CASO CLÍNICO/CASE REPORT

Hemifacial Myokymia as the Presenting Feature of Multiple Sclerosis

Mioquimias Periorais como Forma de Apresentação de Esclerose Múltipla

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Abstract

The clinical presentation of multiple sclerosis (MS) is variable and can pose a diagnostic challenge. A case of unilateral perioral myokymia as the revealing feature of MS is reported. We present the case of a 43-year-old male, observed in our outpatient clinic with a two-week history of continuous involuntary wavelike movement across the left side of his face. Neurological examination revealed left-sided facial myokymia with perioral involvement and hemifacial spasm. Brain magnetic resonance imaging (MRI) showed multiple white matter hyperintensities on T2/FLAIR, suggestive of demyelination, involving periventricular, juxta-cortical regions, and the corpus callosum.

Cerebrospinal fluid (CSF) analysis showed CSF-specific oligoclonal bands. The diagnosis of relapsing-remitting multiple sclerosis was made according to the McDonald Criteria from 2017. This case illustrates an unusual presentation of MS and highlights the importance of awareness of possible MS in young adults with facial myokymia, more so when continuous and with perioral involvement.

Resumo

A apresentação da esclerose múltipla (EM) é heterogénea e é um desafio de diagnosticar. Reporta-se um caso clínico de mioquimias periorais unilaterais como forma de apresentação de EM. Apresenta-se o caso de um homem, 43 anos, observado em consulta de neurologia por um quadro com duas semanas de movimentos ondulantes, contínuos da hemiface esquerda. No exame neurológico destacava-se a presença de mioquimias, continuas, da região perioral esquerda e hemiespasmo facial. Ressonância magnética crânio-encefálica (RM-CE) revelou múltiplas lesões hiperintensas na ponderação T2/FLAIR, da substância branca, localizadas à região periventricular, justa-cortical e corpo caloso, sugestivas de doença desmielinizante.

Punção lombar com presença de bandas oligoclonais tipo 2. Pelos critérios de McDonald (2017), admitiu-se o diagnóstico de EM surto-remissão. O presente caso ilustra uma apresentação rara de EM e realça a importância de investigação adicional em doentes jovens com mioquimias faciais, particularmente contínuas e localizadas à região perioral.

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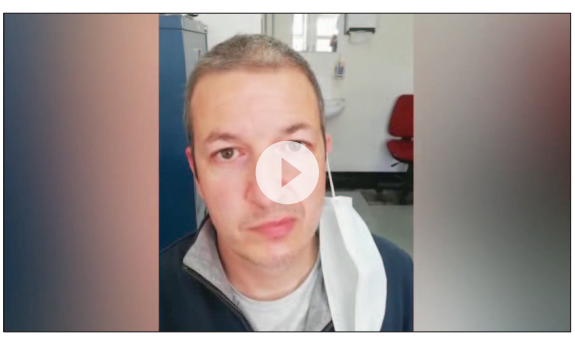
Introduction

The clinical presentation of multiple sclerosis (MS) is variable and can pose a diagnostic challenge. Hemifacial myokymia and hemifacial spasm have rarely been reported as the presenting features of MS and may easily be missed.[1-3] Here, we report a rare case of a male with continuous hemifacial myokymia as the presenting feature of MS.

Case Report

A 43-year-old man with a personal history of major depressive disorder under escitalopram 10 mg for 10 years presented to our outpatient clinic with a two-week history of involuntary wavelike movement across the left side of his face. The general examination was normal, and the neurologic examination revealed continuous, spontaneous left-sided facial myokymia, especially around the mouth, and a left-sided hemifacial spasm (**see supplemental video**). Right voluntary facial movements were preserved. He also had generalized hyperreflexia and imbalance performing tandem gait. Laboratory analyses were unremarkable. Cerebrospinal fluid (CSF) analysis showed CSF-specific oligoclonal bands. Brain magnetic resonance imaging (MRI) described multiple white matter hyperintensities on T2/FLAIR, suggestive of demyelination, involving periventricular, juxtacortical regions and the corpus callosum (**Fig. 1**) Unfortunately, gadolinium was not administered. The spinal cord MRI was normal. The diagnosis of relapsing-remitting multiple sclerosis was made according to the McDonald Criteria from 2017. The patient was treated with methylprednisolone 1 g a day for 3 days and recovered completely after 1 month. He also started peginterferon beta-1a. A follow-up brain MRI was performed three months after the initial study and revealed a new lesion in the dorsolateral portion of the left pons (**Fig. 2**), which was hyperintense on T2/FLAIR and had no enhancement after gadolinium administration.



