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The Perimetric Protuberance-Periosteal Chondroma

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Preface

Periosteal chondroma is an exceptional, benign, gradually progressive cartilaginous neoplasm. Additionally designated as juxtacortical chondroma, sub-periosteal chondroma or soft tissue chondroma, the neoplasm was initially chronicled by Lichtenstein and Hall in 1952 [1]. Characteristically, the lesion arises adjacent to or upon the surface of bone cortex within the sub-periosteal region. The neoplasm arising from hyaline cartilage frequently erodes bone cortex.Periosteal chondroma is appropriately diagnosed with imaging studies and cogent histological features. Morphological distinction of periosteal chondroma from low-grade chondrosarcoma may be challenging. Cytofluorometric analysis of deoxy ribonucleic acid (DNA) ploidy may assist segregation of periosteal chondroma from chondrosarcoma.

Disease Characteristics

An estimated below < 2% of chondromas are constituted by periosteal chondromas.Periosteal chondroma arises prior to 30 years and usually occurs in children and adults between 10 years to 20 years. The neoplasm demonstrates a male predominance or an equivalent gender distribution [2,3]. Periosteal chondroma commonly emerges upon the bony surface of clavicle or ribs. Tumefaction appears beneath periosteal membrane of metaphysis of long bones as the proximal humerus, femur and tibia or upon miniature, tubular bones of hand and feet. Tumefaction may arise within zones lacking a periosteal covering such as the femoral neck. Concomitant bony erosion of the vertebral column, clavicle and costal cartilage may ensue. Lesions within the pelvic cavity are infrequent and may be symptomatic, palpable and fixed [2,3]. Periosteal chondroma commonly emerges through configuration of sub-periosteal cartilage wherein tumefaction is not concordant to the metaphyseal plates [2,3]. Although participant genomic mechanisms remain obscure, genetic rearrangements of chromosome 12 are associated with periosteal chondroma [2,3].

Clinical Elucidation

Periosteal chondroma commonly manifests aswollen, painful, palpable tumefaction although asymptomatic, painless nodules can also be discerned. Tumefaction can arise as a firm, nontender nodule within diverse tumour sites as the distal thigh and is accompanied by uninvolved superimposed cutaneous or neural structures [3,4]. Incriminated subjects may depict low back pain, features of sciatica, gluteal muscle atrophy and claudication. Low back pain is accompanied by sensory and motor impairment [3,4]. Typically, localised swelling is accompanied by moderate, prolonged pain wherein it is contemplated that pain arising within low-grade, cartilaginous neoplasms may indicate aggressive biological behaviour.Compression of abuttingdigital nerves may ensue [3,4].

Histological Elucidation

On gross examination, periosteal chondroma depicts periosteal nodules frequently associated with foci of intrinsic calcification [3,4].Multiple, irregular nodules and islands of hyaline cartilage are demarcated by strips of normal bone marrow and are partially or completely enveloped by sheets of lamellar bone [3,4]. A common morphological pattern encountered is lobules of immature hyaline cartilage with interspersed miniature chondrocytes and extension from superimposed periosteum into adjacent cortical bone. Tumefaction demonstrates a lobular configuration contributed by proliferating mature chondrocytes with miniature, uniform nuclei. Mitotic activity is absent [4].

Tumefaction composed of benign hyaline cartilage is superimposed with periosteum or reactive bone. Neoplasm can be hyper-cellular with variably myxoid regions and occurrence of bi-nucleate cells. Tumour infiltration into medullary cavity or circumscribing soft tissue is absent. On morphological assessment, lobules of hyaline cartilage are composed of chondrocytes along with foci of endochondral ossification and calcification. Tumefaction is hypercellular although cytological atypia is absent [4,5]. Periosteal chondroma exhibits a prominent lobular arrangement of hyaline cartilage which extends from the periosteum into adjacent cortical bone.

Typically, tumefaction is hypo-cellular although foci of enhanced cellularity, nuclear pleomorphism, bi-nucleated or multinucleated tumour cells may be observed. A hyper-cellular neoplasm along with nuclear atypia may be challenging to differentiate from diverse malignant cartilaginous or bony conditions [4,5]. Tumour cells are frequently bi-nucleate and depict cellular and nuclear pleomorphism. Periosteal chondroma depicts a structural genomic rearrangement of chromosome 12q13-15 (HMGA2 / HMGI-C) [5].



Figure 1: Periosteal chondroma depicting a cartilaginous neoplasm with spatial relation with the medullary compartment, bine cortex and periosteum [6].



Figure 2: Periosteal chondromadelineating a cartilaginous neoplasm confined to the metaphysis of long bones [7].



Figure 3: Periosteal chondroma exhibiting hyaline cartilage with mature chondrocytes and vacuolated cytoplasm disseminated in a myxoid and fibrotic stroma [7].



Figure 4: Periosteal chondroma delineating aggregates of vacuolated cartilaginous cells admixed within a myxoid and fibrous tissue- rich matrix [8].



Figure 5: Periosteal chondroma enunciating fragments of woven bone intermingled with vacuolated chondrocytes and a fibrotic stroma [9].



