Young Adult With A Rare Presentation Of Hand Schuller Christian (HSC) Disease

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INTRODUCTION:

Langerhans cell histiocytosis (LCH) is characterized by proliferation and infiltration of bone marrow derived histiocytes. It encompasses a spectrum of disease which localizes to bony lesion or involve multisystem with disseminated visceral involvement. In this case, we highlight an extremely rare presentation of HSC in bilateral femur with extra-skeletal involvement in an adult.

CASE REPORT:

A 34 year old lady with infertility and cranial diabetes insipidus secondary to pituitary adenoma was referred to our clinic for chronic left hip pain for 1 year. There was no preceding trauma nor constitutional symptoms. No tenderness elicited clinically and range of motion of the left hip joint was normal. Serum calcium and phosphate level, full blood count, liver function test, renal profile, parathyroid hormone (PTH) level and inflammatory markers were within normal limits. X-ray pelvis and femur revealed eccentric lytic lesion with sclerotic rim and narrow zone of transition at sub-trochanteric level in bilateral femur. Core needle biopsy of the lesion was done to confirm the diagnosis. Histopathology examination (HPE) showed lamellar bone with dense collection of eosinophils admixed with histiocytes and neutrophils. The histiocytes were immunopositive for CD 1a and S-100 protein which was suggestive of Langerhans cell histiocytosis. Left femur intra-lesional steroid injection was done which resulted in regression of the lesion and improved her symptoms significantly.

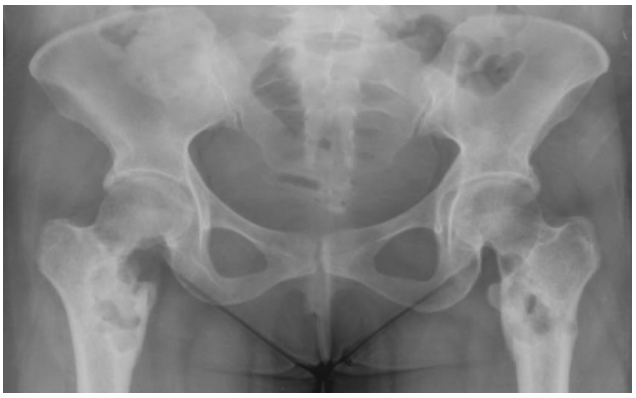

Figure 1: X- ray Pelvis (AP view) shows eccentric lytic lesion at sub-trochanteric level in bilateral femur surrounded by sclerotic rim with narrow zone of transition.

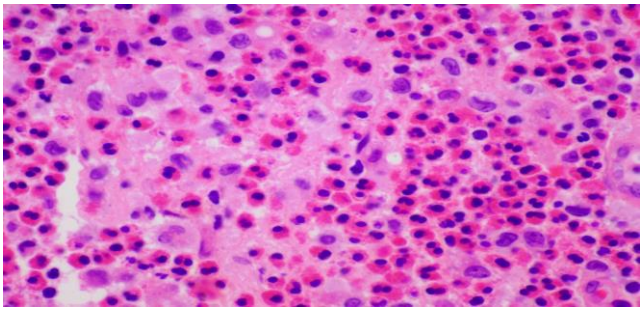

Figure 2: HPE (Haematoxylin and eosin stain, x600) shows dense infiltrate of eosinophils admixed with histiocytes.

CONCLUSION:

HSC disease is a rare form of LCH which is scarcely reported in the literature. Hereby, we would like to reiterate that HSC should be considered in the differential diagnosis of patients presenting with polyostotic lytic lesions with disseminated systemic involvement. High clinical suspicion with multidisciplinary evaluation is crucial to expedite the diagnosis and management of HSC.

REFERENCE:

1. Khung S, Budzik JF, et.al. Skeletal involvement in Langerhans cell histiocytosis. Insights Imaging. 2013;4:569-579.