Chondromyxoid Fibroma

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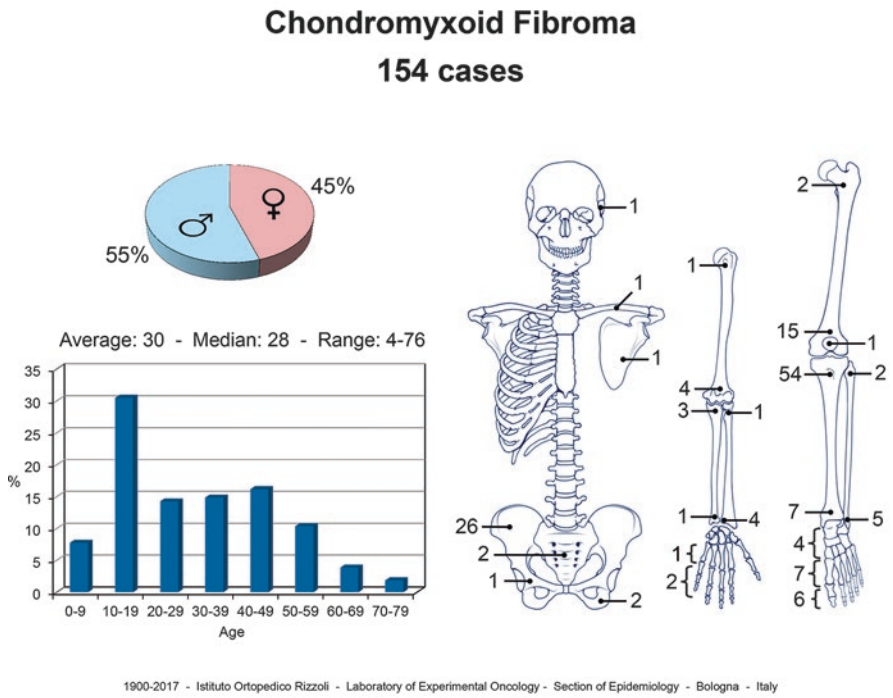
Definition: Benign cartilaginous tumor made of lobulated, fibromyxoid, and chondroid tissues.

Epidemiology: It is definitely rare (0.5% of all bone tumors, about 2% of all benign neoplasms) and prefers 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 metaphysis of long bone; it can invade the epiphysis, especially in adults. Preferred sites: proximal tibia (30% of cases), small bones of the foot, pelvis. Rare in the upper limb and in the trunk besides pelvis.

Clinical: Usually a slow-growing lesion with a relatively long history of mild to moderate pain, sometimes associated with bland local swelling. Occasionally asymptomatic and discovered as incidental finding on radiographs.

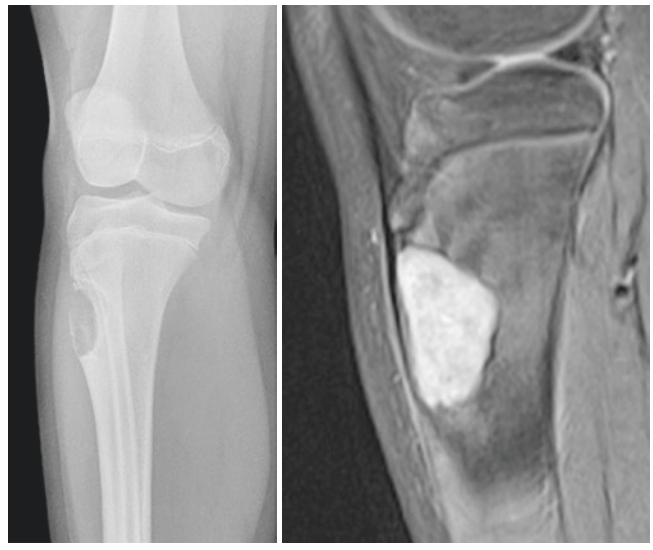
Imaging: Small (generally ≤5 cm), metaphyseal, and eccentric radiolucent defect, usually with the long axis parallel to the bone of origin; sometimes, especially in small bones, fusiform expansion of its entire contour. Sharply marginated by a lobulated shell of endosteal reactive bone because of a sclerotic rim. The cortex is usually cancelled, 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 (intermediate-high in T2, low-intermediate in T1).

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. In the centers of the lobules, spindle–stellate cells in a myxoid background are present; the center of lobule is hypocellular, while at the periphery a condensation of polyhedral cells is evident, often with multinucleated giant cells. Mitotic figures are not common. Cellular atypia may be present (15–20% of cases).

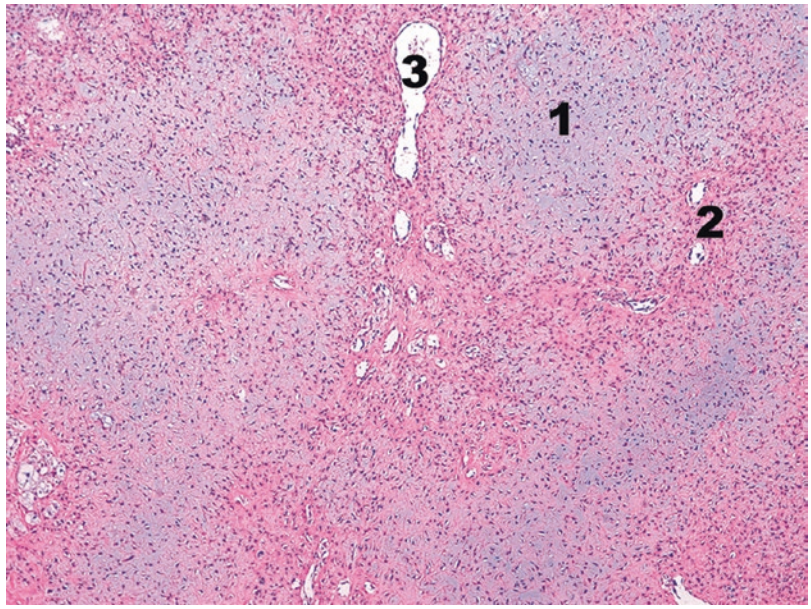
Course and stage: The tumor grows slowly and is generally small; at presentation usually stage 2 and occasionally stage 3.

Treatment: Aggressive curettage is usually indicated and frequently curative for either stage 2 or stage 3 tumors; recurrence rate probably lower than GCT of bone and chondroblastoma. Consider en bloc resection for recurrent lesions and expendable bones.

Key points	
• Clinical	Mild symptoms
• Radiological	Lobulated, subperiosteal lytic lesion
• Histological	Myxochondroid lobules of cartilage surrounded by more cellular bundles with vessels
• Differential diagnosis	Histiocytic fibroma (at imaging)



Radiograph and sagittal T2W image. Metaphyseal cortical well-limited lytic lesion in a 13-year-old boy



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. (2) Dark periphery of the lobuli. There are more densely cellular areas composed of well-stained cells with plump nuclei. (3) Numerous ectatic blood vessels run through these peri- and interlobular bands

Selected Bibliography

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