

Osteofibrous dysplasia of clavicle clinically mimicking chronic osteomyelitis

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Abstract

Osteofibrous dysplasia or ossifying fibroma is an uncommon benign fibro-osseous lesion of childhood, commonly described in the maxilla and the mandible. Among long bones, it usually presents in the tibia as a painless swelling or anterior bowing. Ossifying fibroma of clavicle has never been reported in English literature, to the best of our knowledge. Here, we would like to present an unusual case of osteofibrous dysplasia of clavicle clinically mimicking chronic osteomyelitis.

Key words: Chronic osteomyelitis; clavicle; ossifying fibroma; osteofibrous dysplasia

Introduction

The term “osteofibrous dysplasia” is synonymous with ossifying fibroma and Campanacci’s disease.^[1,2] It is not considered as a true neoplasm, but rather as a fibrovascular defect.^[3] The exact cause of this rare benign fibro-osseous lesion is still an enigma. This distinct entity is commonly seen in the first two decades almost always before puberty, with a slight male predilection. These lesions are more frequent in the maxilla and mandible than in long bones. Among the latter, tibia is the most common site of its occurrence, followed by fibula, radius, ulna, and humerus.^[4-6] Its occurrence in clavicle has never been reported. The usual clinical presentation is that of a painless swelling, incidentally noticed by the parents, along the shin bone or abnormal bowing of tibia.^[2] There has been no report of this lesion mimicking osteomyelitis

clinically. Surgery, if required, is delayed until puberty and it usually requires nothing more than marginal excision and bone grafting.^[7,8] This case report brings forth the unusual presentation of osteofibrous dysplasia of the clavicle mimicking a case of chronic osteomyelitis clinically, which was successfully treated by *en bloc* excision of clavicle.

Case Report

A 14-year-old post-pubertal girl presented with complaints of pain and swelling of her left collar bone which was of insidious onset and gradually progressive over a period of 1 year. There was history of fever on and off, along with history of intermittent serous discharge from a wound over the swelling over the past 6 months. There was a history of biopsy being done outside which was an inadequate sample to be commented upon. On examination, the patient was febrile. There was a diffuse swelling of the left clavicle extending from the sternal end up to the left shoulder. Skin over the swelling showed a healed biopsy scar mark. There was a discharging sinus adjacent to the scar mark. On palpation, there was local rise of temperature. The swelling was tender, ill-defined with bony hard consistency. There were four discrete, non-tender lymph nodes palpable, two each, in the axillary group and the cervical group, largest of them measuring 0.5 cm × 0.5 cm in dimension. Left shoulder range of motion was painful and globally

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restricted. A provisional diagnosis of acute on chronic osteomyelitis was made. The erythrocyte sedimentation rate was high (40 mm/h). The total and differential blood leukocyte counts revealed leukocytosis with a neutrophilic preponderance. Mantoux test was equivocal. The wound discharge was subjected to bacterial and acid-fast bacilli staining and culture, which subsequently turned out to be sterile, adding on to the diagnostic dilemma.

Plain radiographs revealed a diffuse periosteal thickening encircling the clavicle extending from its sternal end to lateral third [Figure 1]. Shoulder joint was normal. Computed tomography (64-detector CT scanner, Lightspeed® VCT, GE Healthcare, Buckinghamshire, UK) showed irregularity and sclerosis of left clavicle along with heterogeneous ossification [Figure 2]. Magnetic resonance imaging (3.0 Tesla MR scanner, Magnetom Verio, Siemens, Erlangen, Germany) showed altered heterogeneous T2 signal within the marrow of the clavicle. The cortical delineation was ill-defined at places with gross periosteal thickening [Figure 3]. Tc99m labeled methylene diphosphonate (MDP) bone scan showed evidence of active bony inflammation in the left clavicle. A repeat biopsy showed immature woven bone with characteristic osteoblastic rimming in a background of fibrovascular stroma. On immunohistochemistry utilizing pan cytokeratin antibody stain, isolated cells in the fibrous stroma were

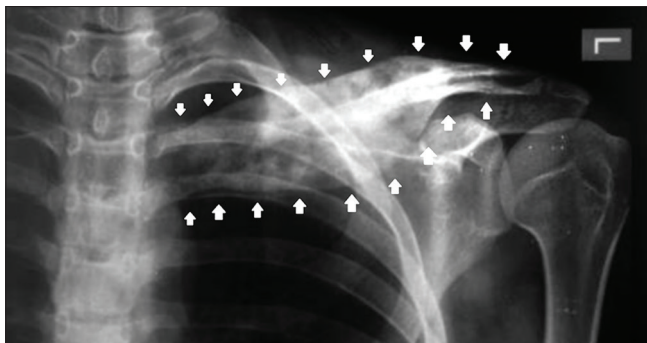


Figure 1: Plain radiograph showing a diffuse periosteal thickening of the left clavicle

