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Periosteal Chondrosarcoma: A Case Report and Literature Review

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Abstract **Case Report**

Periosteal chondrosarcoma, also known as juxta-cortical chondrosarcoma, is a rare malignant cartilaginous tumor arising from the external surface of the bone [1, 2]. Imaging features are often specific. Recognizing periosteal chondrosarcoma and differentiating it from other surface tumors is of capital importance because the prognosis is excellent after adequate local surgery alone. Metastasis is late and very rare [3]. We report the case of a young 23 year old, who consulted for left thigh pain evolving for 3 months without inflammatory signs or deterioration of the general condition. A conventional radiographs, computed tomodensitometry and magnetic resonance imaging were performed and were in favor of periosteal chondrosarcoma. The patient underwent a biopsy which confirmed the diagnostic.

Keywords: Periosteal chondrosarcoma, CR, CT, MRI.

INTRODUCTION

Periosteal chondrosarcoma, also known as juxtacortical chondrosarcoma, is a rare malignant cartilaginous tumor arising from the external surface of the bone [1, 2], accounting for less than 2% of all chondrosarcoma and 0.2% of all bone tumors [4]. Imaging features are often specific, detection and characterization is usually done by conventional radiographs (CR), followed by magnetic resonance imaging (MRI) [5].

CASE REPORT

A 23 year old patient who presented for left thigh pain evolving for 3 months without inflammatory signs or deterioration of the general condition. The conventional radiographs showed irregular medial cortical thickening of the left femoral shaft. The CT showed a well-circumscribed tumor lying on the outer surface of the cortex, containing calcific densities characteristic of cartilage tumors associated with irregular thickened cortical, with no evidence of medullary invasion. MRI demonstrated a sharply delineated mass at the bone surface, it had a bright signal on T2 weighted, slight peripheral and septal enhancement after Gadolinium injection, without intramedullary extension. The patient underwent a biopsy which confirmed the diagnostic.

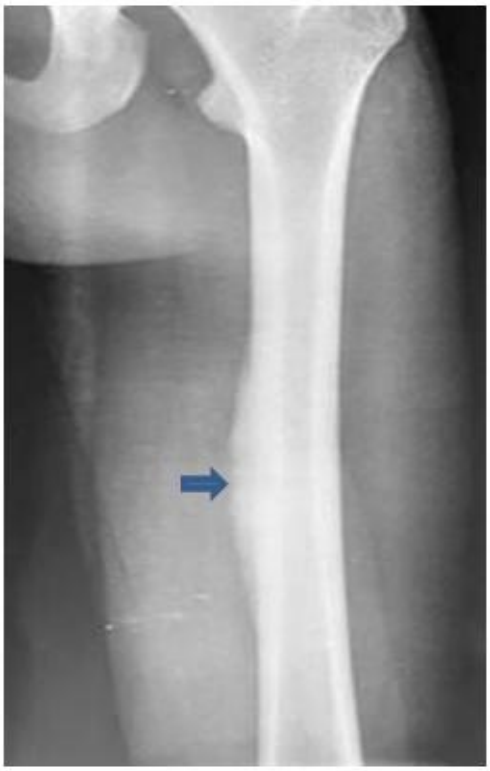

Fig-1: The conventional radiographs showed irregular medial cortical thickening of the left femoral shaft

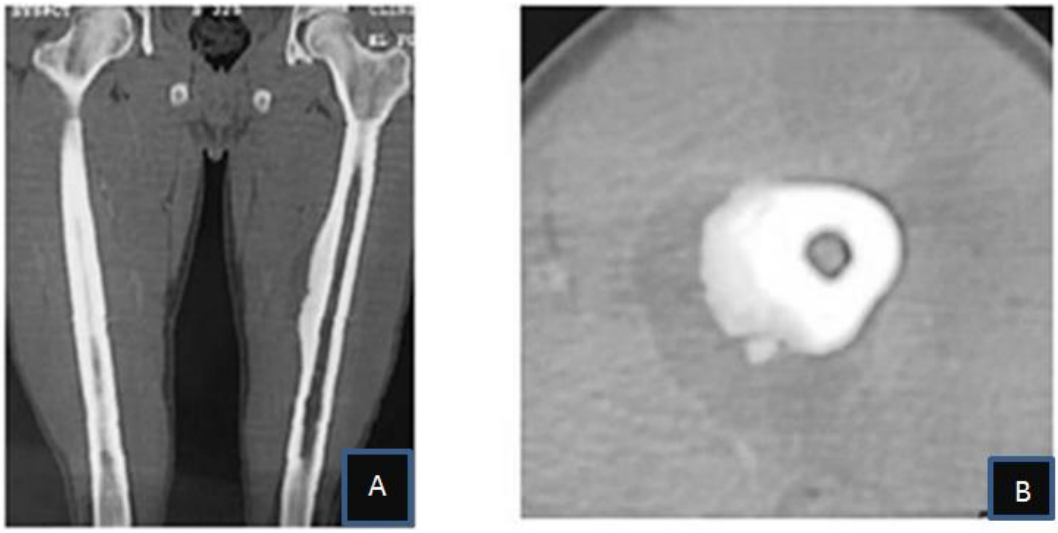

Fig-2: The CT showed a well-circumscribed tumor lying on the outer surface of the cortex associated with irregular thickened cortical. A: coronal section, B: axial section

