Keywords: Concentric sclerosis; Magnetic resonance imaging

A 38-year-old woman had a one-month history of progressively dysphasia, dysphagia and mild cognitive impairment. Previous medical history was unremarkable. Neurologic examination revealed a pseudobulbar palsy with a moderate dysarthria and dysfunction of cranial nerves IX, X and XII. Laboratory investigations disclosed a normal complete blood cell count, biochemistry and the results of the serologic tests including antineutrophil cytoplasmic antibodies and antinuclear antibodies, besides an elevated C-active protein level (13.1mg/l) and an increased erythrocyte sedimentation rate (80 mm/hr). The results of cerebrospinal fluid examination were normal with an immunoglobulin G index of 0.412 and negative antinuclear antibodies, besides an elevated C-active protein level. The complete blood cell count revealed no evidence of hematologic malignancies. Cranial magnetic resonance imaging (MRI) demonstrated multiple lesions in a concentric ring pattern consisting of alternating layers of higher and lower signal intensity in the both centrum semiouale (Figure 1). The lesions were typically irregular concentric areas of iso and low signal in T1-weighted imaging (Figure 1a) while iso and high signal in T2-weighted imaging (Figure 1b). There was no or mild focal, peripheral enhancement in the lesions with contrast enhancement in the (Figure 1c). Restricted diffusion in the outer rings was demonstrated in axial diffusion-weighted imaging (Figure 1d). The typical pattern is consistent with a diagnosis of Baló’s concentric sclerosis (BCS), which was first described by Marburg in 1906, and was advanced by Józef Baló as a rare variant of multiple sclerosis in 1928 [1]. The patient was treated with intravenous methyl prednisolone (1000 mg/day) for 5 days followed by a reducing dose of oral prednisolone. Her condition improved dramatically. On follow-up examination two years later, she was completely recovered with normal neurological examination and no further relapses were reported. Unfortunately, no further MR images of this patient are available.

BCS occurs more in young adults and is more common in Asiatic populations than in Caucasoids. It is histopathologically characterized by larger white matter lesions with concentric alternating layers of myelin preservation and loss, which present with onion-like appearance with a minor mass effect on MRI [2,3]. It may occur as solitary lesions, along with the plaques typically seen in multiple sclerosis. In the differential diagnosis, other demyelinating lesions such as acute disseminated encephalomyelitis and gliomas should be considered. Clinicians used to regard its prognosis as similar to that of Marburg multiple sclerosis, which has a monophasic rapidly progressive course with a fatal outcome. Recently, an increasing number of cases have been described as having prolonged survival, spontaneous remission, or relapsing–remission [4,5].

References