16. Drake Jr M. Migraine as an organic cause of monocular diplopia. Psychosomatics. 1983;24:1024—7.
17. Headache Classification Subcommittee of the International Headache Society. The International Classification of Headache Disorders. 2nd ed. Cephalalgia. 2004;24 Suppl. 24:1—160.
18. Dichgans M, Mayer M, Uttner I, Brüning M, Muller-Höcker J, Rungger G, et al. The phenotypic spectrum of CADASIL: clinical findings in 102 cases. Ann Neurol. 1998;44:731—9.
19. Jourel A, Dodick D, Parisi J, Cecillon M, Tourniere-Lasserve E, Bousser M. De novo mutation in the Notch3 gene causing CADASIL. Ann Neurol. 2000;47:388—91.
20. Menendez Gonzalez M, Garcia Fernandez C, Anton Gonzalez C, Navarro Izquierdo R, Garcia Martinez A, Blazquez Menes B. Un caso de CADASIL por mutación de *novo*. Rev Neurol. 2005;41:503—5.
21. Posada IJ, Ferrero M, Lopez-Valdez E, Goñi-Imizcoz M. Tratamiento con galantamina en la demencia asociada a CADASIL. Rev Neurol. 2008;47:299—300.
22. Keverne JS, Low WC, Ziabreva I, Court JA, Oakley AE, Calaria RN. Cholinergic neuronal deficits in CADASIL. Stroke. 2007;38:188—91.
23. Mesulam M, Siddique T, Cohen B. Cholinergic denervation in a pure multiinfarct state. Observations in CADASIL. Neurology. 2003;60:1183—5.

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Contrast uptake by anterior roots in acute motor axonal neuropathy☆

Captación de raíces anteriores en la neuropatía aguda motora axonal

Dear Editor:

Guillaume—Barré syndrome (GBS) is an acute immune-mediated disease affecting the peripheral nervous system. It can be divided into different subtypes according to clinical, immunological, neurophysiological, and pathological criteria.[1]

GBS includes at least 3 patterns: acute inflammatory demyelinating polyneuropathy (AIDP), acute motor axonal neuropathy/acute motor-sensory axonal neuropathy (AMAN and AMSAN), and Miller Fisher syndrome.

AMAN is a pure motor form of GBS which is frequently associated with the presence of certain anti-ganglioside antibodies and preceded by an infection with *Campylobacter jejuni*. This clinical form may present clinical characteristics that delay diagnosis.

We present the case of a man, aged 29, who came to the emergency department due to acute tetraparesis predominantly affecting the upper limbs and associated with back pain. The patient reported having had diarrhoea one week before. He claimed not to have experienced autonomic dysfunctions or sensory disorders. The neurological examination revealed normal reflexes and asymmetric tetraparesis predominantly affecting right-sided distal regions and the upper limbs.

Contrast-enhanced MR imaging of the spinal cord revealed enhancement limited to the anterior nerve roots (Figs. 1 and 2).

The patient's Achilles reflex subsequently disappeared. The CSF study showed albuminocytological dissociation. The neurophysiological study revealed a decrease in motor evoked potential amplitude with no changes in velocity or latencies and normal sensory nerve conduction. Conduction block was not detected. F-waves displayed normal persistence and latency in the upper and lower limbs. The needle study showed abundant spontaneous muscle activity in proximal and distal muscles of the upper and lower limbs with a reduced recruitment pattern. These neurological findings were compatible with exclusively motor and axonal impairment. Serology tests for *C. jejuni* were positive (1/1000). Tests were negative for anti-ganglioside antibodies GM1, GM2, GD1a, GT1B, and GQ1b. The patient was diagnosed with AMAN and treated with immunoglobulins (2 g/kg body weight), after which symptoms improved.

Three months after onset of symptoms, he presented distal weakness predominantly affecting the upper limbs (4/5 on the Medical Research Council scale).

AMAN is a form of GBS that exclusively affects motor function of the peripheral nervous system and causes axonal impairment while sparing myelin.[1] It seems that there is a relationship between AMAN and a prior infection with *C. jejuni*. This bacterial species has surface lipooligosaccharides that work as antigens and possess structures similar to those of some peripheral nerve gangliosides.[2] In these cases, neurophysiological studies show motor impairment with signs of axonal damage. However, some authors, such as Berciano et al., have shown that the pathological basis of this disease is demyelination even if neurophysiological studies are unable to demonstrate this finding in some cases. Demyelination is primarily radicular with secondary wallerian degeneration.[3]

Different studies have shown contrast uptake by nerve roots in acute polyradiculoneuropathies[4—8] or even by cranial nerves.[4] This uptake seems to be due to the absence of perineurium in preforaminal nerve roots and to a compromised blood—nerve barrier.[9] It has been suggested that this uptake is related to more severe pain and a poorer prognosis. Doctors decided to perform an MRI scan due to the initial asymmetry of the symptoms and because reflexes that were initially present disappeared during the course of the disease. The presence of normal reflexes or even hyperreflexia has been described in other patients with AMAN.[10] Although

