Balo's Concentric Sclerosis Mimicking Tumor on Magnetic Resonance Imaging in a Young Patient

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ABSTRACT: Balo's concentric sclerosis (BCS) is a rare demyelinating disease known as Multiple Sclerosis (MS) lesion type III. It is a disease of the white matter of the brain characterized by a round lesion with variable concentric myelinated and demyelinated layers, appearing as "onion bulb." We present a case of BCS and discuss the imaging findings and management strategies of this disease. A 26-y-old male developed headache, weakness, and numbness of limbs. Magnetic resonance imaging (MRI) showed concentric lamellar like demyelinating lesions at the subcortical regions. The patient's neurological symptoms were consistent with the MRI findings.

KEYWORDS: Balo's concentric sclerosis, demyelinating, multiple sclerosis, magnetic resonance imaging, pseudotumoral lesion

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Introduction

BCS is a primary inflammatory central nervous system demyelinating disease that is considered a rare, radiographically and pathologically distinct variant of MS. BCS was described for the first time in 1906 by Marburg, and in 1928, Josef Balo reported a case of an autopsy with demyelinated lesions as encephalitis periaxialis concentrica.[1,2] BCS is frequently seen in patients at the age of 4 to 56 y-old. The incidence in men is higher than in women. BCS lesions are common in the white matter of the cerebral hemispheres. Other less common locations reported are in the cerebellum, brainstem, optic chiasm, and spinal cord.[1-3]

Case Report

A 26-y-old right-handed man was admitted to the Emergency Department with a bilateral headache, accompanied by numbness, weakness, and clumsiness in the limbs during the past week. He reported a severe headache worse before sleep and when waking in the morning. He always felt numbness in the arms and legs, concomitant with weakness and clumsiness in his hands. Physical examination revealed left pronator drift, bilateral Hoffmann's sign (+), decreased muscle strength, and increased reflexes in upper and lower extremities both sides (left > right). There was mild numbness in both upper and lower limbs bilaterally. He noted similar symptoms 3 mo ago, which resolved spontaneously after a few days. The medical history of the patient and his family were unremarkable. Laboratory tests, including complete blood count, liver function, kidney function, erythrocyte sedimentation rate, vitamin B12 level, and metabolic profile were within normal limits. Brain MRI showed 3 well-circumscribed concentric lamellar lesions in the bilateral frontal and the right parietal-temporal with slight perilesional edema and no mass effect. The lesions demonstrated alternate bands of hypointense to isointense on T1-weighted (T1W) image, isointense to hyperintense on T2-weighted (T2W), and fluid attenuated inversion recovery (FLAIR) images. The lesions were unrestricted diffuse on diffusion-weighted imaging (DWI) and apparent diffusion coefficient (ADC) images. The lesions showed striking but incomplete ring enhancement after contrast administration (Figure 1). Magnetic resonance spectroscopy (MRS) revealed characteristic spectra of BCS with choline peak increased and *N*-acetylaspartate peak decreased (Figure 2). The typical concentric pattern detected on MRI was consistent with a diagnosis of BCS. The patient was treated with intravenous (IV) methylprednisolone acetate at a dose of 1200 mg/d for 3 d, then 600 mg/d for the next 6 d, and then tapered with oral prednisone over 4 wk, starting at 60 mg. His symptoms improved significantly after the first 3 d of treatment. His neurologic examination returned to almost normal after 2 wk. On a 6-mo

