A 41-year-old man with progressive cerebellar ataxia

Teaching Neuro*Images*

Neurology

Resident & Fellow Section

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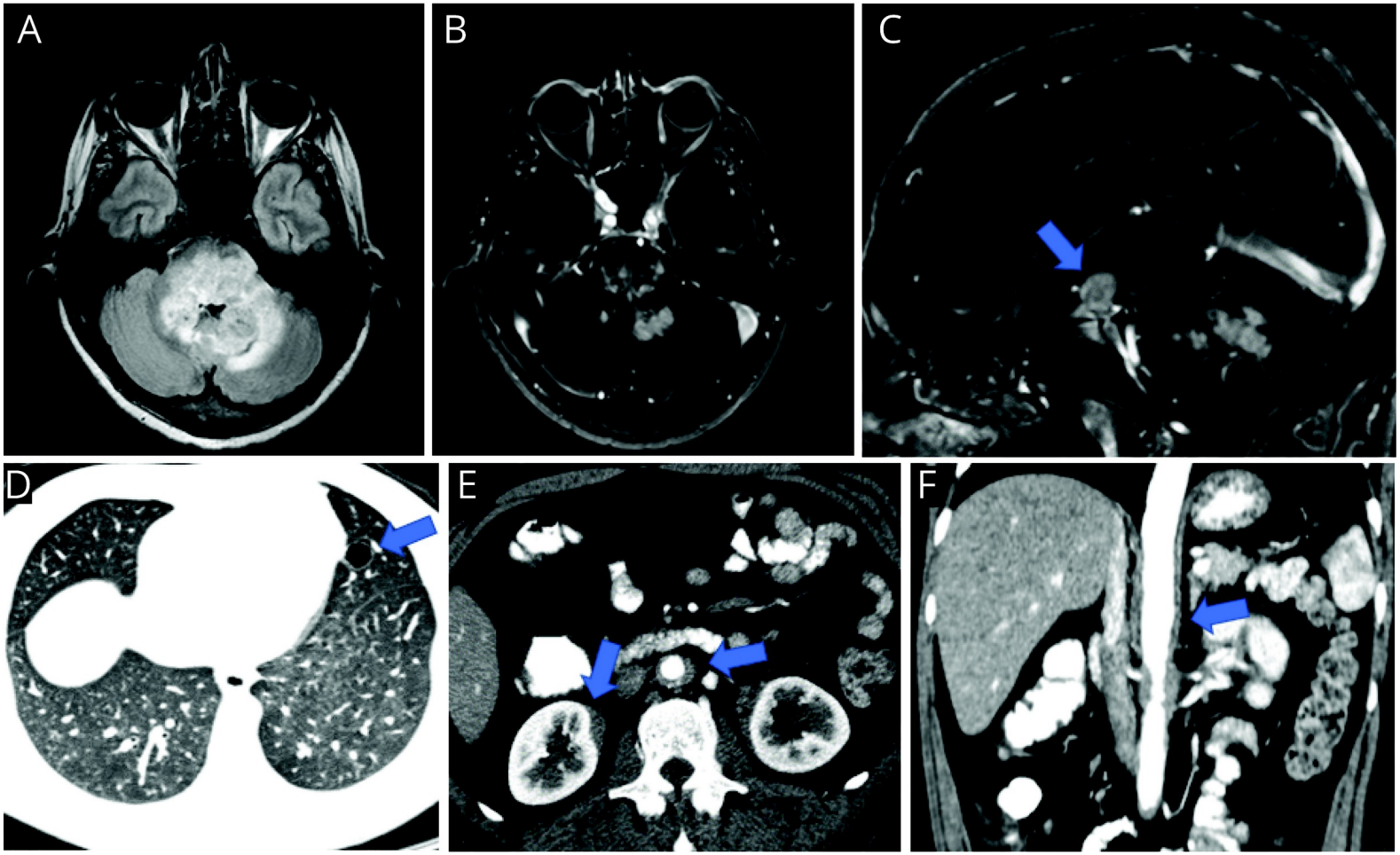
Vignette

- A 41-year-old man with progressive vertigo and unsteadiness for the previous two months.
- Neurological examination was remarkable for a global cerebellar syndrome.
- Brain MRI demonstrated infiltrative lesions of the pons, cerebellar peduncles, and pituitary (Figure).
- With a hypothesis in mind, the investigation progressed with chest-abdomen-pelvis CT and bone scintigraphy (figure).

