Radiology Case Reports ■■ (2017) ■■–■■

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journal homepage: http://Elsevier.com/locate/radcr

Neuroradiology

Progression of tumefactive demyelinating lesion in a child demonstrated with MRI

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ARTICLE INFO

Article history:
Received 9 September 2017
Received in revised form 29 October 2017
Accepted 30 October 2017
Available online

Keywords:
Tumefactive demyelinating lesion
Brain tumor
MR diffusion
MR imaging perfusion
MR imaging spectroscopy

ABSTRACT

Tumefactive demyelinating lesions (TDLs) are atypical presentations of various demyelinating diseases. They can mimic brain tumors in their clinical and radiological features and usually respond favorably to corticosteroid therapy. We report a case of a 17-year-old girl with a single TDL suddenly increasing in size even under steroid therapy. She underwent very strict follow-up examinations with conventional magnetic resonance and diffusion-weighted imaging, perfusion-weighted imaging, proton-magnetic resonance spectroscopy. The behavior of the lesion during the different follow-up sessions posed a diagnostic challenge as it expanded its size during the final examination, in stark contrast to what we forecast. Diagnosis of TDL was initially hypothesized, but the aggressive behavior of the lesion required biopsy.

Introduction

Tumefactive demyelinating lesions (TDLs), also named "demyelinating pseudotumors," are rare subsets of demyelinating manifestations. The exact pathogenesis of TDLs is not clearly understood [1–4]. They can occur in isolation, as part of multiple sclerosis (MS), acute disseminated encephalomyelitis (ADEM), or neuromyelitis optica spectrum disorder (NMOSD) [5]. MS accounts for most cases of TDLs [3,6]. TDLs might be single or multiple and may appear simultaneously at onset or sequentially. When they appear as single mass lesion, they might be mistaken for brain tumors [1–3]. Clinical manifestations vary from asymptomatic lesions to headache, cognitive abnormalities, mental confusion, impaired consciousness, aphasia, apraxia, cerebellar symptoms, visual field defects, or

Informed consent from the subject of this report was obtained.
Competing Interests: The authors have no conflicts of interest to disclose.
Funding sources: This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.
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https://doi.org/10.1016/j.radcr.2017.10.026

seizures [1,2,4]. Usually, there is no previous history of similar episodes and they may appear as the first demyelinating event in the course of MS [4,7,8]. Magnetic resonance imaging (MRI) is a critical tool in the diagnosis of this entity, although the overlapping similarities between the imaging appearances of TDLs with the imaging appearances of brain neoplasms, such as primary central nervous system (CNS) lymphoma and high-grade glioma, often leads to surgical biopsy [1,2,9,10]. In general, TDLs respond well to steroids, reducing in size on follow-up imaging [6,11,12]. However, in many cases, the diagnosis remains uncertain and, therefore, biopsy is indicated [9–12].