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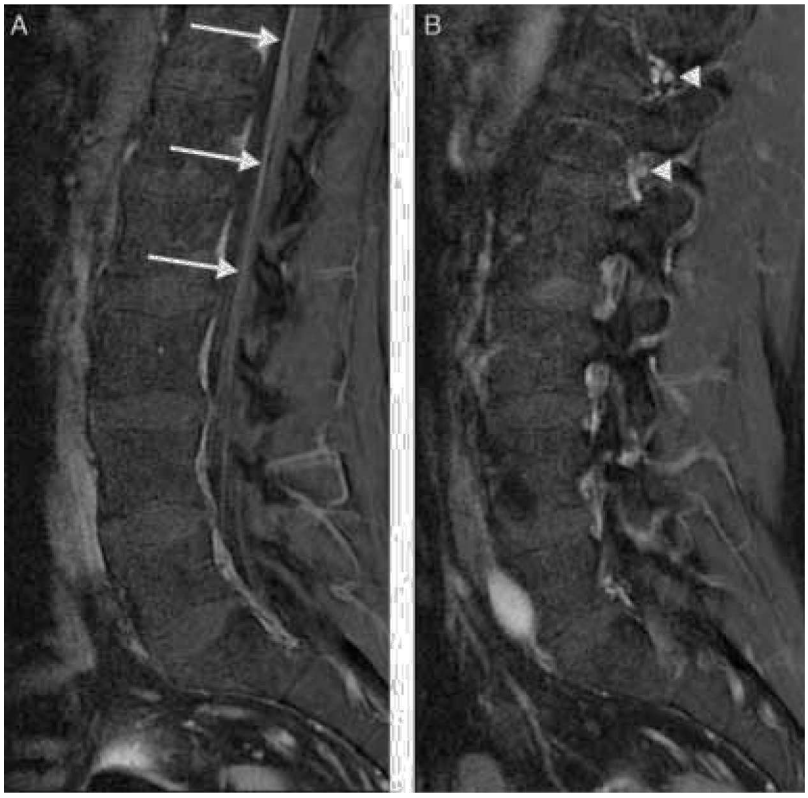

Figure 1 Fat-suppressed T1-weighted MR images with gadolinium contrast. (A) Marked enhancement of the cauda equina roots showing thickening (arrows). (B) MRI of the right intervertebral foramina showing enhancement of L1 and L2 (arrowheads).

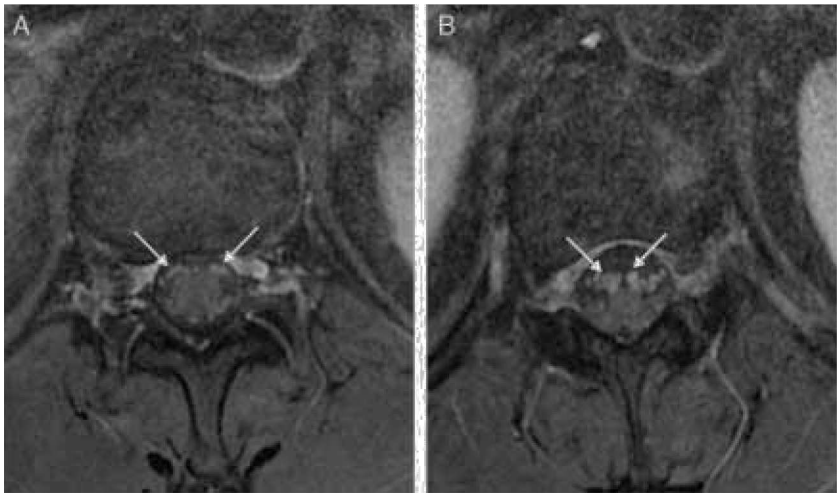

Figure 2 Fat-suppressed T1-weighted axial MRI showing enhancement of the anterior nerve roots in D12 and L1 and in the conus medullaris.

the MRI is not necessary to establish a diagnosis of AMAN or others forms of GBS, it may be useful for differential diagnosis of acute tetraparesis.

References

1. Hughes RA, Cornblath DR. Guillain—Barré syndrome. Lancet. 2005;366:1653—66.
2. Yuki N. Ganglioside mimicry and peripheral nerve disease. Muscle Nerve. 2007;35:691—711.
3. Berciano J. Patologia axonal en el síndrome de Guillain—Barré: una fisiopatologia compleja. Neurologia. 2003;18:121—31.
4. Yikilmaz A, Doganay S, Gumus H, Per H, Kumandas S, Coskun A. Magnetic resonance imaging of childhood Guillain—Barre syndrome. Childs Nerv Syst. 2010;26:1103—8.
5. Zuccoli G, Panigrahy A, Bailey A, Fitz C. Redefining the Guillain—Barré spectrum in children: neuroimaging findings of cranial nerve involvement. AJNR. 2011;32:639—42.
6. Wada M, Kato T, Yuki N, Hosoya T, Moriai S, Kurita K, et al. Gadolinium-enhancement of the spinal posterior roots in acute sensory ataxic neuropathy. Neurology. 1997;49:1470—1.

