A 42-year-old Thai woman developed polyuria and increased thirst as well as 5-kg weight loss over 6 months. She also noticed periobital swelling of both eyelids. Physical examination showed yellowish-brown and symmetrically distributed lesions over both upper eyelids and the infraorbital regions (Picture 1). The diagnosis of Erdheim-Chester disease (ECD) was made by xanthogranulomatosis from skin biopsy and bone scan study demonstrating symmetrical uptake in long bones of extremities (Picture 2). Water deprivation test and pituitary imaging also revealed central diabetes insipidus (Picture 3). DDAVP was started with clinical improvement.

ECD is an idiopathic, progressive non-Langerhans cell histiocytosis characterized by bilateral, symmetrical sclerosis of the metaphyseal and diaphyseal regions of long bones; especially in distal femur and proximal tibia and fibula. It also involves the viscera including kidneys, lungs and the central nervous system (1). Pathologic and radiographic changes in the long bones are pathognomonic. Clinically manifestations range from asymptomatic focal bones lesions...
Sagittal T1-weighted image shows a marked thickening of mucoperiosteal lining in the sphenoid sinus and absence of normal bright spot SI in the posterior pituitary.

to systemic and fatal disease with visceral involvement. A wide range of treatment modalities have been used to manage patients with ECD including conservative observation or systemic glucocorticoids, radiation therapy and chemotherapy.

Reference


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