Subject and methods

A 17-year-old Caucasian girl came to the emergency room of our hospital with vertigo, aphasia, and a difficulty in moving her right leg, first experienced 5 days earlier. A neurologic examination revealed right brachiocrural hemiparesis (MRC2/5), reduction of right tendon reflexes, and hypoesthesia of the right lower limb. She had no medical history and mentioned a tetanus vaccine 3 months earlier. Brain computed tomography showed a large hypodense lesion, without mass effect, in the left periventricular and paratrigonal zone. A brain MRI was immediately performed, which showed a left large periventricular and paratrigonal area of signal abnormality, hypointense on T1-weighted images, hyperintense on T2-weighted and fluid-attenuated inversion recovery images, measuring $40 \times 50 \times 30$mm, with minimal perifocal edema and no mass effect (Fig. 1A). Diffusion-weighted imaging (DWI) revealed areas of restricted diffusion within the lesion, with low apparent diffusion coefficient (ADC) values (Fig. 1B and C). After Gd-contrast administration, no enhancement was seen; only on a very delayed scan (after 15 minutes) a faint enhancement could be appreciated (Fig. 1D). Dynamic susceptibility contrast and perfusion-weighted imaging (PWI) revealed an augmented relative cerebral blood volume (rCBV) (Fig. 2A and B), relative cerebral blood flow (rCBF), and reduction of mean transit time. Proton multivoxel magnetic spectroscopy (PMRS) showed an increase in GLN/Cr, GSH/Cr, a peak of lactate, and reduction of mI/Cr (Fig. 2C). Suspecting a demyelinating lesion, the study was extended to the spine, which revealed a small, peripheral non-enhancing, medullary lesion at D5 level. Based on these imaging features, a diagnosis of TDL was carried out. Low-grade glioma was considered a less probable differential diagnosis. A screen for against aquaporin 4-IG, GAD-SSA-SSB, N-methyl-d-aspartate (NMDA) myelin oligodendrocyte glycoprotein-IG, anti-nuclear, anti-neutrophil cytoplasmic as well as anti-cardiolipin antibodies, angiotensin-converting enzyme, lysozyme, and C-reactive protein yielded negative results or normal values. An infection screen, including human immunodeficiency viruses, treponemal, and Borrelia serology tests yielded negative results or results within the normal ranges. The cerebrospinal fluid (CSF) was clear and showed normal biochemistry and cell counts. Fluorescence-activated cell-sorting analysis revealed no atypical cells. Oligoclonal bands were positively restricted to CSF. Somatosensory and visually evoked potentials were normal. Under the presumptive diagnosis of TDL, treatment with methylprednisolone (1000 mg/d, intravenously) was started, which prompted a clinical response. After 1 week of therapy, a follow-up MRI revealed consistent, unchanged dimension of the lesion, although the values of the ADC seemed widely increased and there was no enhancement (even 1 hour after injection). PWI continued to show an increase in rCBV and rCBF but on a less significant scale than the previous examination. PMRS confirmed higher Gln/Cr, GSH/Cr, Cho/Cr with the reduction of mI/Cr and N-Acetylaspartate/Cr. The results of the neurologic examination improved (brachiocrural hemiparesis MRC 4/5), and oral prednisone therapy was continued (50 mg/d for 2 weeks, after 25 mg/d), although it must be noted the MRI performed 6 weeks later showed an unexpected progression of the lesion. As a matter of fact, it had increased in size, extending to the parietal and in the temporal lobe, with mass effect and midline shift. Also, the perilesional edema was extended (Fig. 3A). The lesion showed necrotic components and intense, inhomogeneous enhancement (Fig. 3B). ADC values (Fig. 3C) were high, and PWI showed a reduction of perfusion parameters (Fig. 3D). PMRS confirmed the spectral profile of the previous examinations. (All results are reported in Table 1.) At this point, a biopsy was necessary to make a certain diagnosis because a neoplastic nature of the lesion could not be excluded. Histology showed demyelination, extensive macrophage invasion (CD68+), gliosis, and necrosis. The patient is currently under interferon β1a (44 μg 3 times for week). The last follow-up MRI showed no new lesions, and the neurological examination is stable.

Discussion

A TDL is defined on MRI by the presence of large brain mass (≥2.0 cm in diameter) with edema and mass effect. The lesion more commonly involves the supratentorial compartment, mainly white matter tracts, in a periventricular distribution [1,2,4]. When there are multiple periventricular white matter lesions involving the major white matter tracts, such as corpus callosum or brachium pontis, associated to the presence of spinal cord lesions, the diagnosis of MS is straightforward [13]. However, a solitary, inhomogeneous lesion can pose a considerable diagnostic challenge. The use of a contrast agent is of limited benefit because any pathologic process associated with disruption of the blood-brain barrier can result in enhancement on MRI [12–17]. Primary and metastatic tumors often manifest as rounded, well-circumscribed, nodular ring enhancement lesions with different sizes, surrounded by a variable amount of vasogenic edema [16]. In comparison with tumors, TDLs have lesser mass effect and edema, relating to plaque size with incomplete or open ring enhancement [12] but the conventional MRI appearance cannot be specific. Advanced MRI techniques, such as DWI, PWI, PMRS, may improve the diagnosis of solitary brain lesions [11,12]. In this study, we have observed and analyzed the evolution of a TDL, under corticosteroid therapy, with conventional and advanced MRI techniques. In the case analyzed, the initial MRI showed areas of restricted diffusion, with decreased ADC in the left paratrigonal and periventricular areas, without the classical peripheral distribution [12]. Several recent case studies have reported reduced ADC values in acute demyelinating lesions