Imaging: Figure 1



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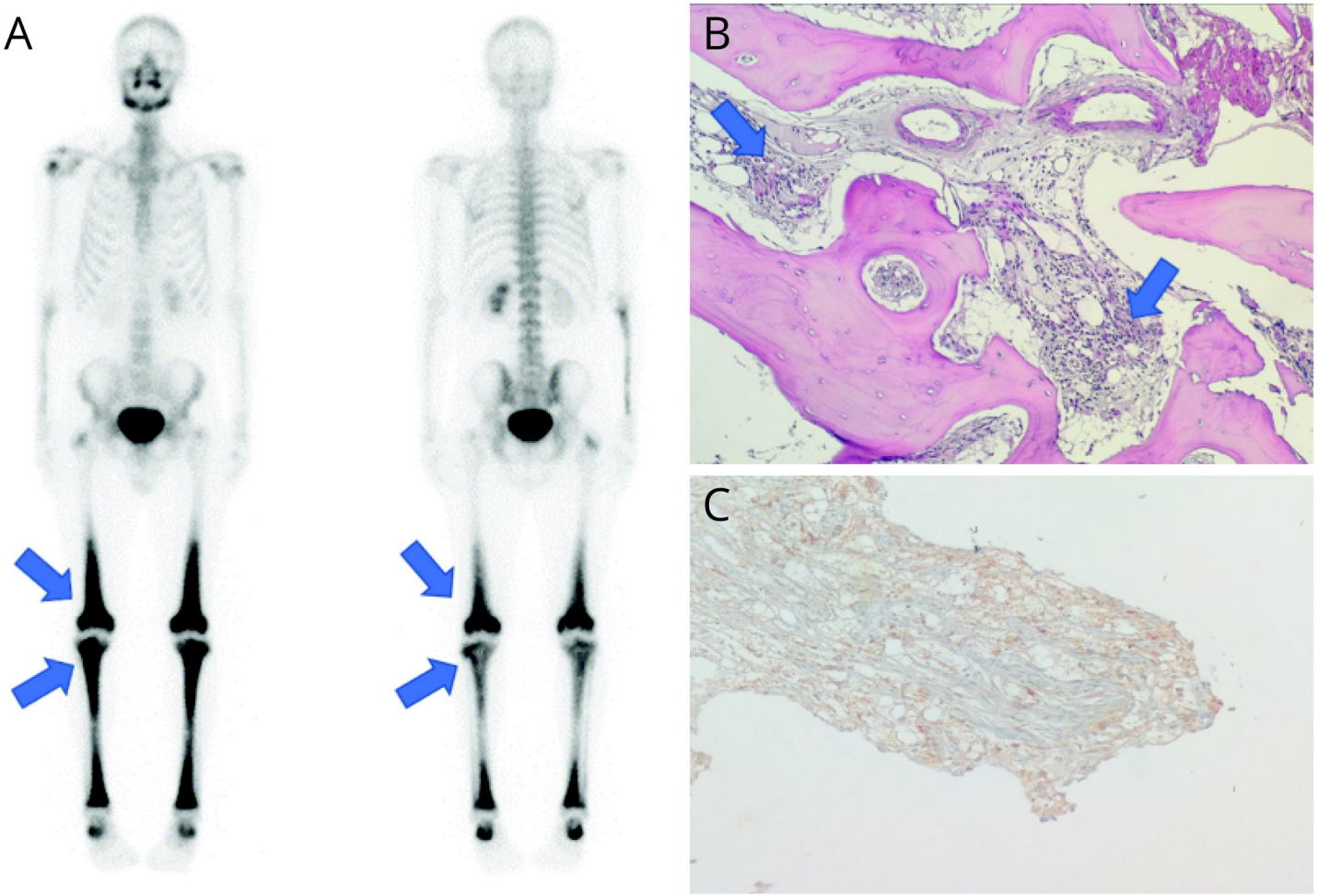
Imaging: Figure 2



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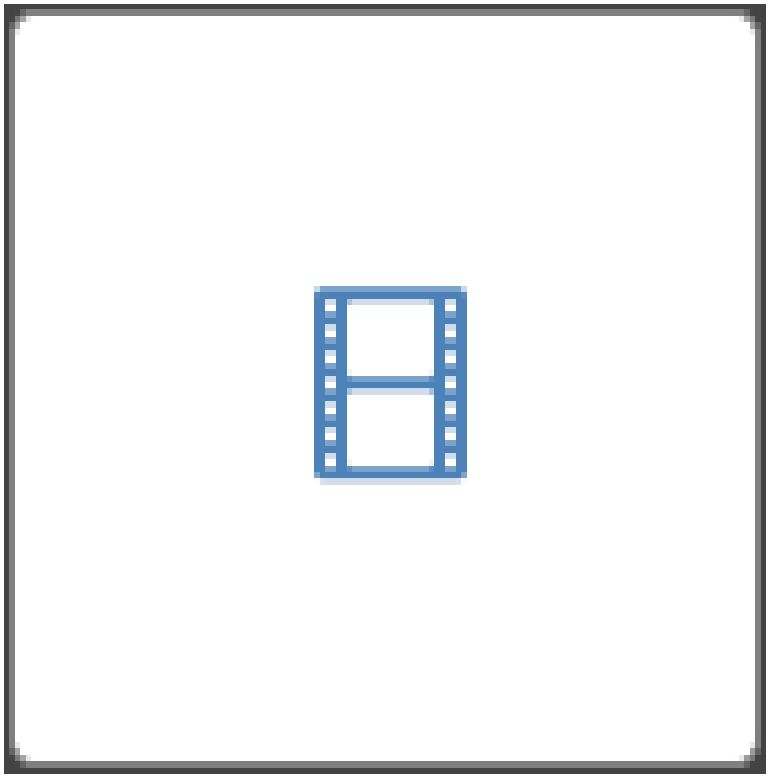
Imaging: Video



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

- The findings at MRI led us to think about the possibility of a histiocytosis, which made the investigation progress with other tests aimed at this hypothesis.
- The exams showed pulmonary, aortic, renal and bone involvement that were compatible with histiocytosis.
- We could confirm the diagnosis without CNS biopsy: histopathological evaluation of a tibial fragment demonstrated intense chronic lymphohistiocytic inflammatory infiltrate in between intact bone trabeculae and positive CD68 immunostain, compatible with Erdheim-Chester disease (ECD).
- ECD is a rare disorder characterized by the infiltration of non-Langerhans histiocytes in multiple tissues, mainly bone, but with CNS involvement in around 40% cases.

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