Chapter 23
Chondromyxoid Fibroma

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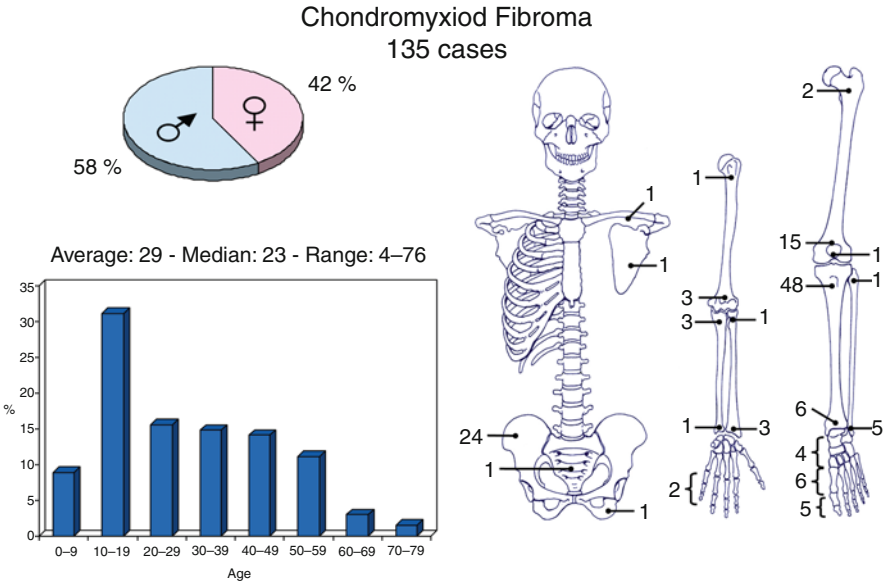
Definition: Benign cartilaginous tumor made of lobulated, fibromyxoid, and chondroid tissue.

Epidemiology: It is definitely rare (0.5 % of all bone tumors, about 2 % of all benign neoplasms) and prefers the male sex by 1.5–1. Generally seen between 5 and 30 years of age, it has a predilection for the second and third decades of life.



