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Devic disease as a differential diagnosis for spinal cord tumors: may a biopsy be needed?

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Introduction: Devic's disease, also known as neuromyelitis optica, is a rare demyelinating disease affecting the spinal cord and the optical nerves. The underlying pathological process is probably an immunological mechanism of tissular damage, and an associated immunoglobulin whose target antigen is aquaporin 4 is described. The main differential diagnosis are intrinsic spinal cord tumors and other types of myelitis. The present work discusses the importance of including Devic`s disease in the ethiological hypothesis of spinal lesions and the possible need for a biopsy.

Methods: Report of an illustrative case and literature review.

Results: A 13 year old previous healthy female patient was admitted in September 2012 at a pediatric tertiary hospital presenting acute onset and progressive asymetric paraparesis and partial left visual loss. Neurological examination showed anisocoria (left pupil greater than the right one), absence of photomotor reflex, optic nerve papilla edema seen by fundoscopy and grade II paraparesis, although asymetric. Sensitivity tests were doubtful. Brain MRI was normal. A spinal MRI showed a hyperintense spinal lesion from T2 to T6 with an associated syringomyelia. CSF sampling revealed a 1/80 positivity for aquaporin 4, with no further abnormalities. Although Devic`s disease was suggested, there was no significant clinical or radiological response after a metilprednisolone curse and two weeks of treatment with azathioprin, as would be expected. There was even a slight increase in the size of the lesion at a second MRI. Given this diagnostic doubt, the patient was submitted to a biopsy through a laminectomy and a median myelotomy at T3 level, considering a probable spinal cord tumor. Histological examination showed no tumoral lesion, but an inflamatory process with a predominance of histiocytes. There was no additional neurological deficit after surgery. These results redirected the hypothesis into a demyelinating disease. After restarting immunomodulating therapy, slightly progressive clinical improvement was noted.

Conclusions: Demyelinating diseases such as Devic's are to be included in the differential diagnosis of rapid progressive intrinsic spinal cord lesions with syringomyelia. It may mimetize a spinal cord tumor. Spinal cord lesions may pose diagnostic challenges that require obtaining tissue for histological examination through a biopsy. A spinal cord biopsy can be safely performed and it may be usefull in determining diagnosis.