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Pituitary

A rare cause of central diabetes insipidus with arthritis (Erdheim-Chester disease)

Mahmood Shakir Khudhair and Gelal Abdali Altae

Department of Medicine, College of Medicine, ALNahrain University, Baghdad, Iraq

Mr. Haider is a 26-year-old resident doctor who presented with intermittent fever relieved by NSAIDs fatigue, weakness, loss of libido, fever with no rigor, night sweat, bone, joint pain and knee swelling. He lost weight.

He was diagnosed with central chronic D.I for the last 3 years after several weeks history of polyurea and thirst without identifiable cause which follows elbow fracture with trivial trauma and was taking nasal desmopressin as required. His last symptoms were followed a Covid 19 attack. Physical examination reveals high fever 39 °C, pallor, thin built, no thyroid or lymph node enlargement. Chest examination is normal with no organomegaly on abdominal examination. Knee examination shows swollen red tender joint. Initial investigation shows ESR 110 mm/hr. CRP 10 mg/dl

Negative screening for tuberculosis, hepatitis, HIV, vasculitis and Brucellosis.

Normal kidney and liver function tests and electrolytes. Mild hypochromic anemia. Normal free T4 and TSH, prolactin and cortisol.

Normal SHBG

Low morning testosterone twice 60 ng/dl. and 64 ng/dl. FSH and LH normal

Normal Dexa scan.

Thus till now there is feature of hypogonadism, D.I and arthritis.

Due to low testosterone MRI pituitary requested and reveals small size pituitary gland, absent post bight spot with mild thickening of the pituitary stalk with no mass lesion.

Suspicion of inflammatory or infiltrative lesion of pituitary was raised.

For proximal tibial lesion bone biopsy was done and revealed marked fibrosis with infiltration of bone marrow by foamy cells with multinucleated cells positive for CD 68 and negative for CD 1a by immunohistochemistry. Further evaluation by PET scan confirms the diagnosis

and shows multifocal hypermetabolic symmetrical bone lesions most intense around elbows and knees consistent mostly with Erdheim Chester disease (ECD).

Patient was treated with testosterone injections with improved libido and consultation with oncologist was done to arrange the need for chemotherapy.

Molecular and genetic test is not available in Iraq and a sample was sent abroad.

To our knowledge this is the first reported case of ECD in Iraq.

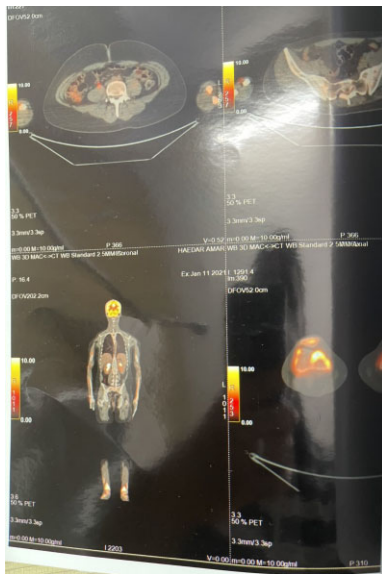


Figure 1. PET image.

