Atypical Presentation of Fibrous Dysplasia - A Case Report

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Abstract
Fibrous dysplasia is a benign intramedullary disease in which area of trabecular bone is replaced by fibrous tissue containing flecks of osteoid and woven bone. Most common sites of involvement are long bones of lower extremity and base of skull. We here report a case of atypical presentation of fibrous dysplasia involving proximal radius in a 35-year-old female presented with pain and swelling with pathological fracture of proximal radius. Open core biopsy and MRI were suggestive of aneurysmal bone cyst. Treatment was done with extended curettage and filling the cavity with cancellous and cortical bone graft. Histological examination of material revealed fibrous dysplasia with secondary cystic lesion.

Keywords: Fibrous dysplasia, Osteolytic lesion, benign bone tumor

Introduction
Fibrous dysplasia is a developmental anomaly of bone formation that may exist in a monostotic or polyostotic form. The hallmark is replacement of normal bone and marrow by fibrous tissue and small, woven spicules of bone. Fibrous dysplasia can occur in the epiphysis, metaphysis, or diaphysis. Some syndromes associated with polyostotic form of fibrous dysplasia like McCune-Albright syndrome & Mazabraud syndrome.

Case Report
A 38-year-old female patient presented with pain in the right forearm for past 10 years. Her pain had intensified in the last 4 months due to fall 4 month back.

Examination revealed bulbous, fusiform swelling of 5×3 cm in size at right proximal forearm with normal range of motion at elbow and wrist. Rest of her general physical and systemic examination was unremarkable. Her routine blood tests, including ESR were normal. Roentgenogram revealed a wide cystic cavity with a lytic appearance causing cortical expansion in proximal aspect of right radius without involvement of elbow joint having fracture line in proximal radius as shown in Figure 1. NCCT of right elbow with forearm was obtained, which revealed expansile lytic lesion of proximal radius involving metaphysis and proximal diaphysis measuring approx 6X3 cms. There is thinning and ballooning of cortical margins. Matrix does not show any calcification. Transverse fracture of radius was noted distal to the lesion. CT opinion was Giant cell tumour as shown in
Figure: 2. MRI of right elbow with forearm was obtained, which revealed well defined expansile lesion in proximal radius appearing isointense on T1 and hypointense on T2 with focal hyperintensity with post contrast enhancement suggestive of Aneurysmal bone cyst as shown in figure 3. **Core Biopsy** was done which showed blood filled cavities suggestive of aneurysmal bone cyst as shown in figure 4.

**Treatment**

Open curettage and biopsy was done through henery’s approach to proximal radius, tumour site was exposed. A bone window was created on volar cortex. The mass was taken out through extended curettage. Non haemorragic, greyish white rubbery material was curettaged out and sent for histopathology. Subsequently, the bone defect created during curettage was filled with autograft from iliac crest bone. Histological examination of the material revealed fibro-osseous lesion of bone (fibrous dysplasia) with secondary cystic component as shown in Figure 6.

Figure : 1 Lateral view of Elbow showing lytic lesion in proximal radius  
Figure : 2 CT scan of Elbow showing expansile lytic lesion with fracture of proximal radius  
Figure : 3 MRI of elbow showing Isointense lesion on T1 and Hypointense lesion on T2

Figure : 4 Core Biopsy showing blood filled cavities suggestive of Aneurysmal bone cyst
Figure : 4 Histopathological examination showing blood filled spaces on Core Biopsy

Discussion

Fibrous dysplasia is a rare benign lesion of bones. Malignant transformations into highgrade-fibro- or osteosarcomas is very rare. Most lesions are monostotic, asymptomatic and identified incidentally. Most common involves long bones like femur, tibia, humerus and ribs. Polyostotic form usually occurs in association with syndromes like McCune-Albright syndrome & Mazabraud syndrome.

On X-ray characteristic ground glass appearance, with reactive peripheral sclerosis, bone expansions, indentations of the inner cortical area is seen. In case of fibrous dysplasia pathological fractures can occur. Curettage and the filling up of individual foci have gained importance in the management of monostic form. But in the case of the polyostotic form bisphosphonate therapy is used⁴. Involvement of proximal radius like in this case is very rare.

Conclusion

Fibrous dysplasia although most commonly involves long bones of lower limbs and cranium but should be considered in differential of any osteolytic lesion. Therefore, this case is a good example of the fact that fibrous dysplasia though rare must be taken into account when using differential diagnosis as one of the cystic lesion.

References

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