A Rare Recurrent Chondromyxoid Fibroma of the Proximal Tibia in an Adolescent Female

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Abstract

Introduction: Lytic lesions of the bone are often seen in children and young adults. These are usually benign but sometimes are locally aggressive bone tumors. Lesser known causes of lytic lesions are chondromyxoid fibroma (CMF) and its recurrence is very rare. The accepted treatment for CMF is surgical curettage with bone grafting and the review of the literature shows that recurrence rate is 3–22%.

Case Report: We present a rare case of an adolescent girl of 14 years who presented with a recurrent bone tumor of the proximal tibia, after 4 years of the primary curettage. It turned out to be a CMF. She presented with swelling over the right upper leg for the past month. It was gradual in onset, progressive, and non-radiating in nature. There was no history of trauma. She had had a similar complaint 4 years back in 2016, for which she was operated elsewhere, where curettage and synthetic bone substitutes were used for filling the bone cavity. She was taken up for surgery for extended curettage and bone cementing. The histopathological examination was suggestive of CMF. Subsequent follow-up, she reported complete resolution of symptoms with a pain-free range of knee movements and no radiological signs of recurrence.

Conclusion: The accepted treatment for CMF is surgical curettage with bone grafting, but the recurrence rate is 3–22%.

Keywords: Chondromyxoid fibroma, benign lesions, tibia, recurrence.

Introduction

Lytic lesions of the bone are often seen in children and young adults. These are usually benign but sometimes are locally aggressive bone tumors. These lesions are often asymptomatic and are diagnosed incidentally on radiographs or when these present with symptoms of pain, swelling, and pathological fracture [1]. The commonly found benign osteolytic lesions in younger individuals include unicameral and aneurysmal bone cysts and fibrous dysplasia [2,3]. However, these conditions need to be differentiated from the lesser-known causes of lytic lesions such as giant cell tumor , chondromyxoid fibroma (CMF), fibrocartilaginous dysplasia , osteofibrous dysplasia ,desmoplastic tumor and browns tumor as mentioned in Table 1 [2-9].

We present a rare case of an adolescent girl of 14 years who presented with a recurrent bone tumor of the proximal tibia, after 4 years of the primary curettage. It turned out to be a CMF.

Case Report

A 14-year-old female presented with swelling over the right upper leg for the past month. It was gradual in onset, progressive, and non-radiating in nature. There was no history of trauma. She had had a similar complaint 4 years back in 2016, for which she was operated elsewhere, where curettage and synthetic bone substitutes were used for filling the bone cavity (Fig. 1 and 2). The histology was suggestive of chondromyxoid fibroma (CMF).

Four years after the first surgery, this young lady presented with a recurrence of pain and swelling at the operated site. The plain radiographs confirmed the presence of a large lytic lesion in the proximal tibia with lobulations, but the cortices seemed intact (Fig. 3). Magnetic resonance imaging of the right knee joint (T2-weighted images) showed well-defined multiple cystic lesions arising from the anterior aspect of proximal meta-diaphysis of the tibia. The lesion had irregular margins; the overlying cortex was intact with no definite soft-tissue involvement and periosteal thickening or reaction was seen (Fig. 4).

Treatment

She was taken up for surgery for extended curettage and bone cementing after obtaining informed consent from the parents and explaining all the risks, benefits, and alternatives of the operation. The lesion was approached after excising the previous scar. A cortical window was created anterolaterally, and the tumor tissue was curetted out thoroughly using fiber-optic cable light. The walls of the bony cavity were then cleaned using a high-speed burr. The cavity was then treated with 10% phenol for 15 min. After

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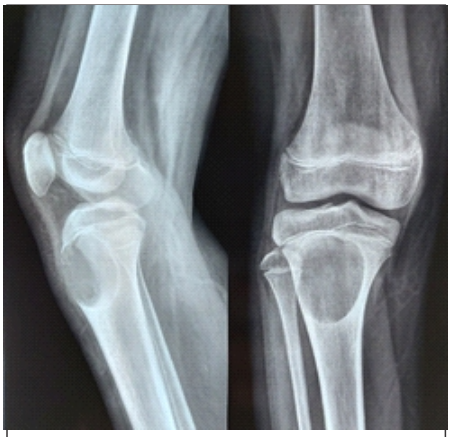

Figure 1: Plain radiograph (AP and lateral views) showing a large lytic lesion in the proximal tibia.

