LETTERS TO THE EDITOR

Unilateral eyelid myokymia as a form of presentation of multiple sclerosis\textsuperscript{*},\textsuperscript{**}

Mioquimias palpebrales unilaterales como forma de presentación de una esclerosis múltiple

Dear Editor:

Myokymias are localised involuntary contractions that are wavelike or vermicular and propagate through affected striated muscle.\textsuperscript{1–3} They are caused by simultaneous or sequential activations of 1, 2, or more motor units of a muscle. An electromyography reading shows spontaneous muscle activity with different motor units producing brief, repetitive discharges of action potentials in rhythmic or semi-rhythmic bursts. This is followed by a short (0.5 to 3 seconds) and frequently irregular interval of electric silence before the following myokymic discharge occurs.\textsuperscript{4,5}

When myokymia occurs in the face, the most frequently affected muscle is the orbicularis oculi. Myokymia in this muscle results in small, visible contractions of part of the eyelid, typically the lower eyelid. Eyelid myokymias (EM) tend to be transient and self-limiting within a few days of onset. They present in young, healthy subjects with no associated diseases.\textsuperscript{3,6,7} We describe a patient in whom the presence of continuous EM resulted in a diagnosis of multiple sclerosis (MS).

The patient was a 34-year-old woman with no relevant medical history or drug use. She was examined for abnormal movements compatible with myokymias in the right lower eyelid which had been occurring for 2 months. The symptoms initially presented sporadically during the day in episodes lasting from a few minutes to an hour. As the weeks passed, their frequency and intensity had increased and muscle twitches had become continuous. The first neurological examination revealed generalised hyperreflexia; all other findings were normal.

Given the intensity of the myokymias, the patient began treatment with carbamazepine (800 mg/day), but the drug had to be discontinued quickly due to the appearance of a maculopapular rash and elevated transaminases.

The symptoms resolved spontaneously 9 weeks after their onset. A week after EMs had become continuous, doctors performed a brain MRI which showed multiple lesions in supratentorial white matter in both hemispheres, semi-oval centres, and the juxtaventricular zone, plus a lesion in the posterior right frontal lobe (Fig. 1). A complete blood study including serology and autoimmune markers yielded normal results. CSF analysis indicated the presence of oligoclonal bands, although they were not found in serum. A second cranial MRI taken at 4 months revealed new lesions, including several with gadolinium uptake (Fig. 2). Based on these findings, the patient was diagnosed with MS.

Facial myokymias that occur continuously and affect all the muscles on one side of the face have often been described in association with different types of lesions of the ipsilateral pontine tegmentum,\textsuperscript{1–6,9} especially tumours (gliomas or metastasis),\textsuperscript{1,5} cysticercosis,\textsuperscript{6} and MS. In MS, myokymias may occur either throughout the duration of the disease\textsuperscript{2,6,9,10} or appear as its first symptom.\textsuperscript{6} Other entities with which EM is less frequently associated are subarachnoid haemorrhage,\textsuperscript{11} multiple system atrophy, and Guillain-Barré syndrome.\textsuperscript{2,3,5} In MS, facial myokymias tend to be self-limiting in the course of a few weeks (typically between 2 weeks and 6 months)\textsuperscript{2,5,10} and they rarely last as long as a year.\textsuperscript{9} However, myokymias secondary to malignant tumours typically last several years and resolve with treatment of the neoplasia.\textsuperscript{1,5,10}

Unlike facial myokymias, those limited to the eyelid only tend to appear in healthy young subjects. They are associated with stress, fatigue, exercise, and excessive caffeine use.\textsuperscript{3,7,12} On rare occasions, they may be associated with underlying disease or extend to other facial muscles. In a typical presentation, they are unilateral and transient, appearing in episodes lasting a few minutes at a time over the course of many days or weeks. It is very uncommon for EMs to appear continuously during periods spanning months, and they would still be considered benign even in such cases.\textsuperscript{3,7} However, available evidence is scarce. There is only 1 study evaluating persistent long-term EM,\textsuperscript{3} and this study found that only 1 of its study population of 15 developed clinically defined MS. It concluded that associations between long-term EM and other neurological diseases are uncommon. As we were unable to identify any of the trigger factors described for EM in our patient, and the condition


\textsuperscript{**} Part of this study was presented in poster format at the 15th annual meeting of the Catalan Society of Neurology.
persisted, we performed 2 cranial MRI scans and a lumbar puncture which resulted in a diagnosis of MS.

