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Case Report

A Rare Case of Fibrous Dysplasia of Clavicle

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Abstract

Fibrous dysplasia is a rare entity 0f long bones and these lesions are uncommon in childhood, generally illustrated in the mandible and the maxilla. Tibia is among the long bones in which it typically appears as a painless lump or anterior bowing. Here, we would like to report a rare case of fibrous dysplasia of clavicle in a 15 year old female who presented to us with painful swollen area over her left clavicle region. On radiograph there was visible lytic lesion in the middle 3rd of left clavicle with subtle periosteal reaction. Histopathological features suggested benign fibro-osseous proliferation, favouring fibrous dysplasia. To the best of our knowledge, there are very few cases of fibrous dysplasia affecting clavicle to be reported in the literature.

***Keywords:** Fibrous Dysplasia, Clavicle, Osteolytic Lesion; Ossifying Fibroma, Histopathology*

Introduction

Fibrous dysplasia is a non-neoplastic uncommon condition of unknown etiology affecting the long bones (1). Majority of lesions fibrous dysplasia appears on the anterior cortex of tibia. In the literature it is still not considered a true neoplasm, but rather as a fibrovascular defect (2). Osteofibrous dysplasia was first described by Frangenheim in 1921 (3) after that several cases affecting the tibia have been reported (4-6). There are few reports describing involvement of ipsilateral fibula (5,6), lesions in humerus (7) and in radius (8). The exact etiology of this lesion is still not clear. Most commonly it is seen in the first two decades of life and almost always before the puberty. Osteofibrous dysplasia is found most commonly in maxilla and mandible than in long bones. In long bones, tibia is most frequent site which is involved, followed by fibula, radius, ulna and humerus. Clavicle has rarely been reported as a site of occurrence of osteofibrous dysplasia lesion. The case report we present a rare case of fibrous dysplasia lesion in mid clavicle area in a 15 year old girl.

Case Report

A 15 year old post-pubertal girl presented with pain and swelling over her left clavicle region which was incidious onset without any history of trauma and gradually progressing over a period of 6 months. There was no other history suggestive of weight loss or constitutional symptoms. On clinical examination the swelling was diffuse, 27 mm x 44 mm x 25 mm in size, originating from midshaft Clavicle, nonmobile, not adherent to overlying skin. Overlying skin was normal. The swelling was tender, ill-defined with bony hard consistency. Left shoulder range of movement was painful to some extent with some restriction due to pain in clavicle on abduction beyond 100 degrees. Rest of shoulder joint examination was normal. There was no tenderness over the left shoulder joint.

There was no significant axillary or cervical lymphadenopathy. Distal neurovascular status was intact. The total and differential blood leukocyte count, CRP and erythrocyte sedimentation rate were within the normal range. Plain radiographs of left clavicle (Figure 1) anteroposterior view revealed a thinning out of superior and inferior cortices of midshaft area of left clavicle with ill-defined cortical outline at places and there was a clearly well-defined lytic lesion. There are some radiolucent areas and intervening calcified septae. Computed tomography showed expansile lytic lesion in middle 1/3rd of left clavicle causing destruction of underlying bone along with subtle periosteal reaction. On post contrast, there is no obvious contrast enhancement. These CT findings were suggestive of possibility of aneurysmal bone cyst or ewings sarcoma and suggested for histopathological correlation. Magnetic resonance imaging (Figure 2) showed lobulated expansile abnormal signal intensity mass which appeared hyperintense on T2 and STIR with hypointense on T1 sequence involving middle 1/3rd of left clavicle with internal fluid level. After multidisciplinary team meeting the decision was taken to plan for curettage and grafting of the lesion. After proper consenting of the patient and explaining parents about the risks and benefits of the procedure, patient was taken to theatre and under general anaesthesia the curettage and grafting was done. Skin incision was given over the midshaft clavicle area just above the lesion, deeper tissue dissection done and lesion was reached. Then, a small bone window created over the superior cortex portion over the lesion after drilling three to four times (Figure 3). The lesion was thoroughly curetted and the curetted material sent for histopathology. The bone cavity in clavicle was filled with hydroxyapatite crystals. Haemostasis was maintained and wound was closed in layers. Postoperative period was uneventful. Post-operatively patient recovered well and sutures were removed on day 14 and the immediate post-operative x-ray looked satisfactory (Figure 4). For first 2 weeks post surgery patient was placed on shoulder immobilizer. After suture removal, gradual range of movements of shoulder were started. Patient was advised not to lift any heavy objects for at least 6 to 8 weeks post surgery.