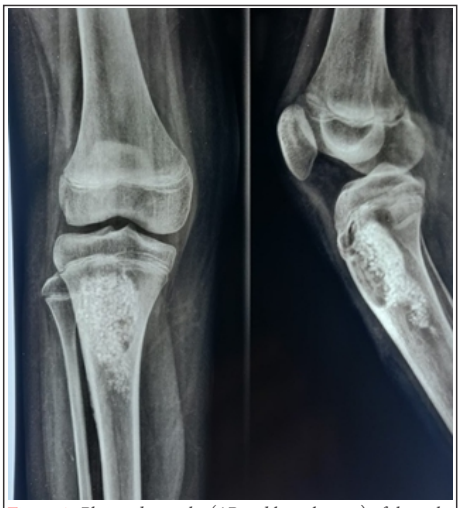

Figure 2: Plain radiographs (AP and lateral views) of the right knee joint, showing bone substitute filling of the upper tibial lesion.

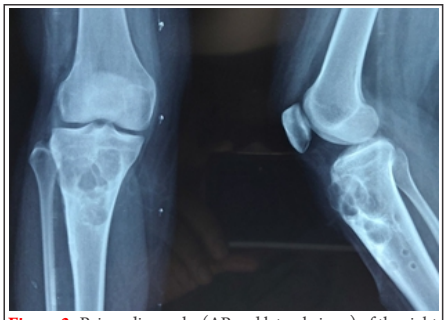

Figure 3: Pain radiographs (AP and lateral views) of the right knee joint showing recurrence of a multiple lobulated lytic lesion in the proximal tibia.

extended curettage, the cavity was filled up with gentamycin-loaded bone cement (Palacos-GR) (Fig. 5).The removed tumor tissue was firm to solid in consistency, and yellowish-red in color, with no blood in the cavity.

The histopathological examination was suggestive of CMF (Fig. 6). The tissue showed a mixture of chondroid, fibrous, and myxoid elements in varying proportions. Numerous hypocellular lobules of poorly formed hyaline cartilage were seen composed of chondroblasts with abundant pink cytoplasm and myxoid tissue with fibrous septae containing spindle cells and numerous osteoclasts like giant cells. These lesions were more cellular at the periphery of the nodules. Tumor cells present in lacunae in myxoid areas, stellate in myxoid areas with long delicate cell processes that approach other cells. Focal regions of tumors were noted close to blood clots. Marked degenerative changes and hyalinized changes were seen. No significant increase in mitotic activity or necrosis was observed. No direct osteoid or bone formation by tumors cells. No rims of woven bone or osteoblastic rimming were seen in multiple examined sections.

The patient was kept non-weight-bearing for 6 weeks and gradual weight-bearing until 3 months. The range of motion of the knee was started from the beginning. At 6-month follow-up, she had a full range of painless motion of the knee.

Discussion

CMF is a benign and extremely rare bone tumor accounting for less than 1% of benign and malignant bone tumors [10, 11]. It is a benign cartilaginous tumor which was described in 1948 by Jaffe and Lichtenstein [12, 13, 14]. CMF predominantly affects adolescents and young adults in the second or third decade of life [10, 13, 15]. The CMF is most commonly seen in the lower extremity, particularly the proximal end of tibia. About 95% of cases of CMF are seen in metaphyseal regions of the long bones. The clinical presentation varies according to the area involved and is associated with long-standing history of non-specific symptoms such as pain and edema. Usually, CMF is a slow-growing tumor and detected incidentally on routine radiography. There is a long history of chronic local pain (85%), swelling and edema (65%) with palpable soft-tissue mass and restricted movements in symptomatic patients [11, 12]. CMF should be differentiated from the other osteolytic lesions in younger adults, by clinical presentation, radiological and histopathological findings, as presented in Table 1, of the differential diagnosis of such lesions [2-9].

Radiologically, the CMF presents as an eccentric, lobulated, expansile lytic lesion with well-defined scalloped or lobulated margins and sclerotic bone formation. Radiographic calcification is noted in 10% of cases [11]. Partial cortical erosion and septations are noted in a few cases [3], like in our case.

The microscopic appearance of CMF is a mixture of fibrous, myxoid, and cartilaginous elements with several nodules composed of rounded areas of myxoid or chondroid tissue. Stellate to spindle-shaped cells are noted in the center of lobule with characteristic high cell density toward the periphery of lobules. Sometimes, multinucleated giant cells are also noted in the periphery [14]. These features were similar to our case on the histopathological examination (Fig. 6).

