Preface

Osteochondromyxoma is an extremely exceptional, benign, congenital bone neoplasm delineating an admixture of chondroid and osteoid stroma. The tumefaction appears as a component of Carney complex. Osteochondromyxoma demonstrates extensive myxoid alterations intermingled with a chondroid and osteoid matrix.

Osteochondromyxoma may be denominated as an osteochondroma associated with myxoid components [1]. Although an uncommon manifestation, the benign neoplasm is contemplated as a definitive clinical diagnostic criterion of Carney complex, a syndrome also designated as familial lentiginous or multi-organ tumour syndrome. Classically, osteochondromyxoma is associated with Carney complex and associated lentigines or diverse, unusual conditions and may be designated as a "Carney bone tumour" [1]. Precise determination of benign osteochondromyxoma is essential as the concordant Carney complex may be fatal.

Osteochondromyxoma can be adequately discerned on account of pertinent, singular sites of tumour emergence, clinical symptoms and radiographic appearance which is diverse from associated myxoid bone lesions such as chondroblastoma, chondromyxoid fibroma, rhabdomyosarcoma, myxoid chondrosarcoma or neurofibroma.

Disease characteristics

Generally, osteochondromyxoma incriminates an estimated 1% of subjects with Carney complex. The neoplasm is discerned in childhood, usually before < 2 years. Typically, osteochondromyxoma appears prior to < 2 years and may arise as a congenital lesion although no age of tumour emergence is exempt [2]. Tumefaction arising within adults are distinct, independent lesions. Usually, primary bone tumours occurring within early childhood are uncommon. Nevertheless, osteochondromyxoma is an exceptionally discerned diagnostic criterion of Carney complex [2,3].

Osteochondromyxoma is generally situated within the facial bones, nasal sinuses, nasal bones, skull, sellar region, ribs, diaphysis of long bones as the tibia or radius, thoracic region and vertebral column. The neoplasm originates from bony cortices and is commonly discerned within the nasal region, tibia and radius [2,3].

Osteochondromyxoma is concurrent with inactivating mutations of tumour suppressor gene PRKAR1A situated upon chromosome 17q22-24 and is an uncommon, diagnostic component of Carney complex. Osteochondromyxoma possibly arises from deranged mesenchymal stem cells entitled to engender osteoblasts. Pertinent mutation arises on account of hyper-stimulation of protein kinase A (PKA) associated with elevated cellular levels of cyclic AMP (cAMP) [2,3].

**Clinical elucidation**

Typically, the neoplasm arises as a painless nodule, although may be discovered incidentally within a skeletal survey conducted for determining Carney complex [4,5]. Osteochondromyxoma delineates swelling, discomfort and localized inflammation of soft tissues wherein clinical symptoms are contingent to tumour magnitude and location [4,5].

Clinical symptoms of the skeletal neoplasm may be nonspecific. Additionally, osteochondromyxoma may occur as an isolated entity.

Characteristically, osteochondromyxoma delineates a painless tumefaction associated with clinical symptoms arising due to oedema and tumour mass effect. Symptoms contingent to Carney complex such as cardiovascular and endocrine anomalies or spotty cutaneous pigmentation may ensue. The benign osteochondromyxoma may simulate an osteosarcoma and demonstrate brisk, localized tumour infiltration although distant metastasis is absent [4,5].

**Histological elucidation**

On gross examination, osteochondromyxoma appears as a well-circumscribed, non-encapsulated, gelatious, yellowish, cartilaginous, haemorrhagic neoplasm which may erode the superimposed bone cortex. Macroscopically, a well-circumscribed, calcified, greyish/white, occasionally encapsulated bony or cartilaginous tumefaction is discerned. Tumour mass and matrix of circumscribing bone structures are variable although tumefaction is confined within the periosteum. Erosion of encompassing bone and infiltration of soft tissue is delineated. The neoplasm demonstrates gelatinous areas along with an admixture of cartilaginous and bony areas [4,5].

Tumefaction is minimally to moderately cellular and demonstrates erosion of cortical bone with infiltration of surrounding soft tissue. Bland tumour cells configure sheets and lobules which are admixed within a myxomatous, cartilaginous and osseous matrix with foci of hyaline fibrous tissue [4,5].

