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Optical neuropathy in Lewis-Summer syndrome: A casual relationship?*

Neuropatía óptica en un síndrome de Lewis-Sumner: ¿una asociación casual?

Dear Editor:

It was with great interest that we read the clinical case published in your journal under the title "Optic neuropathy in Lewis-Sumner syndrome: Presentation of a case".¹ We expected to read about a nosological link between central demyelinating disease in optic neuropathy and the peripheral demyelinating disease that characterises Lewis-Sumner syndrome (LSS). At the very least we thought the authors would have envisaged this possibility, upon finding both pathologies in the same patient. Instead, we found a short (but interesting) review of neuropathies with conduction block mentioning both multifocal motor neuropathy (MMN) and LSS. The differences between the two syndromes may not have been sufficiently emphasised. LSS is currently considered an asymmetric or multifocal type of chronic inflammatory demyelinating polyneuropathy (CIDP). Unlike in MMN, the sensory fibres are affected, finding anti-GM1 antibodies is uncommon, and a significant number of cases of this disease respond favourably to corticosteroid treatment.^{2,3} Nevertheless, this therapeutic response has been called into question by a recent study proposing that LSS be clearly distinguished not only from MMN, but also from CIDP.⁴ Lastly, regarding the case mentioned in the letter by Sánchez Ferreiro and Barreiro González, impairment of the central nervous system has been described in LSS, but as far as we know, not in MMN.^{5–7} In the case recently mentioned in *NEUROLOGÍA*,¹ the authors describe the co-presence of LSS and retrobulbar optic neuritis in the same

patient. However, neither of the pathologies is completely illustrated: no EMG results are given, and the pathogenesis of the optic neuritis is not clearly explained, since the authors list no vascular or autoimmune tests and do not specify the anatomical structure in which the magnetic resonance scan revealed demyelinating lesions. Above all, the authors' intent to establish an aetiopathogenic association between the two illnesses is never fully clarified. We therefore wonder if Sánchez Ferreiro and Barreiro González, in their letter to the editor, meant only to report the presence of two unusual pathologies in the same patient (pathologies which, furthermore, were not correctly described), or if they meant to suggest that the co-presence of two demyelinating diseases might be due to more than a mere chance. If the latter is true, the authors have unfortunately failed to transmit the interesting points of this case and their possible hypothesis.

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Selective immunoglobulin A deficiency is exceptionally associated with multiple sclerosis[☆]

El déficit selectivo de inmunoglobulina A excepcionalmente se asocia a esclerosis múltiple

Dear Editor:

It was with great interest that we read the article by Remolina López et al.¹ on one of the few cases in the literature of IgA deficiency in association with multiple sclerosis. In fact, we could only find one other case, that of a Japanese patient, in PubMed.² We would like to call attention to the rareness of this association and its possible aetiopathogenic implications.

Selective IgA deficiency is the most common primary immunodeficiency, and its prevalence varies among different ethnic groups. In our own region, our team found the highest prevalence of this deficiency in the general population in the Western world: 1 in 163 individuals.³ According to the current study protocol for multiple sclerosis, we measure serum Ig levels (IgG, IgA, and IgM), the kinetics of intrathecal immunoglobulin synthesis, and oligoclonal bands in cerebrospinal fluid in almost every patient. Therefore, we do not believe that immunoglobulin deficiencies go undetected or under-diagnosed in this disease. We studied 183 cases of multiple sclerosis in our series of patients, and we have not found any cases of IgA deficiency in the past 18 years. This contrasts with the high prevalence of IgA deficiency in the general population (0.61%) and in conjunction with numerous autoimmune diseases such as coeliac disease (4.1%), systemic lupus erythematosus (1.56%), thyroiditis (2.69%), and diabetes mellitus (4.70%).⁴ The absence of an association between multiple sclerosis and selective IgA deficiency may have aetiopathogenic implications. It could indicate a different genetic substratum or a different environmental trigger from those involved in the autoimmune diseases mentioned above. The lack of IgA in secretions fosters antigenic overstimulation, especially in the digestive

tract. It is unlikely, however that the digestive tract would play an important role in triggering multiple sclerosis. In this regard, it is particularly revealing that in myasthenia gravis, another autoimmune disease of the nervous system, we also find no association with selective IgA deficiency.⁵ Given the above, we believe that the case described by Remolina López et al. should be considered an exception based on the statistical evidence, and due to the high prevalence of both diseases. Discovering a possible causal association or a common origin for the diseases seems less likely.

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