

# ACTA SCIENTIFIC CLINICAL CASE REPORTS

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Editorial

# Besiege and Beleaguer-Perisoteal Chondroma

### Anubha Bajaj\*

Department of Histopathology, Panjab University/A.B. Diagnostics, India

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Perisoteal chondroma emerges as an exceptionally discerned, benign cartilaginous neoplasm which adheres to surface of bony cortex and is situated subjacent to the periosteum. Additionally designated as juxtacortical chondroma or subperiosteal chondroma, tumefaction represents as a well demarcated, dome shaped lesion confined to superficial bone surface.

Neoplasm appears composed of benign hyaline cartilage wherein a distinct communication with medullary cavity appears to be lacking. Tumefaction comprises of < 2% of chondromas. Commonly, paediatric population and young adults, preponderantly < 30 years are implicated. A male preponderance is encountered with male to female proportion of 1.5:1 [1,2].

Periosteal chondroma is frequently confined to small bones of hands and long bones of appendicular skeleton, especially proximal metaphyseal or diaphyseal region of humerus and femur [1,2].

Of obscure aetiology, periosteal chondroma depicts chromosomal mutations of IDH1 and IDH2 genes. However, consistent cytogenetic anomalies appear absent. Besides, a subset of periosteal chondromas delineate genetic mutations within IDH genes. Additionally, diverse cytogenetic anomalies are encountered [1,2].

Clinically, a miniature neoplasm of magnitude < 3 centimetres is observed. Typically, lesion is painless and may induce a swelling or palpable tumour mass [2,3].

Upon frozen section examination, mature hyaline cartilage is observed. Grossly, a lobular, well demarcated, waxy, blue/grey neoplasm of magnitude < 5 centimetres is encountered. Foci of calcification are encountered [2,3].

Tumour bone appears 'buttressed' upon the lateral neoplastic edge. Foci of medullary invasion appear absent [2,3].

Upon microscopy, the well demarcated lesion appears disparate from subjacent sclerotic bone. Underlying cortical bone may demonstrate focal erosion although tumour permeation is absent. Neoplasm depicts a distinct lobular architecture and is encased by an intact layer of attenuated periosteum [3,4].

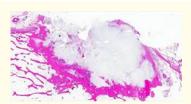
Generally minimal, tumour cellularity appears variable. Chondrocytes appear devoid of cytological atypia. Occasionally, few neoplasms appear cellular and exhibit significant nuclear pleomorphism or bi-nucleation. Besides, a component of spindle shaped cells may be observed [3,4].

Neoplastic invasion of circumscribing soft tissue or medullary canal appears absent [3,4].

Fédération Nationale des Centres de Lutte Contre le Cancer (FN-CLCC) grading system [3,4]



**Figure 1:** Periosteal chondroma composed of bland cartilaginous cells with minimal nuclear atypia. Foci of calcification are discerned [7].



**Figure 2:** Periosteal chondroma expounding lobular architecture and an amalgamation of uniform chondrocytes with minimal atypia, encompassed within a layer of attenuated periosteum [8].

### **Tumour differentiation**

- Score 1: Sarcomas resembling normal tissue
- Score 2: Sarcomas with defined histological differentiation
- Score 3: Undifferentiated sarcomas or sarcomas of uncertain histologic differentiation

Mitotic count as discerned within 10 successive high power fields (HPFs) within significantly mitotically active areas

- Score 1: 0 9 mitoses
- Score 2: 10 19 mitoses
- Score 3: ≥ 20 mitoses

#### Tumour necrosis

- Score 0: Absence of necrosis
- Score 1: < 50% necrosis
- Score 2: ≥ 50% necrosis

Tumour grade is comprised of a total figures of tumour differentiation, mitotic count and tumour necrosis and is denominated as

- Grade 1: 2 to 3 points
- Grade 2: 4 to 5 points
- Grade 3: 6 to 8 points

Score 1 Well differentiated liposarcoma, leiomyosarcoma, fibrosarcoma, chondrosarcoma, MPNST

Score 2 Myxoid liposarcoma, MFH, chondrosarcoma, conventional fibrosacoma, MPNST, leiomyosarcoma, angiosarcoma, well differentiated malignant HPC or solitary fibrous tumour, typical storiform/pleomorphic MFH

Score 3 Round cell liposarcoma, pleomorphic liposarcoma, dedifferentiated liposarcoma, poorly differentiated fibrosarcoma, poorly differentiated MPNST, epithelioid MPNST, malignant Triton tumour, conventional malignant HPC, giant cell/inflammatory MFH, poorly differentiated/pleomorphic/epithelioid leiomyosarcoma, synovial sarcoma, embryonal/alveolar/pleomorphic rhabdomyosarcoma, mesenchymal chondrosarcoma, poorly differentiated/epithelioid angiosarcoma, extraskeletal osteosarcoma, Ewing's sarcoma, ASPS, ES, malignant rhabdoid tumour, clear cell sarcoma, undifferentiated sarcoma.

**Table 1:** Tumour Differentiation Score- Updated Fédération Nationale des Centres de Lutte Contre le Cancer system (3,4).

MPNST: Malignant peripheral nerve sheath tumour, MFH: Malignant fibrous histocytoma, HPC: hemangiopericytoma, ES: Epithelioid sarcoma, ASPS: Alveolar soft part sarcoma.

Periosteal chondroma requires segregation from neoplasms as periosteal chondrosarcoma, periosteal osteosarcoma, parosteal osteosarcoma, bizarre parosteal osteochondromatous proliferation, periosteal chondromyxoid fibroma or soft tissue chondroma.

Appropriate tumour ascertainment is contingent to specific radiological features [5,6].

Tumour is comprised of well defined tumefaction confined to juxta-cortical region. Neoplasm is imbued with 'popcorn' or 'ring-like' focal calcification, a feature which is pathognomonic of cartilaginous tumours.

Plain radiographs delineate a distinct soft tissue tumefaction with saucerization or scalloping and sclerosis of subjacent bony cortex along with protrusion of tumour perimeter [5,6].

Computerized tomography (CT) appears advantageous for identification of scattered intra-tumour calcification and absence of intramedullary neoplastic extension [5,6].

Magnetic resonance imaging (MRI) characteristically depicts a well circumscribed tumour confined to juxta-cortical region.

T1 weighted magnetic resonance imaging delineates a tumefaction with intermediate signal intensity whereas T2 weighted imaging expounds a neoplasm with enhanced signal intensity [5,6].

Periosteal chondroma may be appropriately managed with comprehensive surgical excision of the lesion. Additionally, cogent procedures as intralesional, marginal or en bloc surgical resection may be beneficially employed. Surgical extermination with precise marginal excision and neoplastic curettage appear appropriate for eradication of pre-diagnosed, confirmed lesions [5,6].

The essentially benign neoplasm demonstrates estimated tumour reoccurrence at  $\sim$ 3.6% [5,6].

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- 7. Image 1 Courtesy: Orthobullets.com.
- Image 2 Courtesy: Atlas in genetics and cytogenetics in Oncology.