain. Regarding the localization of the presumed lesion, semiology suggested a fascicular or peripheral involvement, however neuroimage was not informative. In fact, features suggesting a nuclear palsy as bilateral eyelid ptosis or contralateral right rectus palsy[2] were not found. All the etiologic investigation performed at that time was normal. So, we could exclude or at least consider as highly improbable the diagnosis of aneurysm, diabetic or vasculitic stroke of the nerve, mesencephalic stroke, tumor and infectious (borrelia, syphilis, tuberculosis) or sarcoidosis meningitis. Also, she had no history suggestive of ophthalmoplegic migraine or trauma. Some authors have underlined that minor head trauma can cause an isolated III CN palsy in the absence of abnormal findings on brain MRI[3] which was not the case of our patient. MS criteria[4] was developed later, so retrospectively the III CN nerve palsy was interpreted as a clinically isolated syndrome (CIS). Isolated CN palsy is a rare finding in MS, occurring in 1.6 % of patients and involving in decreasing order of frequency the CN VI, VIII, VII, III and IV[5]. This hierarchy of frequency is due to the anatomic variability seen among the different CN. Namely, the length of the intra-axial traject (less length in CN III, IV and VIII), the density of the fascicular nerves (CN III and VIII have diffuse fascicules)[5] and the amount of myelin exposed in the fascicular traject (less in CN IV)[6]. The largest published series demonstrated that the isolated CN palsy was always associated with brainstem involvement, either as a lesion seen on MRI (1.5 T) or as a dysfunction disclosed in electrophisiologic tests[5].

This study and others[7] proved that brainstem auditory evoked potentials and blink reflex abnormalities are highly sensitive on detection of brainstem demyelinating lesions some of which are not amenable to MRI detection. So, in the light of this, we suggest that the most probable localization involved in the ophthalmoparesia of the patient currently reported is fascicular mesencephalic. This cannot however be confirmed as long as neurophisiologic tests were not performed by lack of any clinical indication at that time. The MRI performed with a 1.5 T field could justify the underdetection of a small brainstem lesion inasmuch as studies demonstrated that high field MRI such as 3 T provides a significantly higher detection rate of inflammatory brain lesions[8]. In addition, literature reports only one MS patient with isolated III CN palsy caused by peripheral nervous system involvement who differently from our patient had pupilar involvement and enhancement of the cisternal portion of the CN III on MRI[9]. To the best of our knowledge only two other patients were reported in English-language literature showing an isolated CN III paresis as the presenting sign of MS[10,11]. As both observations were performed more than 15 years ago, we aim with this case report to provide an updated insight on this rare manifestation of MS.

In conclusion, a CIS indicating a pathological process involving the CN may constitute a hard diagnostic challenge for the clinician. This hypothesis should be considered in a young woman presenting with an isolated III CN palsy even with a normal MRI if all other other possible diagnosis were excluded. From an accurate identification of the first MS attack depends a timely initiation of the disease-modifying treatment.

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