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

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Disclosure
The authors report no disclosure relevant to the manuscript. Go to Neurology.org/N for full disclosures.

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References

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