

Erdheim-Chester Disease in a 49 Year Old Man

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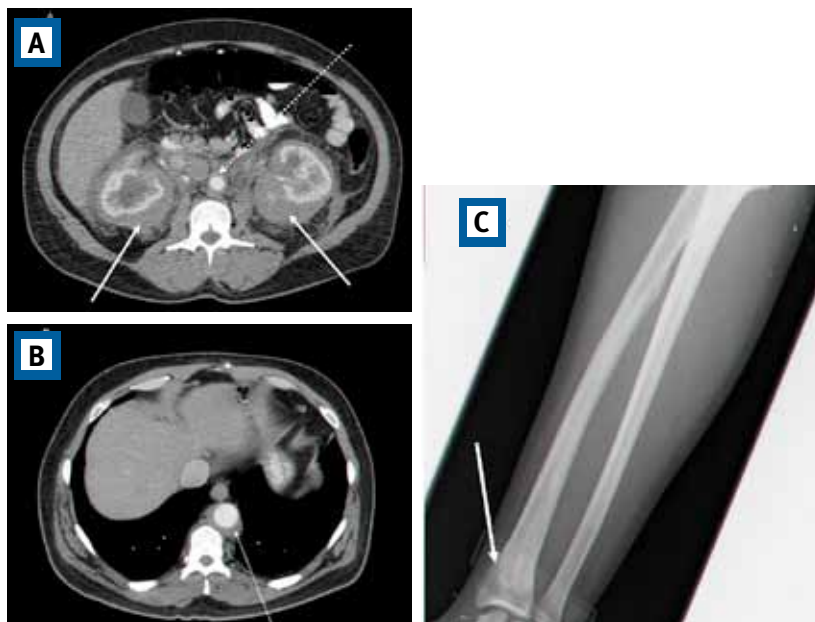
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A 49 year old male with non-specific abdominal pain underwent computed tomography scan, which demonstrated bilateral perirenal soft tissue infiltrates with mild hydronephrosis [Figure A] and periaortic infiltrates [Figure A & B, dotted arrows]. Skeletal studies showed symmetric osteosclerotic lesions in the distal radius [Figure C, arrow]. Blood tests revealed anemia (11.4 g/dl), thrombocytosis (427,000/mm³), elevated erythrocyte sedimentation rate (84/hr) and C-reactive protein (40, normal < 0.8). Liver, renal, electrolytes and serological tests were all normal. Biopsy of the perirenal mass demonstrated foamy histiocytic aggregates (CD68 positive, S-100 negative) indicating macrophage but not dendritic/Langerhans cell lineage histiocytosis, establishing the diagnosis of Erdheim-Chester disease [1,2].

Erdheim-Chester disease is a rare non-Langerhans cell histiocytosis of unknown etiology, with bone, perirenal (“hairy kidneys”) and periaortic (“coated aorta”) infiltrates. Cardiac, orbital, pituitary (presenting as diabetes insipidus), pulmonary and skin infiltrations were also reported [3].

Clinical manifestations and prognosis depend on the organs involved. Patients with systemic involvement without treatment have a poor prognosis with a mean survival of 2–3 years after diagnosis [4]. Corticosteroids, vincris-



Abdominal CT scan demonstrating bilateral perirenal soft tissue infiltrates with mild hydronephrosis [A, arrow] and periaortic infiltrates [A & B, dotted arrows]. Skeletal studies demonstrating symmetric osteosclerotic lesions in distal radius [C, arrow]

tine, interferon-alpha, imatinib mesylate (Glivec[®], Novartis, Switzerland) and bisphosphonates were reported to be beneficial in the treatment of patients with Erdheim-Chester disease [5]. Our patient was treated with interferon-alpha and bisphosphonates, which led to the normalization of his abnormal blood tests (hemoglobin, thrombocytes, erythrocyte sedimentation rate, C-reactive protein) and to the partial resolution of his bone, perirenal and periaortic lesions.

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