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Balo's concentric sclerosis: a case report

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ABSTRACT

Balo's concentric sclerosis is a rare variant of multiple sclerosis which is characterized by lesions consisting rings of demyelination alternating with areas of relatively preserved myelin. These pathological changes are reflected by characteristic magnetic resonance imaging findings. We present the imaging findings of a case of BCS who presented with an acute neurological disturbance, showed a dramatical recovery following corticosteroid therapy, and remained free of relapse for more than 1 year. Although previously considered as a fulminant and fatal disease, characteristic MRI findings of Balo's concentric sclerosis enable earlier diagnosis and treatment, and better prognosis.

Keywords: Balo's concentric sclerosis, multiple sclerosis, demyelination

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Balo's concentric sclerosis (BCS) is a rare variant of multiple sclerosis which is characterized by a lesion formed by alternating rings of demyelination and relatively preserved myelin, reminding an onion ring appearance. Previously BCS was regarded to be an invariably fatal disorder. However, with the advent of magnetic resonance imaging (MRI), the earlier diagnosis and management of the disease offered a better prognosis [1]. On MRI, acute BCS lesions are seen as T2-hyperintense lamellae surrounding a T2-hyperintese core which is called 'storm centre'. The areas of active demyelination within the lesion show restricted diffusion, and the peripheral aspect of the lesion enhances following gadolinium administration [2]. Correct characterization of the lesion enables accurate diagnosis and allows prompt treatment of BCS. Most of the patients who develop symptomatic BCS are shown to make a substantial or complete recovery following convenient treatment [1]. We herein present the imaging findings of a case of BCS who presented with an acute neurological disturbance, showed a dramatical recovery following corticosteroid therapy, and remained free of relapse for more than 1 year.

CASE PRESENTATION

A previously healthy 25-year-old woman presented with acute onset of headache, dysarthria and bilateral upper extremity weakness. MRI demonstrated two round lesions with well-defined borders in the right frontoparietal region and in the corpus callosum (Figure 1). The lesions showed a concentric lamellar appearance on all pulse sequences. The concentric rings were; hypointense and isointense on T1-weighted images (Figures 2a and 3a), hyperintense and isointense on T2-weighted images (Figures 2b and 3b). There was neither surrounding edema nor mass effect accompanying the lesions. On diffusion weighted imaging (DWI); the right

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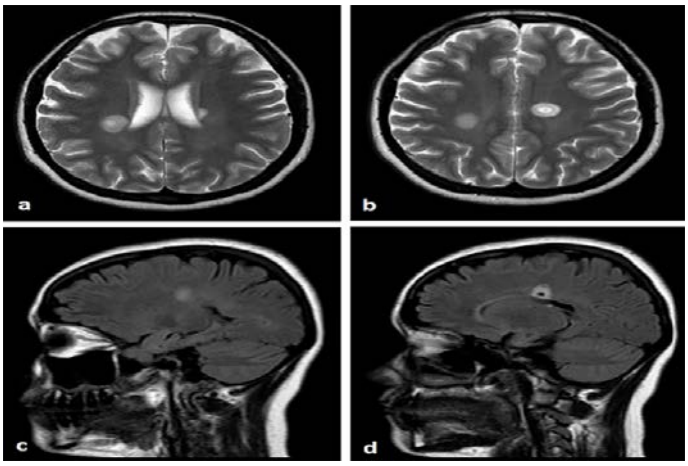

Figure 1. Axial T2-weighted (a, b) and sagittal FLAIR (c, d) images demonstrate a right frontoparietal (a, c), and a callosal (b, d) lesion which show a concentric lamellar appearance, consistent with Balo's concentric sclerosis.