Cranial MRI study of our patient was unable to show any lesions specific to the ipsilateral pons. Some cases of hemifacial myokymias are described in which cranial MRI scans did not show typical lesions in the pons.\(^1,4,9\) One possible reason is that the lesion causing facial myokymia in MS tends to resolve when clinical symptoms have stopped appearing.\(^9\) This was observed in a study that used cranial MRI to re-examine patients in whom typical lesions had previously been detected and whose symptoms had resolved. In 6 of the 8 patients, the lesions had disappeared within periods ranging from 1 week to 44 months.\(^7\) A cranial MRI was performed on our patient a week after her referral to our unit. Another possible mechanism could be hyperexcitability of the intraaxial part of facial nerve fibres due to possible functional deafferentation of inhibitory pathways of the facial nerve nucleus caused by demyelinating lesions on the supranuclear level.\(^3,9,13\) As a result, myokymias would not necessarily have to be caused by lesions in the pons.

In conclusion, we feel that while EM is a benign entity, it may be indicative of an underlying neurological disease, such as MS, if it persists over an extended period of time. We recommend conservative treatment for these patients at first, and elimination of known triggers such as caffeine, tobacco, and alcohol. If the EM continues, imaging studies should be completed in order to rule out any underlying lesions.

References


Usefulness of procalcitonin and C-reactive protein in acute meningitis in the emergency department

Utilidad de la procalcitonina y la proteína C reactiva en las meningitis agudas en urgencias

Dear Editor:

It was with great interest that we read the article by Jiménez Caballero et al. relating a descriptive analysis of cases of viral meningitis and their different characteristics in children and adults. We have been studying all diagnosed cases of central nervous system infections (CNSI) cared for by our emergency department (ED) for many years. In these cases, immediate and appropriate action has a crucial effect on the microbiological diagnosis, prognosis, and morbidity and mortality. However, one of the most important decisions made by an ED providing care to patients with acute meningitis (AM) is determining whether the meningitis is bacterial (BM) or viral (VM). This information allows professionals to administer the proper antimicrobial drug as soon as it is needed.3,4 We believe that cooperation between the ED doctor, the on-call neurologist, and the microbiology department is essential (quality of care is the result of teamwork with each member of the team performing properly). In the ED, doctors should immediately consider the possibility of AM and discuss possible diagnoses (in priority order) with the neurologist and microbiologist when necessary. Among other reasons, this is because the amount of cerebrospinal fluid (CSF) is limited.4 After lumbar puncture has been performed, and depending on the CSF’s cytochemical profile, (pleocytosis, spinal fluid protein concentration, spinal fluid glucose concentration), lactate levels, and opening pressure, doctors should then order a microscopic examination, Gram staining, and bacterial antigen test (pneumococcal, meningococcal, etc.) while awaiting results from the CSF culture and any necessary blood cultures. In addition, as stated by Laguna del Estal,5 polymerase chain reaction (PCR) may prove to be a relatively rapid (results in 24 hours) and very useful tool for identifying isolated cases or outbreaks in the ED. This technique is being incorporated in clinical practice in EDs.6 Nevertheless, in cases with an unclear CSF profile, prior use of antibiotics (partially-treated AM), no results from the Gram stain, or symptoms of sepsis (S), severe sepsis (SS), or septic shock (SSH), procalcitonin (PCT) and C-reactive protein (CRP) may be useful biomarkers in the ED. This test, which requires a small blood sample and can be processed in 20 minutes, is helpful in distinguishing BM from VM and in deciding which blood cultures are needed, how best to administer early treatment with targeted antimicrobial drugs, and which department should admit the patient.5,7 We believe that biomarkers help predict whether or not the systemic reaction in AM is due to a bacterial infection, as this may be more difficult to determine in some cases than others.8 With that view in mind, we retrospectively included all adults diagnosed with AM between 1/1/2006 and 1/1/2010 who had undergone a PCT and CRP analysis (a total of 39 cases). AM was pronounced bacterial if the bacteria or its antigens could be isolated in CSF and viral if it met the criteria described by Jiménez Caballero et al.1 We used our laboratory’s normal reference values: 0 to 8 mg/L for CRP and PCT < 0.5 ng/mL. We determined PCR by using the quantitative method of enzyme immunoassay with a sensitivity of 1 mg/L (VITROS CRP slide for biochemistry). PCT was first measured with an immunochromatographic PCT-Q method (BRAHMS-PCT®) with semiquantitative evaluation, followed by ELECSYS (BRAHMS PCT®) with quantitative evaluation (these latter results were included in the intervals corresponding to the semiquantitative measurement). We used SPSS software version 15 for Windows (t-test for normal distributions and the χ² test for proportions) in the comparative analysis. P < .05 was considered statistically significant. Table 1 shows results (the most frequently isolated strains) that suggest that levels of PCT > 2 ng/mL (P < .05) may be useful in distinguishing between bacterial and viral AM, and that values as low as PCT 10 ng/mL (and CRP > 90 mg/L)

References


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