

Case report

MRI features of Balo's concentric sclerosis

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Abstract. We report MRI findings in a 56-year-old woman with Balo's concentric sclerosis (BCS) who initially presented with a progressive hemiparesis. MRI showed two lesions with a concentric pattern in the left frontoparietal region and a laminated, arcuate pattern in the right frontal region. These patterns were best seen in post-contrast images and were consistent with BCS. In addition, there were several small cerebral multiple sclerosis-like plaques. The clinical symptoms improved and the MR findings regressed after corticosteroid therapy. The patient had completely recovered 12 months later, except for mild right hand numbness. MRI showed further regression of the lesions, but the concentric pattern was still present. This case demonstrated that BCS can run a benign prolonged course and may persist for a long time. Concentric or laminated contrast enhancement in the acute phase may suggest that bands of demyelination in BCS occur synchronously rather than successively.

Balo's concentric sclerosis (BCS) is a rare demyelination disorder characterized pathologically by concentric lesions of alternating demyelinated and myelinated bands in the white matter [1, 2]. Most cases of BCS have been diagnosed on post-mortem examination. However, several cases have been diagnosed antemortem by MRI [3–10]. We describe MR findings in a patient with BCS exhibiting distinctive contrast enhancement patterns, and discuss the relationship with the clinical course and pathogenesis.

Case report

A 56-year-old woman was admitted to hospital with a 2-week history of progressive weakness in the right limbs, especially the leg. Physical examination showed mild right central facial palsy and right hemiparesis. Slight slurring of speech and gait disturbances were also noted. There were no sensory disturbances. MRI (Figure 1) showed two large contiguous lesions with vague concentric architecture in the left frontoparietal region. There was mild perifocal oedema and local mass effect. Another wedge-shaped lesion with a lamellar pattern was noted in the right frontal subcortical region. In addition, there were several plaque-like lesions in both thalami, the right basal ganglion and the periventricular area on proton density and T_2 weighted images. After intravenous administration of gadolinium, the nodule in the left frontoparietal region

exhibited conspicuous concentric ring enhancement, and the right frontal subcortical lesion demonstrated laminated enhancement. Some of the plaque-like lesions also showed dot enhancement (Figure 1). Based on the MR findings, a presumptive diagnosis of BCS was made. Stereotactic CT-guided biopsy of the largest concentric lesion showed mild gliosis, foamy histiocytes and perivascular cuffing of lymphocytes, compatible with an acute demyelinating process.

The patient was initially treated with oral prednisolone 60 mg day⁻¹. The right hemiparesis markedly improved after 2 weeks of treatment. The gait disturbances, slurred speech and left central facial palsy completely recovered. The dose of prednisolone was then gradually reduced to 30 mg day⁻¹. The right hemiparesis had resolved within 1 month of treatment. The second MR scan showed a reduction in the size and number of the lesions. The gadolinium enhancement of the lesions also decreased, and only slight peripheral enhancement was seen in the left frontoparietal lesions. The prednisolone dose was further reduced to 5 mg day⁻¹. 3 months after treatment, the patient developed numbness and clumsiness of the right hand. The third MR scan showed further reduction in size of the white matter lesions in the left frontoparietal regions with loss of contrast enhancement. However, a new, small, non-enhancing lesion in the left midbrain was found. Prednisolone therapy was resumed at 15 mg day⁻¹. The clumsiness gradually resolved within 1 year of treatment and the numbness slightly improved. The fourth MRI showed further regression of the white matter lesions but the concentric pattern of the left parietal lesion was still present (Figure 2).

Received 2 October 1998 and accepted 23 November 1998.

