Subtrochanteric periosteal chondroma: A case report and tumor overview

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Abstract

We report a rare case of subtrochanteric periosteal chondroma in a 16 years boy. The purpose of this presentation is to stress the importance of a proper clinico-radiological and histopathological diagnosis of this lesion. Awareness of the features of this lesion helps to prevent overtreatment of this benign condition, since its differentiation with malignant lesions may be extremely difficult, even at histology; and since conservative surgery with complete excision of the lesion leads to permanent cure.

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INTRODUCTION

Periosteal or juxtacortical chondroma is an uncommon benign chondroid tumor that arises under or on the periosteum on the surface of cortical bone. Because of its histological and radiological appearance, the tumor is often misdiagnosed with resultant radical overtreatment [1,5,6]. Ever since its first distinctive description by Lichtenstein and Hall (1952), it has not been described in the subtrochanteric region of the femur [1-8]. The purpose of this presentation is to report such a rare lesion at the subtrochanter region and to stress the distinctive features of this tumor for proper diagnosis and prevention of its overtreatment.

CASE REPORT

A 16 years old boy presented with swelling and pain at the medial aspect of his right proximal thigh for the last 18 months. He had noticed an increase in the swelling and pain for the last 2 months and correlated it to a bicycle trauma. On inspection there was fullness at the medial aspect of his proximal thigh with overlying normal skin. On palpation, there was a 3.5cm x 2cm nontender, firm, bony hard swelling with well defined margins in the proximal fourth of medial thigh. It appeared in continuation with femur. There was terminal restriction of abduction and external rotation, without any distal neurovascular involvement. Systemic examination was normal and there was no other swelling anywhere else. Radiographs and a CT scan revealed scalloping of peripheral cortex, overhanging margins with variable calcification (Figs. 1, 2a, 2b) Hematological and other radiological examination were insignificant. A Fine Needle Aspiration (FNAC) revealed benign cartilaginous cells with no signs of infection or malignancy. A probable diagnosis of a benign chondroid tumor was made based on these features.

A marginal excision with the rim was done through the Ludloff's approach. Peroperatively, there was a dull white mass under the periosteum with intact cortex. The histopathological report was consistent with a periosteal chondroma (Fig.3). There has been no recurrence till the last followup at 35 months (Fig.4) except a keloid formation at the incision site, which the patient has accepted.

Fig. 1: AP X-rays showing scalloping of peripheral cortex, overhanging margins with variable calcification at the subtrochanteric region.

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DISCUSSION

Periosteal chondroma is a relatively uncommon, benign cartilaginous tumor of periosteal origin and its importance lies in its characteristic radiologic and pathologic appearance, which should be in assistance in the differentials of eccentric lesions of the bone. Although it can occur in any age, it is usually seen in the second and third decade of life with a male to female ratio of 2:1 [8]. Though mostly occurring in the tubular bones of the hand and feet and at the osseous insertions of tendons and ligaments, they have also been reported elsewhere and the proximal humerus is the most favoured long bone [1,8]. The totipotent primitive cells, unrelated to the growth plate, and located in the periosteum are considered the histologic origin of this tumor [1,5].

The initial complaint is most commonly often that of a painless localized swelling, which may increase even after skeletal maturity. When pain is present, it is usually described as dull and aching, and tumor may be tender to palpate. Associated soft tissue swelling is present in upto 1/3 of the cases. Many a times, it is an incidental radiographic finding [1]. Plain radiographs show a lesion upto 4 cm that causes saucerization or scalloping of the cortex. A well circumscribed sclerotic rim of reactive bone is present underlying the lesion, with a well defined margin between the tumor and bone. The cortex is eroded but remains intact. Periosteal new bone may form buttresses overhanging the edges of the lesion. Calcification or ossification of the cartilaginous matrix is not uncommon and it does not infiltrate the medulla or the surrounding tissue [1,2]. No expansion but a pathological fracture through the excised area has been reported in literature [1,7]. A CT shows the same findings with an isodense soft tissue mass with stippled calcification. A typical MR shows a sharply delineated subperiosteal lobulated mass at bone surface, consisting of a matrix with bright signal, bordered by hypointense lining on T2 weighted images, hypo to isointense signal relative to muscle on T1 weighted images, and presenting predominantly peripheral contrast enhancement without intramedullary extension [8].

