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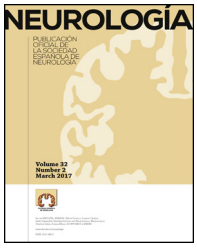

ORIGINAL ARTICLE

Differential diagnosis and prognosis for longitudinally extensive myelitis in Buenos Aires, Argentina☆

E. Carnero Contentti[a,*], J.P. Hryb[a], F. Leguizamón[b], J.L. Di Pace[a], J. Celso[b], E. Knorre[b], M.B. Perassolo[a]

[a] *Consultorio de Neuroinmunología, Servicio de Neurología, Hospital Dr. Carlos G. Durand, Ciudad Autónoma de Buenos Aires, Argentina*
[b] *Servicio de Neurología, Hospital General de Agudos Dr. Teodoro Álvarez, Ciudad Autónoma de Buenos Aires, Argentina*

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KEYWORDS
Longitudinally extensive transverse myelitis;
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Prognosis;
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Abstract
Introduction: Longitudinally extensive transverse myelitis (LETM) has classically been grouped with the full or limited neuromyelitis optica spectrum disorders (NMOSD). However, differential diagnosis reveals a wide range of aetiologies.
Objective: To report on differential diagnosis and prognosis for LETM observed in a group of patients in Buenos Aires, Argentina.
Patients and methods: Cross-sectional and retrospective multicentre study in two hospitals in Buenos Aires from June 2008 to June 2014. Inclusion criteria: medullary syndrome associated with spinal cord lesion extending for 3 or more contiguous spinal segments in magnetic resonance imaging (MRI). Clinical, radiological, and biochemical data were collected and subjects were rated on the Winner—Hughes Functional Disability Scale (WHFDS) at 3 months.
Results: We evaluated 27 patients, 74% of whom were women; mean age was 35.22 years. The NMO-IgG antibody test was performed in 66.6% and oligoclonal band testing in 71%. NMO-IgG seropositivity was found exclusively in NMOSD patients (75%). Brain MRI was normal in 59.2% and revealed a mean of 7.9 affected spinal segments. Differential diagnoses revealed NMOSD (37%), idiopathic LETM (22.2%), lupus (11.1%), tumour (11.1%), dural fistula (7.4%), acute disseminated encephalomyelitis (7.4%), and a single case of multiple sclerosis (3.7%). Patients with lesions to $\geq$7 spinal segments showed poor recovery at 3 months ($P < .001$); these cases were associated with neoplastic, vascular, idiopathic, and lupus-related aetiologies.
Conclusions: The most frequent causes of LETM in our cohort were NMOSD followed by idiopathic cases. Neoplastic, vascular, lupus-related, and idiopathic LETM may constitute a critical group with a distinct prognosis and other treatment needs.

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* Corresponding author.
E-mail address: junior.carnero@hotmail.com (E. Carnero Contentti).

PALABRAS CLAVE
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Anticuerpo antiaquaporina-4;
Pronóstico;
Diagnósticos diferenciales

Diagnósticos diferenciales y pronóstico de las mielitis longitudinales extensas en Buenos Aires, Argentina

