Case Report

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Fibrous Dysplasia with Extensive Cartilagenous Differentiation: A Rare Case Report

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Abstract

Fibrous Dysplasia with extensive cartilaginous component is a rare variant of Fibrous Dysplasia. The median age of presentation is 30 years. It presents as a well defined expansile, lytic lesion of long bones. Most commonly involving the proximal femur. Histopathologically, it shows lobules of mature appearing cartilage within proliferating fibro-osseous component. Differentiating of FD with extensive cartilaginous component from other benign and malignant cartilaginous tumours is of utmost importance. We are presenting this case here for its rare presentation in the proximal humerus of an elderly patient.

Keywords: Fibrous Dysplasia; Fibrous Dysplasia with extensive cartilaginous component.

Introduction

Fibrous Dysplasia (FD) is a rare developmental anamoly of bone.[1] It is a genetic non inherited disorder which occurs due to missense mutation in chromosome 20.[2] It is a benign expansile lytic lesion involving the medulla of longbones. Proximal femur is the most common site in the long bones. It can also occur in skull, jaws and also sometimes ribs.

Fibrous dysplasia(FD) can occur at any age with majority seen between 4 to 30 years age group, with equal gender distribution. It can occur as monostotic, polyostotic or craniofacial forms.[3] Polyostotic form is associated with Mc cune – Albright syndrome. Vast majority presents as asypmtomatic monostotic lesions that are diagnosed incidentally or can present with pain and pathological fractures. FD with extensive cartilaginous differentiation in proximal humerus is an rare variant of fibrous dysplasia. It is also known as Fibrocartilagenous dysplasia(FCD).[4]

FCD presents as an intramedullary expansile lesion that can be radiodense to radiolucent depending on the relative proportions of the fibrous and osseous component.

Histopathologically FD is characterized by branched irregular trabeculae of woven bone in a fibrous stroma.[5] However FCD shows varying proportions of lobules of mature cartilage amidst the proliferating fibro osseous components.

Awareness about this entity is necessary to differentiate it from other benign or malignant cartilaginous bone tumours.

Case Presentation

A 60 year old female presented to the opd with complaints of shoulder pain for past 6 months. MRI of right shoulder showed a well defined lytic lesion in the head and neck region of humerus. CT guided biopsy of the lesion was done. Histopathological examination showed bland appearing chondrocytes in deep blue chondroid matrix with no significant increase in cellularity. Possibility of Enchondroma was suggested. Curettage of the lesion was done. Histopathological examination of the curetted fragments showed a lesion with mixture of

cartilaginous and fibro osseous component. (Fig. 1) Fibroosseous component is composed of branching and anastamosing irregular woven and lamellar bone in a fibrous stroma. (Fig. 2) Some of them, were rimmed by osteoblasts. (Fig. 3) Cartilagenous component is composed of islands of mature hyaline cartilage. Focal aneurismal bone cyst like areas are also seen. (Fig4) No pleomorphism/ mitoses/necrosis are seen. Features were suggestive of Fibrous Dysplasia With Extensive Cartilagenous Component (Fibrocatilagenous dysplasia). Immunohistochemistry carried out showed MIB labeling index as 1% in highly proliferative areas. CK and SATB2 did not highlight the lesional cells, (Fig. 5 and 6) there by ruling out the possibility of Chondrosarcoma.

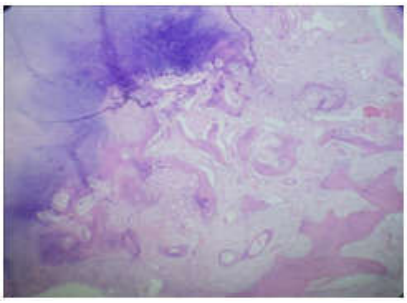

Fig. 1: FCD(10x) showing both cartilagenous fibrous components.

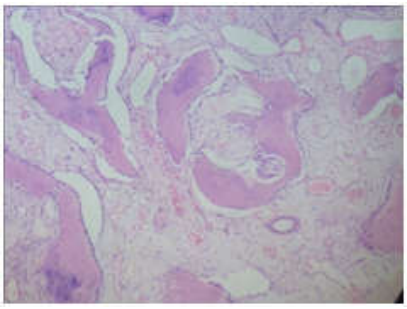

Fig 2: FCD(10x) showing with branched trabeculae of bone.

