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Case report

Baló's concentric sclerosis with spontaneous remission and favorable prognosis

Yin-Xi Zhang [a], Gao-Li Fang [b], Jin-Long Tang [c], Qi-Lun Lai [d,*]

[a] Department of Neurology, Second Affiliated Hospital, School of Medicine, Zhejiang University, Hangzhou, 310009, China
[b] Department of Neurology, Zhejiang Chinese Medicine and Western Medicine Integrated Hospital, Hangzhou, 310003, China
[c] Department of Pathology, Second Affiliated Hospital, School of Medicine, Zhejiang University, Hangzhou, 310009, China
[d] Department of Neurology, Zhejiang Hospital, Hangzhou, 310013, China

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ABSTRACT

Introduction: Baló's concentric sclerosis (BCS) is a rare type of central nervous system demyelinating disorder. Most patients with BCS are treated with corticosteroids, and spontaneous remission has seldom been described.
Case presentation: A 46-year-old man presented with a subacute-onset headache and memory loss. Brain magnetic resonance imaging (MRI) revealed multiple onion-shaped ring lesions with mild enhancement in the outermost ring. A brain biopsy revealed significant myelin loss. The diagnosis of BCS was established based on the MRI results and pathological findings. Interestingly, the patient recovered almost completely without immunotherapy, with repeated brain MRI at the 1-year follow-up showing an obvious reduction in the extent of the lesions.
Conclusion: Neurologists should improve the recognition of the typical MRI features of BCS to avoid unnecessary biopsies. Although rare, spontaneous remission can be observed in clinical practice.

1. Introduction

Baló's concentric sclerosis (BCS) is a rare type of central nervous system (CNS) demyelinating disorder. Some patients may be misdiagnosed with brain tumors and undergo brain biopsy or surgical resection. Patients with BCS generally present with either acute or subacute neurological deterioration, and most require immunotherapy [1,2]. Few reports on spontaneous remission of BCS have been published. Herein, we report the case of a patient diagnosed with BCS who achieved almost complete recovery without immunotherapy.

2. Case presentation

A 46-year-old man presented to our hospital with subacute-onset headache and memory loss for 4 months, with significant deterioration including the development of disorientation and incoherent speech half a month prior. He denied having fever or any history of malignancy. Neurological examination on admission revealed only mild cognitive deficits, including calculation and memory impairments. Initial laboratory studies, including complete blood count, basic metabolic and immunological panels, and

* Corresponding author. Department of Neurology, Zhejiang Hospital, No.12 Lingyin Road, Hangzhou, 310013, China.
E-mail address: laiqilun@126.com (Q.-L. Lai).

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tumor markers, were unrevealing. Chest computed tomography and lymph node ultrasonography revealed no abnormalities. Brain magnetic resonance imaging (MRI) revealed multiple onion-shaped ring lesions with mild enhancement in the outermost ring (Fig. 1A–D). Magnetic resonance spectroscopy (MRS) of the right frontal lesion revealed a significant increase in the choline-to-N-acetyl-aspartate (Cho/NAA) ratio (10.7), as well as a moderate increase in the lactate peak (Fig. 1E). Cerebrospinal fluid (CSF) analysis revealed positive unmatched oligoclonal bands, with normal pressure, cell counts, and protein levels. No evidence of infection was detected in the serum or CSF. Cell-based assays to test for serum and CSF aquaporin-4 and myelin oligodendrocyte glycoprotein antibodies were negative.

Histopathological examination of the brain tissue obtained from the right frontal lesion demonstrated significant myelin loss, and infiltrated with a large number of foam-like histiocytes and scattered reactive astrocytes (Fig. 2A–D). The diagnosis of BCS was established based on the MRI appearance and pathological findings, and intravenous methylprednisolone followed by a tapering scheme was recommended. However, the patient refused to receive immunotherapy. Interestingly, the patient's clinical symptoms gradually resolved without treatment. Six months later, the patient had almost fully recovered and returned to work. At the 1-year follow-up, the patient remained stable, and repeated brain MRI revealed an obvious reduction in the extent of the lesions (Fig. 1F). At 2-year follow-up, no recurrence was reported.

3. Discussion

The classical manifestations of BCS are similar to those of intracerebral mass lesions and include headache, cognitive deficits, behavioral changes, muteness, urinary incontinence, seizures, aphasia, and hemiparesis [2]. Most patients are young adults, with onset generally occurring between 20 and 50 years of age, while a slight female predominance has also been observed [2].