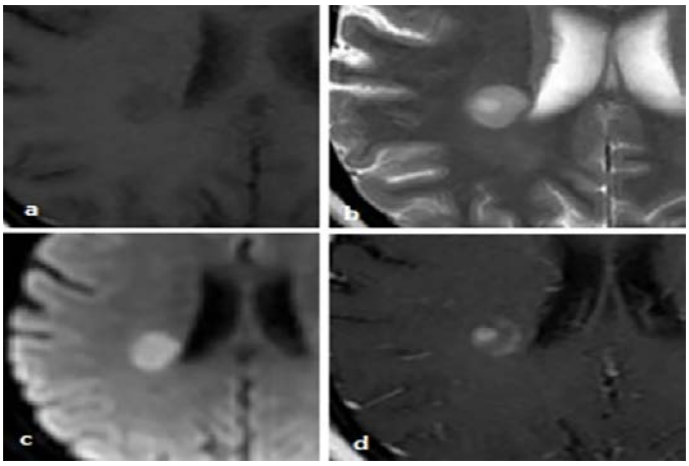

Figure 2. The right frontoparietal lesion is iso/hypointense on T1-weighted (a), iso/hyperintense on T2-weighted (b), hyperintense on diffusion weighted (c) images, and its T2-hyperintens areas show enhancement after gadolinium administration (d).

frontoparietal lesion was completely hyperintense (Figure 2c), whereas hypointense central core of the left callosal lesion was surrounded by a hyperintense ring (Figure 3c). T1 hypointense rings of the right frontoparietal lesion showed enhancement following gadolinium administration (Figure 2d), whereas the callosal lesion showed a minimal enhancement which was difficult to distinguish (Figure 3d). The whole spine MRI was normal. The patient was diagnosed as having BCS and given a high-dose oral corticosteroid therapy. She showed a significant improvement in her clinical symptoms in the first 7 days following the treatment and a complete clinical recovery was obtained in a 2 months' time.

Nine months after the initial MRI, a follow-up MRI was performed. Both lesions exhibited remarkable reduction in size. The lamellae seemed shrunken on both T1 and T2-weighted images (Figure 4). T2-hyperintense rings changed to be focal, milimetric, irregular hyperintense areas (Figure 4b). There was neither contrast enhancement nor restriction of diffusion in either lesions. The repeat whole spine MRI was normal. The patient has had no more clinical relapses at 12 month follow-up and is presently asymptomatic.

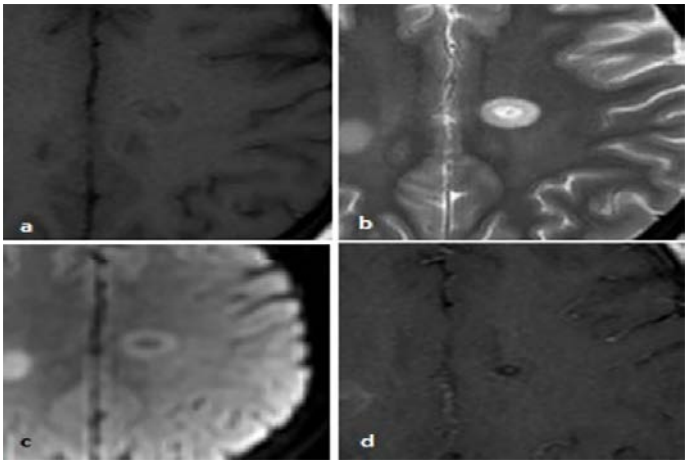

Figure3. Callosal lesion is iso/hypointense on T1-weighted (a) and iso/hyperintense on T2-weighted (b) images. On diffusion weighted image (c), hypointense central core of the lesion is surrounded by a hyperintense ring. Following gadolinium administration, the lesion shows a minimal enhancement which is difficult to distinguish (d).

DISCUSSION

Cerebral white matter oligodendrocyte loss and demyelination are histopathological characteristics of BCS. Cortical grey matter is typically reserved. The lesions characteristically show alternating lamellar pattern of demyelinated and relatively preserved white matter, reminding a union bulb appearance. They usually present as a solitary mass, less commonly, multiple lesions may be detected. BCS lesions occur predominantly in the cerebral white matter, but also basal ganglia, pons, and cerebellum lesions have been reported [3, 4].