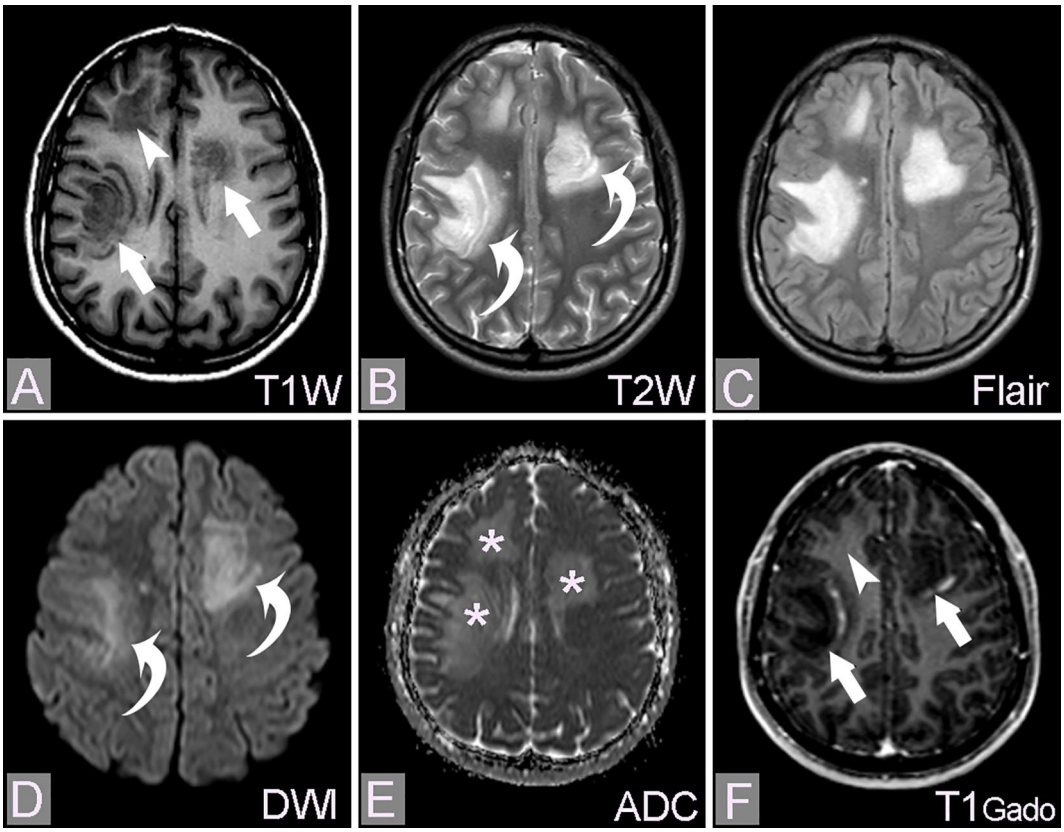

Figure 1. Pseudotumoral MS with typical lesions of BCS shows concentric lamellae (arrows), whereas more atypical Balo-like lesions display more complex shapes (head arrows): (A) T1W image shows hypointense lesions in the right frontal-parietal-temporal and left frontal, (B, C) lesions have a high signal intensity on T2W and FLAIR images, (D, E) lesions are unrestricted diffuse seen on the ADC image (asterisks). Note that the high signal intensity of lesions on the DWI image is an artifact due to T2 shine-through effect (curved arrows), and (F) a contrast-enhanced T1W image reveals a characteristic enhancement as a broken ring (arrows).

follow-up examination, patient was doing well with no evidence of dissemination of the disease.

Discussion

BCS is regarded as a rare variant of MS. It is characterized by demyelinating disorder of the central nervous system and is diagnosed by pathological features and clinical manifestations. The term "concentric" springs from lesion morphology the same as "onion-like" in which layers of demyelinated and myelinated tissue arrange concentrically and alternately. BCS symptoms include headaches, muscle pain or spasms, weakness, paralysis, dysphasia, dysarthria, or cognitive impairment,[4,5] such as in our case.

BCS clinical features are found similarly to MS. In histopathology, BCS is distinguished from MS by lesion pattern and lesion size. Typical MS-related demyelinating lesions are small (mainly less than 10 mm), often with an ovoid appearance, whereas BCS lesions are usually much larger.[6] According to the literature case reports, BCS usually occurs with a solitary lesion. The difference in our case is that BCS presents with many large-sized lesions bilaterally.