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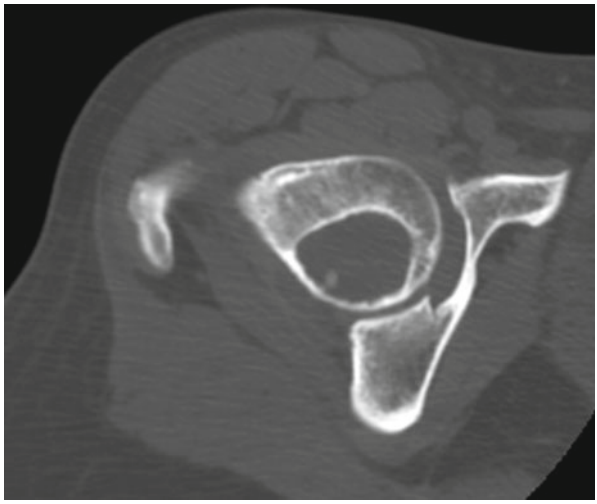
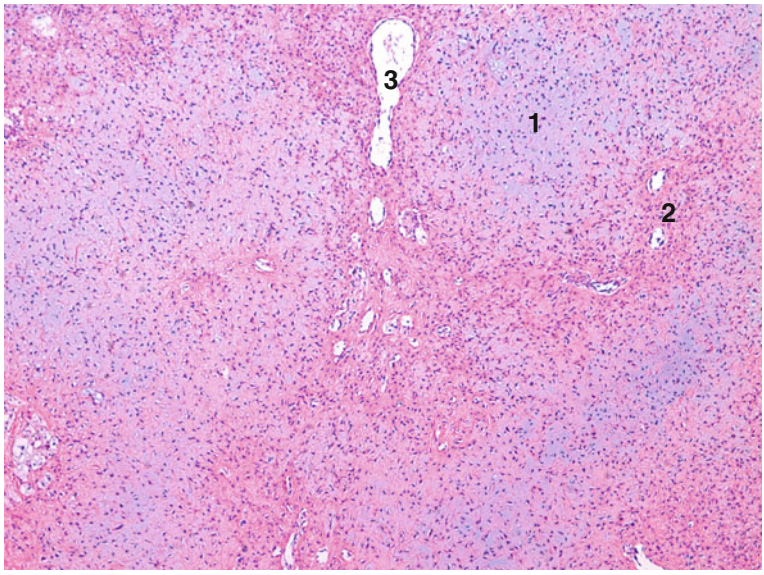
Location: Typically located in the long bone metaphysis, it can invade the epiphysis, especially in adults. Preferred sites proximal tibia (30 % of cases), small bones of the foot, and pelvis. Rare in the upper limb and in the trunk besides pelvis.
Clinical: Long-standing mild to moderate pain sometimes associated with bland local swelling is the typical scenario, because of the rather slow growth. Occasionally asymptomatic and discovered as incidental finding on roentgenograms.
Imaging: Small (generally <5 cm), metaphyseal, and eccentric radiolucent defect, usually with the long axis parallel to the bone of origin and occupying one half to two thirds of the diameter of the shaft; sometimes, especially in small bones, fusiform expansion of its entire contour. Sharply marginated by a lobulated shell of endosteal reactive bone because of a sclerotic rim. The cortex is usually canceled, with the tumor "bubbling out" into soft tissue. Little and peripheral periosteal reaction, chronic in nature and similar to that of periosteal chondroma. Intratumoral calcification is unusual. Isotope scan: moderately hot and corresponding to radiographic extent. CT: metaphyseal, eccentric, and subperiosteal, heavily marginated radiolucency, usually without any mineralization. MRI: homogenous intratumoral signal.
Histopathology: Gross: the tumor is rubbery soft, distinctly lobular, and clearly separated from the surrounding bone. Tissue is whitish or bluish and semitranslucent in chondromyxoid areas, tannish red in the undifferentiated and vascularized zones. Histology**:** lobular pattern better appreciated at low power, a light center, and a dark periphery. Basic elements stellate cells (blue) and myxoid background; center of lobule is hypocellular, periphery hypercellular. While the smaller lobules are exclusively or prevalently dark, the larger lobules are predominantly light, the dark part being limited to a narrow layer at the periphery of the lobules and in between them. These dark bands are absent where the lobules fuse together. Mitotic figures are not common. Cellular atypia may be present (15–20 % of cases). Occasionally, chondroblastoma-like cells at periphery.
Course and Stage: The tumor grows slowly and is generally small, at presentation usually stage 2 and occasionally stage 3. Few reports of malignant transformation.
Treatment: Aggressive curettage is usually indicated and frequently curative for either stage 2 and stage 3 tumors; recurrence rate probably lower than GCT and chondroblastoma. Consider en bloc resection for recurrent lesions and expendable bones.

Key Points

Clinical	Mild symptoms
Radiological	Lobulated, subperiosteal lytic lesion
Histological	Lobules of myxoid cartilage surrounded by more cellular bundles with vessels
Differential diagnosis	Histiocytic fibroma (at imaging)

TC: the lesion is well limited by a sclerotic line, metaphyseal, and eccentric, and contains small calcifications (usually non-detected on radiographs)





Histologic characteristics are particularly evocative at low power view. This shows the typical lobular architecture of the tumor. (*1*) Clear center of the lobuli. They are composed of spindle-stellate cells interspersed in an abundant and fluid ground substance. These areas represent the more mature phase of the tumor and are avascular. (*2*) Dark periphery of the lobuli. There are more densely cellular areas composed of well stained and usually not very pleomorphic cells with plump nuclei. Numerous and ectatic blood vessels run through these peri- and interlobular bands. These areas represent the younger more proliferative and undifferentiated phase of the tumor. (*3*) Blood vessels

Selected Bibliography

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