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Clinical Letter

Pediatric Baló Concentric Sclerosis Response to Dimethyl Fumarate

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Baló concentric sclerosis (BCS) is an atypical idiopathic inflammatory demyelinating disorder initially described by Josef Balo in 1928.[1] BCS is characterized by concentric rings of alternating demyelinated and normally myelinated tissue.

Patient Description

This 17-year old left-handed, previously healthy white male presented with acute-onset left hemiparesis. His examination was notable for dysarthria and left-sided findings of distal greater than proximal hemiparesis, facial droop, reduced vibratory sense, and hyporeflexia. Magnetic resonance imaging showed a T2-hyperintense enhancing targetoid lesion and two T2-hyperintense nonenhancing lesions in periventricular and subcortical territories (Fig 1); magnetic resonance imaging of the spine was normal.

The initial differential diagnosis with this imaging included primary central nervous system lymphoma, BCS, and infection. Cerebrospinal fluid showed oligoclonal bands with an elevated Immunoglobulin G index; aquaporin 4 antibodies were negative. Extensive infectious evaluation including human immunodeficiency virus, tuberculosis, toxoplasmosis, syphilis, cryptococcus, bartonella, coccidioidomycosis, histoplasmosis, blastomyces, coccidioides, and aspergillosis was negative. Oncologic studies including immunoglobulin heavy locus rearrangement, flow cytometry, and non-gyn cytology were unrevealing. Because of his classical imaging finding for BCS, two other lesions typical for multiple sclerosis (MS), and inflammatory cerebrospinal fluid, he was diagnosed with MS.

He was treated acutely with five days of high-dose intravenous methylprednisolone and then discharged with a 30-day oral prednisone taper. Follow-up imaging showed resolution of enhancement in his BCS lesion (Fig 2) with no evidence of disease progression after four months, but he still required an ankle and foot orthotic and cane for ambulation. Because of his diagnosis of MS, large BCS lesion, young age, and normal premorbid function, we initiated aggressive disease-modifying therapy with dimethyl fumarate (DMF). He was then lost to follow-up without repeat hospitalizations for disease flares.

Discussion

This young man with BCS had a good outcome, which we propose is due to his age and treatment. Because children with BCS tend to do well, it has been postulated that the developing brain and immune system are more resilient to this specific demyelinating injury.[2]

In addition, treatment with maintenance DMF, a therapy that has never been previously described for BCS, was shown in this case to be feasible in patients with limited health literacy and follow-up because it is oral and relatively well tolerated. There are conflicting theories about the pathophysiology of BCS.[3] One study found extensive aquaporin-4 and connexin loss in BCS without associated autoantibodies, suggesting an astrocytopathy that is not antibody mediated.[4] This finding helped inform our decision to use DMF, which has been shown to have a more broad anti-inflammatory effect primarily on T cells with less of an impact on the humoral immune response.[5,6]