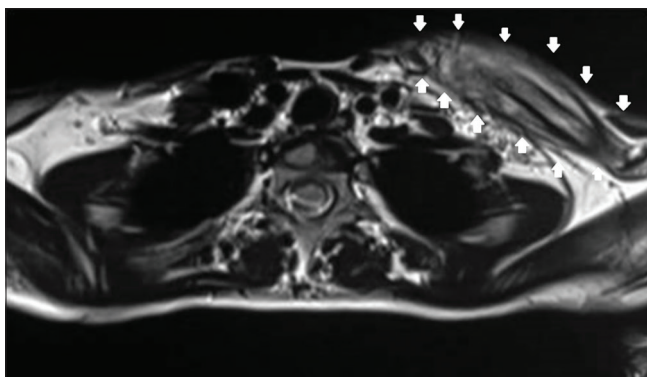


Figure 3: MRI, T2-weighted (axial section) showing heterogeneous signal within the marrow and grossly thickened periosteum encircling the clavicle. The cortical outline is ill-defined at places

cytokeratin (AE1/AE3 + CK-1) positive. Both histopathology and immunohistochemistry helped in clinching the final diagnosis-ossifying fibroma [Figure 4]. The patient had progressive worsening of symptoms-increase in swelling size along with active signs of inflammation in spite of starting empirical antibiotics -warranting a surgical intervention. As the entire clavicle was found involved by the disease process, excision of the lesion en masse with the clavicle in toto was done [Figure 5]. Postoperative period was uneventful.

Discussion

The most frequent radiographic picture of osteofibrous dysplasia is that of an eccentric intracortical lytic lesion in tibia without any associated periosteal reaction or soft tissue extension.^[2,6] The lesion is well defined by a rim of sclerosis. The overlying cortex may be expanded and thinned out. There might be associated pathological bowing and/or pathological fracture of the involved bone. In our case, there was a diffuse periosteal thickening of the clavicle with ill-defined cortical outline at places.

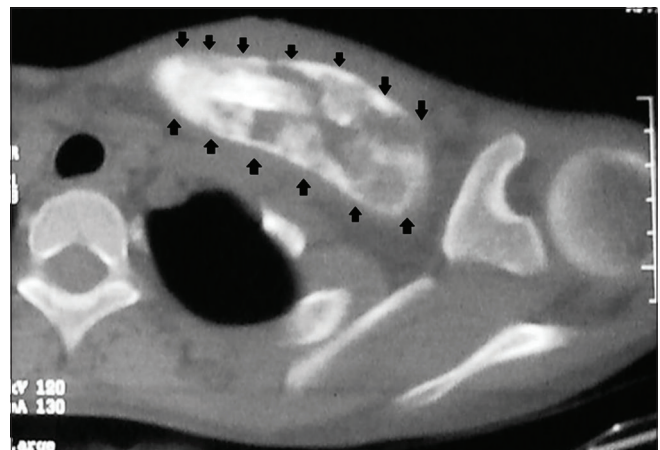


Figure 2: CT scan bony window (axial section) showing heterogeneous ossification with non-ossified soft tissue surrounding the left clavicle

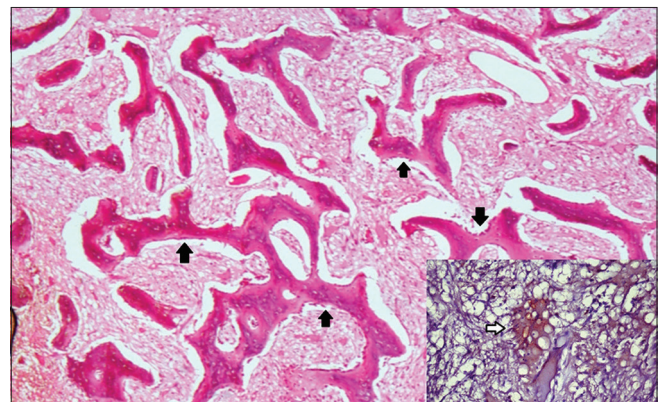


Figure 4: Histopathologic photomicrograph showing immature woven bone in a background of benign fibrovascular stroma. The immature woven bone shows osteoblastic rimming which is characteristic of osteofibrous dysplasia. (Inset) Immunohistochemistry showing isolated cytokeratin (AE1/AE3 + CK-1) positive cells in the stroma