Supplemental video. Neurological examination revealed continuous, spontaneous left-sided facial myokymia, especially around the mouth, and a left-sided hemifacial spasm.

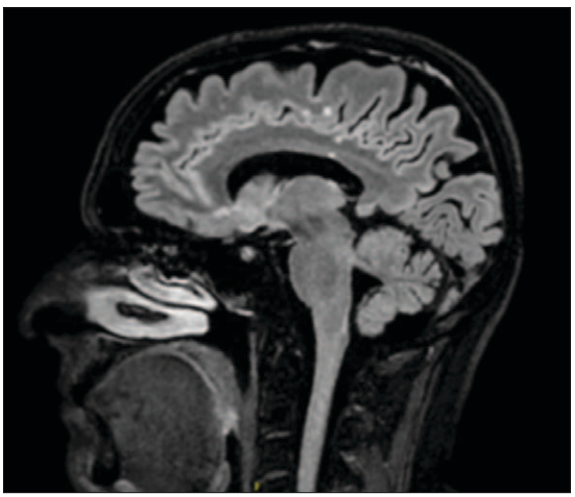


Figure 1. Brain MRI: sagittal T2 FLAIR FS images showing multiple white matter hyperintensities on T2/FLAIR, suggestive of demyelination, involving juxtacortical regions and the corpus callosum.

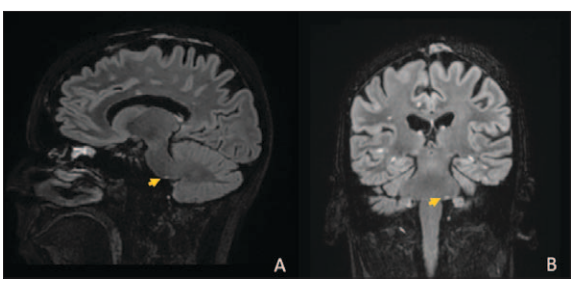


Figure 2. Follow-up Brain MRI: sagittal (A) and coronal (B) T2 FLAIR FS images showing a hypersignal in the dorsolateral aspects of the left pons (yellow arrows).

Discussion

We report a rare case of MS manifested by hemifacial myokymia and hemifacial spasm. Facial myokymia is defined as an involuntary movement disorder of the musculature supplied by the facial nerve and characterized by spontaneous undulating, vermicular movements that spread throughout the face.[3] The prevalence of facial myokymia in MS is uncertain, however in a recently published article that evaluated 2260 patients with MS, 1.28% had facial myokymia in the course of the disease.[4] Although eyelid myokymia is recognized as a common and usually benign symptom, associated with stress, caffeine intake, fatigue, or exercise, strict unilateral continuous myokymia, especially with perioral involvement, as in our patient, suggests the presence of a structural pontine lesion, warning the need for brain imaging.[5] Unilateral myokymia may be associated with brainstem lesions,

namely tumors, cysticercosis, Guillain-Barre syndrome, or MS.[6] Myokymia may be the presenting feature of MS,[2,3,7-10] as in our case, or may correspond to relapse in a patient with an established multiple sclerosis diagnosis undergoing immunomodulatory or immunosuppressive treatment.[11,12] Also, it is well known that facial myokymia seems to be related to dorsolateral tegmentum pontine lesions concerning the ipsilateral post-nuclear portion of the facial nerve, particularly lateral to its genu.[2] In our case, the initial brain MRI failed to identify the pontine lesion, later described in the follow-up MRI. We hypothesized that different acquisition MRI techniques with different slices thickness and different sensitivity accrual could explain the absence of the tiny brainstem lesion on the first brain MRI scan. Moreover, the "clinic-radiological paradox" in MS is a well-known entity, that has been described as a mismatch between clinical assessment and visible lesions on MRI.[13]

The clinical outcome of patients with facial myokymia and hemifacial spasm associated with multiple sclerosis is uncertain. However, facial myokymia is usually self-limited and most patients recover in less than 12 months. Also, myokymia seems to respond well to the relapse treatment with corticosteroids or symptomatic treatment with gabapentin, carbamazepine, and botulinum toxin.[1,14,15] Nevertheless, the hemifacial spasm may persist for longer periods and require multiple treatments, such as botulinum toxin.[1,14]

In summary, this case illustrates an unusual presentation of multiple sclerosis and highlights the importance of awareness of possible multiple sclerosis in young adults with persistent facial myokymia, especially those with perioral involvement. ■

Contributorship Statement / Declaração de Contribuição

MM: First draft of the manuscript, conception and final approval.

FS;IF;MVB: Critical review and final approval.

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