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MONOSTOTIC FIBROUS DYSPLASIA--A CASE REPORT

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BACKGROUND
Fibrous dysplasia of bone is a fairly common, well-recognized, locally circumscribed, slowly progressing, benign disorder of fibro-osseous tissue.[1] Certain endocrinopathies and abnormal pigmentations of the skin may form a part of the total disease process. Three basic forms of the disease are currently recognized: Monostotic form, Polyostotic form, Polyostotic form with associated endocrine abnormalities.[2] According to Ippolito and colleagues, in monostotic FD, the most commonly appearing site is the femur.[3] Tibia, humerus, rib, clavicle and craniofacial skeleton are the next in order of frequency.

CLINICAL PRESENTATION

A 14 yr old, female patient presented with history of pain in the right hip, right thigh for 6 months and limping. Initially the pain was intermittent in nature which progressed and now the pain is persistent for the last 1.5 months. No history of trauma was there.

OBJECTIVES & MATERIAL AND METHOD

Objective of this report is to describe a case of a patient with monostotic FD involving the proximal end of right femur.

The patient was evaluated using X-RAY followed by CEMRI. CT was done for academic purpose. The diagnosis was confirmed by excisional biopsy.

IMAGING FINDINGS

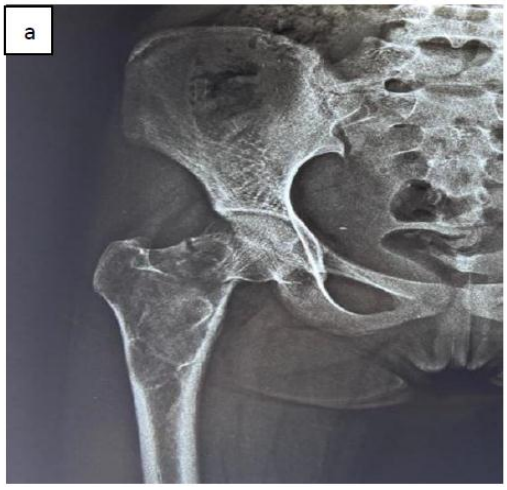
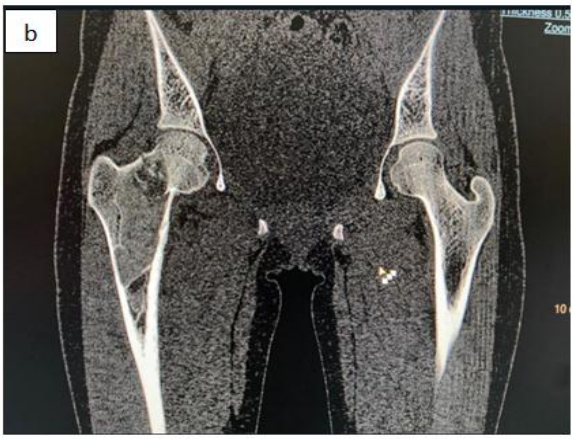
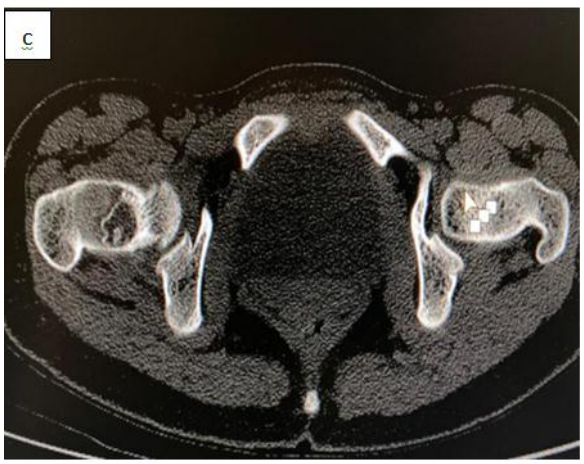


Fig.a- X-ray right hemipelvis (AP-view) with right hip joint demonstrates mildly expansile, well-circumscribed, ground-glass like density involving neck of femur, trochanteric region and proximal shaft of right femur in meta diaphyseal location. Evidence of endosteal scalloping seen. No e/o periosteal reaction seen. Pathological fracture noted in neck of femur.