On computed tomography (CT) images, some patients may have normal findings; some may have low attenuation lesions. BCS usually shows 1 or more lesions typically located in the white matter layer of both cerebral hemispheres. Recent studies indicate that there is an increase in concomitant BCS and MS lesions. There is a theory that they can represent a continuous process of the same disease.[2,5,6]

MRI is the most effective modality used to diagnose BCS with the characteristic of different intensified concentric rings representing alternating areas of demyelination and myelination. Acute reactions involving demyelination demonstrate hyperintense concentric rings on T2W but hypointense on T1W, while preserved myelinated zones appear as isointense rings on both sequences. Several studies indicate that a peripheral enhancement on T1W post-contrast of the lesions at sites of the increased blood-brain barrier is responsible for active demyelination.[1,7,8] Massive MS lesions are considered a tumefactive lesion or pseudotumor on MRI with hyperintensity on T2W and FLAIR images. Similar to MS, an acute lesion in BCS may demonstrate restricted diffusion on DWI and ADC images. MRI reveals more findings in BCS and MS than CT's.[4,7,9] MRS is very important, it is the key to help diagnose BCS,[1] just like in our case.

Hardy TA recommended that testing for AQP4-IgG and MOG-IgG should be performed when an atypical pseudotumoral demyelinating lesion is identified, and brain biopsy may be avoided if there is close clinical and radiological follow-up.[8,10] In

