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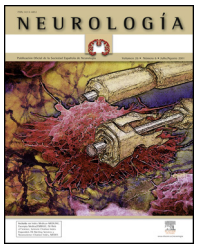

ORIGINAL ARTICLE

Lesser-known myelin-related disorders: Focal tumour-like demyelinating lesions☆

J.A. Jiménez Arango[a,b,*], **C.S. Uribe Uribe**[b], **G. Toro González**[c]

[a] *Johns Hopkins University, Baltimore, United States*
[b] *Universidad de Antioquia, Medellín, Colombia*
[c] *Instituto Nacional de Salud, Universidad Nacional de Colombia, Bogotá, Colombia*

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Abstract
Introduction: Focal tumour-like demyelinating lesions are defined as solitary demyelinating lesions with a diameter greater than 2 cm. In imaging studies, these lesions may mimic a neoplasm or brain abscess; as a result, invasive diagnostic and therapeutic measures may be performed that will in some cases increase morbidity. Our aim was to analyse and characterise these lesions according to their clinical, radiological, and pathological characteristics, and these data in addition to our literature review will contribute to a better understanding of these lesions.
Methods: This descriptive study includes five cases with pathological diagnoses. We provide subject characteristics gathered through reviewing their clinical, radiology, and pathology reports.
Results: Patients' ages ranged from 12 to 60 years; three patients were female. The time delay between symptom onset and hospital admission was 3-120 days. Clinical manifestations were diverse and dependent on the location of the lesion, pyramidal signs were found in 80% of patients, there were no clinical or radiological signs of spinal cord involvement, and follow-up times ranged from 1 to 15 years.
Conclusion: Brain biopsy is the gold standard for the diagnosis of demyelinating tumour-like lesions; however, their clinical features, along with several magnetic resonance imaging features such as open ring enhancement, venular enhancement, the presence of glutamate in spectroscopy, and others may be sufficient to differentiate neoplastic lesions from focal tumour-like demyelinating lesions.

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* Corresponding author.
E-mail address: jorge.neurologia@gmail.com (J.A. Jiménez Arango).

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PALABRAS CLAVE
Enfermedades autoinmunes desmielinizantes;
Esclerosis múltiple;
Lesiones desmielinizantes focales seudotumorales

Enfermedades menos conocidas de la mielina: lesiones desmielinizantes focales seudotumorales

Resumen
Introducción: Las lesiones desmielinizantes focales seudotumorales se definen como lesiones solitarias, desmielinizantes, con diámetro superior de 2 cm. Estas pueden imitar mediante el estudio imagenológico una neoplasia o absceso cerebral, lo que lleva a medidas diagnósticas y terapéuticas invasivas en algunos casos, incrementando la morbilidad. Nuestro objetivo fue analizar y caracterizar estas lesiones clínica, radiológica y patológicamente, lo que, sumado a la revisión de la literatura, aportará al entendimiento de este tipo de trastornos.
Métodos: En este estudio descriptivo, se reportan 5 casos con diagnóstico patológico. Mediante la revisión de informes relacionados, clínicos, radiológicos y patológicos, se resumen las características de los sujetos.
Resultados: La edad de los pacientes osciló entre los 12 y los 60 años, 3 pacientes fueron de género femenino. La latencia de los síntomas hasta admisión hospitalaria fue entre 3 y 120 días, las manifestaciones clínicas fueron diversas y dependientes de la localización de la lesión, en el 80% de los pacientes se encontraron signos piramidales y no se encontraron clínica o imagenológicamente lesiones de la médula espinal; el seguimiento de los pacientes abarca desde un año hasta 15 años.
Conclusión: La biopsia cerebral es el estándar de oro para el diagnóstico de las lesiones desmielinizantes seudotumorales; no obstante, las características clínicas, junto con varias características de la resonancia magnética, tales como el realce en anillo abierto, el realce venular y la presencia de glutamato en la espectroscopia, entre otras, pueden ser satisfactorias en la diferenciación de las lesiones desmielinizantes focales seudotumorales de lesiones neoplásicas.

Introduction

Primary demyelinating disorders of the central nervous system (CNS) are a set of entities that include acute disseminated encephalomyelitis (ADEM), acute haemorrhagic leukoencephalopathy, neuromyelitis optica (Devic disease), and several types of multiple sclerosis (MS). The latter includes chronic MS (Charcot type), acute MS (Marburg type), myelinoclastic diffuse sclerosis (Schilder type), and concentric sclerosis (Baló type). Charcot or classic MS is the most frequent and best-known variant, and although its prevalence is low in tropical regions,[1,2] it is currently on the rise. However, there is a rare demyelinating disease that presents as a large area of focal demyelination (>2 cm) associated with a mass effect which can imitate a tumour or cerebral abscess in an imaging study. The literature refers to this disease as 'demyelinating pseudotumour', 'tumour-like demyelinating lesions', and 'swollen demyelinating lesions'.

Although magnetic resonance imaging (MRI) has good sensitivity to detect these lesions, it is not a specific test for demyelinating pseudotumour. A neuroradiological study showing a large lesion located in the deep white matter with a significant mass effect, which clinically manifests as signs of increased intracranial pressure, is very indicative of a CNS tumour. In rare cases, a demyelinating disease can manifest with atypical symptoms and images that indicate a brain tumour. These focal tumour-like demyelinating lesions (FTDL) may pose diagnostic challenges for both doctors and radiologists.

The difficulty involved in diagnosing a focal tumour-like demyelinating lesion often leads to a surgical biopsy, which is currently the gold standard for a definitive diagnosis.

We report five cases in which diagnosis was confirmed by a pathology study. After reviewing related clinical, radiological, and pathological reports, we summarise their diagnostic characteristics.

Methods

This descriptive study presents five cases with a diagnosis confirmed by pathology study. We reviewed patient clinical histories to extract clinical data such as presentation profile, clinical manifestations, treatment applied, and response to treatment. Patients' clinical outcomes were also recorded. Characteristics of the lesions on the magnetic resonance images were reviewed, and we describe microscopic findings from the lesions where possible.

Results

Age at which lesions appeared ranged from 12 to 60 years; 3 of the 5 patients were women. Time from symptom onset to hospital admission ranged from 3 days to 4 months. Clinical manifestations varied greatly and were dependent on lesion location; however, 80% of the patients presented pyramidal signs. No spinal cord lesions were detected by clinical signs