

## Soggy and Muculent - Chondromyxoid Fibroma

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Chondromyxoid fibroma is designated as an infrequent, benign, cartilaginous neoplasm commonly incriminating proximal or distal segments of long bones. Upregulation of GRM1 is pathognomonic feature associated with chondromyxoid fibroma although molecular assay remains superfluous for ascertaining chondromyxoid fibroma.

Morphologically, distinct zonal architecture is exemplified. Localized tumour reoccurrence is common although malignant metamorphosis is extremely exceptional.

The exceptional, benign, chondroid neoplasm commonly emerges within young adults between second decade to third decade although no age of tumour emergence is exempt. Neoplasm is frequently diagnosed > 30 years. A slight male predominance is observed [1,2].

Majority of chondromyxoid fibromas arise within metaphyseal region of long tubular bones. Tumour may extend to epiphyseal line or infrequently about the articular surface. However, neoplasms exclusively confined to epiphyseal region are absent [1,2].

Classic site of neoplastic emergence is upper one-third of tibia. Besides, distal femur, small bones of hands and feet, pelvis or sacrum may be incriminated. Vertebral column, calvarium, base of skull, mandible or sternum may infrequently display the neoplasm [1,2].

Of obscure aetiology, majority (~90%) of chondromyxoid fibromas demonstrate upregulation of glutamate receptor gene GRM1 situated upon coding region of chromosome 6 through recombination with numerous partner genes [1,2].

Typically, chondromyxoid fibroma exhibits progressive pain of extensive duration. Besides, affected extremity enunciates bony swelling and restricted range of motion [1,2].

Neoplasms occurring within flat bones or small bones manifest as tumefaction associated with occasional pain. Additionally, short tubular bones of hands and feet with minimal circumscribing soft tissue may demonstrate restricted range of motion [1,2].

Cytological examination exemplifies moderately cellular smears demonstrating fragments of fibrillary, metachromatic myxochondroid tissue. Fragments of spindle-shaped cells appear variably intermingled with osteoclast-like giant cells. Nuclear atypia is variable and the lesion may be misinterpreted as a malignant neoplasm. Detection of focal calcification may be challenging upon cytology [1,2].

Grossly, a lobulated tumefaction with an uninterrupted periosteum is observed. Cut surface is firm, glistening and exhibits variable myxoid areas. Focal calcification may be challenging to discern upon gross examination [1,2].

Upon microscopy, tumefaction exhibits a lobulated architecture. Neoplastic lobules appear subdivided by fascicles of mononuclear, spindle-shaped cells intermingled with multinucleated giant cells. Neoplastic lobules demonstrate hypo-cellular centric zone alternating with hyper-cellular periphery [1,2].

Intervening stroma is variably myxoid to chondroid thereby representing diverse stages of cartilaginous development [1,2].

Morphologically, neoplasm enunciates a zonal architecture constituted of lobules of myxoid to chondroid tissue commingled with spindle-shaped and multinucleated giant cells. Variable foci of coarse calcification may ensue [1,2].

Neoplastic lobules demonstrate stellate cells disseminated within myxoid ground substance. Tumour cells reside within lacunae of chondroid zones [1,2].

Neoplastic cells delineate abundant, variably eosinophilic cytoplasm with bipolar to multipolar cytoplasmic projections along with elliptical to spindle-shaped nuclei. Significant nuclear pleomorphism and nucleoli are occasionally discerned [1,2].

Periphery of tumour lobules display spindle-shaped, fibroblast-like cells commingled with scattered, multinucleated, osteoclast-like giant cells. Mitotic activity is minimal [1,2].

Intervening stroma exemplifies coarse calcification, especially within neoplasms occurring in elderly subjects and unusual locations. Focal lymphocytic infiltrate is common. Hemosiderin pigment deposition may be discerned [1,2].

Focal necrosis, cystic change or degenerative alterations are uncommon. Few (~10%) neoplasms demonstrate aneurysmal bone cyst-like foci [1,2].

Upon ultrastructural examination, neoplastic cells demonstrate cytoplasmic processes, intracytoplasmic glycogen and thickened nuclear membrane [1,2].

**Figure 1:** Chondromyxoid fibroma depicting chondroid cells with minimal atypia confined to lacunae interspersed within myxochondroid matrix [5].

**Figure 2:** Chondromyxoid fibroma depicting spindle-shaped fibroblastic cells admixed with benign chondroid cells embedded within lacunae confined to myxochondroid ground substance [6].

Chondromyxoid fibroma can be appropriately discerned in the absence of cogent immunohistochemistry. However, chondromyxoid fibroma is immune reactive to S100 protein and SOX9 whereas periphery of tumour lobules are immune reactive to smooth muscle actin(SMA). Chondromyxoid fibroma is immune non reactive to CD34, RUNX2 or cytokeratin [3,4].

Chondromyxoid sarcoma requires segregation from neoplasms such as central chondrosarcoma, chondroblastoma, osteochondroma, aneurysmal bone cyst, giant cell tumour, non ossifying fibroma or metaphyseal fibrous defect [3,4].

Appropriate discernment of chondromyxoid fibroma mandates integration of radiological and histological features. Tumefaction can be challenging to discern upon fine needle aspirates, core needle biopsy or miniature surgical tissue samples [3,4].

Upon plain radiography, chondromyxoid fibroma characteristically demonstrates an eccentric, lytic lesion circumscribed with sharply defined, sclerotic or scalloped intramedullary perimeter [3,4].

Neoplasm confined to long bones demonstrate eccentric, lytic, radiolucent, metaphyseal lesions enveloped by well defined, sclerotic or scalloped intramedullary perimeter [3,4].

Neoplasm seldom expands to incriminate a growth plate. Associated foci of secondary, aneurysmal bone cyst may be observed.

Computerized tomography (CT) is optimal for detecting intrinsic mineralization of the neoplasm [3,4].

Upon T1 weighted magnetic resonance imaging (MRI), isointense or hypo-intense images are observed. T2 weighted imaging enunciates a heterogeneous, hyper-intense image [3,4].

Enhanced uptake of fluorodeoxyglucose (FDG) is observed upon positron emission tomography (PET) [3,4].

Tumefaction confined to small bones and flat bones delineate lytic, loculated, expansible lesions. Neoplasm may be locally destructive. Lesions confined to ribs appear fusiform with attenuation of superimposed bony cortex [3,4].

Tumours confined to sacrum and vertebral column demonstrate cortical discontinuity with extension into adjacent soft tissue or spinal canal and require demarcation from site-specific, malignant neoplasms [3,4].

Comprehensive surgical extermination of neoplasm or en bloc tumour resection is a recommended mode of therapy.

Tumours unamenable to exhaustive surgical eradication may be subjected to adjuvant radiation following surgical intervention [3,4].

Complete disappearance of neoplasm subsequent to radiofrequency ablation is documented.

Simple curettage of neoplasm is associated with tumour reoccurrence (15%). Distant tumour metastasis is absent.

Malignant metamorphosis may ensue following radiation therapy [3,4].

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5. Image 1 Courtesy: Pathology outlines.
6. Image 2 Courtesy: Springer link.