

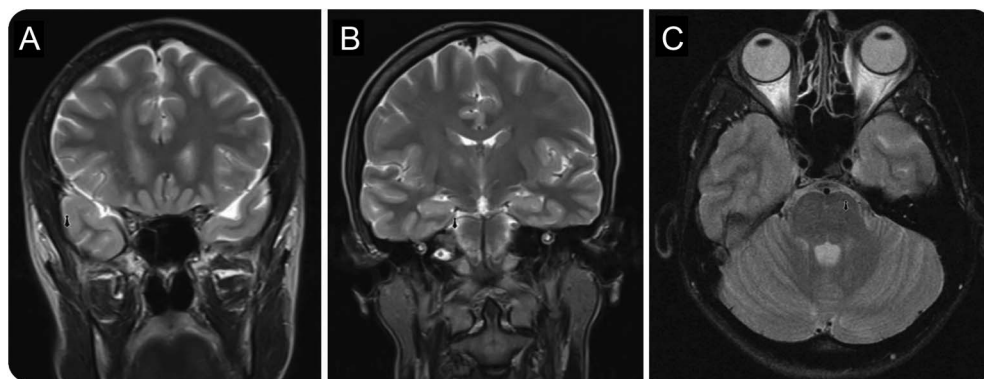
Linear scleroderma en coup de sabre presenting with positional diplopia and enophthalmos

Figure 1 Physical examination findings



(A) Hypopigmented frontoparietal morphea right of midline. (B) Right enophthalmos due to suspected orbital fat fibrosis; also inferior lagophthalmos.

Figure 2 Brain and orbital MRI



(A) Coronal T2-weighted MRI demonstrates nonenhancing hyperintensities in the right frontal lobe and (B) right thalamus. (C) Axial T2-weighted MRI orbits reveals right enophthalmos.

A 28-year-old woman with alopecia since childhood presented with 4 years of progressive intermittent diplopia and right eye getting “stuck” with upward gaze. Maddox rod test revealed a subtle intermittent right hypotropia and right enophthalmos (figure 1). Asymptomatic T2 hyperintensities were noted on MRI (figure 2). Scalp morphea was confirmed pathologically. Formal rheumatologic evaluation was otherwise unremarkable. Linear scleroderma en coup de sabre presenting as positional diplopia, unilateral enophthalmos, and focal neuroimaging findings is rare and is suspected to represent a chronic local inflammatory process.^{1,2} The brain lesions tend to develop adjacent to the skin atrophy.¹

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