COMPLEX HYPERKINETIC MOVEMENT DISORDERS

An unusual presentation in multiple sclerosis

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Movement disorders (MD) in patients with multiple sclerosis (MS) has been few reported. The most common MD associated with MS are tremor and dystonia, but parkinsonism, myoclonus, chorea, ballism, paroxysmal dyskinesias and hemifacial spasm have been more rarely found in MS patients[1,2]. Complex hyperkinetic movement disorders (CHMD) is the term used to describe patients who present concomitantly more than one type of MD, although there is not always consensus as to the type of motor symptomatology[3].

We report on the first case of CHMD in a Brazilian patient with MS.

CASE

A 21-year old woman presented with acute left facial paresis and weakness in right upper limb. At age 27, she developed a sudden episode of visual loss, dysphagia, dysarthria, dizziness, incoordenation, loss of balance, weakness in left lower limb and gait difficulty. The diagnosis of MS relapse was made (EDSS score was 5.0), brain magnetic resonance imaging (MRI) demonstrated demyelinating abnormalities which fulfill the criteria for MS, and after treatment with intravenous methylprednisolone the symptoms partially remitted.

However, during the following two years she presented progressive visual loss, weakness and postural instability associated to incapacitating involuntary movements, which restricted her to wheelchair (EDSS score was 8.0), despite treatment with interferon beta-1a (44 μg tiw). Neurological examination showed nystagmus; anarthria; facial paresis; muscle strength 4 in upper limbs and 3 in lower limbs (MRC score); arms and legs spasticity; brisk reflexes in the right leg; bilateral Babinski's sign; and CHMD characterized by no rhythmic tremor at rest (frequency of 3–4 Hz) which intensified during action in upper limbs (predominantly left), lower limbs (particularly in left) and jaw, cervical dystonia (spasmodic torticollis), ballistic movements in upper limbs, and left hemifacial spasms.

The clinical features were thought to be consistent with secondary progressive form of MS and treatment with mitoxantrone was started (total dose of 80 mg for 9 months) which no reduce her disease progression (EDSS score was 9.5 after one year).

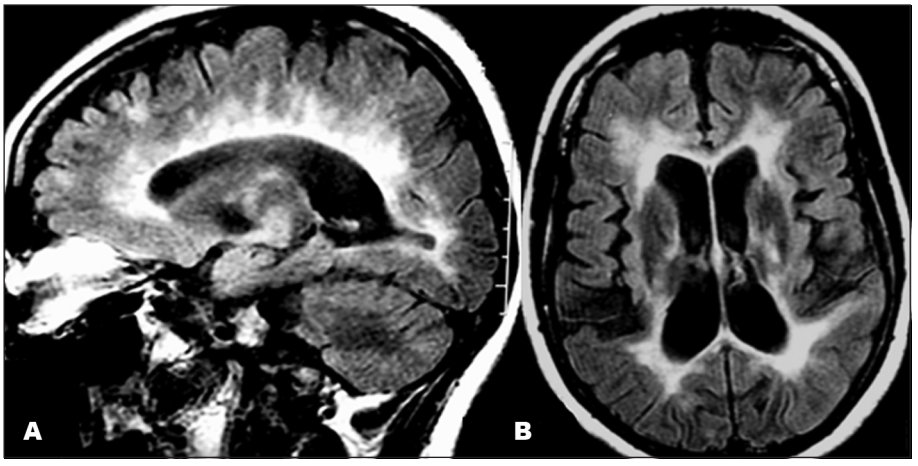

Figure. Brain MRI in sagittal [A] and axial [B] FLAIR sequence.

TRANSTORNO DE MOVIMENTO HIPERCINÉTICO COMPLEXO: UMA APRESENTAÇÃO INCOMUM EM ESCLEROSE MÚLTIPLA

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CHMD was managed with levodopa (125 mg tid) that was later followed by use of clonazepam (6 mg/day) and biperiden (12 mg/day), but no significant improvement occurred.

Follow-up brain MRI demonstrated brain atrophy, basal ganglia alterations, multiple high-signal-intensity areas on FLAIR and low-signal-intensity areas on T1-weighted images in the periventricular and subcortical white matter of the brain hemispheres as well as in the brainstem and thalamus, and post-gadolinium sequences did not show any contrast enhancement (Figure). In addition, diffusion tensor MRI showed a marked reduction in fractional anisotropy values in white matter lesions.

All studies were done following informed consent.

DISCUSSION

In our patient, CHMD was characterized by tremor, dystonia, ballism and hemifacial spasm, but this concomitant MD presentation is extremely unusual by MS patient.

Several types of tremor can be observed in MS patients, such as cerebellar tremor, which has a dominant intentional component, action (postural or intention), rest and Holmes' tremor[1,4]. Tremor can occur in 25.5% to 58% of MS patients, usually as action tremor, which can be disabling only in 3% to 15% of patients[4-6]. In MS, true rest tremor can be observed in 1% of patients and Homes' tremor was described only in case reports[4,7]. Our patient presented with Holmes' tremor, which was characterized by the presence of no rhythmic tremor at rest with a low frequency (< 4.5 Hz) which intensified during movement[1,7]. MS lesions in brainstem could cause damage in cerebellothalamic and nigrostriatal fiber tracts explaining the Holmes' tremor in our patient[7].

MS causing dystonia may probably be under recognized etiology because is a rare manifestation of MS which can be observed only in 11% of cases[2,4]. Spasmodic torticollis (cervical) is the most common dystonia in these patients, but other focal dystonia also have been found[2,4]. MS involvement of the midbrain or spinal cord has been suggested in the dystonia pathogenesis[2,4].

The involvement of basal ganglia (mainly in the thalamic or subthalamic region) and their connections fibers, by demyelinating plaques, have been found causing ballism in MS patients[8-14]. MS with thalamic and subthalamic demyelinating lesion may be the cause of ballism in 5.8%[13]. Ballism can occur as hemiballism, monoballism or biballism, but MS still is a very rare etiology for ballism[8-17].

Hemifacial spasm is considered an extremely rare MD manifestation in patients with MS. Hemifacial spasm is possible due to lesions of MS in the brainstem, but only few case reports have described this MD in MS patients[18].

The concomitance of ballism and tremor was previously described, as rare MS presentation, but others MD association which could result in CHMD in the secondary progressive MS patient, similar our case, has yet not been reported[15,16]. Our clinical observations with this patient suggest: MS plaques may produce CHMD and MS should be included in the differential diagnosis of patients presenting CHMD.

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