

Chapter 5

Histiocytic Fibroma

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Synonyms: Non-ossifying fibroma, Metaphyseal fibrous defect, Fibrous cortical defect, Fibrous xanthoma

Definition: It is a common benign lesion essentially consisting of histio-fibroblastic tissue, most frequently originating eccentrically, in the metaphyseal portion of a long bone, in a skeletally immature individual.

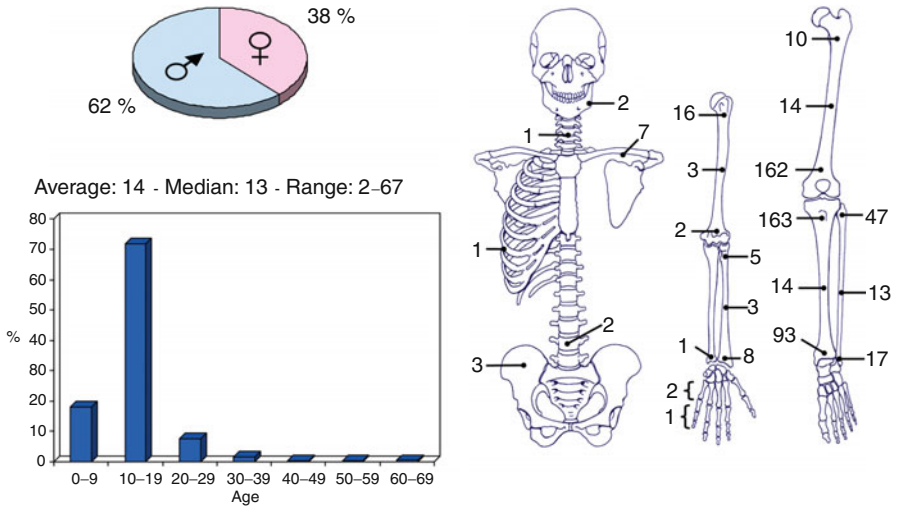
The tendency to remain asymptomatic and spontaneously resolve in most instances and the common relationship with tendon and ligament insertions in proximity of the growth plate suggest a posttraumatic or developmental defect producing faulty ossification rather than a true neoplasm. It is not infrequent to see two or three lesions in either one or both lower extremities of the same patient. A different and very rare clinical scenario is the association of numerous multiple histiocytic fibromas, more extensively involving the skeleton, with extraskeletal abnormalities. This condition known as Jaffe-Campanacci syndrome will be discussed separately.

Epidemiology: Overall, it is a very frequent lesion, the incidence of which has been estimated around 30 %. However, patients are in most instances asymptomatic, and the true incidence is therefore underestimated. There is slight male sex predominance. It is typical in childhood and adolescence, being rare before 5 and after 20 years of age.

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Histiocytic Fibroma

563 cases (including 18 multicentric cases)



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Localization: It is generally localized in the metaphysis at first and then displaced toward the diaphysis. It starts intracortically/subperiosteally. Most of the lesions are located around the knee and ankle. It is rare in the proximal femur and upper limb, exceptional in the trunk, hand, and foot. Two or three lesions may appear in the same or both lower limbs (e.g., distal femur and proximal tibia).

Clinical Aspects: Diagnosis is usually based on a radiogram obtained for unrelated reasons (usually trauma), histiocytic fibroma being the most classic and common lesion diagnosed as incidental finding. Rarely, pathologic fractures may occur in larger lesions (more than 1/2-2/3 of the bone cross section, particularly for the distal tibia and fibula).

Imaging: Standard X-rays are usually diagnostic. The defect is metaphyseal, intracortical, and/or subperiosteal. Generally lobulated, its inner boundaries are surrounded by a rim of bone sclerosis. The osteolytic image may appear multilocular due to corrugations of the bony wall. The cortex is sometimes attenuated, rarely slightly expanded due to chronic periosteal reaction.