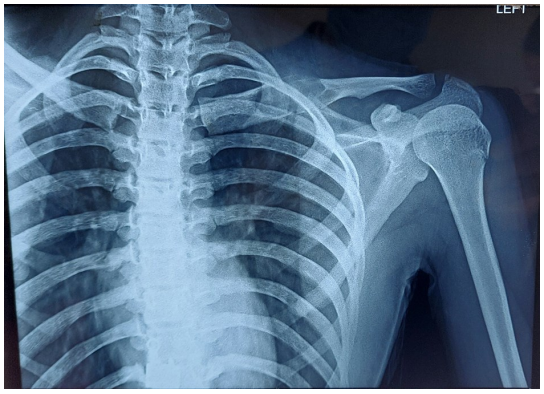

Figure 1: *Pre-operative plain radiograph showing the lesion in the left Clavicle.*

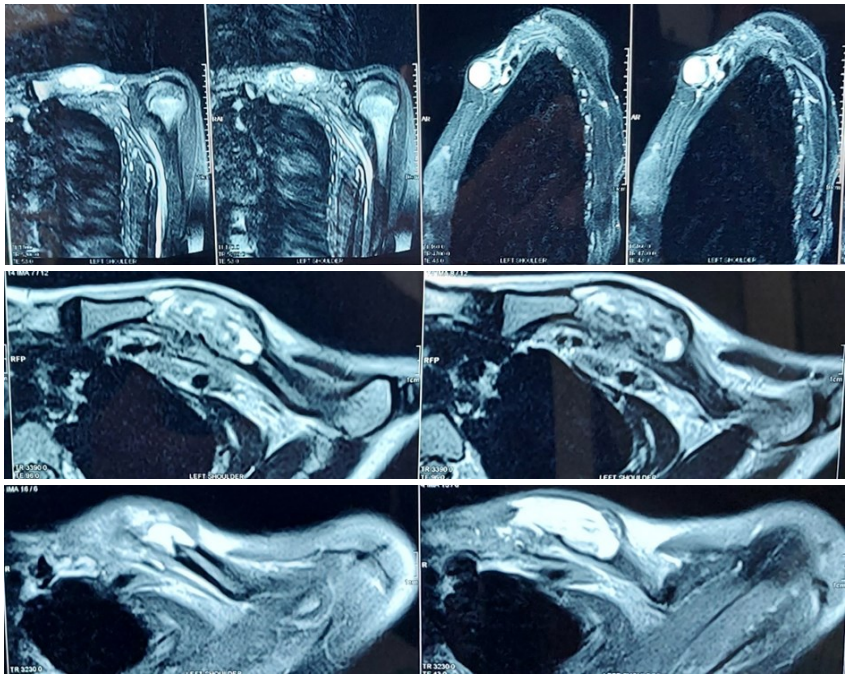

Figure 2: *Pre-operative MRI images showing the nature of the lesion in the left Clavicle.*