On microscopy, osteochondromyxoma is a neoplasm of variable cellularity delineating an abundant myxoid, cartilaginous and partially osteoid matrix. Tumour cells are designated as chondroblast-like and osteoblast-like. Mitotic figures are occasional [4,5].

Tumour capsule is composed of parallel, 10 cells to 15 cells thick layers of spindle-shaped mesenchymal cells. Tumour cells are imbued with an eosinophilic cytoplasm and columns of nuclei. Non encapsulated neoplasms are devoid of a distinct demarcation between the neoplasm and circumscribing soft tissue. Also, soft tissue inflammation is exceptional. Lobular sheets of chondroid tissue, osteoid, polymorphic cellular component and hyaline bands are observed. The exceptional, benign, locally aggressive neoplasm is devoid of bluish, immature bony trabeculae [5,6].

Tumefaction is composed of an admixture of mesenchymal cells, basophilic myxoid foci and mucopolysaccharide ground substance. Tumour cellularity is contingent to quantifiable myxoid matrix. Osteoid, bone, collagen fibres, immature and mature cartilage or bands and nodules of hyaline, fibrous tissue are disseminated within the neoplasm. Tumour cells are organized within well-defined sheets, occasional poorly defined cellular whorls or demonstrate a disorganized, micro-lobular or macro-lobular tumour pattern [5,6].

Enlarged lobules are imbued with multiple, miniature, sinusoidal vascular articulations and display a definitive cellular perimeter. Accumulated myxoid matrix engenders a dissociation of miniature, irregular cellular aggregates into delicately adhered strands and cords of cells with a reticular configuration. Cellular and nuclear atypia or tumour necrosis are infrequent [5,6].

Majority of tumour cells are polygonal, stellate and bipolar. Cytoplasmic vacuoles and occasional cytoplasmic inclusion bodies usually surround myxochondroid areas. Chondroblast-like and osteoblast-like cells engendering osteoid are scattered within the tumour parenchyma [5,6]. Miniature foci of mature adipose tissue cells are observed. Tumour cell nuclei are uniform, well preserved, chromatic and vesicular with miniature, discernible nucleoli. Nuclear atypia is infrequent [5,6].

---

**Citation:** Anubha Bajaj. “The Muculent Ossein- Osteochondromyxomas”. EC Clinical and Experimental Anatomy 4.4 (2021): 01-07.
Immune histochemical elucidation

Tumour cell cytoplasm can be stained by periodic acid-Schiff’s (PAS) stain wherein intense staining of matrix is within minimal foci. Cellular cytoplasm and matrix are stained by colloidal iron [6,7]. Movat’s pentachrome stain can segregate tumour components as myxomatous (faint blue), cartilaginous (blue, green or olive), hyaline (yellow) and osseous (yellow, scarlet or brick red) [7]. Tumour cells are immune reactive to vimentin and occasionally to S-100 protein. Immune reactivity to collagen II is focal and moderate wherein immune reactivity to collagen IV is minimal [6,7].

Differential diagnosis

Osteochondromyxoma requires a segregation from diverse neoplasms such as

- Chondromyxoid fibroma which is a neoplasm which depicts a distinctive, lobular architecture along with peripherally disseminated, cellular foci of chondrocytes and stellate cells alternating with centric, minimally cellular myxoid areas and foci of cystic degeneration. Tumour cells are spindle-shaped or stellate with spherical to elliptical nuclei and indistinct, eosinophilic cytoplasm. Foci of necrosis or mitotic activity are absent whereas foci of coarse calcification are variable [7,8].

- Myxoid chondrosarcoma exhibits a multinodular architecture wherein fibrous tissue septa segregate the neoplasm into accumulates of abundant myxoid or chondromyxoid matrix incorporating tumour cells. Tumour cells are uniform, imbued with eosinophilic to vacuolated cytoplasm, delicate cytoplasmic processes, spherical to elliptical nuclei and inconspicuous nucleoli. Tumour cells configure cords, miniature clusters and a complex trabecular or cribriform pattern. Spindle-shaped cells are commonly discerned. Tumour stroma is hypo-vascular and mitotic activity is minimal [7,8].

