Basic and advanced imaging of a case of Balò's concentric sclerosis

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DESCRIPTION

In January 2012 a 51-year-old woman was admitted to our department for a slow increasing right hemiparesis with right brisk deep tendon reflexes. She suffered from Sjogren syndrome, fibromyalgia and hepatitis B virus (healthy carrier).

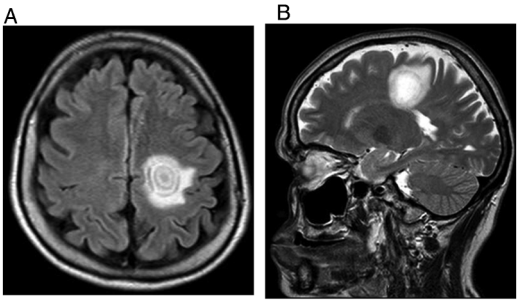

Figure 1 At diagnosis. (A) Axial fluid attenuated inversion recovery sequence and (B) sagittal T2-sequence evidence the typical concentric ring lesion in the white matter.

Conventional MRI scans (figure 1A,B) were typical for Balò's sclerosis.[1]

The patient rejected a brain biopsy. In order to better define the lesion, we therefore performed advanced radiological techniques such as diffusion, spectroscopy and perfusion.

In the diffusion sequences (figure 2A) a concentric ring of unrestricted diffusion appeared clearly distinguishable.[2]

The spectroscopic image (figure 2B) of the lesion documented increased choline peak and decreased N-acetyl aspartate peak, but normal peaks in the normal appearing white matter near the lesion.[1]

The perfusion analysis (figure 2C), based on four different regions of interest (figure 2D), suggested a decreasing gradient of perfusion from the centre to the periphery of the lesion, supporting the hypothesis that the centre of the ring corresponds anatomically to a deep venous vessel.[3] Authors are not aware of previous report of perfusion studies in Balò's sclerosis.

Cerebrospinal fluid analysis detected oligoclonal bands, without anti-aquaporin-4 antibodies.

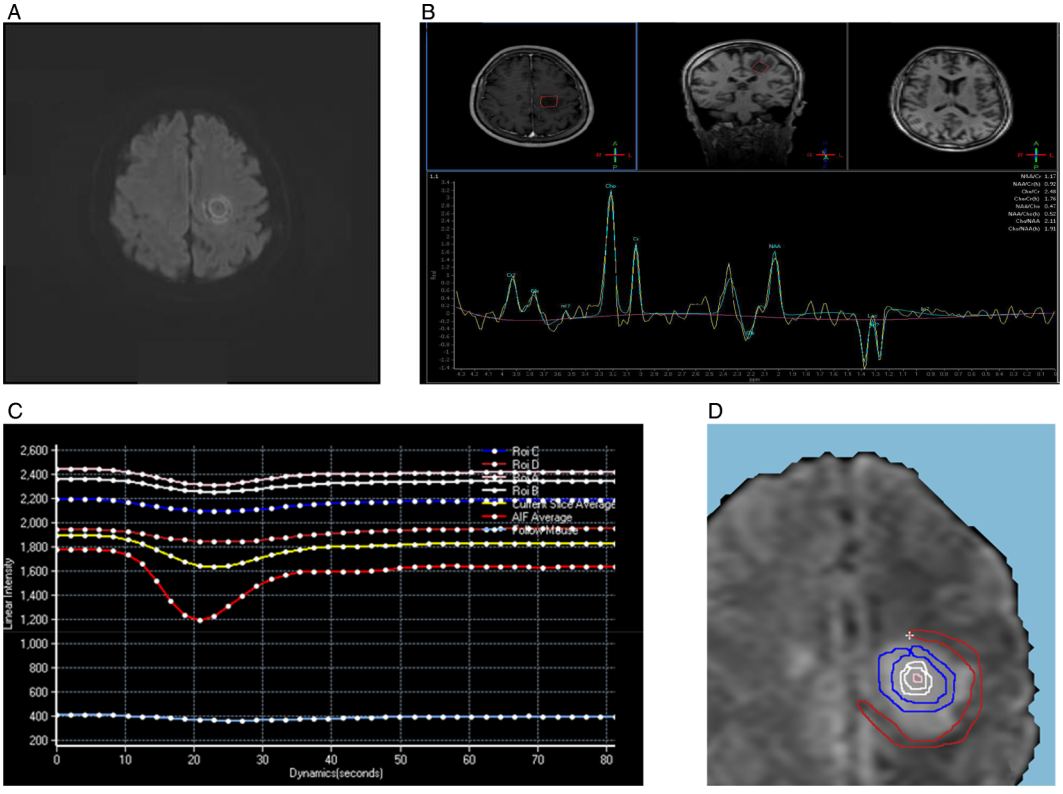

Figure 2 At diagnosis. (A) Diffusion shows a restricted peak depicting the classical ring of demyelination and remyelination. (B) Spectroscopy shows a high ratio Cho/N-Acetylaspartate, double pick of lactate-lipids. (C) Perfusion MRI analysis, based on four regions of interest depicted in D (A, pink, B, white, C, blue, D, red) suggests that the central layers of the ring are more perfused compared to the peripheral ones.

To cite: Ripellino P, Stecco A, Leone MA, *et al*. *BMJ Case Reports* Published online: [*please include* Day Month Year] doi:10.1136/bcr-2012-008413

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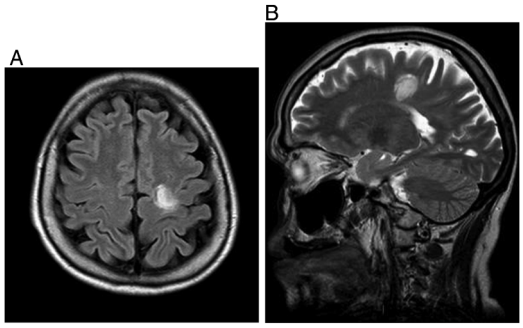

Figure 3 At 8 months follow-up (after treatment). (A) Axial fluid attenuated inversion recovery sequence and (B) sagittal T2-sequence evidence the reduction in the lesion volume compared to figure 1.

High-dose intravenous methylprednisolone was started (1 g/day for 10 days) and followed by oral prednisone (1 mg/kg/day for 2 months, then slowly tapered in 2 months) with concomitant lamivudine as antiviral prophylaxis. The right paresis fully recovered after 2 weeks and has not relapsed after 11 months.

The 8 months follow-up MRI showed a reduction in the lesion volume and absence of new demyelinating lesions (figure 3A,B).

Learning points

- Balò's concentric sclerosis is a rare demyelinating disease presenting with a concentric ring in the white matter. The clinical and radiological features can mimic other diseases such as primary central nervous system lymphoma, low-grade glioma or stroke.
- A brain biopsy should be obtained whenever possible. However, nowadays advanced neuroimaging studies (spectroscopy, MRI diffusion and perfusion) seem to be a reliable tool for the diagnosis.
- Balò's sclerosis could respond very well to high-dose steroids alone or in combination with other immunosuppressive treatments (eg, plasma exchange).

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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