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FIBROUS DYSPLASIA - A BENCH TO BIOPSY DISCUSSION

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A nine yrs old girl presented to the Outpatient Dept with pain in left leg since last 6 months. It was associated with pain in the hip and aggravated by movement. No associated history of pigmentation over the trunk or body, abnormalities in growth or similar symptoms in other parts of the body. Xray of the left leg revealed a translucent smooth lesion in the proximal part of femur with endosteal scalloping and cortical thinning in the neck of left femur (Fig 1).

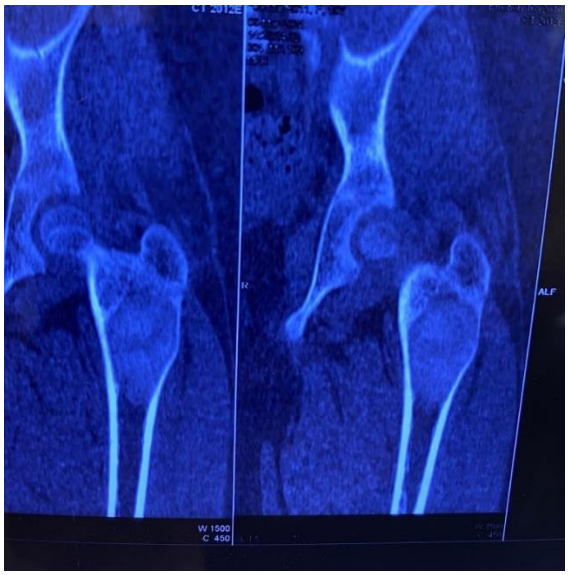

Fig 1: Xray of the hip shows a translucent smooth lesion in the proximal part of femur

No similar lesions or symptoms were seen in other parts of axial and appendicular skeleton. The patient was operated and sample processed for Histopathological examination. Hematoxylin and Eosin (H&E) stained sections from the specimen showed irregular osteoid deposition in a Chinese letter pattern within a fibrous stroma. The curvilinear osteoids were lined by osteocytes and no osteoblastic rimming was noted (Fig 2,3)

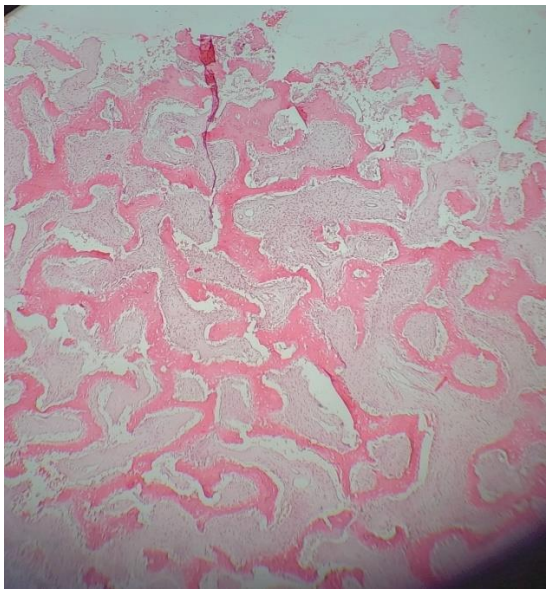

Fig 2: Histopathological examination of the lesion - Low power view H&E stained section shows mosaic pattern of fibrous tissue and irregular bone formation

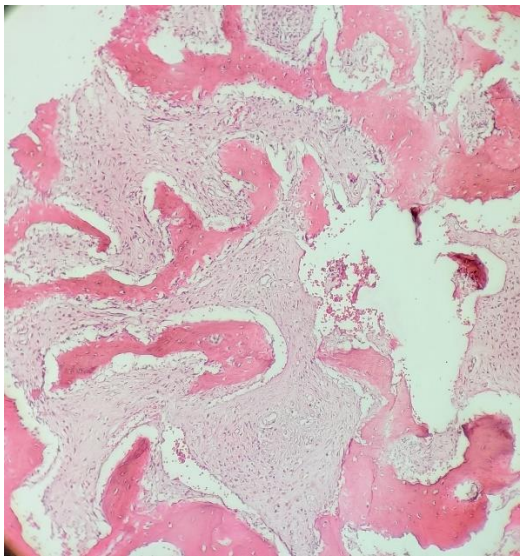

Fig 3: High power view H&E stained section shows irregular osteoid deposition (lined by osteocytes) in a Chinese letter pattern within a fibrous stroma

Discussion:

Fibrous dysplasia is a non-malignant disease characterized by transformed osteogenesis leading to an intramedullary fibro-osseous proliferation with fibrous and osseous tissue components being present in varying degrees.(1) It is found in a monostotic or polyostotic form and depends on whether only one single bone or multiple bones are affected. However, there is no evolution from the monostotic to the polyostotic form. (2)

Fibrous dysplasia was first defined by the American bone pathologist Louis Lichtenstein in 1938 and the clinical, radiological and histological spectrum of findings have been later characterized by him and his colleague Henry Louis Jaffe in 1942.(3) It is a condition affecting one, several or many bones, severe cases of which may present

abnormal pigmentation of skin, premature sexual development, hyperthyroidism or other extra-skeletal abnormalities. It is caused due to somatic, gain-of-function mutations in *GNAS.*[4] The mutations affecting the stimulatory alpha subunit of G protein (Gs) have been discovered in dysplastic bone lesions. The histological analysis of these lesions revealed that the mutations in Gs alpha caused aberrations in cells of the osteoblastic lineage and therefore in the bone matrix. Further in vitro analyses revealed that the abnormal deposition of immature bone matrix in fibrous dysplasia is an effect of decreased differentiation and increased proliferation of osteoblastic cells. Finally, the signalling pathway involved in these osteoblastic abnormalities has been understood. It is now apparent that the constitutive elevation in cAMP level.[5]

Monostotic FD cases form about 80% of patients with FD. The most common site of monostotic FD are the ribs, skull and femur. Most of the bony lesions become non-silent and clinically significant by the age of 10 years, with almost no new lesions appearing after the age of 15.[6] Malignant change of FD lesions is a rare complication, accounting for up to 2.5% of cases. Malignant transformations to Osteosarcoma, Fibrosarcoma, Chondrosarcoma and Malignant fibrohistiocytoma have been reported. [7]

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