

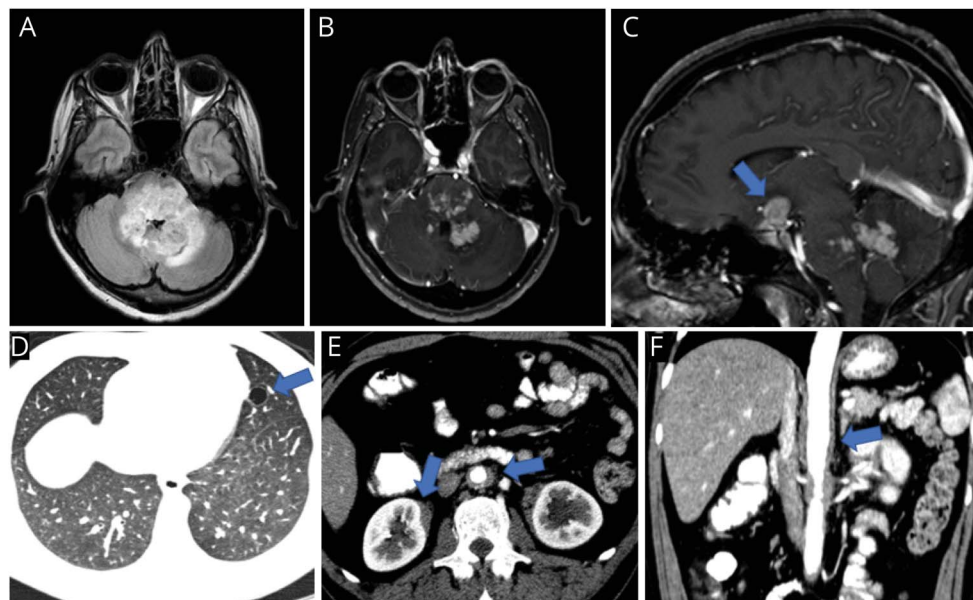
Teaching Video NeuroImages: Multisystemic Erdheim-Chester Disease Presenting as a Cerebellar Ataxia

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Figure 1 Brain MRI and Chest/Abdomen/Pelvis CT



Brain MRI demonstrates infiltrative lesion in the pons and cerebellar peduncles: (A) fluid-attenuated inversion recovery hyperintense and (B, C) T1/gadolinium-enhancing. An enhancing pituitary lesion was also present (arrow in C). CT images demonstrate a pulmonary cyst and posterior ground-glass pattern (D) and perirenal and periaortic soft tissue (E, F).

A 41-year-old man was admitted to the neurology ward due to progressive vertigo and unsteadiness for the previous 2 months. Neurologic examination was remarkable for a global cerebellar syndrome. Investigation with brain MRI led to the hypothesis of a histiocytosis due to infiltrative lesions of the pons, cerebellar peduncles, and pituitary. Therefore, investigation progressed with chest/abdomen/pelvis CT, bone scintigraphy, and a tibial biopsy that confirmed the diagnosis of Erdheim-Chester disease (figures 1 and 2, video).

Erdheim-Chester disease is a rare disorder characterized by the infiltration of non-Langerhans histiocytes in multiple tissues, mainly bone, but with CNS involvement in around 40% of cases.^{1,2}

Study Funding

No targeted funding reported.

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Video

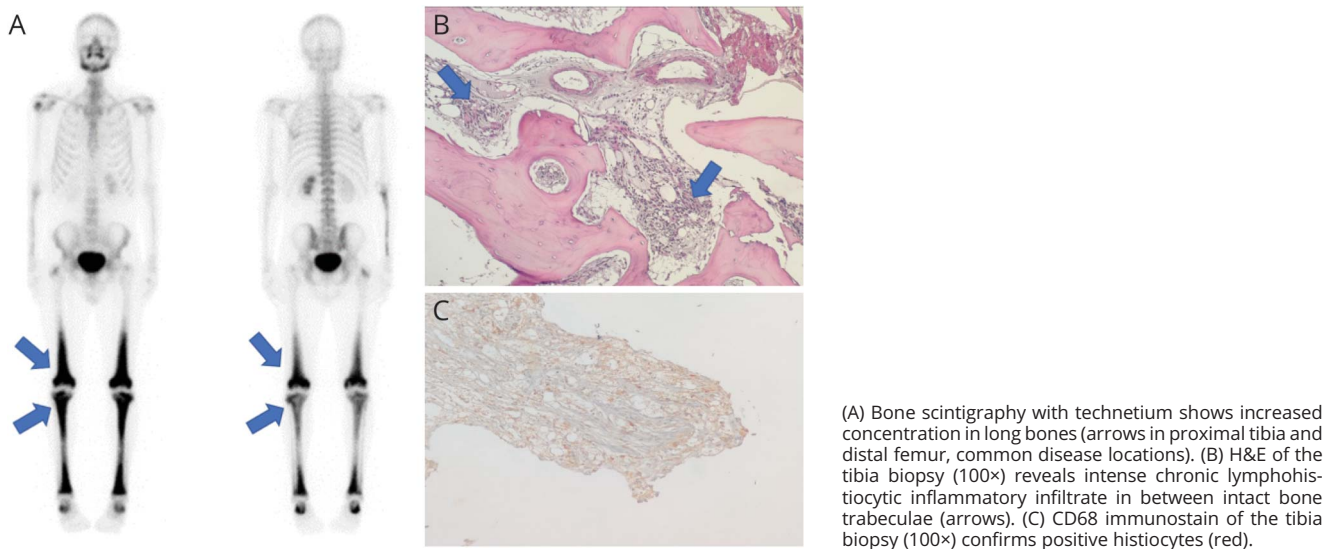
Teaching slides

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Figure 2 Bone Scintigraphy and Tibial Biopsy Histopathology



(A) Bone scintigraphy with technetium shows increased concentration in long bones (arrows in proximal tibia and distal femur, common disease locations). (B) H&E of the tibia biopsy (100×) reveals intense chronic lymphohistiocytic inflammatory infiltrate in between intact bone trabeculae (arrows). (C) CD68 immunostain of the tibia biopsy (100×) confirms positive histiocytes (red).

Disclosure

The authors report no disclosure relevant to the manuscript. Go to [Neurology.org/N](https://www.neurology.org/N) for full disclosures.

Appendix Authors

Name	Location	Contribution
Marcelo Houat de Brito, MD	Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas	Design and conceptualization of the study; data collection, analysis, and interpretation; drafting the manuscript
Marcos Castello Barbosa de Oliveira, MD	Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas	Data collection, analysis, and interpretation; revision of manuscript
Yuri Reis Casal, MD	Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas	Data collection, analysis, and interpretation

Appendix (continued)

Name	Location	Contribution
Andre Neder Ramires Abdo, MD	Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas	Data collection, analysis, and interpretation; revision of the manuscript
Leandro Tavares Lucato, MD, PhD	Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas	Data collection, analysis, and interpretation; revision of the manuscript
Mateus Mistieri Simabukuro, MD	Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas	Data collection, analysis, and interpretation; revision of manuscript

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