Pathologically, the lesions are grossly nearly always ovoid or lenticular and lay under the periosteum. The chondroid matrix is recognized by its dull white or blue white color. Histologically, immature cartilage cells are the predominant tissue, lying on an osseous layer formed by the enchondral ossification of tumor tissue and the underneath reactive cortical bone. A fibrous capsule
always marks the external boundary of the tumor. Though there is increased cellularity, cellular pleomorphism and binucleate cells within the benign tumor, no mitotic figures are found [1,7].

Since the classical diagnostic radiographic features as described by deSantos et al are present in only one third of the cases, the differential diagnosis must include several other peripheral cortical lesions, both tumor and tumor like entities [1,2]. Chondrosarcoma, either central or periosteal, is perhaps the most important lesion to be differentiated. They are generally larger (more than 5 cm), occurring in older population, and may also have a soft tissue extension. Although scalloping and sclerosis can be similar, periosteal chondrosarcoma tend to permeate the underlying bone and forms no reactive bone underlying the cortical erosion, best shown by CT. Even isotope scans tend to be 'hotter' for chondrosarcoma. However, grade 1 periosteal chondrosarcoma can mimic periosteal chondroma histologically. Here adequate but conservative local resection, rather than radical surgery is the treatment of choice. A distinction between the two is possible once permanent sections have been made and then a definite surgery can be planned [1].

Parosteal osteosarcoma is diaphyseal, and contains the spicules of bone extending out from the cortex. The malignant cells, which may be spindle shaped and may produce recognizable osteoid, typically are found in the peripheral parts of the chondroid lobules that make the bulk of the tumor [1]. Rarely Ewings sarcoma shows saucerization of the exterior surface of the cortex. However, these are completely lytic, with soft tissue infiltration and without sharp margination. Their clinical course is faster too. Synovial chondromatosis arises in an articular site and does not show scalloping of the bone. Parosteal desmoid never shows the typical radiographic features of periosteal chondroma [1]. Osteochondromas are most common in maturing skeleton. It is radiologically diagnosed when a tumor contains a dense osteoid formation in the cortex with its medulla continuous with the bone of its origin. Chordomas can be locally destructive, but are characteristically heterogenous with low density on CT. Similarly lymphomas and osteolytic metastasis can produce local destruction, but are also of relatively low density and they rarely calcify. Clinical features and a bone scan will differentiate them. Aneuysmal bone cyst may show faint radiolucency with sclerotic rim and rudimentary trabeculae. However, no popcorn calcification, osteosclerosis or saucerization is seen. Myositis ossificans generally shows a wider area of osteosclerosis with zonation, completely encircling a radiolucent mass. Paraarticular pigmented villonodular synovitis may produce erosion, but not scalloping or overhanging edges. A fibrous cortical defect, even when subperiosteal, never grows in the soft tissue. Lesions like neurofibroma, ganglion cyst or others may erode the cortex, simulating periosteal chondroma but no calcification or overhanging edges are present.

The treatment of an asymptomatic periosteal chondroma that is latent in activity is observation. For painful lesions, complete excision with a marginal resection along with the intact rim of underlying bone is curative [1-8]. When a significant amount of cortex is involved, bone grafting is recommended and the extremity be protected until complete bone healing occurs. The recurrence rate is less than 4% and it happens when the rim is not taken out. No malignant transformation or metastasis or multiple lesions have been reported [1-8], but has been twice described to occur synchronously with other chondroid tumours [3,4].

CONCLUSIONS

Periosteal chondroma is a rare benign cartilaginous tumor, which can be cured by a simple marginal excision along with the intact rim of underlying bone. Differentiation from a malignant lesion is, therefore, crucial to prevent more extensive unnecessary surgery due to ominous histological or radiological features.

REFERENCES