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IMAGES IN NEUROLOGY

Orbital apex syndrome associated with diffuse hypertrophic pachymeningitis in isolated neurosarcoidosis

Síndrome da fissura orbitária superior associada a paquimeningite hipertrófica difusa na neurosarcoidose isolada

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A 43-year-old woman reported acute onset of right retro-orbital headache followed by ptosis, paralysis of extraocular muscles, and subsequently amaurosis, characterizing an orbital apex syndrome[1]. Brain MRI showed diffuse pachymeningitis with leptomeningeal and parenchymatous involvement (Figure 1). CSF analysis was unremarkable. ACE and

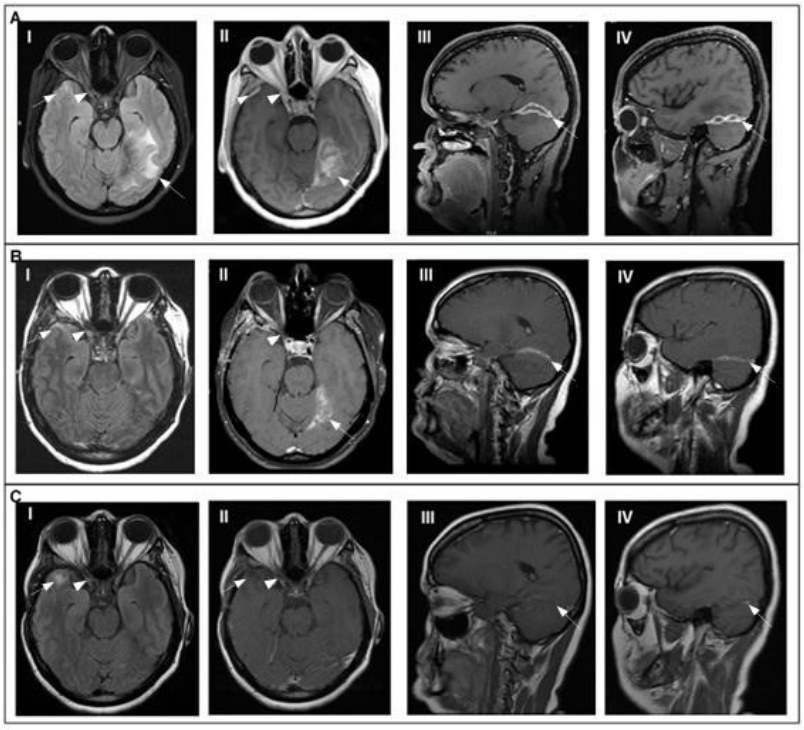

Figure 1. Brain MRI at disease onset (A); two months after steroid pulse therapy (B); and one year after treatment (C). At disease onset (A), there is a cortical and subcortical FLAIR hyperintensities (arrows in A.I) in the anterior right temporal pole and left

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temporal lobe. Those areas show T1 contrast enhancement (A.II) and highlight the pachymeningitis involving right temporal pole and parasellar region, including the orbital apex (A and B, arrow heads). Sagittal T1 with contrast (A.III and A.IV) show pachymeningitis in the left cerebellar tentorium (arrows), with subtle leptomeningeal involvement, characterized by sulci enhancement shown in the axial plane (arrows in A.II). Two months after pulse therapy (B) there is a reduction in the FLAIR hyperintensity and cortical/sulci enhancement in the left temporal lobe and a decrease of pachymeningeal thickening and enhancement. One year after pulse therapy (C) there is an almost complete resolution of meningeal abnormalities with a residual focal FLAIR hyperintensity in the right temporal pole, in the biopsy site.

IgG4 levels were normal. However, investigation was done under oral steroid use. Lymphoproliferative and exocrine gland disorders were excluded. Lung scintigraphy and CT were normal. Meningeal biopsy disclosed diffuse non-caseous granulomatosis. Intravenous steroid pulse improved ocular movements (Figure 2), but amaurosis persisted.

Neurologic manifestations represent less than 10% of sarcoidosis cases[2]. Isolated neurosarcoidosis is even rarer[3].

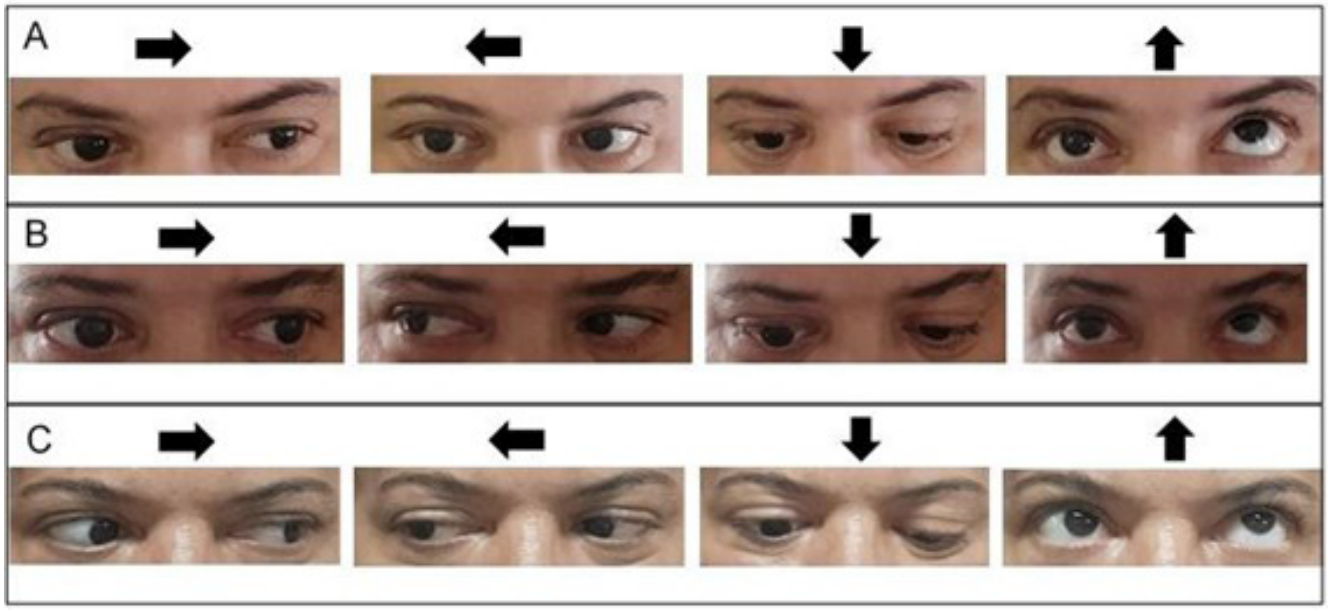

Figure 2. Evaluation of ocular motricity of the right eye (RE): A. Around 3 weeks after pulse intravenous methylprednisolone therapy. B. Two months after pulse therapy, a mild improvement, especially in abduction of RE. C. One year after pulse therapy, expressive improvement of horizontal motricity and remaining palsy of vertical gaze, particularly downward.

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