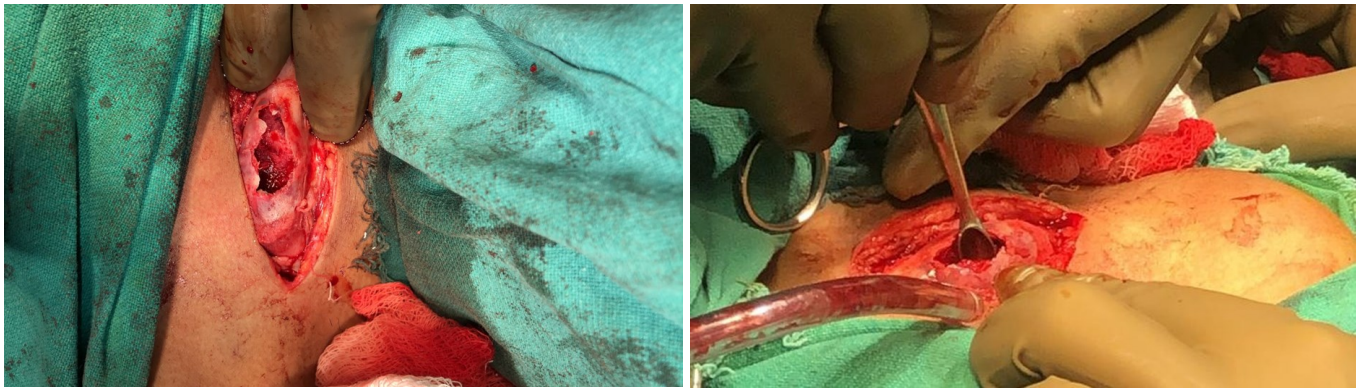

Figure 3: *Intra-operative picture showing the bone window over the left Clavicle and the curettage being done.*

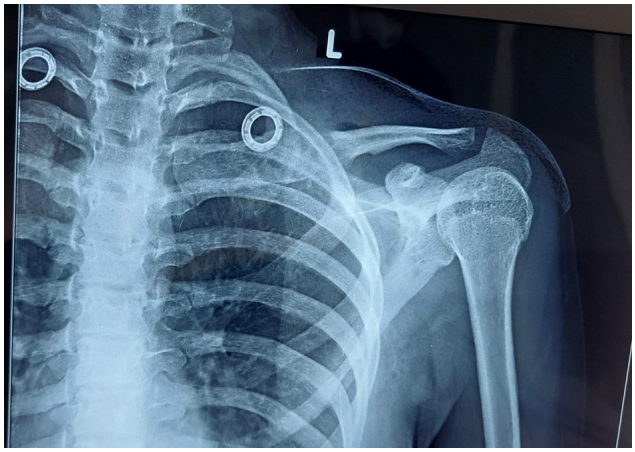

Figure 4: *Immediate Post-operative plain radiograph showing the adequate filling up of hydroxyapatite crystals within the left Clavicle after curettage.*

On histopathological examination of slides prepared from lesion revealed presence of cellular proliferation of plump spindle cells with production of collagen. Cells showed no mitosis, no cytologic atypia. Islands of woven bone seen with mineralization, osteoblastic rimming is not seen in woven bone. Areas of aneurysmal bone cyst like changes also seen with presence of telangiectatic blood filled spaces. Spaces are not lined by endothelium. Spindle cells proliferation seen. These histopathological features were suggestive of benign fibro osseous proliferation, favoring fibrous dysplasia with aneurysmal bone cyst like changes.

At 8 weeks post surgery, patient was able to perform all activities of daily living without any discomfort. Patient was followed up after 3 months with a normal X-ray with satisfactory consolidation of midshaft left Clavicle (Figure 5). In her recent x-ray which was around 8 months post operatively showed good remodeling of lesion (Figure 6).