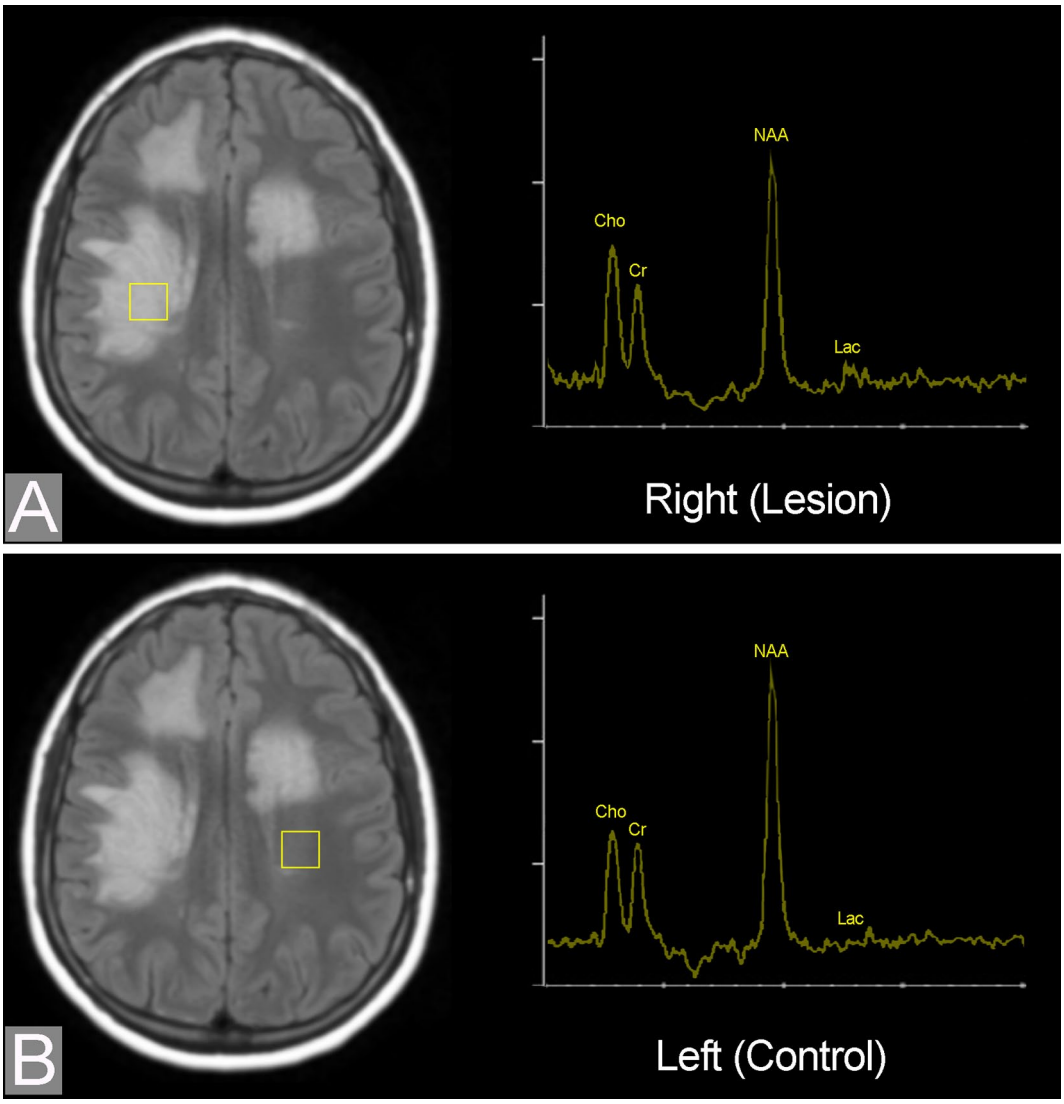

Figure 2. Single-voxel MRS of BCS: (A) spectroscopy imaging of the lesion reveals choline peak (Cho) slightly increased; *N*-acetylaspartate peak (NAA) decreased; small lactate doublet peak (Lac) slightly increased, (B) normal spectroscopy imaging of the contralateral side to compare.

our case, AQP4-IgG and MOG-IgG were not tested because laboratory equipment was unavailable. Fortunately, the BCS lesion found in MRI is typical, so that our patients evaded brain biopsy.

Due to the small number of patients, there are no effective therapeutic trials for BCS. In some reviews or case reports, patients are usually treated with high-dose intravenous steroids in the acute phase and tapered down gradually. Some authors recommend methylprednisolone 500 to 2000 mg/d IV for 3 to 10 d. Tapering doses of oral steroids following IV steroid treatment usually leads to good clinical responses. The majority of patients have excellent recovery following IV steroids.[11,12]

In some patients with poor or no response to steroids, other treatments such as plasma exchange, IV immunoglobulin, cyclophosphamide, and immunosuppression could be considered.[8-10] Plasma exchange therapy has been studied but applied only to worsen or incompletely recovered patients. However, a recent retrospective study indicates that it is not really effective against BCS disease.[13] Plasma exchange, rituximab, and cyclophosphamide have been used for expanding lesions causing physical disability, but the effect of these drugs is quite slow.[1,5,14]

Longer-term immunotherapy can be necessary for some patients to prevent relapse. The factor that predicts relapse in Baló-like lesions is oligoclonal bands of IgG restricted in the cerebrospinal fluid. If the serum AQP4-IgG or MOG-IgG is positive then chronic immunosuppression with prednisolone and other agents, such as rituximab or mycophenolate, should be considered. Some case reports revealed poor results of Baló-like lesions with various therapies. Therapy with a B-lymphocyte-depleting agent such as rituximab or ocrelizumab proved to be effective for MS and other atypical demyelinating conditions as BCS. Natalizumab and glatiramer acetate are similarly effective in case reports.[10,14]

Immune modulation therapy may also be combined with steroids therapy. Although BCS lesions could be life-threatening

according to earlier literature, recent cases and our case show an apparent, complete recovery, which may be the result of early diagnosis and favorable response to steroids. Rarely, decompressive hemicraniectomy may be required in cases with profound mass effect and potential herniation.[10-15]

Conclusion

BCS's essential features are interleaving circles of demyelinated and myelinated axons. Autopsy was used to diagnose BCS historically, but recently, MRI has become the conventional diagnostic method in the absence of pathological confirmation.

Author Contributions

The authors contributed equally.

Informed Consent

Written informed consent was obtained from the patient for publication of this case report.

Ethical Approval

This report was prepared in accordance with the ethical standards of the institutional ethics committee and with the 1964 Helsinki Declaration. Our institution does not require ethical approval for reporting individual cases or case series.

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