Pathology: Tissue is compact, rubbery soft, tan-brown in color, sometimes containing yellow (foam cells) or dark (hemosiderin) areas. Dense network of cellular typically plump spindle cells with a prominent storiform pattern and with scattered

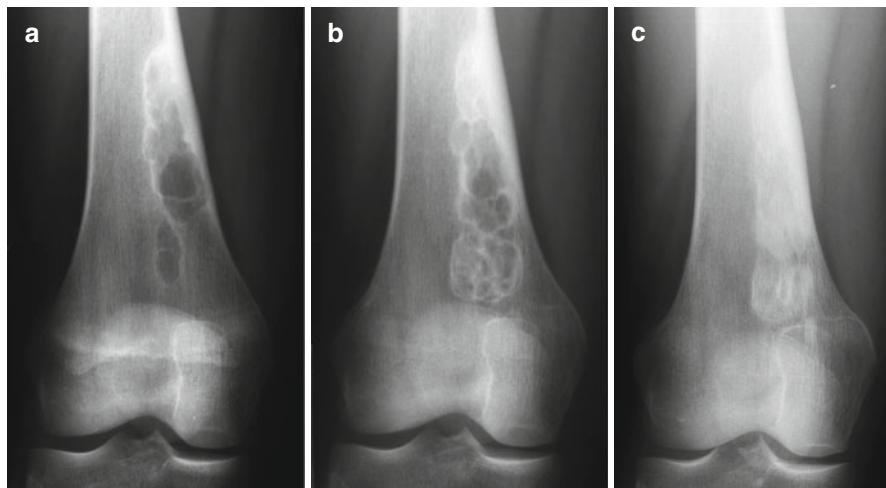
multinucleated giant cells are evident. Intra- and extra-cytoplasmatic hemosiderin and lipid-loaded foam cells are common findings.

Course and Staging: Tumor growth stops at skeletal maturity and very frequently even before. The lesion then tends to ossify slowly. Stage is initially 1 or 2 and then constantly becomes 1.

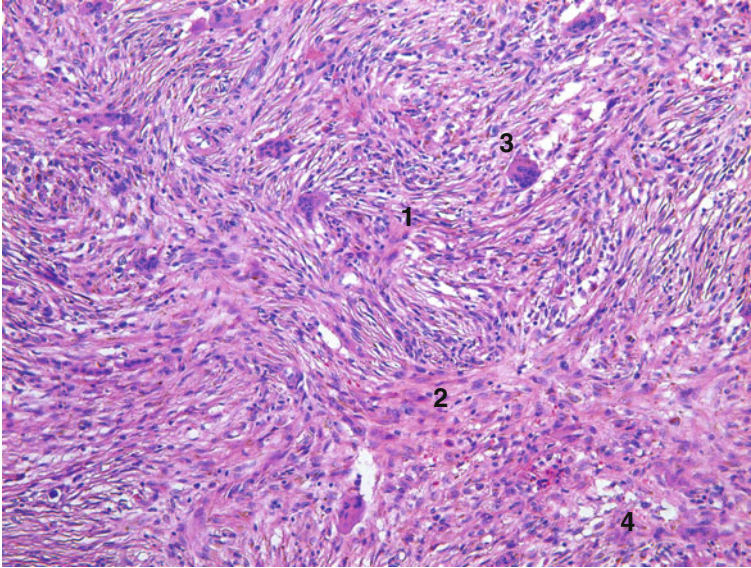
Treatment and Prognosis: The majority of histiocytic fibromas do not require treatment; diagnosis can usually be made on clinico-radiographic features avoiding biopsy. Occurrence of a pathologic fracture per se is not an indication for surgery, as fractures heal essentially in a normal fashion. Curettage and bone grafting are rarely needed for large lesions, possibly associated with internal fixation for displaced fractures.

Key Points

Clinical	Incidental findings
Radiological	Eccentric, lobulated, subperiosteal, or intracortical
Histological	Bland spindle cells without atypia and with foam cells
Differential diagnosis	Chondromyxoid fibroma (at imaging)



Radiographs of the knee. The lesion is lytic, metadiaphyseal, centered on the cortex, and limited by a sclerotic line. As it was painless, diagnosis of histiocytic fibroma was sure, and biopsy was not performed. With time ossification that slowly completely regressed appeared in the proximal (older part) lesion. (a) 1997 (b) 2000 (c) 2003



(1) Compact histio-fibroblastic tissue arranged in imbricated and whorled bundles (storiform pattern). (2) Great number of slightly plump and well-stained nuclei; rare and normal mitotic figures (active areas of the lesion). (3) Small and sparse giant cells, less numerous cells and more abundant collagenous bundles (florid stage). (4) Foam cells (regressive phenomena of the tumor)

Selected Bibliography

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