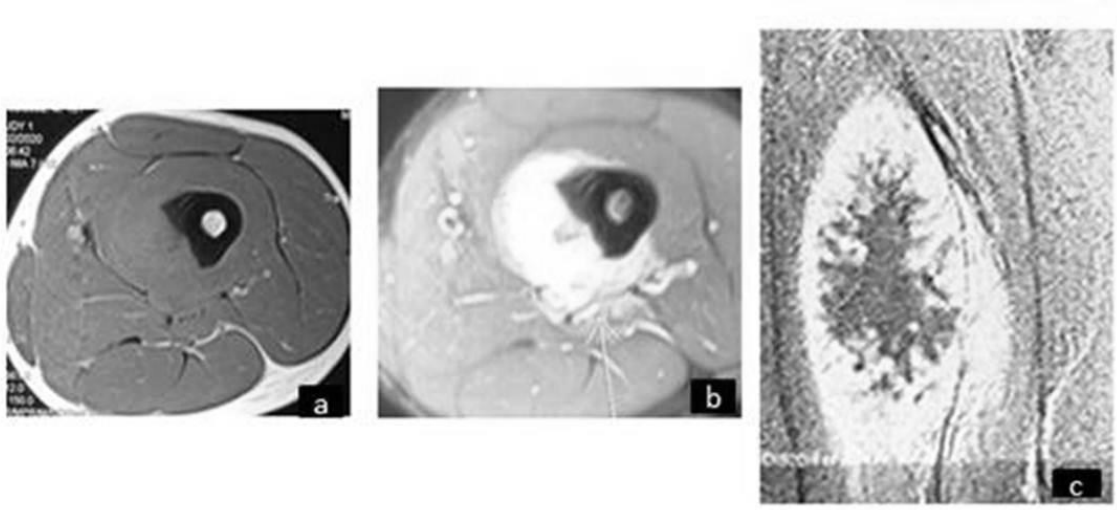

Fig-3: MRI demonstrated a sharply delineated mass at the bone surface, it had a iso- signal on T1 weighted (a), a bright signal on T2 weighted STIR, (b), slight peripheral and septal enhancement after Gadolinium injection (c: Sagittal contrast-enhanced T1-weighted) , without intramedullary extension

DISCUSSION

Periosteal chondrosarcoma, also known as juxta-cortical chondrosarcoma, is a rare malignant cartilaginous tumor arising from the external surface of the bone [1, 2], accounting for less than 2% of all chondrosarcomas and 0.2% of all bone tumors [3]. The mean age of PC is about 32 years (range, 20 to 40 years) [6], the location mainly exists in the limbs, including distal femur, proximal tibia and proximal humerus [6].

Clinically, pain and swelling are usually the presenting features [7]. Often, the tumor runs a long, indolent course [8].

Radiographically, periosteal chondrosarcomas appear as a soft tissue mass with sharply defined borders that may contain popcorn, spotty, peripheral, or ring-and-arc calcifications characteristic of cartilaginous tumors [9], and the underlying cortex shows saucer-shaped erosion in most cases [10]. At the margins of the tumor, there is often periosteal buttressing because of the chronic periosteal reaction. However, perpendicular striations, the most characteristic feature of periosteal osteosarcoma, is absent in almost all cases of periosteal chondrosarcoma [10].

MRI reveals a sharply delineated mass at the bone surface. The tumor matrix has low to intermediate signal intensity on T1- weighted sequences. On T2-weighted sequences, the tumor has a bright signal and associated lobulated structure with hypointense septa. Low-signal, punctate foci of mineralization can be seen on both T1- and T2-weighted sequences. There is peripheral and septal enhancement after gadolinium injection (9). MRI is superior to all other imaging techniques for detecting intramedullary abnormalities and soft-tissue extensions [11].

CT is superior for demonstrating characteristic overhanging bone spicules [5]. CT more clearly depicts the well-delineated overhanging shelf of bone around the proximal and/or distal ends of the exophytic component of a benign juxta-cortical lesion [12]. Furthermore, CT is more sensitive than CR and MRI in depiction of subtle intra-lesional chondroid calcifications [5].

Histologically, periosteal chondrosarcoma consists of solid lobules of hyaline cartilage with foci of myxoid stroma and chondroid cells with a variable degree of atypia. Nodules of the tumor can invade surrounding soft tissues [13, 14]. Periosteal chondrosarcoma and periosteal chondroma can show overlapping radiological and histological features [15]. A size exceeding 50 mm and/or invasion of the underlying cortical bone have been reported as criteria for diagnosing periosteal chondrosarcoma [16], Periosteal chondrosarcomas smaller than 50 mm have been reported in these cases [17], the presence of cortical invasion and/or soft tissue extension favours malignancy [15].

Chemothrapy and radiotherapy are not effective treatments of the PC. Wide surgical resection with a sufficient margin remains the recommended therapy of this type of bone tumor [4].

The prognosis for patients with periosteal chondrosarcoma is good compared with those with central chondrosarcoma of the same histologic grade [1, 9]. Invasion of the medullary cavity is not frequent; a few authors have described periosteal chondrosarcoma with medullary invasion [1]. Bone metastasis is even rare; there have been a few studies about bone metastasis of chondrosarcoma [4]. Lung metastasis, even regional lymph node metastasis, have been reported in published literature [4, 14]. The majority of lung metastasis have inadequate surgical treatment of the primary lesion and often accompany with local reccurence [4].

CONCLUSION

Periosteal chondrosarcoma is a rare primary low-grade surface tumor of bone that often has specific radiologic features. Because prognosis and survival are excellent after adequate local surgery, this lesion must be recognized and differentiated from other surface tumors.

Abbreviations

PC: periosteal chondrosarcoma
CR: conventional radiographs
MRI: magnetic resonance imaging
CT: computed tomodensitometry

Conflict of Interest: All authors state that they have no conflicts of interest.

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