Resumen
Introducción: Las mielitis longitudinales extensas (LETM) fueron clásicamente relacionadas con los trastornos del espectro de la neuromielitis óptica (NMOSD) tanto definidas como limitadas. Sin embargo, los diagnósticos diferenciales incluyen una amplia gama de etiologías.
Objetivo: Comunicar los diagnósticos diferenciales y el pronóstico de LETM observados en un grupo de pacientes en Buenos Aires, Argentina.
Pacientes y métodos: Estudio multicéntrico retrospectivo transversal realizado en 2 hospitales de Buenos Aires desde junio del 2008 hasta junio del 2014. Criterios de inclusión: síndrome medular asociado a una lesión en la médula espinal con una extensión de 3 o más segmentos vertebrales contiguos en la resonancia magnética (RM). Datos bioquímicos, radiológicos y clínicos fueron evaluados. Asimismo, se aplicó la escala de discapacidad funcional de Winer-Hughes (WHFDS) a los 3 meses.
Resultados: Se evaluó a 27 pacientes, el 74% mujeres, edad (media): 35,22 años. NMO-IgG se realizó en el 66,6% y las bandas oligoclonales en el 71%. NMO-IgG se observó exclusivamente en pacientes con NMOSD (75%). La RM de encéfalo fue normal en el 59,2% y la media de segmentos afectados en RM espinal fue 7,9. Los diagnósticos diferenciales encontrados fueron: NMOSD (37%), idiopática (22,2%), lupus (11,1%), tumores (11,1%), fístula dural (7,4%), encefalomielitis diseminada aguda (7,4%) y esclerosis múltiple (3,7%). Los pacientes con $\geq$ 7 segmentos afectados tenían peor WHFDS ($p < 0,001$) y se asoció a etiología tumoral, vascular, lupus e idiopática.
Conclusiones: En nuestra cohorte, NMOSD seguidos por idiopática, fueron las causas más frecuentes de LETM. Las LETM tumorales, vasculares, lupus e idiopáticas pueden representar un grupo crítico con diferente pronóstico y tratamiento.

Introduction

Longitudinally extensive transverse myelitis (LETM) is defined as spinal cord lesions affecting at least 3 spinal cord segments and resulting in hyperintensities on sagittal T2-weighted magnetic resonance imaging (MRI) sequences.[1–3] Though rare, LETM has severe clinical consequences. A systematic evaluation to establish the aetiological diagnosis and assign early treatment are therefore essential, not only to improve short- and long-term functional outcomes but also to prevent further inflammatory-demyelinating attacks to the spinal cord and/or central nervous system.[3–5] At present, LETM is mainly associated with neuromyelitis optica (NMO) and neuromyelitis optica spectrum disorders (NMOSD).[1–6] However, differential diagnosis includes a number of conditions: non-NMO autoimmune inflammatory disorders (acute disseminated encephalomyelitis [ADEM], multiple sclerosis [MS], etc.), systemic autoimmune diseases (systemic lupus erythematosus [SLE], Sjögren syndrome, sarcoidosis, etc.), infections and parainfections (viral, bacterial, parasitic), paraneoplastic disorders (mainly secondary to CRMP5 antibodies), neoplasms (astrocytoma, ependymoma, spinal cord metastases, etc.), metabolic disorders (vitamin B_{12} or copper deficiency), and vascular diseases (stroke, dural fistulas), among others.[3] In 2002, the Transverse Myelitis Consortium Working Group proposed a set of diagnostic criteria for idiopathic myelitis for those cases in which aetiology could not be established.[1,2]

Given the scarcity of data from our region, our purpose is to evaluate aetiological diagnoses, clinical and paraclinical characteristics, and functional outcomes at 3 months in a cohort of patients with LETM in Buenos Aires, Argentina.

Patients and methods

We conducted a multicentre cross-sectional retrospective descriptive study of patients examined in 2 public hospitals in Buenos Aires (Hospital Carlos G. Durand and Hospital Teodoro Álvarez) between June 2008 and May 2014 by reviewing their clinical histories. We included all patients meeting the inclusion criteria (Table 1) and evaluated their demographic, clinical, paraclinical, radiological, and aetiological characteristics and functional progression at 3 months assessed on the Winner—Hughes Functional Disability Scale (WHFDS).[7] Demographic data included sex and age at onset. We also checked for any motor, sensory, or autonomic disorders at the time of the neurological examination. Immunological studies, coagulation tests, and blood tests to screen for infection were conducted when there was clinical suspicion of immunological, coagulation, or infectious diseases, according to the validated international criteria for each aetiology.[1,4,8–15] Likewise, serum aquaporin-4 (AQP4) antibodies (NMO-IgG) were determined by indirect immunofluorescence (confocal microscopy) on monkey cerebellum sections. Oligoclonal bands (OCB) in cerebrospinal