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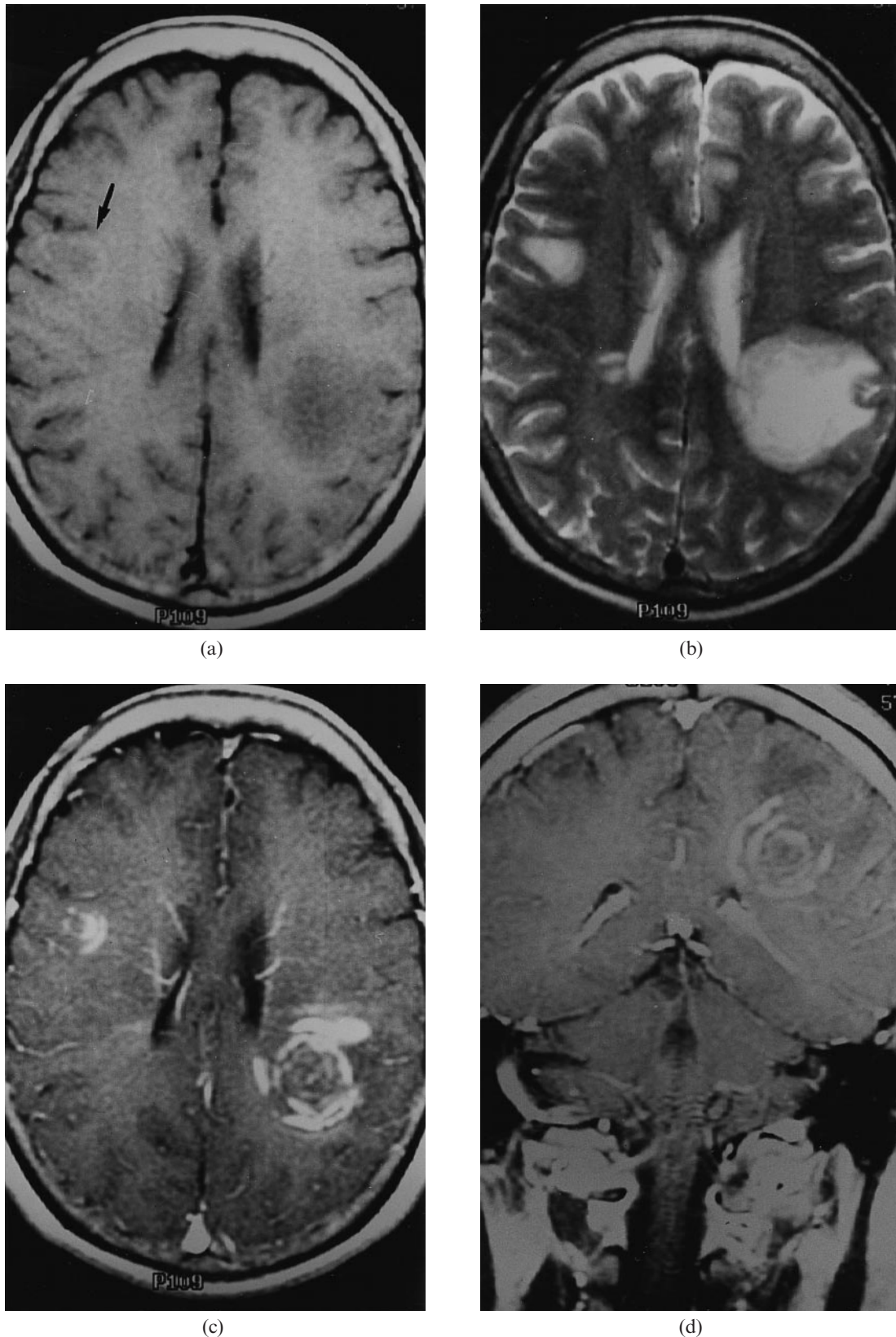


Figure 1. MRI 17 days after the onset of symptoms. (a) Axial T_1 weighted image and (b) T_2 weighted image shows nodular lesions in the left parietal and right frontal subcortical region (arrow). Two small plaques were also noted in the right periventricular region. (c) After contrast enhancement, axial T_1 weighted image shows enhancement of concentric rings in the left frontoparietal lesions, laminated arcuate enhancement of the right frontal subcortical lesion, and dot enhancement of the right periventricular lesions. (d) The striking concentric enhancement of the left parietal lesion is also seen on the coronal contrast enhanced T_1 weighted image.

