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Juxtacortical Chondroma: A Rare Entity Involving the Phalanges

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Abstract **Case Report**

Periosteal chondroma or juxtacortical chondroma is a rare benign bone tumor of cartilaginous origin. It arises from the periosteum, particularly that of the long tubular bones. We present an interesting case of periosteal chondroma involving the phalanx of the right hand presenting as a palpable mass in a 22-year-old male. The slowly growing mass lesion was investigated in our institute. Characteristic imaging findings on the radiograph and MRI were correlated with histopathological findings to confirm the diagnosis. Establishment of benignity of the lesion guided the surgical management of the patient.

Keywords: Periosteal chondroma, Juxtacortical chondroma, chondroma, phalanges, benign bone tumour.

INTRODUCTION

Periosteal or juxta cortical is a rare benign cartilaginous tumor involving the periosteum and cortex of the metaphysis of the long tubular bones. It accounts for <2% of cartilaginous tumors. Phalangeal involvement is relatively rare. They present as painless phalangeal masses with a slight male predilection. It can mimic other bone lesions which involve the phalanges, including malignant masses such as chondrosarcoma. Histopathological findings along with characteristic radiological findings such as saucerization of the outer cortex of bone without medullary involvement can help ascertain the diagnosis of a benign lesion, thus, avoiding unwarranted radical surgery. We report a case of periosteal chondroma in a young male to revisit the characteristic imaging findings of this rare entity.

CASE REPORT

A 22-year-old male presented with a slowly growing painless swelling involving the medial and dorsal aspect of the proximal phalanx of the little finger of the right hand. There was no limitation of movement of joints. There was no redness and tenderness associated with the swelling. There was no other significant medical history. No other similar swelling was present the body. On physical examination, a small palpable, firm, immobile, and non-tender subcutaneous mass was noted in the right 5th proximal phalanx. The overlying skin color was normal with no color changes. Skin adhesion to the tumor was absent. Pre-operative blood workup was within normal limits.

The patient was referred to our department for diagnostic imaging. AP and Lateral view radiograph of the right hand were performed which showed a lytic lesion involving the meta-diaphysis of the proximal phalanx of 5th digit of right hand causing saucerization of the underlying cortex with mild surrounding solid periosteal reaction. Few areas of calcifications were seen within. MRI showed a well-defined lobulated lesion, iso to hypointense on T1 weighted images and hyperintense on T2 weighted images displaying heterogenous post contrast enhancement. There was saucerization of the underlying cortex seen, however no obvious cortical breech was seen. It showed diffusion restriction. Few foci of blooming were seen on

susceptibility weighted images suggestive of calcifications.

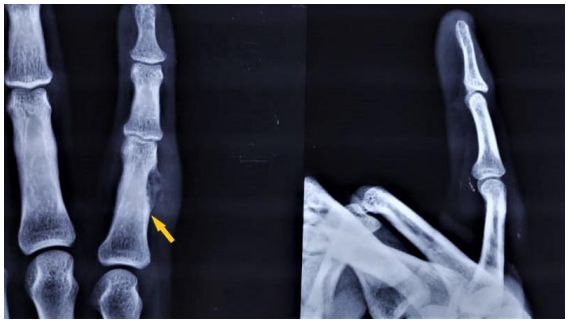

Fig-1: AP and Lateral radiograph of the right little finger shows a lytic lesion with soft tissue component involving the medial aspect of the meta-diaphysis of the proximal phalanx. There is cortical buttressing saucerization seen with associated ring and arc calcification within the soft tissue.

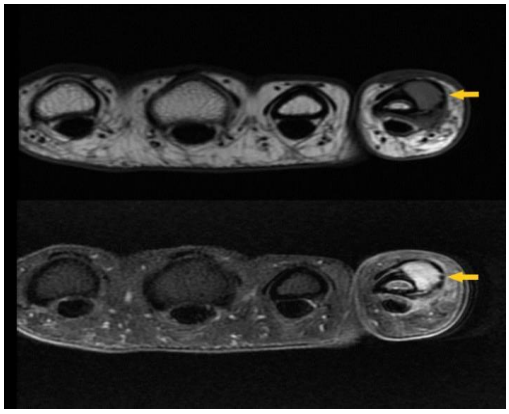

Fig-2: Axial T1WI and PDFS images show a well-defined lobulated lesion causing thinning of the cortex without any obvious cortical breech

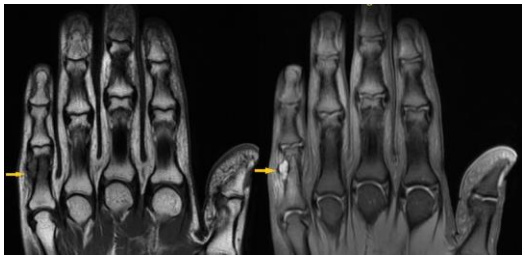

Fig-3: Coronal T1WI and PDFS images show a well-defined lobulated lesion causing saucerization of the cortex without any obvious cortical breech