With an average age at onset of 34 years (range;

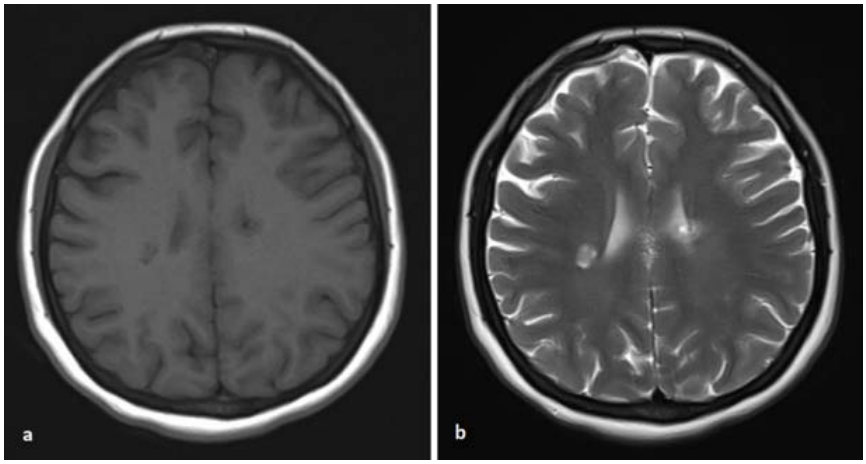

Figure 4. Follow-up MRI shows remarkable reduction in size of both lesions. The lamellae seems shrunken on both T1 (a) and T2-weighted (b) images. T2-hyperintense rings of acute stage appears to be changed to focal, milimetric, hyperintense areas (b).

3-62 years), BCS mainly occurs in young adults. The most common symptoms of the entity are similar to those of any intracerebral mass: headache, seizures, cognitive deficiency, behavioral changes, aphasia, hemiparesis and urinary incontinence. Some patients with BCS can present with classic focal symptoms of multiple sclerosis, [1]. Similar to the most of the previously reported cases of BCS, our patient presented with headache, dysarthria, and bilateral upper extremity weakness.

Histopathological characteristics of the BCS lesions are typically reflected on MRI imaging. In acute lesions; T1-weighted images show alternating isointense (areas of preserved myelin) and hypointense (areas of demyelination) concentric rings. In a similar fashion, T2-weighted images show hyperintense rings (areas of demyelination) surrounding a T2 hyperintense core. In the sites of demyelination rings, increased blood brain barrier permeability and inflammation causes enhancement following gadolinium administration [1, 2]. Both lesions of our patient showed these characteristic findings of an acute BCS lesion, except for the relatively reduced gadolinium enhancement in the callosal lesion. This might be due to either the older age of this lesion than the right frontoparietal one, or relatively preserved blood brain barrier permeability (Figure 3d). In the follow-up MRI, neither of the lesions enhanced, suggesting the lack of active inflammation within the lesions.

DWI can be used for the confirmation of the presence of BCS. On DWI, areas of active demyelination show restricted diffusion, whereas myelinated areas show fascilitated diffusion. Thus, on DWI, BCS lesions demonstrate alternating rings of high and low signal intensity, representing demyelinated and myelinated areas, respectively [2, 5, 6]. In our case; the right frontoparietal lesion was completely hyperintense on DWI (Figure 2c), whereas hypointense central core of the left frontal lesion was surrounded by a hyperintense ring (Figure 3c), probably reflecting the two different stages in which the two lesions were.

The characteristic magnetic resonance spectroscopy pattern of acute BCS lesions is; a decreased N-acetylaspartate peak (reflecting neuronal damage), elevated choline and lipid peaks (suggesting increased membrane turn-over), and elevated lactate peak (consistent with impaired aerobic metabolism) [7, 8]. On fluorodeoxyglucose positron emission tomography (FDG-PET), BCS lesions do not show increased uptake, while the main differentials of BCS including other acute demyelinating lesions and high-grade neoplasms do. This makes FDG-PET a useful adjunct to MRI in characterization of equivocal lesions [1, 8].

For the treatment of acute BCS lesions, corticosteroids and plasma exchange are recommended as the first and second line therapy respectively. Although some authors recommend multiple sclerosis disease-modifying therapy as an ongoing therapy in patients who have developed a

BCS lesion, a standard ongoing treatment guide has not been established yet. The prognosis of the patients with BCS lesion varies. Complete clinical and radiological recovery is possible; however, aggressive BCS lesions could be fatal. [1, 9]. Fortunately, after the corticosteroid therapy, our patient showed a dramatical recovery and remained asymptomatic for the following 1 year.

CONCLUSION

Although previously considered as a fulminant and invariably fatal disease, characteristic MRI findings of BCS enable earlier diagnosis and treatment, and better prognosis.

Informed consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of interest

The authors declared that there are no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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