

neurological dysfunction can be seen in up to 60% of the cases and headache, impaired level of consciousness, focal deficits, seizures, or coma may be present. The intensity of the neurological involvement is quite variable and is frequently reversible. Skin rash is seen in 33% of cases and is mainly observed on the chest, neck, axillae, and on the oral and conjunctival mucosae. These skin lesions usually resolve within one week.

Examination of the *fundus oculi* will usually reveal multiple cotton-wool exudates, oedema, and retinal bleeding around the optic nerve, all of which is secondary to multiple infarcts of the nerve fibres.^{6,7}

The diagnosis is made on the basis of clinical findings and the most widely used diagnostic criteria are those put forth by Gurd.¹ Cerebral MRI is useful to demonstrate typical findings, such as diffuse hyperintense foci in the long TR sequences located in subcortical or periventricular white matter and *centrum semiovale*. Some of these lesions present restriction on the DWI sequence, corresponding to the ischaemia-related cytotoxic oedema. The GRE sequence may show low signal, spotty foci compatible with micro-haemorrhages in several regions. Furthermore, MRI can help to rule out other trauma processes, such as diffuse axonal lesion, contusion or haematoma.⁸ It also aids in establishing the prognosis of brain damage, since some studies have shown that the number of lesions on the MRI correlates with the GCS score and that disappearance of the brain lesions is related to the resolution of the neurological symptoms.⁹

In conclusion, the presence of neurological decline in a patient with multiple fractures, especially 24–72 h following trauma, should lead us to suspect FES. Findings on the cerebral MRI and the ophthalmoscopy are useful in making the diagnosis and ruling out other aetiologies. Despite the extensive lesions present on neuroimaging studies, prognosis may be favourable, as was the case of our patient.

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Optic neuropathy in Lewis–Summer syndrome: Presentation of a case[☆]

Neuropatía óptica en un síndrome de Lewis-Summer: a propósito de un caso

Sir,

Multifocal motor neuropathy is an immunomediated condition characterized by weakness and muscular atrophy, absence of sensory and pyramidal signs, in which weakness is caused by a multifocal block of conduction in motor nerves.¹ It has been associated with high titres of antibodies to ganglioside, especially anti-GM1, and it responds

adequately to treatment with high doses of intravenous immunoglobulin (IVIG).^{2–5} In 1985, Lewis et al. described 5 patients with multifocal acquired demyelinating sensory and motor neuropathy (MADSAM) (Lewis–Miller syndrome) basically affecting the upper limbs, with multifocal conduction block. Subsequently, Parry and Clarke described a similar condition but with pure motor compromise, similar to and confusing with amyotrophic lateral sclerosis. This entity was given the name multifocal motor neuropathy (MMN). This was followed by observations on the association of this condition with high titres of anti-GM13 antibodies, and evident recovery of muscle strength in most patients with the administration of IVIG. Segmentary demyelination implies damage to the myelinic membrane or Schwann's cell, with less important involvement of the axon. It usually appears in immunologically mediated demyelination or in alterations of myelin metabolism. Myelin may also be affected through myelinotoxic agents or mechanically by compression. This kind of patient poses problems when it comes to achieving a correct diagnosis: is it a case of axonal or demyelinating neuropathy? And if we reach a diagnosis of demyelinating

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neuropathy, is it acquired or hereditary? These questions must be answered in the clinical course, electrophysiological analysis, study of the cerebrospinal fluid, immunological tests, biopsy of the nerve and genetic studies.

Axonal polyneuropathies are usually treatment-resistant, probably because the treatment is inadequate or because the process of recovery through regeneration is very slow or incomplete, habitually associated with metabolic or toxic diseases. For this reason, achieving the definition of a polyneuropathy as demyelinating after ruling out hereditary causes suggests a pathology capable of being treated. Acquired demyelinating neuropathies represent a heterogeneous group, generally immunologically mediated. Chronic demyelinating inflammatory polyneuropathy is the most common cause.²

We report the case of a 61-year-old male attending the emergency department due to loss of visual acuity in his right eye over a period of 48 h. We conducted a complete ophthalmological examination in which visual acuity in that right eye allowed finger-counting while the left eye had 10/10. An afferent pupil defect was observed, while nothing abnormal was found on examination of the *fundus oculi* by means of mydriasis. The patient was therefore suspected of having retrobulbar optic neuritis and ER staff requested an analysis and GSV, all completely normal, and a campimetry which showed a generalized depression in sensitivity in the right eye, while the left eye was completely normal. He was initially prescribed boluses of corticosteroids and we started to study the patient with the neurology department. During anamnesis, the patient reported the presence of progressive muscle weakness, especially in the upper body, lasting for years, and that he had been operated for a pinched nerve in an upper limb without any improvement, but which he had ignored. The diagnosis was approached thanks to the electromyographic study (in which we were to find nerve conduction block in two or more nerves, altered sensory conduction velocity and abnormal sensory responses. It is difficult to reach a diagnosis with respect to other polyneuropathies, particularly with multifocal motor neuropathy, because it is complicated to demonstrate conduction block).^{6,7} The magnetic resonance scan requested revealed demyelination lesions. A lumbar puncture was performed, with the protein level that was found to be normal and a complete analysis showed no evidence of AntiGM1.²⁻⁴ He achieved scant or no recovery of his eyesight, even though the bolus steroid treatment was continued as it has been shown to be effective in some cases, and treatment was also begun with immunoglobulins and the patient's course has been monitored.

The difficulties in diagnosing demyelinating polyneuropathies range from the clinical picture to the electrophysiological pattern,^{6,7} thus requiring the performance of additional complementary examinations or even inadequate or unnecessary treatments. On the other hand, all the international diagnostic criteria must be taken into account with caution, individualizing each case; although they show high specificity, they do not present great sensitivity, leaving patients without adequate treatment.

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