Alternating rings of myelin preservation or remyelination and myelin loss, commonly localized in the supratentorial regions, are characteristic pathological findings of BCS [3]. Initially, BCS could only be diagnosed on autopsy [1,3]. However, early diagnosis can now be made using non-invasive techniques, while the distinctive onion-ring or whorled appearance helping distinguish it from other demyelinating lesions [2]. In a recent cohort of patients with BCS reported by Jolliffe et al. [4], brain biopsy/autopsy were performed

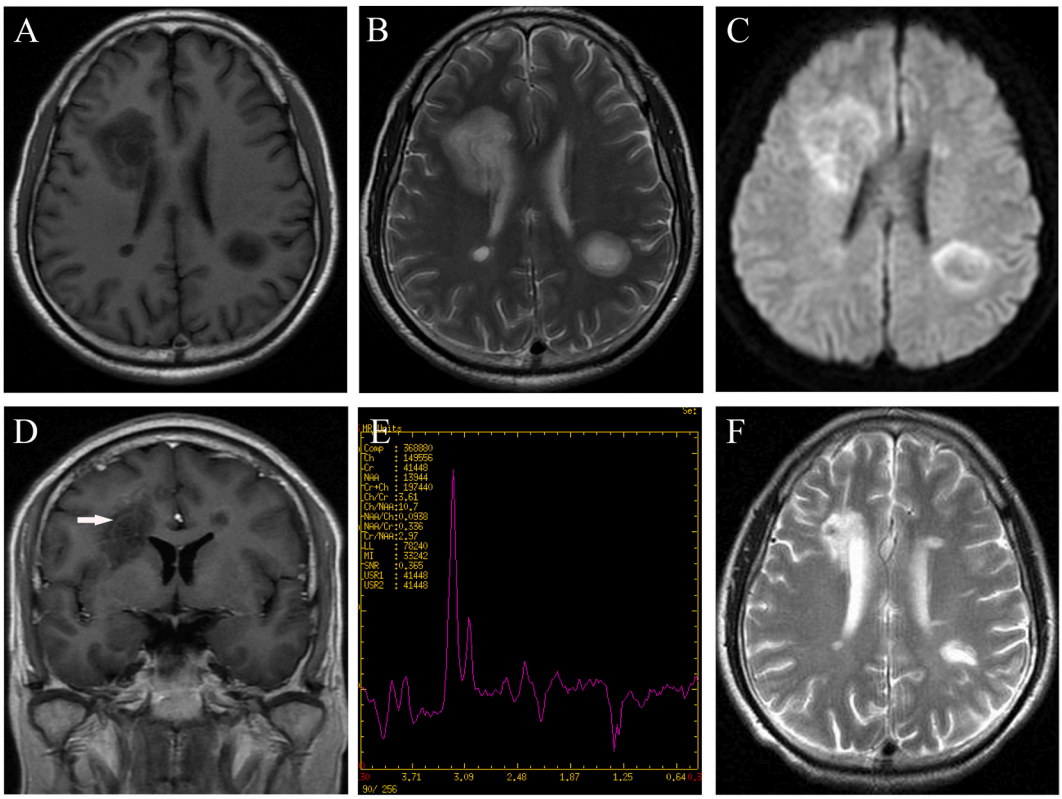

Fig. 1. Brain magnetic resonance imaging findings of the patient. (A–D) Brain magnetic resonance imaging (MRI) showing multiple T1-hypointense and T2-hyperintense multilayered ring-like lesions in the white matter, with diffusion restriction and mild enhancement appearing (white arrow) in the outermost ring at disease onset. (E) MR spectroscopy of the right frontal lesion showing a significant increase in the choline-to-N-acetyl-aspartate ratio, and a moderate increase in the lactate peak. (F) Brain MRI showing an obviously reduced extent of the lesions at 1-year follow-up. (A) Axial T1-weighted imaging, (B, F) Axial T2-weighted imaging, (C) Axial diffusion-weighted imaging, (D) Coronal gadolinium-enhanced T1-weighted imaging, and (E) MR spectroscopy.