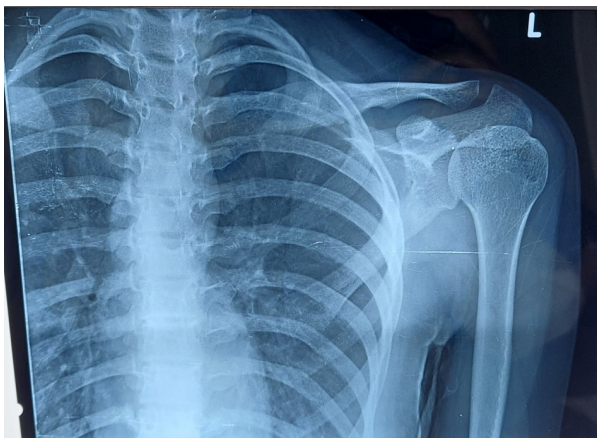

Figure 5: *3 months Post-operative follow up check X-ray of left Clavicle.*

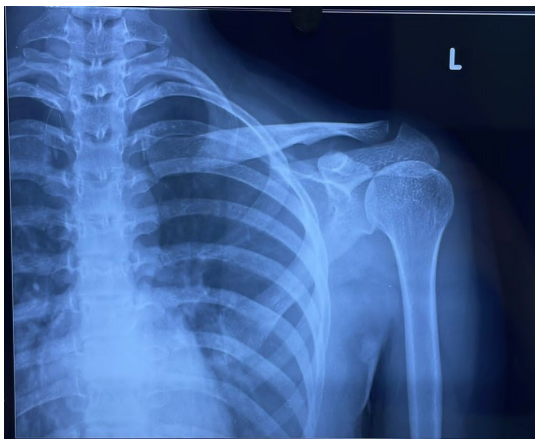

Figure 6: *8 months Post-operative follow up plain radiograph of showing good consolidation and remodelling of midshaft left Clavicle.*

Discussion

Fibrous dysplasia is one of the rare non-neoplastic disorder. We report here a patient who is 15 year old female with this lesion in clavicle. Usually, it is observed in a child under the age of 10 years with peak incidence in the age group of 1 -5 years. Although there are reports in literature about adult patients diagnosed with fibrous dysplasia with the oldest patient of 39 years (4).

Causes for osteofibrous dysplasia are still unclear. There is a school of thought mentioning that it may arise from fibro-vascular defect. In a paper by Johnson et al (2) a relationship between osteofibrous dysplasia and adamantinoma was explained. As per his research article, fibrous dysplasia occurs because of the defect of Haversian canal while the adamantinoma is the consequence of anomaly of intramedullary vasculature.

The most common plain radiograph presentation of fibrous dysplasia is an eccentric intracortical lytic lesion in tibia with no associated soft tissue extension or periosteal reaction (8,9,10). It is a well defined lesion with a rim of sclerosis. There may be expansion or thinning out of the overlying cortex. Pathological bowing and/or pathological fracture of the involved bone might also be present sometimes. In our case, there was a thinning out of superior and inferior cortices of midshaft area of left clavicle with ill-defined cortical outline at places and there was a clearly defined lytic lesion.

The natural history of presentation of fibrous dysplasia is unpredictable. It may regress naturally or may develop swiftly on skeletal maturity with time. We did curettage and grafting with hydroxyapatite crystals for our case. Non-surgical treatment is usually suggested till skeletal maturity by few authors but surgical extraperiosteal marginal resection has also been mentioned in the literature, (11) as there are high recurrence rate reported for these lesions following curettage and bone grafting. In our case, we haven't seen any signs of recurrence till now.

Adamantinoma, non-ossifying fibroma and fibrous dysplasia are the close differential diagnoses of fibrous dysplasia which can be differentiated from each other based on their salient clinical, radiological, and histopathological characteristics. Osteofibrous dysplasia is mostly seen after the first decade of life with involvement of tibia, femur and/or ribs with typical intramedullary ground glass appearance on plain radiographs (12). Adamantinoma is a malignant neoplasm found in the tibia usually after the first decade, and radiologically it differs from fibrous dysplasia by its typical intramedullary involvement, periosteal reaction and soft tissue extension in the absence of pathological fracture (13). Histologically it shows hyperchromatic epithelial islands of malignant cells. Metaphyseal location is typical for non-ossifying fibroma on radiology while histology shows scattered multinucleate giant cells and storiform pattern of spindle cells. Some of the other differentials includes chronic recurrent multifocal osteomyelitis (CRMO), SAPHO (synovitis, acne, palmo plantar pustulosis, hyperostosis, and osteitis) syndrome, eosinophilic granuloma, vascular tumours, Ewing's sarcoma, and osteoblastoma (10,14,15). CRMO most commonly involves medial third of clavicle in all age groups, and it appears on plain radiographs as a sclerotic expansile lesion (13), and apart from clavicular involvement, there is distinctive involvement of the sterno clavicular joint, sterno costal joint, and ossification of the costo clavicular ligaments in SAPHO syndrome. Histopathologically is the key to differentiate ossifying fibroma from CRMO and SAPHO, as ossifying fibroma will show only nonspecific inflammatory changes (16).

