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Teaching NeuroImages: Erdheim-Chester disease (polyostotic sclerosing histiocytosis)

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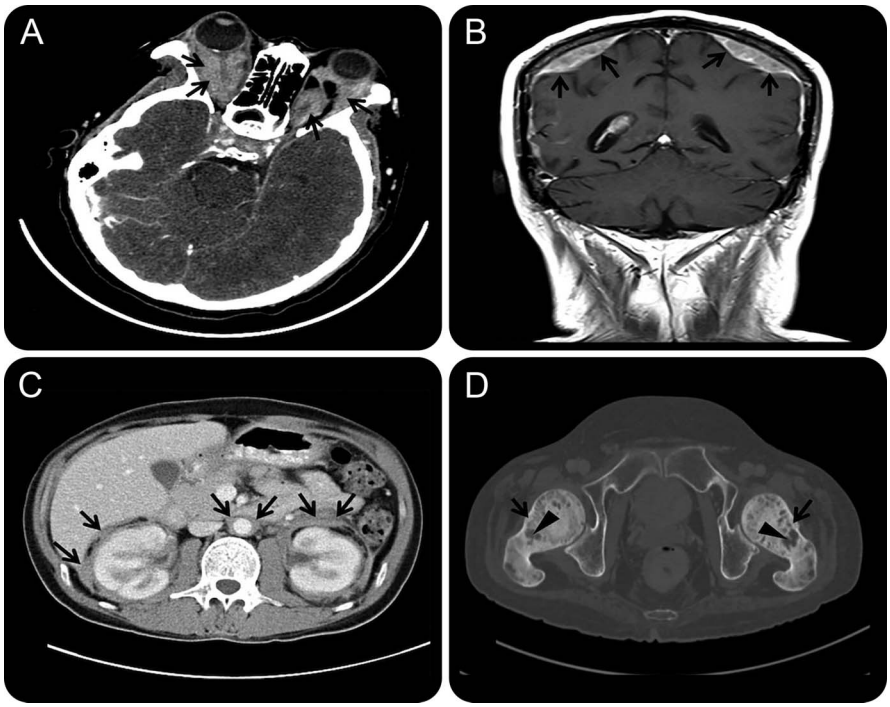
Figure Radiographic findings



(A) Soft tissue infiltration in bilateral retro-orbital spaces (arrows). (B) Brain MRI demonstrates enhancement of pachymeninges (arrows). (C) Abdominal CT illustrates encasement of bilateral kidneys and aorta by infiltrates (arrows). (D) Pelvic CT shows mixed osteolytic (arrowheads) and osteosclerotic (arrows) lesions of femoral heads.

A 34-year-old man presented with progressive visual loss, diplopia, headache, intermittent fever, and significant weight loss for 7 months. Physical examination revealed bilateral visual loss and exophthalmos with complete ophthalmoplegia. Imaging demonstrated pachymeningeal enhancement, soft tissue infiltration in the retro-orbital spaces and multiple organs including kidneys and abdominal aorta, and mixed osteosclerotic and osteolytic lesions in bilateral femoral heads[1] (figure). Pathologic study of the retro-orbital lesion revealed foamy histiocytes negative for CD1a and S100,[2] consistent with Erdheim-Chester disease (ECD). The patient was treated with IV methylprednisolone followed by cyclosporine with minimal benefit. Four months later, he died due to sudden unexplained death during sleep. An autopsy was not performed. ECD is a rare, non-Langerhans histiocytosis characterized by chronic histiocytic infiltration of multiple organs. Common manifestations include bone pain, diabetes insipidus, exophthalmos, and xanthelasma.[1] The nervous system is affected in 25%–50% of patients and can be involved in the extra-axial and intra-axial compartments.[1,2] Standard treatment has not been established.

AUTHOR CONTRIBUTIONS

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