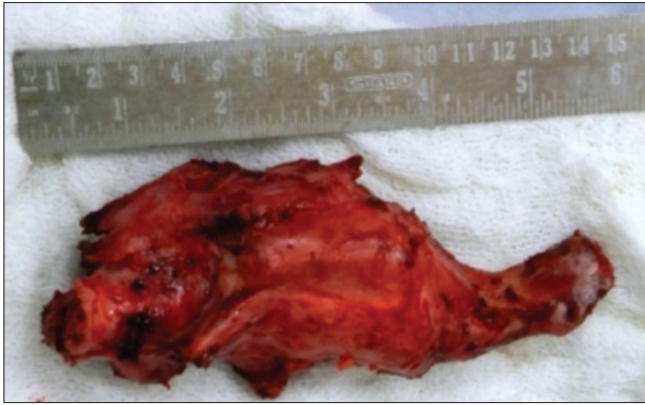


Figure 5: Gross photograph of surgically resected clavicle

The close differential diagnoses of osteofibrous dysplasia include adamantinoma, monostotic fibrous dysplasia, and non-ossifying fibroma, which can be differentiated from each other based on their salient clinical, radiological, histopathological, and immunohistochemical characteristics. Fibrous dysplasia is seen after the first decade involving femur and/or ribs with typical intramedullary ground-glass appearance on radiographs.^[9] There is neither osteoblastic rimming on histology nor cytokeratin positivity on immunohistochemistry. Sweet *et al.* differentiated osteofibrous dysplasia from fibrous dysplasia based on immunohistochemistry-scattered, isolated cytokeratin (AE1/AE3 + CK-1) positive cells were seen in the stroma of osteofibrous dysplasia, but not in fibrous dysplasia.^[10] Non-ossifying fibroma is typically seen in metaphyseal location on radiology. Histology shows storiform pattern of spindle cells and scattered multinucleate giant cells. Adamantinoma is a cytokeratin-positive malignant neoplasm seen in the tibia usually after the first decade, which radiologically differs from osteofibrous dysplasia by its characteristic intramedullary involvement, soft tissue extension, and periosteal reaction in the absence of pathological fracture.^[11] Histologically it shows hyperchromatic epithelial islands of malignant cells. The other differentials include chronic recurrent multifocal osteomyelitis (CRMO), SAPHO (synovitis, acne, palmo-plantar pustulosis, hyperostosis, and osteitis) syndrome, osteoblastoma, eosinophilic granuloma, Ewing's sarcoma, and vascular tumors.^[12,13] In fact, CRMO is the most common disease process involving the medial third of clavicle in all age groups, radiologically presenting as a sclerotic expansile lesion,^[13] whereas in addition to clavicular involvement, there is distinct involvement of the sterno-clavicular joint, sterno-costal joint, and ossification of the costo-clavicular ligaments in SAPHO syndrome. Both CRMO and SAPHO syndrome can be differentiated from ossifying fibroma histopathologically, which shows only non-specific inflammatory changes.^[14]

Most of the small lesions of osteofibrous dysplasia undergo spontaneous resolution after puberty.^[15] Hence, treatment,

if required, is delayed until puberty. Large painful lesions, lesions with pathological fracture and/or bony deformity are usual indications for surgery. Marginal resection with bone grafting is usually required. These lesions are notorious for their recurrence, especially if operated before skeletal maturity.^[7,8] The present case is unique in this aspect as the lesion was not only noticed 2 years after puberty but also became progressively symptomatic rather than undergoing regression. Also, in our case, involvement of clavicle clinically mimicking osteomyelitis was a unique presentation. Even though our patient had fever, discharging sinus, and leukocytosis, we could not isolate any bacteria on staining and cultures. This could have been negative due to previous antibiotics as the patient was being treated elsewhere for past 6 months as per history. *En bloc* excision of clavicle was planned for in this case after taking various factors into consideration such as the extent of lesion, chronicity of the lesion, failure to respond to antibiotics, and persistence of symptoms like pain, swelling, fever, and wound discharge associated with the lesion.

Conclusion

Osteofibrous Dysplasia can present in an uncommon location like clavicle. It can clinically mimic osteomyelitis. Radiologically, it may be difficult to rule out other differential diagnoses of clavicular enlargement. Histopathology and immunohistochemistry are indispensable for clinching the diagnosis.

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