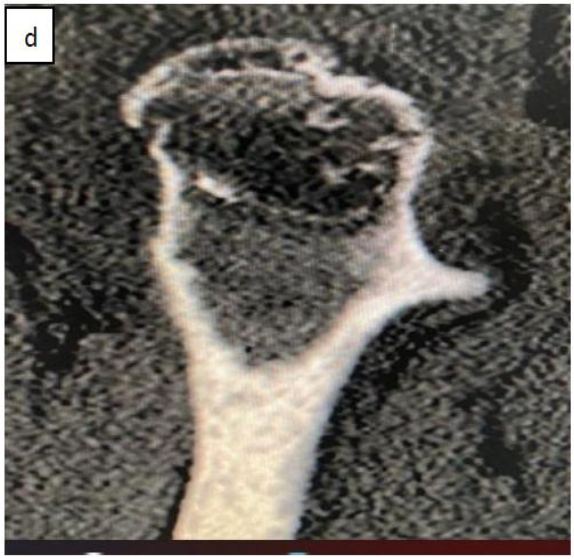
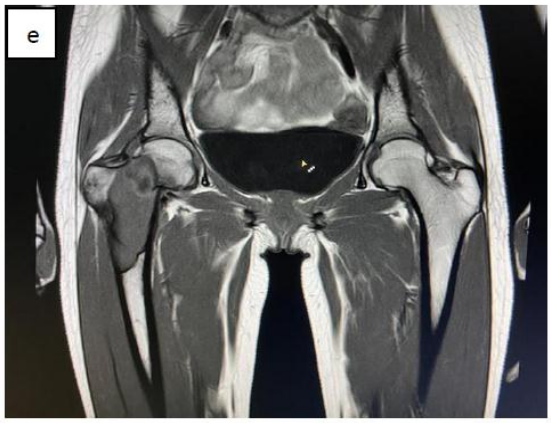
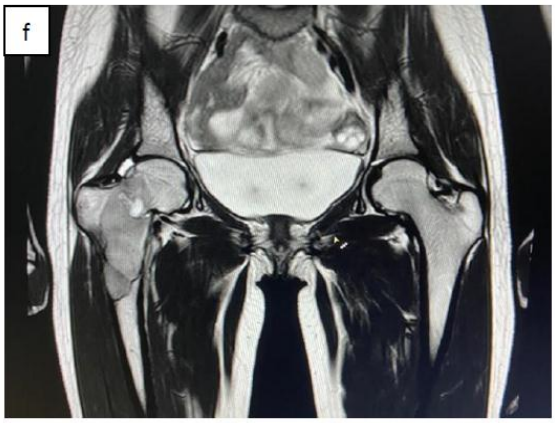


Fig b,c,d- coronal, axial and sagittal NCCT images reveal an expansile lytic lesion with ground glass like density within, in the metadiaphyseal region of right proximal femur associated with endosteal scalloping. There is transcervical fracture of right femoral neck. There is no e/o periosteal reaction.





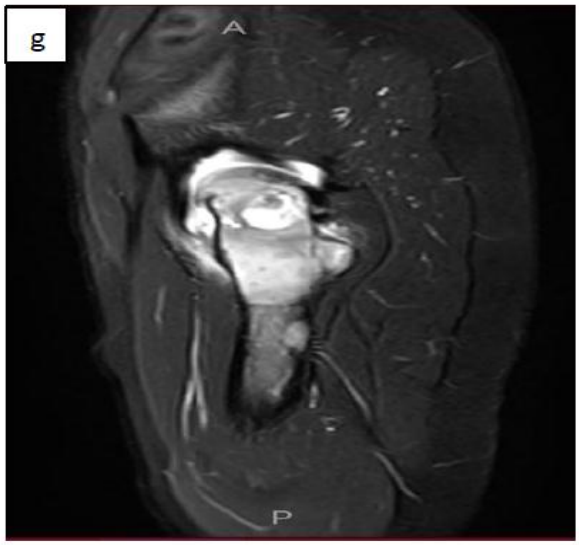
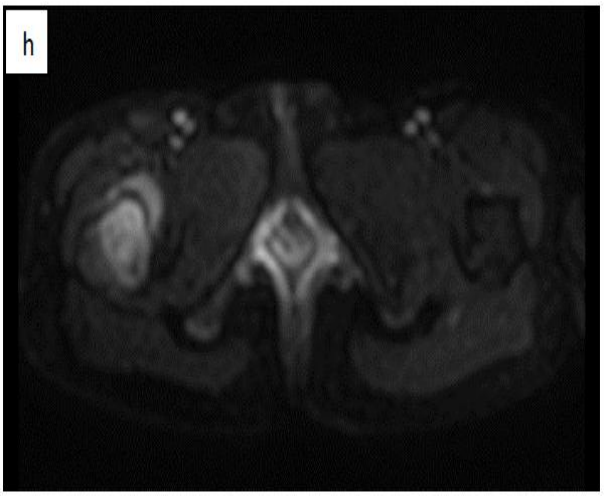