Figure 6: Periosteal chondroma exhibiting vacualated chondrocytes with uniform nuclei and a circumscribing myxomatous stroma [9].



Figure 7: Periosteal chondroma demonstrating a glistening, grey/white, cartilaginous neoplasm with a superimposed fibrotic perichondrium and adipose tissue aggregates [10].



Figure 8: Periosteal chondroma exemplifying clusters of vacuolated chondrocytes with abundant cytoplasm and uniform nuclei admixed within a fibrotic stroma [11].

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Differential Diagnosis

Periosteal chondroma requires a segregation from benign and malignant neoplasms such as osteochondroma, parosteal osteosarcoma, periosteal osteosarcoma, chondrosarcoma and enchondroma [12].

•osteochondroma is a cartilage engendering neoplasm commonly discerned within femur of adolescent subjects. Around 15% of conventional chondrosarcomas arise from pre-existing osteochondroma. Typically, tumefaction displays a dense osteoid within the cortex and medulla along with contiguity with the parent bone. Neoplasm demonstrates a cartilaginous cap composed of mature hyaline cartilage with a fibrous perichondrium. Transition between bone and cartilage resembles the growth plate and delineates endochondral ossification with origin of mature bone. Cartilaginous cap is absent in lesions of long-standing duration [12,13].

•parosteal osteosarcoma is a gradually progressive surface neoplasm originating from extraneous periosteal layer and contributing to roughly 65% of surface osteosarcomas. The low grade, hypo-cellular neoplasm is composed of well formed bony trabeculae, osteoid, cartilaginous componentand a fibrotic, malignant, spindle-shaped cellular stroma. Mitotic figures or osteoclast-like giant cells are absent [12,13].

•chondrosarcoma is a malignant cartilaginous neoplasm commonly discerned within subjectsexceeding > 50 years. Tumefaction is enlarged and is associated with possible extension into abutting soft tissue. Tumour cells produce a cartilaginous matrix. Cellular and nuclear atypia may be focal and minimal. Direct osteoid or bone formation from the neoplastic cells is absent. Intracytoplasmic hyaline globules are commonly discerned in low grade neoplasms. The neoplasm is immune non-reactive to keratin [12,13].

•periosteal chondrosarcoma demonstrates foci of "popcorn" calcification and accumulation of scalloped radiolucencies with a sclerotic perimeter upon radiography [12,13]. •periosteal osteosarcoma is a gradually progressive neoplasm which predominantly arises beneath the periosteum, especially within long bones as the femur. The neoplasm configures new bone and appears as a radiolucent lesion confined to the bone surface. Perpendicular spicules of calcification and a peripheral Codman's triangle may be discerned. Periosteal osteosarcoma is an infrequently discerned variant of osteosarcoma wherein tumour nodules are usually below < 3 centimetre magnitude. Lace-like pattern of malignant osteoid may be delineated although the neoplasm displays a predominance of chondroid areas composed of high-grade, anaplastic cells [12,13].

Investigative Assay

Imaging with computerized tomography (CT) or magnetic resonance imaging (MRI) is beneficial for appropriate discernment of the neoplasm [13,14]. Plain radiographs depict a protrusive tumour edge along with radiolucent shadows and stippled calcification. Characteristically, tumefaction arises from the bony surface, exhibits scalloping of bone cortex and a well-defined perimeter between tumour and bone. Tumour sclerosis is common and envelops the protrusive tumour edges and abutting soft-tissue masses. Variable foci of calcification and ossification are observed [13,14].

Periosteal lesions are nearly 2 centimetres to 4 centimetres magnitude, well demarcated and demonstrate a sharply defined, scalloped extraneous bone cortex [13,14].Radiographically,

tumefaction can depict clear scalloping of bone cortex, protruding tumour edges, tumour sclerosis and calcification or ossification of cartilaginous matrix. However, calcification or ossification of cartilaginous matrix may be absent [13,14]. Computed tomography (CT) demonstrates a solid tumefaction with specks of calcification. Computerized tomography (CT) enunciates an iso-intense soft tissue mass with stippled calcification, focally enhanced signal intensity and associated localized bone destruction [13,14].

Magnetic resonance imaging (MRI) depicts a well-defined, sub-periosteal, lobulated tumefaction situated upon the bone surface. Tumour matrix delineates a bright signal with hypointense layering upon T2-weighted imaging and an isointense signal concordant to adjoining skeletal muscle upon T1-weighted imaging [13,14]. Magnetic resonance imaging (MRI) delineates a lobular, heterogeneous mass which is hypo-intense upon T1weighted imaging and hyper-intense upon T2-weighted imaging [13,14].

Therapeutic Options

Surgical intervention is optimal and ensures enhancement of functional abilities along with amelioration of associated pain [13,14].Periosteal chondroma is appropriately treated by a variety of surgical manoeuvres such as intralesional removal, marginal excision or en bloc resection. Curettage of the tumour can be adopted [13,14]. Marginal resection and curettage are preferably adopted in definitively diagnosed lesions.Alternatively, an extended or en bloc surgical resection can be employed, including eradication of margins of normal bone. Inadequately excised neoplasms may reappear. Estimated proportion of localized tumour reoccurrence is 3.6% [13,14].

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