The accepted treatment for CMF is surgical curettage with bone grafting, but the recurrence rate is 3–22% [5]. Preferable

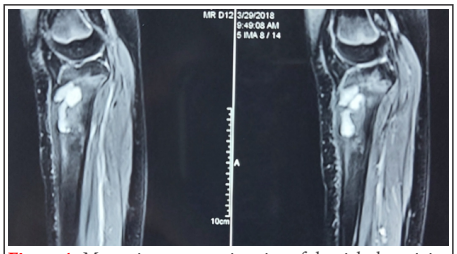

Figure 4: Magnetic resonance imaging of the right knee joint (T2-weighted image) showing a well-defined multiple cystic lesion arising from the anterior aspect of proximal meta-diaphysis of the tibia.

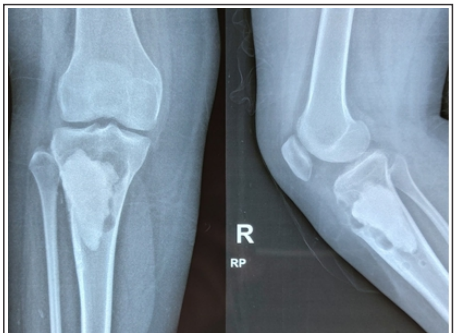

Figure 5: Post-operative radiographs (AP and lateral views) of the right knee joint showing filling of the cavity with bone cement.

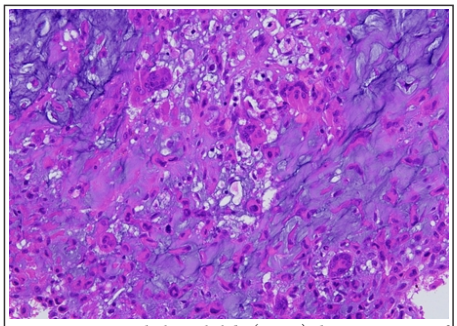

Figure 6: Histopathological slide (200 ×) shows a mixture of chondroid, fibrous, and myxoid elements in varying proportions.

Table 1: Differential diagnosis and characteristic features of benign bone tumors