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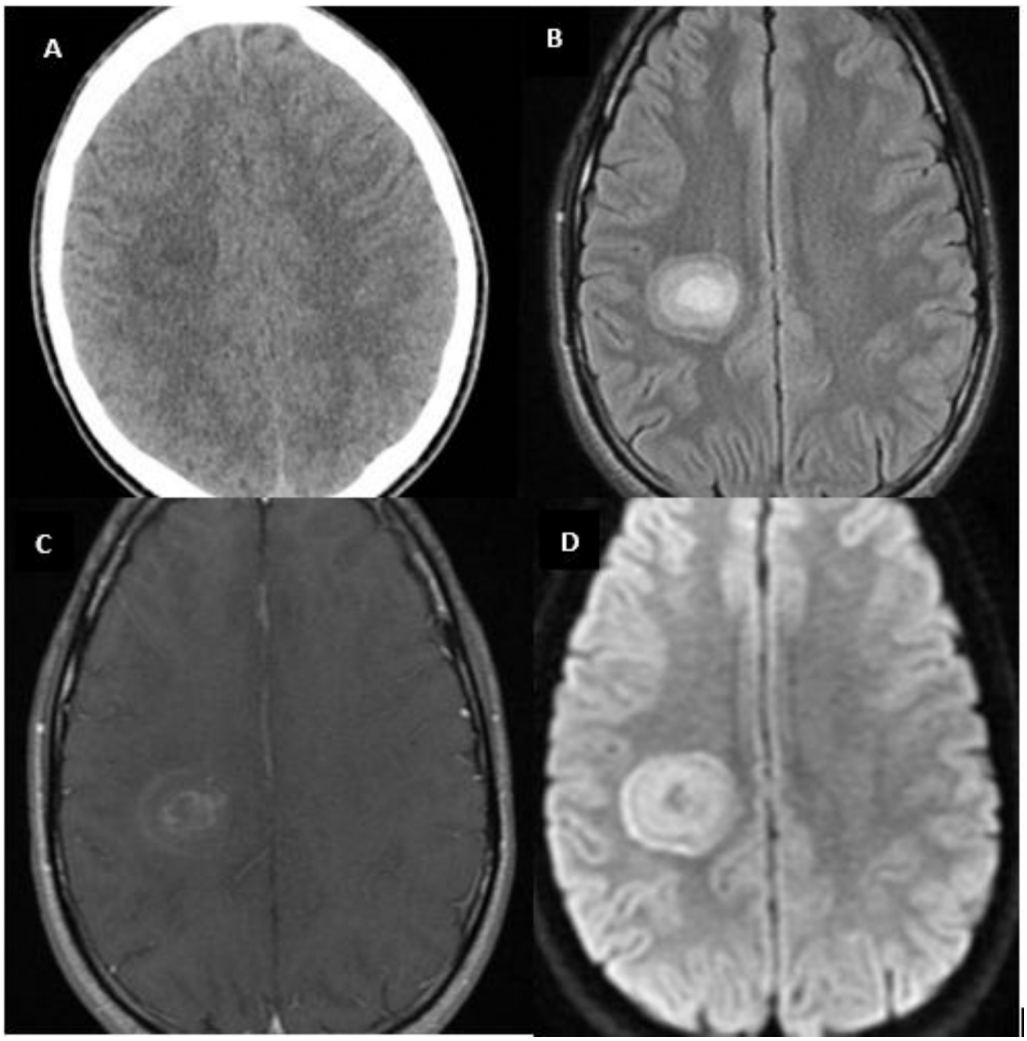

FIGURE 1. (A) Computed tomography of the head without contrast showing targetoid, mass-like 2.8-cm lesion in the right centrum semiovale. Magnetic resonance fluid-attenuated inversion recovery (B), postcontrast (C), and diffusion-weighted imagings (D) showing T2-hyperintense supratentorial white matter lesion with incomplete ringlike enhancement and restriction.

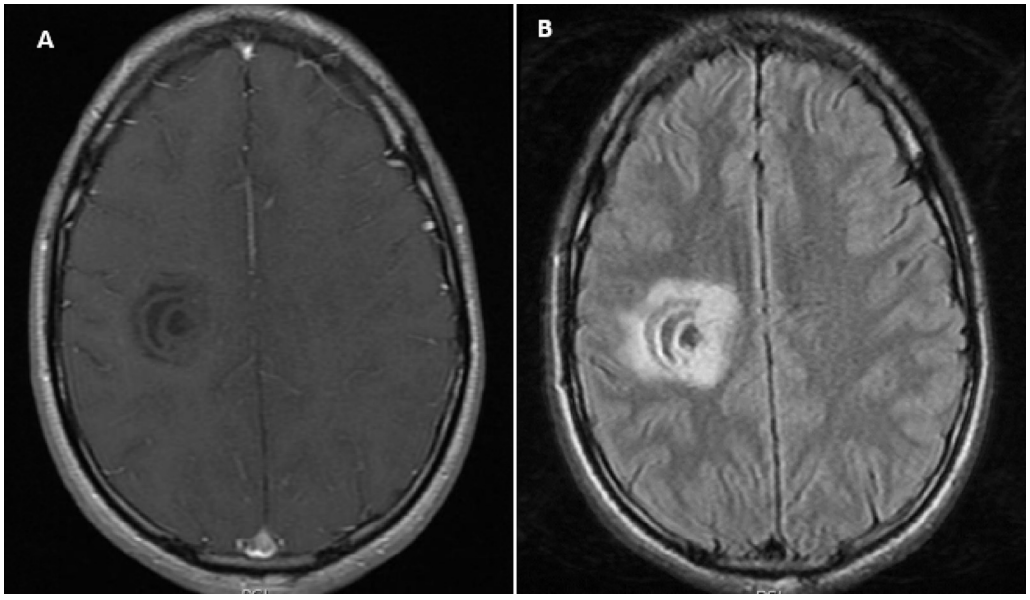

FIGURE 2. Magnetic resonance imaging two months after presentation with postcontrast T1 (A) and fluid-attenuated inversion recovery (B).

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