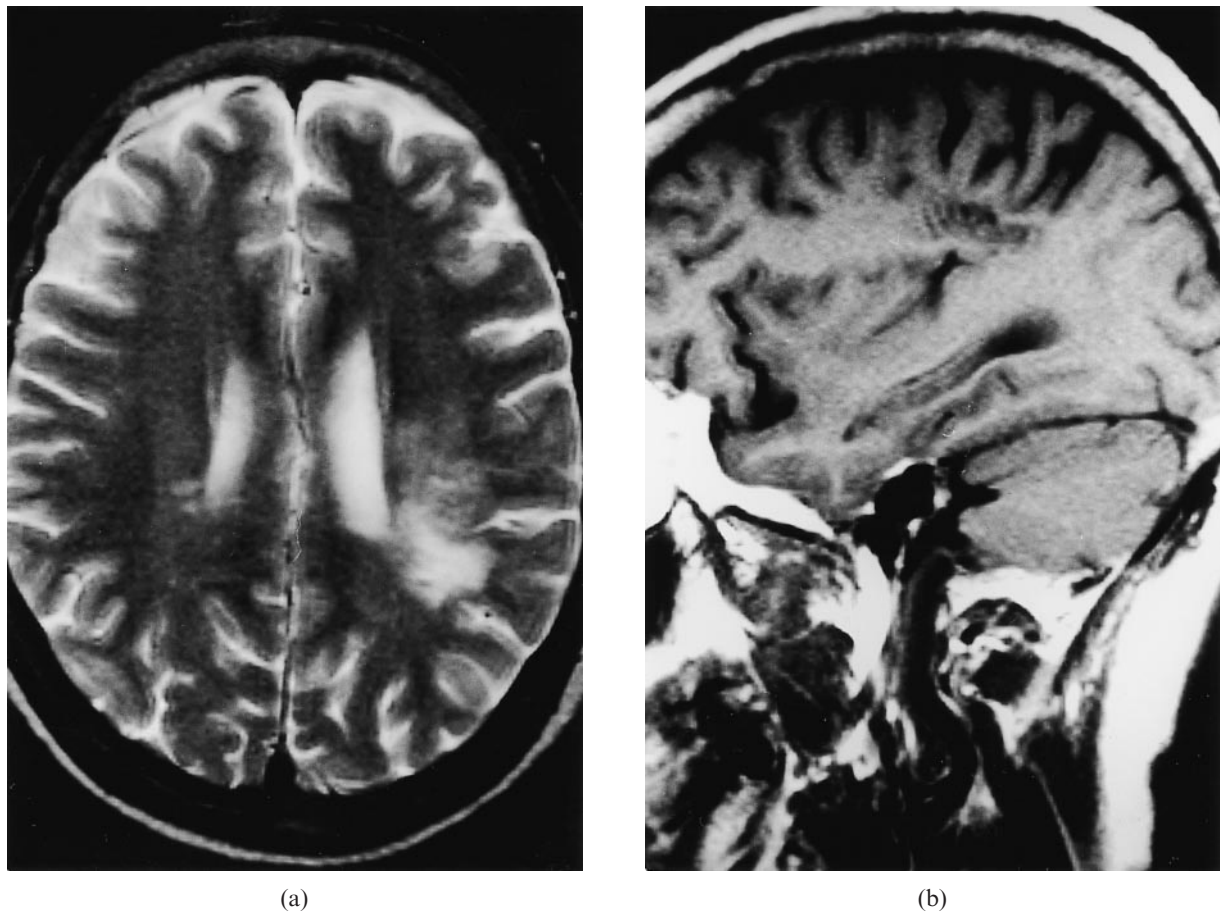


Figure 2. Follow-up MRI 12 months later. (a) Axial T_2 weighted image and (b) sagittal T_1 weighted image show that the concentric pattern of the left parietal lesion is still present.

Discussion

BCS is a rare demyelinating disease. The pathogenesis of the concentric bands remains controversial. Moore et al [11] proposed that these bands may represent areas of remyelination at the borders of successive episodes of acute demyelination. The centre of the lesion is the oldest area, with the concentric rings of demyelination decreasing in age with increasing distance from the centre. However, Yao et al [12] suggested that the myelinated bands may represent early stages of demyelination rather than remyelination.

BCS had been thought to have a slightly higher prevalence among young males and to run an acute, rapidly progressively fatal course [1, 2]. Recently, several patients with a non-fatal form of BCS and the characteristic MR features have also been reported [3–6, 8, 10]. These patients, like the one presented here, remained alive for many months or years after the diagnosis. This may imply that many cases of BCS with a benign course were not recognized before the advent of MRI. Thus, the clinical course of BCS is variable and not so fulminant as previously thought. Clinical and MRI improvement followed prednisolone therapy in this case, although an exacerbation occurred when prednisolone was being tapered.