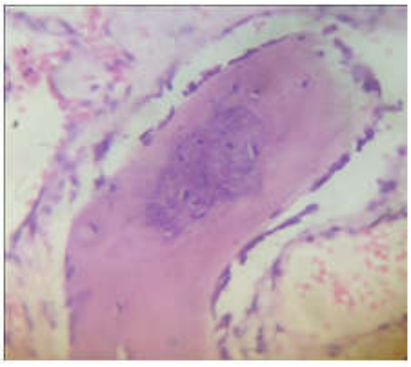

Fig 3: FCD(40x) showing bony trabeculae lined by osteoblasts.

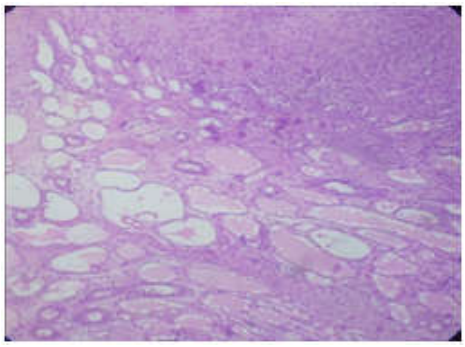

Fig 4: FCD(10x) showing aneurysmal bone cyst like areas.

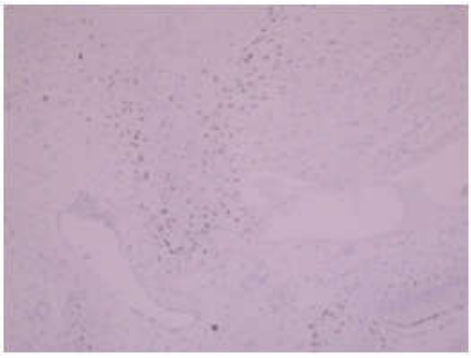

Fig 5: FCD-IHC(40x)SATB2-Negative.

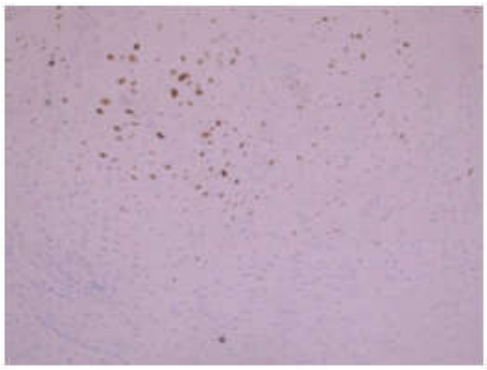

Fig 6: FCD: IHC-CK is also negative.

Discussion

FCD was first reported by Pelzmann et al in 1980 as a subtype of polyostotic FD.[6]

Fibrous dysplasia with extensive cartilaginous differentiation is a rare variant of FD. It is characterized by presence of varying amount of lobules of chondroid tissue amidst proliferating fibro osseous component. Malignant change in FD is very rare.

Histologically differentiating FD with extensive cartilaginous differentiation from benign or malignant cartilaginous tumours of bone is very

important.[7] Enchondroma, fibrocartilagenous mesenchymoma and chondrosarcoma were the closest histological differentials.

Enchondroma is a benign tumour that occurs in tubular bones of hands and feet. However it is rare in old age . Histologically it is composed of benign hyaline cartilage with no fibrous osseous component .

Fibrocartilagenous mesenchymoma is a locally aggressive neoplasm seen commonly in the metaphysis of long tubular bones. However the median age of presentation is 13 years. Histologically fibrocartilagenous mesenchyma shows spindle cell component shows mild hyperchromia.[8]

Chondrosarcoma is the closest differential as it is common in older age group and long bones is the common site. Histologically it shows double nucleated cells with mild to moderate atypia. Permeation of marrow space is an important feature. However proliferating fibrosseous component is not seen.

Conclusion

Fibrocartilagenous Dysplasia in the proximal humerus of an elderly female is a rare presentation. Awareness and knowledge about this entity is essential to differentiate it from other cartilaginous tumours including chondrosarcoma which is an important differential diagnosis. Reporting this rare variant will help us to avoid the diagnostic pitfalls and to analyse the clinicopathological aspects better.

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