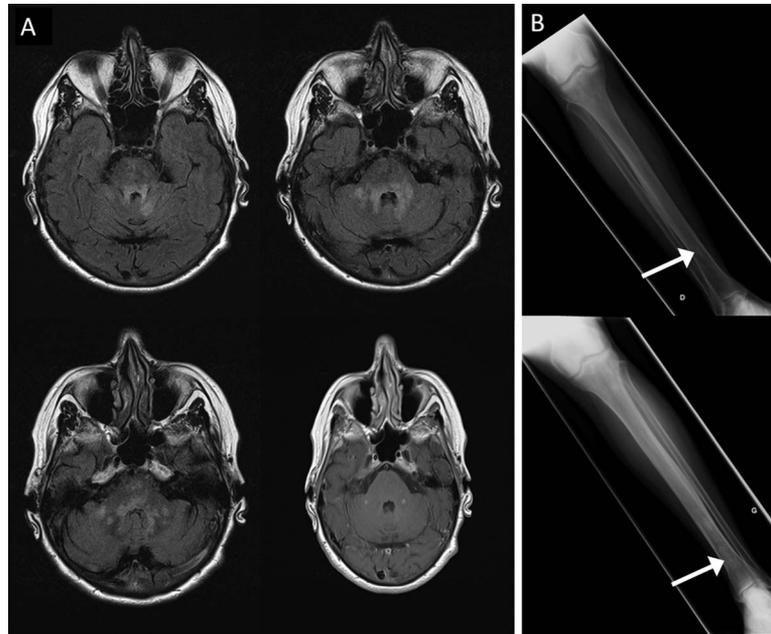


Teaching NeuroImages: Ataxia and diabetes insipidus

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Figure Brain MRI and leg x-rays in Erdheim-Chester disease



(A) Brain MRI shows fluid-attenuated inversion recovery hyperintensities involving cerebellar peduncles and the pons, with punctiform gadolinium enhancement on T1-weighted sequences. (B) Right and left x-rays show cortical osteosclerosis of the 2 tibiae.

A 63-year-old woman presented with recent cerebellar ataxia. Diabetes insipidus had been diagnosed 35 years earlier. Brain MRI showed fluid-attenuated inversion recovery hyperintensities involving cerebellar peduncles and pons, with punctiform gadolinium enhancement on T1 sequences (figure, A). Leg x-rays revealed cortical osteosclerosis of both tibiae (figure, B). Tibia biopsy revealed bone remodeling associated with bone marrow fibrosis and lymphohistiocytic reaction confirming the diagnosis of Erdheim-Chester disease (ECD).

ECD is a non-Langerhans histiocytosis. The most frequent manifestations are skeletal involvement, peri-aortic infiltration, diabetes insipidus, and pericardial involvement. CNS localizations include brainstem, cerebellum, and meninges, and are a major prognostic factor. Interferon- α can be first-line therapy.¹

AUTHOR CONTRIBUTIONS

Romain Lefaucheur: acquisition of data, analysis and interpretation. David Maltête: acquisition of data, analysis and interpretation, critical revision of the manuscript for important intellectual content, study supervision. Julien Haroche: acquisition of data, analysis and interpretation. Alaina Borden: acquisition of data, analysis and interpretation, critical revision of the manuscript for important intellectual content. David Wallon: acquisition of data, analysis and interpretation. Bertrand Bourre: acquisition of data, analysis and interpretation, critical revision of the manuscript for important intellectual content.

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