Of the nine cases of BCS diagnosed by antemortem examinations [3–10, 13], seven were diagnosed solely by the characteristic concentric pattern on MRI [3, 5–10]. In these cases, MRI showed that BCS had concentric hypointense bands on T_1 weighted images and hyperintense bands on T_2 weighted images, alternating with isointense white matter. The hyperintense signals on T_2 weighted images corresponded to concentric bands of demyelination with gliosis and perivascular lymphocytic infiltration. The areas of isointense white matter bands on T_2 weighted images represented spared white matter or areas of remyelination [7]. After gadolinium administration, the lesion shows peripheral contrast enhancement or garland-like enhancement [4, 6, 8–10]. In our case, striking concentric ring enhancement and laminated arcuate enhancement were well demonstrated in the left frontoparietal lesions and the right frontal lesion, respectively. Contrast enhancement in the brain indicates blood–brain barrier disruption and acute disease activity. Thus, concentric ring enhancement of the lesion may reflect that active demyelination in the concentric bands occurs simultaneously. In the light of the present case, bands of demyelination in BCS are likely to occur synchronously rather than successively.

Over time, the MR abnormalities of BCS decrease in size and do not enhance with contrast medium. The duration of the characteristic concentric pattern is variable, ranging from 6.5 weeks to 1 year [3, 6, 8, 9]. Bolay et al [9] suggested that the concentric pattern might be a transient feature in the development of BCS. However, this description does not concur with the findings in our case, where the concentric bands were still visible on MRI more than 1 year after diagnosis.

There is still controversy about whether BCS is a separate demyelinating disease from multiple sclerosis [2, 5, 7, 8]. Interestingly, both concentric lesions and multiple sclerosis-like plaques were found in the MR images of our patient. Another small, new lesion developed in the brainstem during reduction of prednisolone therapy. These findings support the proposal by Yao et al [12] that BCS is a rare variant of multiple sclerosis.

References

1. Balo J. Encephalomyelitis periaxialis concentrica. *Arch Neurol Psychiatry* 1928;19:242-64.
2. Couvrille CB. Concentric sclerosis. In: Vincken PJ, Bruyn GW, editors. *Handbook of clinical neurology*, Vol. 9. Amsterdam: North Holland, 1970:437-51.
3. Spiegel M, Kruger H, Hoffmann E, Kappos L. MRI study of Balo's concentric sclerosis before and after immunosuppressive therapy. *J Neurol* 1989; 236:487-8.
4. Hanemann CO, Kleinschmidt A, Reifanberger G, Freund HJ, Seitz RJ. Balo's concentric sclerosis followed by MRI and positron emission tomography. *Neuroradiology* 1993;35:578-80.
5. Miyata K, Itoyama Y, Kobayashi T, Yasumori K, Goto I. A case of demyelinating disease showing peculiar honeycomb-like and lamellar structure on MRI. *Rhinsho Shinkeigaku* 1990;30:402-6.
6. Korte JH, Bom EP, Vos LD, Breuer TJM, Wondergem JHM. Balo's concentric sclerosis: MR diagnosis. *Am J Neuroradiol* 1994;15:1284-5.
7. Gharagozloo AM, Poe LB, Collins GH. Antemortem diagnosis of Balo's concentric sclerosis: correlative MR imaging and pathological features. *Radiology* 1994;191:817-9.
8. Chen CJ, Ro LS, Wang LJ, Wong YC. Balo's concentric sclerosis: MRI. *Neuroradiology* 1996; 38:322-4.
9. Bolay H, Karabudak R, Tacal T, Onol B, Selekler K, Saribas O. Balo's concentric sclerosis: report of two patients with magnetic resonance follow-up. *J Neuroimaging* 1996;6:98-103.
10. Chen CJ, Ro LS, Chang CN, Ho YS, Lu CS. Serial MRI studies in pathologically verified Balo's concentric sclerosis. *JCAT* 1996;20:732-5.
11. Moore GRW, Neumann PE, Suzuki K, Lijtmaer HN, Traugott U, Raine CS. Balo's concentric sclerosis: new observations on lesion development. *Ann Neurol* 1985;17:604-11.
12. Yao DL, Webster H deF, Hudson LD, Brenner M, Liu DS, Escobar AI, et al. Concentric sclerosis (Balo): morphometric and in situ hybridization study of lesions in six patients. *Ann Neurol* 1994; 35:18-30.
13. Garbern J, Spence AM, Alvord EC Jr. Balo's concentric demyelination diagnosed premortem. *Neurology* 1986;36:1610-4.