Bone tumor	Common age	Sex predilection	Common sites	Clinical presentation	Radiological findings	Histology findings	Preferred treatment	Recurrence rates	Comments
Unicameral bone cyst	1st and 2nd decades	Male:female;	Proximal humerus, proximal femur	Usually asymptomatic, unless a pathological fracture	Centrally located, radiolucent lesion, concentrically expanded cortex, and no cortical destruction	Cyst filled with straw-colored fluid, thin fibrovascular lining	Observation, aspiration/injection(steroids, bone graft substitute) curettage	50% [2]	
		2:01							
Aneurysmal bone cyst	1st and 2nd decades	Slight female predominance	Proximal humerus, proximal femur	Pain	Eccentric expansile radiolucent lesion. Thin cortical shell	Hemorrhagic cavernous spaces, septae of fibroblasts, histiocytes, hemosiderin-laden macrophages, and giant cells	Extended curettage consider pre-operative embolization for pelvic lesions	10–20% [2]	
					Fluid levels on magnetic resonance imaging				
Fibrous dysplasia	1st and 2nd decades	Male=Female	Metaphysis of long bones	Asymptomatic, usually discovered incidentally on plain radiographs or if there is a pathologic fracture	Geographic, eccentric lesion located in metaphysis of long bones multi-lobulated appearance with well-defined sclerotic margins	Bland-appearing spindle cells arranged in a storiform pattern in a collagenous matrix	Observation curettage if large fractures usually treated non-operatively	21% (after curettage and bone grafting) [3]	McCune-Albright syndrome – polyostotic fibrous dysplasia, cutaneous pigmentation, endocrine abnormalities Mazabraud syndrome – polyostotic fibrous dysplasia, intramuscular myxomas
Giant cell tumor	3rd and 4th decades	Slight female predominance	Distal femur, proximal tibia, distal radius	Pain, swelling, pathologic fracture (10–30%)	Eccentrically located in epiphysis	Multinucleated giant cells in a sea of mononuclear cells	Extended curettage resection if residual bone stock inadequate Consider radiation for spinal/sacral tumors Resection of pulmonary metastases	0 to 65 % (Depending on the type of treatment, and local presentation of the tumor) [4]	3% incidence of benign pulmonary metastases
					Purely radiolucent (no matrix formation) Usually no rim of reactive bone Abuts subchondral bone May exhibit cortical destruction with soft-tissue extension	Nuclei of mononuclear cells identical to nuclei of giant cells			
					Metaphyseal in skeletally immature patients				
Chondromyxoid fibroma	2nd and 3rd decades	Slight male predominance	Proximal tibia	Pain	Well-circumscribed bubbly lesion	Lobules of hypocellular myxoid cartilaginous tissue	Extended curettage	3–22% [5]	Important to distinguish from chondrosarcoma
				Can present with painless mass in hands and feet	Thin rim of reactive bone (appearance similar to Non-ossifying fibroma)	Lobules separated by cellular fibrous tissue			
Fibro cartilaginous dysplasia	1st–3rd decades	No sex predilection	Femur, proximal tibia, skull bones, ulna, radius, humerus, and phalanges.	Asymptomatic or may present with pain or pathological fractures	Well-defined intramedullary and expansile lesion	Spindle cells embedded in fibrous stroma along with irregularly shaped trabeculae of immature bone (termed as Chinese letter pattern) in place of normal bone and marrow	Observation, curettage with bone grafting, intramedullary nailing, and *bloc* excision	21% [6]	Associated condition – infantile tibia vara
Osteofibrous dysplasia	1st and 2nd decades	Male = female	Tibia (diaphysis)	Asymptomatic unless pathologic fracture anterior bowing	Multicentric, radiolucent lesions in the cortex of the tibia	Irregular trabeculae with prominent osteoblastic rimming	Observation Fractures usually	64–100%[7]	
						Loose fibrous stroma	Treated non-operatively Surgery for correction of deformity		
Desmoplastic fibroma	2nd and 3rd decades	Male:female	Any	Pain	Radiolucent lesion with cortical erosion frequently with septations	Hypocellular fibrous tissue with abundant collagen	Extended curettage versus wide resection	55% (After curettage);	
		2:01		Pathologic fracture	May have soft-tissue mass			17% (After resection) [8]	
Brown tumor	Adults	Male = female	Any	Bone lesions frequently asymptomatic unless pathologic fracture symptoms of hypercalcemia (nausea, weakness, headaches, generalized bone pain	Diffuse osteopenia	Giant cells, increased osteoclastic activity, marrow fibrosis	Medical Management Surgery for actual or impending pathologic fractures	26.9% (in primary tumor); 46.7% (in recurrent tumor) [9]	Primary hyperparathyroidism usually due to parathyroid adenoma
					Multifocal radiolucent lesions with surrounding reactive bone	Diagnosis usually made by raised serum parathyroid hormone, hypercalcemia and hypophosphatemia			Secondary hyperparathyroidism usually due to chronic renal failure

treatment for CMF of long bones is en bloc resection with allograft and artificial bone grafts [15]. In 1975, Taylor et al. first described vascular fibular bone grafting for it. The advantage of vascularized grafting over non-vascularized grafting is faster union and preservation of circulation [10]. Radiation therapy is indicated in surgically inaccessible tumors [11]. The diagnosis is made after anatomopathological analysis, and the treatment may vary according to the region affected. Di Giorgio et al. [16] reported that treatment using curettage, together with phenolization of the lesion, had a better prognosis. Likewise, Jesus-Garcia Filho [17] stated that treatment consisting of simple resection of the lesion was not as effective as a treatment, including associated adjuvant methods.

Conclusion

In the present case, extended curettage with bone grafting was done considering the young age of the patient. The literature suggests that when these lesions are treated using curettage, this technique should be accompanied by an adjuvant method to eliminate the entire mass of the neoplastic cell. We had opted to fill in the bone cavity of the tumor, with polymethylmethacrylate bone cement, similar to its recommended use in giant cell tumor cases [18], as the family was not in favor of autogenous bone grafting and synthetic bone grafts have failed earlier. The recurrence rate is 3–22% [5].

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Conflict of Interest: NIL
Source of Support: NIL

How to Cite this Article

Goyal AK, Vaish A, Vaishya R, Baweja P| A Rare Recurrent Chondromyxoid Fibroma of the Proximal Tibia in an Adolescent Female | Journal of Bone and Soft Tissue Tumors | September-December 2020; 6(3): 16-19.