

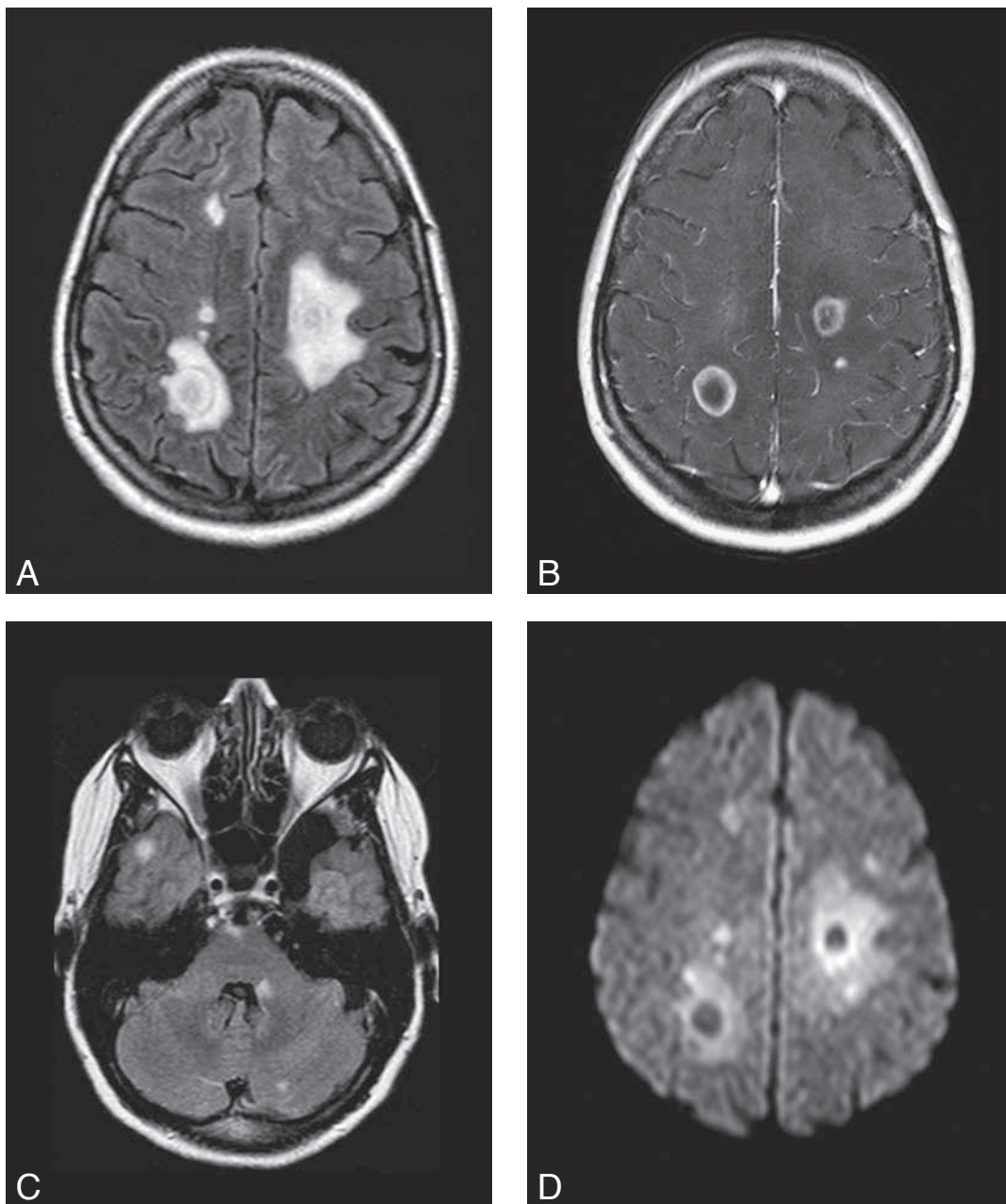
BALÓ'S CONCENTRIC SCLEROSIS

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Key-word: Sclerosis, concentric

Background: A 46-year-old woman was admitted with right-sided weakness, vertigo and nausea. Clinical examination revealed ataxia of the right limbs, diplopia and a counter-clockwise rotatory nystagmus.

MRI of the brain was performed.



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Fig.

1A	1B
1C	1D

Work-up

MRI of the brain (Fig. 1) shows on axial fluid attenuation inversion recovery, FLAIR image (A) two large, subcortical areas of vasogenic edema in the parietal white matter on both sides with hypo-intense concentric rings. There are three additional white matter lesions in the paraventricular region. Axial T1-weighted contrast-enhanced image (B) demonstrates extensive peripheral rim enhancement of the two parietal lesions. On axial FLAIR image (C), multiple white matter lesions in the brachium pontis are observed. Axial diffusion-weighted (DWI) image (D) indicates that there is no diffusion restriction.

Radiological diagnosis

A biopsy of the left contrast-enhanced parietal lesion was performed. Based on its histopathological examination and the MRI findings, *Baló's concentric sclerosis* was diagnosed. Additional neurophysiologic examination and cerebrospinal fluid analysis were consistent with multiple sclerosis.

Discussion

Baló's concentric sclerosis (BCS) was first described by Baló in 1928 as 'encephalitis periaxialis concentrica'. BCS is considered to be a rare variant of multiple sclerosis (MS). Most of the reported cases concerned patients between 20-50 years of age. BCS often has an acute onset and a rapidly progressive and monophasic clinical course. Most commonly reported symptoms include headache, aphasia, cognitive or behavioural dysfunction, hemiparesis, ataxia and seizures.

In the past, the diagnosis of BCS was only made at autopsy and it was therefore considered to be an invariably fatal disease. Since the advent of MRI, it has become increasingly clear that BCS exhibits characteristic radiographic findings, making ante-mortem diagnosis possible and suggesting that the disease may have a relatively benign course. Early

diagnosis may have a significant effect on morbidity and mortality associated with BCS.

BCS lesions are histopathologically characterized by alternating lamellae of demyelinated and normally myelinated white matter, resulting in the typical concentric rings.

There is a striking resemblance between this histopathological manifestation and the MRI features on T2-weighted images of a concentric pattern of hyperintense bands of demyelinated white matter and iso-intense bands of normal white matter. Gadolinium enhanced T1-weighted images show peripheral rim enhancement, suggesting a disruption of the blood brain-barrier and the presence of inflammatory white cells.

In the acute phase, lesions may be accompanied by edema and mass effect. Patients may or may not have additional multiple white matter lesions consistent with classic MS. Three of such white matter lesions were found in the paraventricular region and brachium pontis of our patient. Together with the two characteristic concentric lesions, MRI findings were in retrospect diagnostic for BCS and brain biopsy could have been avoided.

Clinical and MRI differential diagnosis includes acute disseminated encephalomyelitis (ADEM), infections, primary neoplasms and metastatic disease. However, none of these diseases exhibit the characteristic concentric rings of BCS.

Bibliography

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