- Chondromesenchymal hamartoma is composed of well demarcated mature cartilage, myxoid stroma, focal aggregates of osteoclast-like, multinucleated giant cells, aneurysmal bone cyst like areas, spindle-shaped cells and accumulated collagen fibres [7,8].

- Fibrous dysplasia is comprised of immature collagen and immature bone trabeculae configured within a fibrocellular matrix. Osteoblastic maturation arrest contributes to an absence of osteoblastic rimming of bone trabeculae. Therefore, metamorphosis from normal bone to aberrant bone is sudden and abrupt. Fibrous tissue stroma intermingled with bone trabeculae is composed of bland, spindle-shaped cells [7,8]. Cytological atypia and mitotic activity is exceptional. Commingled stroma may depict myxoid and adipose tissue metaplasia. Secondary aneurysmal bone cyst-like alteration can be enunciated [7,8].

Investigative assay

Upon plain radiography, osteochondromyxoma appears as a heterogeneous neoplasm. Radiographic features are contingent to site of the neoplasm. Plain radiographs and cross-sectional imaging demonstrate a “ring-like” or “bubbly” appearance. Tumefaction can appear as a lytic lesion or a mixed, sclerotic and lytic lesion. Osteochondromyxoma may be associated with an expansible tumour evolution with permeation of enveloping tissue [9,10].

Upon plain radiography, osteochondromyxoma is well circumscribed, destructive and depicts focal mineralization of the neoplasm. Tumour countenance can be diverse and contingent to tumour location [9,10]. Tumefaction delineates expansion of bone cortex along with admixture of lucent and sclerotic zones [10].

Plain radiography contributes to assessment of tumour configuration, location, tumour perimeter, transition zone and breach within superimposed bone cortex [9,10].

Upon imaging, osteochondromyxoma requires a segregation from neoplasms such as chondromyxoid fibroma, mesenchymal hamartoma, myxoma, chondrosarcoma with myxoid change and fibrocartilaginous mesenchymoma [9,10].

Computerized tomography (CT) demonstrates a calcified soft tissue density along with an osteoid matrix. Erosion of bone cortex may be occasionally discerned [9,10].

Magnetic resonance imaging (MRI) is a precise methodology for assessing symptomatic bone tumours. Osteochondromyxoma demonstrates an enhanced signal intensity upon T2 weighted imaging. Magnetic resonance imaging (MRI) displays a heterogeneous, mixed-signal intensity upon T1 weighted imaging which is isointense or hypo-intense to adjoining skeletal muscle. Upon T2 imaging, a heterogeneous tumefaction with predominantly enhanced signal intensity is observed. Upon administration of gadolinium contrast, a heterogeneous signal enhancement is exemplified [9,10].

**Therapeutic options**

Osteochondromyxoma can be appropriately managed with comprehensive surgical eradication. Inadequate surgical extermination of the neoplasm may result in localized tumour reoccurrence [9,10].

Comprehensive surgical extermination of osteochondromyxoma ensures appropriate alleviation and is associated with a superior prognosis. As localized tumour reoccurrence is frequently discerned following inadequate surgical resection, neoplastic relapse is common within sites where comprehensive tumour resection is challenging to attain. Distant tumour metastasis is unknown [9,10].

![Figure 1: Osteochondromyxoma appearing as a diagnostic criterion of Carney complex [11].](image-url)
Figure 2: Osteochondromyxoma delineating cogent genomic alterations of tumour suppressor gene PRKAR1A situated upon chromosome 17q22-24 [12].

Figure 3: Osteochondromyxoma demonstrating a heterogeneous neoplasm with a bubbly appearance within the skull bones and sellar region [13].
Figure 4: Osteochondromyxoma depicting a chondroid, osseous and myxoid matrix admixed with sheets and lobules of polygonal or stellate, chondroblast-like and osteoblast-like cells [14].

Bibliography


The Muculent Ossein- Osteochondromyxomas


11. Image 1 Courtesy: Carney complex community.

12. Image 2 Courtesy: Science Direct.

13. Image 3 Courtesy: Quantitative imaging in medicine and surgery.


Volume 4 Issue 4 April 2021
©All rights reserved by Anubha Bajaj.