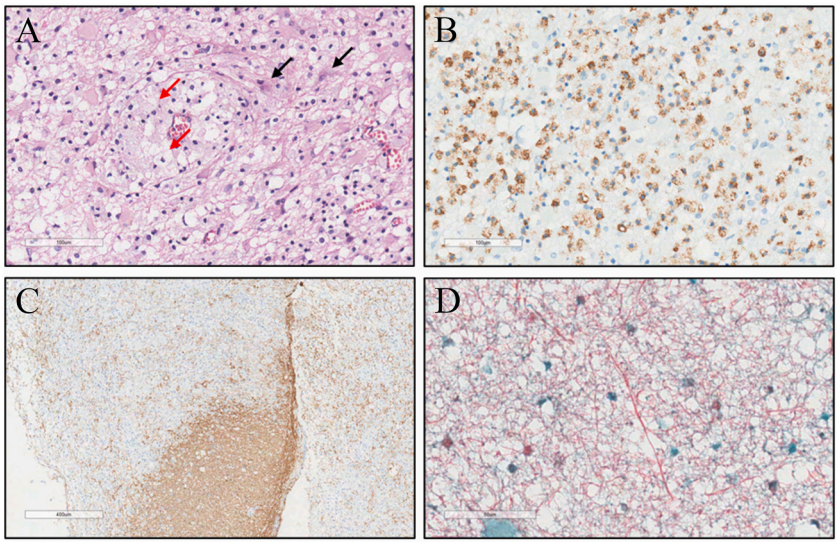

Fig. 2. Brain pathological findings of the patient. (A) Hematoxylin-eosin staining of the brain tissue showing a large number of foam-like histiocytes (red arrow) infiltrating the brain tissue, with scattered reactive astrocytes (black arrow). (B) Immunohistochemical staining for CD68 confirming the extensive histiocyte infiltration. Myelin basic protein staining (C) and chromotropic acid 2R staining (D) showing significant myelin loss.

in only 17/40 patients, and the diagnosis of BCS was therefore based on clinical manifestations and classical MRI features in most patients. For patients with typical MRI manifestations, brain biopsy can be avoided; however, serial MRIs must be performed as follow-up.

MRS of BCS lesions generally revealed moderately increased Cho/NAA ratios, with relatively decreased NAA levels [5]. In contrast, our patient showed a marked increase in Cho/NAA ratio, as well as a moderate increase in the lactate peak, raising the suspicion of a primary or secondary brain neoplasm. Hence, a brain biopsy was performed, and alternative causes, including neoplastic disorders, and histiocytic diseases (such as Langerhans cell histiocytosis), were excluded based on the pathological findings. If a BCS lesion can be diagnosed based on the typical MRI appearance, then a brain biopsy is not required; however, when a lesion is atypical or has certain characteristics indicating the possibility of malignancy, a brain biopsy is necessary.

Given its low prevalence, there is currently no consensus on treatment for BCS. Based on case reports and clinical experience, corticosteroids are the recommended first-line therapy during the acute phase, while other treatment options include plasma exchange and intravenous immunoglobulin [2]. However, our patient refused to receive any of these immunotherapies. The prognosis of BCS has long been considered poor, with a progressive and fatal course lasting several weeks or months [6]. However, as the number of patients with BCS is increasing, the prognosis of this condition has been shown to vary considerably. In one pooled analysis of 68 reported cases, 50 (73.5 %) patients showed a good response to therapy, achieving complete clinical recovery or only mild residual deficits, with no significant effect on daily life [7]. Although more patients with BCS have tended to have a good prognosis in recent years, spontaneous remission without immunotherapy, as demonstrated in our patient, is rare, with only one patient being previously described [8]. The present patient has remained stable and relapse-free during the 2-year follow-up so far, although further follow-up is needed to assess the long-term outcomes.

The relationship between BCS and multiple sclerosis still needs to be determined, with further research needed to determine whether BCS is a rare type of multiple sclerosis, or a distinctive disease entity [9,10]. BCS is generally considered a monophasic disease; however, some patients relapse or develop multiple sclerosis. In a recent cohort of patients with BCS, 50 % (9/18) relapsed during follow-up [4]. Multiple sclerosis disease-modifying therapy may be considered in BCS patients displaying the dissemination in time and space defined in the diagnostic criteria for multiple sclerosis or who experience clinical relapse [2]. Our patient remained stable and relapse-free during the 2-year follow-up; however, further follow-up will be needed to assess his long-term outcomes.

4. Conclusion

BCS lesions are characterized by a distinctive onion-ring or whorled appearance on MRI. Improving the recognition of this characteristic MRI appearance can sometimes prevent unnecessary biopsy, but not in patients with atypical findings. Although rare, the favorable prognosis of our patient indicated that spontaneous remission can occur in some BCS patients.

Ethics statement

Informed consent was obtained from the patient for the publication of all images/data. The case report was reviewed and approved by the Ethics Committee of Zhejiang Hospital for Human Research (approval number: 2024-CA-01).

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Data availability statement

Data will be made available on request.

CRediT authorship contribution statement

Yin-Xi Zhang: Writing – review & editing, Writing – original draft, Investigation, Formal analysis, Data curation, Conceptualization. **Gao-Li Fang:** Writing – review & editing, Investigation, Formal analysis, Data curation. **Jin-Long Tang:** Writing – review & editing, Investigation, Formal analysis, Data curation. **Qi-Lun Lai:** Writing – review & editing, Validation, Supervision, Investigation, Formal analysis, Data curation, Conceptualization.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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