Surgical treatment for fibrous dysplasia is delayed until puberty. Most common indication for surgery are the large painful lesions, lesions with pathological fracture and/or bony deformity. Marginal resection with bone grafting is usually required. These lesions have tendency for recurrence, particularly if surgery is done before skeletal maturity (17). The present case is exceptional in this aspect as the lesion in the left clavicle was not only noticed 3 years after puberty but also it showed progressive symptoms rather than undergoing regression. Therefore, we planned to perform curettage and grafting of the lesion. And patient recovered well after surgery with good remodelling of the hydroxyapatite crystals in the bone. There were no post-operative complications reported by patient and no signs of any recurrence on recent plain radiographs which are 8 months post-surgery.

Conclusion

Fibrous dysplasia is a rare non-neoplastic lesion which can present in an uncommon location such as clavicle. There are several differential diagnosis of these kind of lesions in clavicle. With radiological investigations, it might be challenging to rule out other differential diagnosis of clavicular enlargement. Histopathology is crucial for the confirmation of fibrous dysplasia in rare sites like clavicle.

Conflict of Interest

The authors declare no conflict of interest.

References

1. McCaffrey M, Letts M, Carpenter B, Kabir A, Davidson D, Seip J. Osteofibrous dysplasia: a review of the literature and presentation of an additional 3 cases. Am J Orthop. 2003 Oct;32(10):479-86.
2. Johnson LC. Congenital pseudoarthrosis, adamantinoma of long bone and intracortical fibrous dysplasia of the tibia. J Bone Joint Surg 1972;54A: 1355.
3. Frangenheim P. Angeborene ostitis fibrosa als ursache einer intrauterinen unterschenkelfraktur. Arch Klin Chir. 1921;117:22-9.
4. Sweet DE, Vinh TN, Devaney K. Cortical Osteofibrous dysplasia of long bone and its relationship to adamantinoma. A clinicopathologic study of 30 cases. Am J Surg Pathol. 1992 Mar;16(3):282-90.
5. Campanacci M, Laus M. Osteofibrous dysplasia of the tibia and fibula. J Bone Joint Surg [Am]. 1981 Mar;63(3):367-75.
6. Ishida T, Iijima T, Kikuchi F. A clinicopathological and immunohistochemical study of osteofibrous dysplasia, differentiated adamantinoma, and adamantinoma of long bones. Skeletal Radiol. 1992;21(8):493-502.
7. Schlitter HE. Benign bone tumors with unusual localization & spontaneous fracture. Fortschr Geb Rontgenstr Nuklearmed. 1958 Feb;88(2):195-200.
8. Wang JW, Shih CH, Chen WJ. Osteofibrous dysplasia (ossifying fibroma of long bones). A report of four cases and review of the literature. Clin Orthop. 1992 May;(278):235-43
9. Ozaki T, Hamada M, Sugihara S, Kunisada T, Mitani S, Inoue H. Treatment outcome of osteofibrous dysplasia. J Pediatr Orthop B 1998;7:199-202.
10. Campanacci M. Osteofibrous dysplasia of long bones a new clinical entity. Ital J Orthop Traumatol 1976;2:221-37.
11. Lee RS, Weitzel S, Eastwood EW, Monsell F, Pringle J, Cannon SR, et al. Osteofibrous dysplasia of tibia: is there a need for radical surgical approach? J Bone Joint Surg [Br]. 2006;88-B:658-64.
12. Grabias SL, Campbell CJ. Fibrous dysplasia. OrthopClin North Am 1977;8:771-83.
13. Most MJ, Sim FH, Inwards CY. Osteofibrous dysplasia and adamantinoma. J Am Acad Orthop Surg 2010;18:358-66
14. Park YK, Unni KK, McLeod RA, Pritchard DJ. Osteofibrous dysplasia: Clinicopathologic study of 80 cases. Hum Pathol 1993;24:1339-47
15. Suresh S, Saifuddin A. Unveiling the 'unique bone': A study of the distribution of focal clavicular lesions. Skeletal Radiol 2008;37:749-56.
16. Bjorkstén B, Boquist L. Histopathological aspects of chronic recurrent multifocal osteomyelitis. J Bone Joint Surg Br 1980;62:376-80.
17. Hahn SB, Kim SH, ChoNH, Choi CJ, Kim BS, Kang HJ. Treatment of osteofibrous dysplasia and associated lesions. Yonsei Med J 2007;48:502-10.

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