Multiple cerebral cavernomas in linear scleroderma: an unusual association

Múltiplos cavernomas cerebrais na esclerodermia linear: uma associação incomum

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A 30-year-old female patient presented with recurrent right-sided clonic seizures that had started at the age of 4. Concurrently, her parents noted mild hyperpigmentation, dermal atrophy, and alopecia on the left side of her forehead. She was diagnosed with *en coup de sabre* scleroderma (ECDS), a subtype of linear scleroderma (LS), given the absence of systemic sclerosis symptoms (►**Figure 1**). A brain magnetic resonance imaging (MRI) revealed cavernomas and white matter lesions in the left cerebral hemisphere, ipsilateral to the cutaneous lesions (►**Figure 2**).

Localized facial scleroderma can be associated with neurological complications, as illustrated by our case involving ipsilateral brain cavernomas.[1–3] This diagnosis is presumptive, as histopathological confirmation of the lesions was not obtained. Differential diagnoses should consider small hemorrhages, potentially secondary to vasculopathy, as suggested by the susceptibility-weighted imaging (SWI) MRI findings.[4,5]

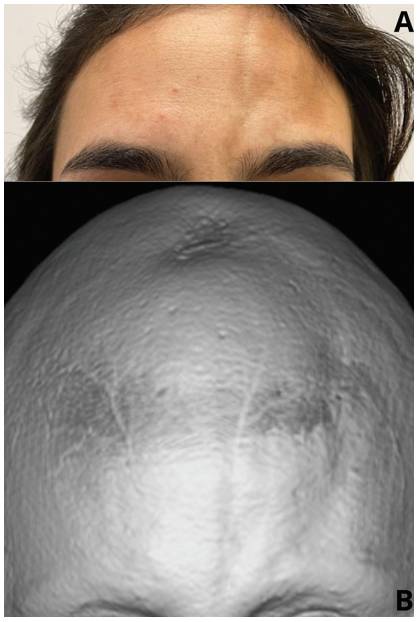

Figure 1 Autoimmune atrophy of the skin and subcutaneous tissue promotes the hallmark of the disease - the *coup de sabre* lesion (A). Three-dimensional reconstruction in volume rendering using brain magnetic resonance imaging (B) additionally demonstrates another *coup de sabre* in the interparietal region.

Author's Contributions

GRT, MESFB, RTNLM, RBR: conceptualization, resources, and writing – original draft; SBAN, supervision; MPMM: supervision and writing – review & editing; JLP, OGPB: supervision.

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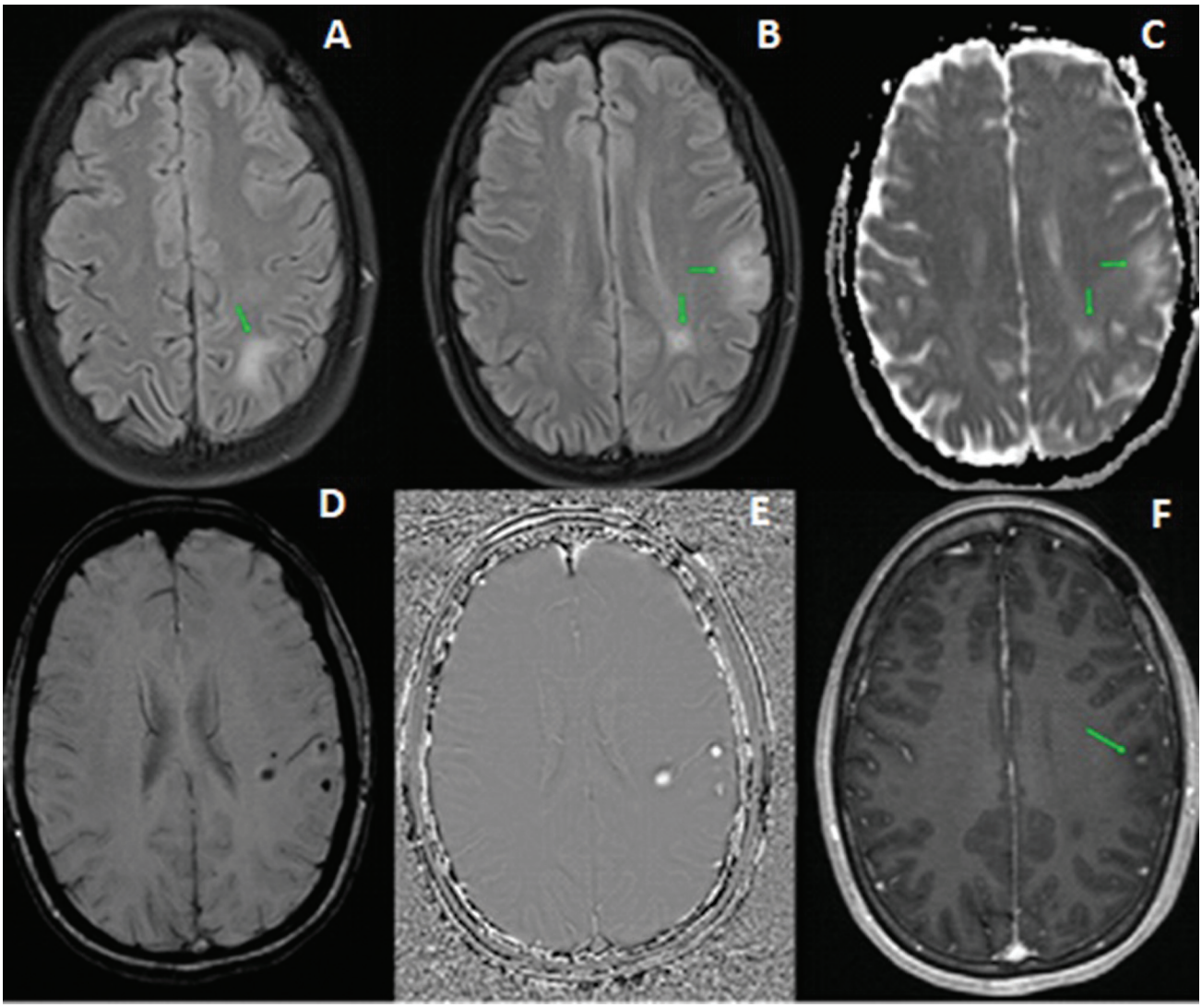

Figure 2 Fluid-attenuated inversion recovery images (A and B) and ADC map (C) shows T2 hypersignal and facilitated diffusion in the subcortical white matter ipsilateral to the skin lesion. Susceptibility-weighted imaging (SWI) (D), SWI phase map (E) and postcontrast T1 spoiled gradient-recalled (SPGR) (F): small subcortical nodules exhibiting blooming on SWI, compatible with cavernomas.

Conflict of Interest

The authors have no conflict of interest to declare.

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