Fig e,f- coronal T1 & T2W images shows a T1 hypo and T2 hyperintense intramedullary lesion involving the neck, trochanter and proximal shaft of right femur causing widening of the medullary canal. Fig g- STIR sagittal image reveals expansion and cortical thinning with minimal surrounding hip joint effusion.



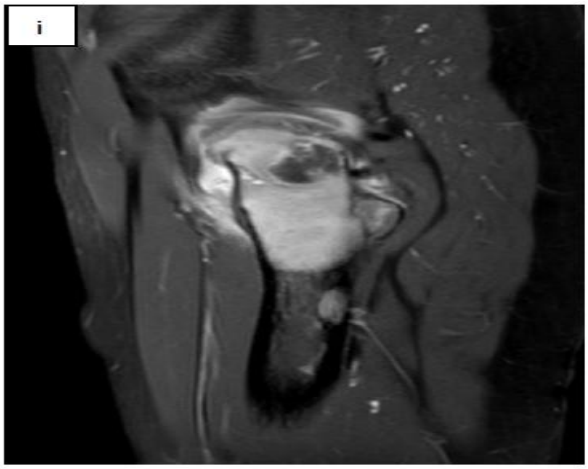
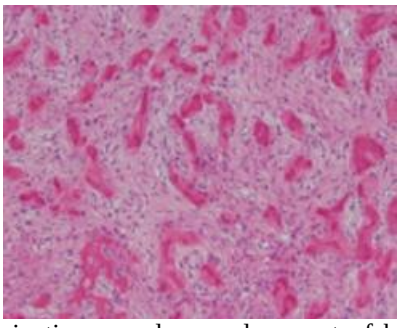


Fig h- showing diffusion restriction on DWI with heterogeneous post contrast enhancement (Fig i).

HISTOPATHOLOGY IMAGE



HPE examination reveals – replacement of bone tissue by loose fibrous tissue with irregular spicules of immature bone (hematoxylin and eosin stain)

DISCUSSION

FD is a common, benign skeletal disorder. The mutation at the Gsα gene at chromosome 20q13.2-13.3 is regarded as the etiologic cause of FD.[4]

In radiographs, FD presents as a well-marginated peripheral sclerotic bone lesion which may be lucent, sclerotic or mixed or may have a ground glass appearance, depending on the amount of bone trabeculae, fibrous elements and calcification.[5] Endosteal scalloping and focal cortical thinning may be present without cortical disruption or periosteal reaction.[6]

CT delineates the extent of skeletal involvement. [7]In MRI the lesions are largely isointense on T1w and heterogeneously hyperintense on T2w with patchy central or rim or homogeneous enhancement on post contrast images.[8] The histopathologic hallmark of the FD is fibrous tissue and immature, spindle-shaped, fibroblast-like cells within the bone marrow.[9]

CONCLUSION

Fibrous dysplasia denotes a subset of benign bone tumours manifesting in childhood or early adult life. These lesions are characterized by age related histological, radiographic and clinical transformations. Hence it is important to incorporate all the clinical, radiographic and pathologic features to diagnose FD.

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