7. Gorson KC, Ropper AH, Muriello MA, Blair R. Prospective evaluation of MRI lumbosacral nerve root enhancement in acute Guillain—Barré syndrome. Neurology. 1996;47:813—7.
8. Vara-Castrodeza A, Tola-Arribas MA, Mendo-González M. Síndrome de Guillain—Barré: hallazgos por resonancia magnética en presentación clínica atípica. Rev Neurol. 2003;36:596—7.
9. Berciano J. Thickening and contrast enhancement of spinal roots on MR imaging in Guillain—Barre syndrome: thoughts on pathologic background. AJNR. 2011;32:E179.
10. Badía Picazo MC, Santonja Llabata JM, Gil Gimeno R, Salvador Aliaga A, Pascual Lozano AM, Láinez Andrés JM. Hiperreflexia en un paciente con síndrome de Guillain—Barré. Rev Neurol. 2004;38:697—8.

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Migraine-triggered hemifacial spasm: Another case study☆

Espasmo hemifacial desencadenado por migraña: un caso más

Dear Editor:

Hemifacial spasm (HFS) is characterised by involuntary tonic or clonic contractions of muscles innervated by the facial nerve. The pathogenesis of this condition is usually attributed to vascular compression at the emergence of the nerve root from the brainstem. This could be due to the appearance of ectopic discharges and the ephaptic transmission of abnormal impulses. Another potential mechanism could be central hyperexcitability with neural impairment in the facial nerve motor nucleus. To date, 5 cases of HFS caused by migraine have been published.[1—3]

We present the case of a woman aged 30 with no relevant personal history who had experienced migraine attacks without aura since adolescence. In the previous year, headache episodes became more intense and were sometimes preceded by visual aura. Pain always presented on the right side of the head; upon reaching its maximum intensity, it was accompanied by involuntary contractions of the periocular ipsilateral muscles. Contractions decreased as pain subsided and only appeared during migraine attacks. These episodes occurred an average of 4 times per month. Initial neurological examination was normal. However, during one of the migraine episodes, we were able to observe the contractions of the right periocular muscles described above. Electromyography (EMG) showed tonic activity of the right orbicularis oculi with high-frequency bursts of increased muscle fibre recruitment (Fig. 1) coinciding with clonic spasms. The study of facial nerve motor conduction and blink reflex showed no relevant abnormalities. Brain MRI and MRA revealed a loop of the right anterior inferior cerebellar artery in contact with the facial nerve (Fig. 2). Treatment with topiramate was prescribed at increasing doses reaching 75 mg per day, which decreased the frequency and intensity of migraine episodes and suppressed the associated muscle spasm.

The temporal relationship between migraine and HFS in the 6 published cases supports the idea of a pathophysiological link existing between those processes. As in prior cases,[1—3] our patient presented episodes of HFS that coincided with maximum pain intensity. Therefore, rather than being a migraine aura phenomenon with positive signs, HFS seems to be a consequence of migraine. Husid raised the possibility that HFS could be due to a mechanism of central hyperexcitability related to migraine or to facial nerve compression caused by the dilation of nearby vessels,

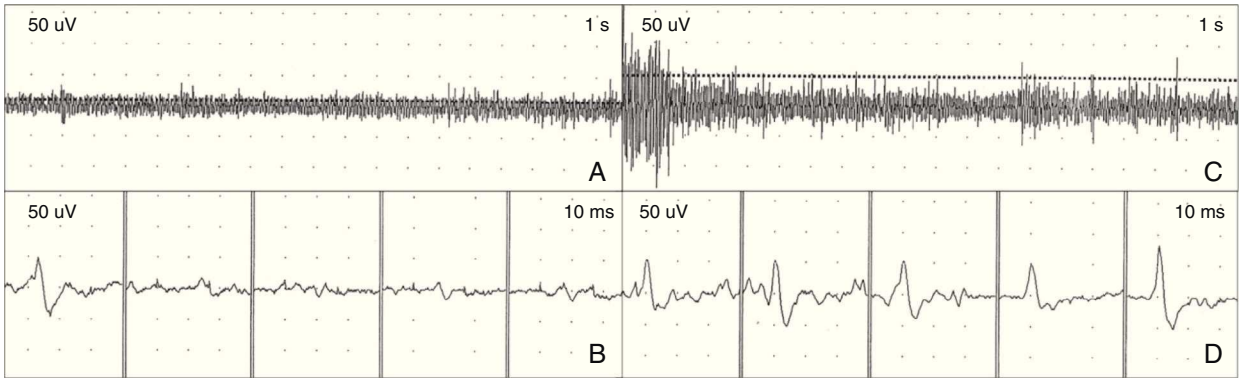

Figure 1 Electromyogram showing spasm of the right orbicularis oculi during a migraine episode. (A, B) Baseline tonic muscle activity coinciding with tonic contraction and reduced palpebral fissure. (C, D) High-frequency bursts of muscle fibre recruitment coinciding with clonic spasms of the upper and lower eyelids.

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