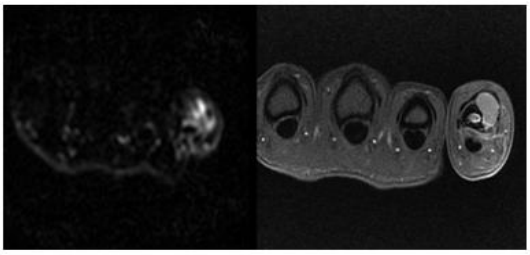

Fig-4: The lesion shows diffusion restriction and mild peripheral post contrast enhancement

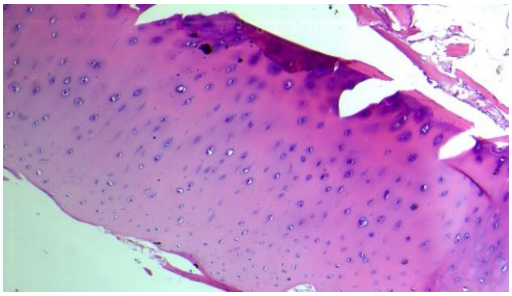

Fig-5: Histological image showing chondrocytes in an abundant hyaline cartilage maxtrix

DISCUSSION

Periosteal chondroma is a slowly growing benign cartilaginous tumor, arising from and developing beneath the periosteum. It was first described by Lichtenstein and Hall in 1952. Jaffe coined the term 'Juxta-cortical chondroma'. It is a rare entity accounting for 1.3 % of cartilaginous tumors [1]. The metaphysis of the long tubular bones is the typical site of involvement, the most commonly the proximal humerus, distal femur, and the phalanges. Takada et al. and Yoshimura et al. found that 28% of the periosteal chondromas affect the hands [2, 3]. Rarely, there may be involvement of clavicle, cervical vertebrae, tibia, radius, ulna, and fibula are more commonly. Males affected are more commonly affected with a male: female ratio of 2: 1, more so during 2nd to 3rd decade of life [4]. Though the pathogenesis of the tumor is unclear, an association of isocitrate dehydrogenase 1 (IDH1) mutations has been implicated in 71% of cases [5]. They are usually small tumors that may reach up to 3-4 cm in size. Most patients present with a painless mass or swelling. The children might complain of mild pain of long duration, with or without an associated bony bump.

Histologically it is similar to enchondroma in appearance, but with more malignant-looking cells, showing binucleation, nuclear enlargement, and increased cellularity. Similar may also be present in low-grade chondrosarcoma, thus emphasizing upon the need for imaging for distinguishing between the two entities.

The characteristic radiographic appearance is an area of scalloping or cauterization of the outer cortex of the metaphysis of the long tubular bones with a well-defined inner margin, surrounding sclerotic periosteal reaction, and overhanging of the superior and inferior margins [4, 6, 7]. Dense lamellar bone is usually seen separating the tumor from the medulla. Chondroid matrix calcifications are seen in 50 % of the cases [8]. Endophytic soft tissue mass is seen in one-third of the patients, which may show medullary extension.

MRI aids conventional radiography in ascertaining the diagnosis. Typically, a well-defined juxta-cortical soft tissue mass lesion, with lobulated

configuration and pressure erosion over the underlying cortex, is seen. It shows low to intermediate signal on T1-weighted images, with a hypointense rim and hyperintense signal on T2-weighted images. T2* images may show hyperintense signal interspersed with low signal, representing the calcifications. It usually demonstrates heterogenous enhancement after intravenous gadolinium administration in a ring and arc pattern [9, 10].

CT is a better imaging modality to demonstrate the overhanging bony margins at the proximal and distal ends of the juxta-cortical lesion. Also for demonstration of chondroid calcifications within the lesion, CT also more sensitive than radiography and MRI[11].

Differential diagnosis consists of other peri- and periosteal lesions. The main differential is bizarre parosteal osteochondromatous proliferation, which is also known as Nora's lesion, which is many times indistinguishable from periosteal chondroma on the basis of clinical, radiographic, and MRI features. Histopathological examination is required to distinguish the two lesions, in which numerous osteoblasts and bizarre chondrocytes are seen in Nora's lesion [12] Periosteal chondrosarcoma generally affects older age groups. Involvement of phalanges is very rare. They have a larger soft tissue component (usually >3.5 cm) as compared to the juxta-cortical chondroma [12]. Other differentials include soft tissue chondroma, non-ossifying fibroma (NOF), cortical neurofibroma, desmoid, and periosteal osteosarcoma.

Surgical resection is recommended for all tumours. Recurrence is usually rare (1.2 %), however incidence is increased in tumours affecting the hand upto 15 %, majorly owing to incomplete excision.

CONCLUSION

Periosteal chondroma is a rare entity, with clinical and histological features overlapping with various benign and malignant entities; however, in conjunction with radiological features on radiograph and MRI, a definitive diagnosis can be reached, giving a good prognosis to the patient after adequate local surgery.

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