Histiocytic Fibroma

11

Laura Campanacci

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

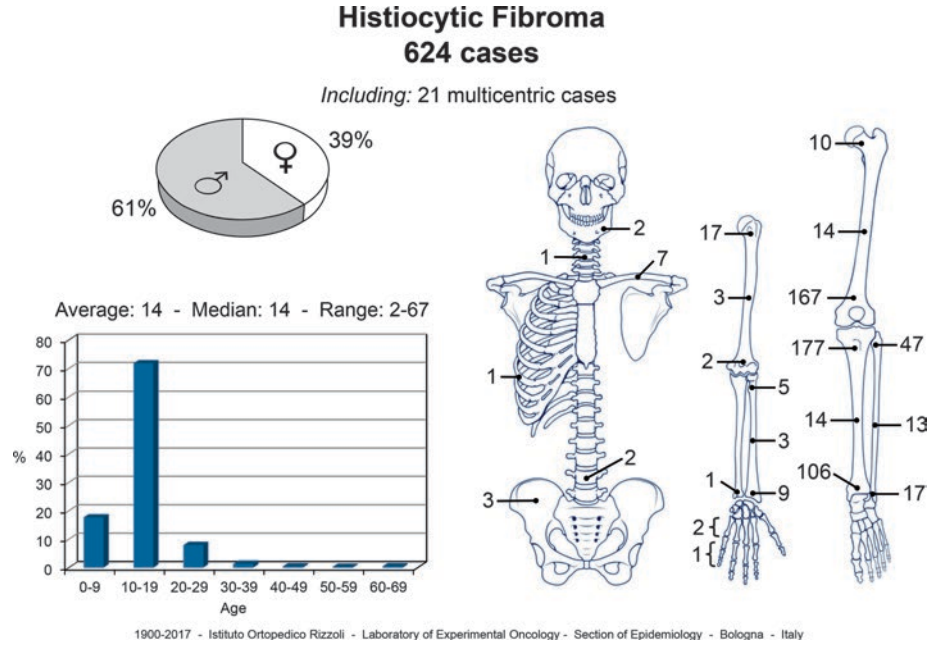
Definition: It is a common benign lesion essentially consisting of histio-fibroblastic tissue, most frequently originating eccentrically, in the metaphyseal portion of a long bone, in a skeletally immature individual.

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.

L. Campanacci (✉)
Orthopedic Oncology Department, IRCCS Istituto Ortopedico Rizzoli, Bologna, Italy
e-mail: Laura.campanacci@ior.it

P. Picci et al. (eds.), *Diagnosis of Musculoskeletal Tumors and Tumor-like Conditions*,
https://doi.org/10.1007/978-3-030-29676-6_11



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).

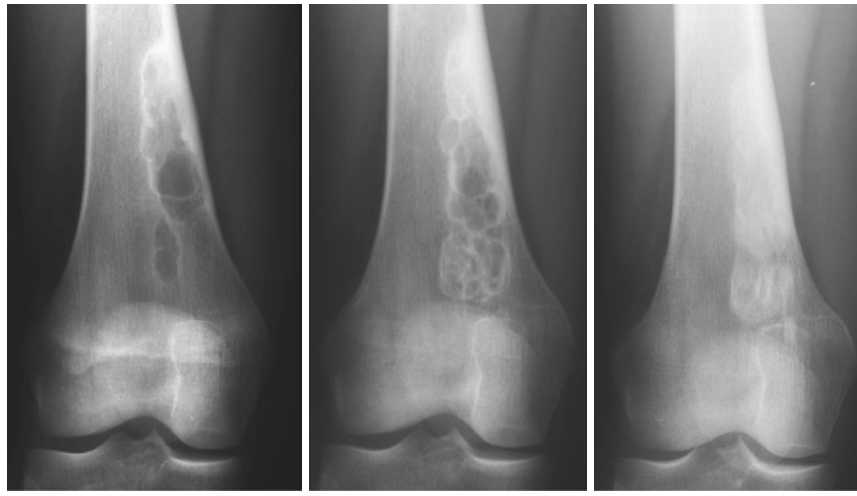
Imaging: Standard X-rays are usually diagnostic. The defect is metaphyseal, intracortical, and/or subperiosteal. Generally lobulated, its inner boundaries are surrounded by a rim of bone sclerosis. The osteolytic image may appear multilocular due to corrugations of the bony wall. The cortex is sometimes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

Histopathology: 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. Intra- and extra-cytoplasmatic hemosiderin and lipid-loaded foam cells are common findings. Mitotic figures, foci of reactive bone, and infiltration in between the host bony trabeculae can be present.

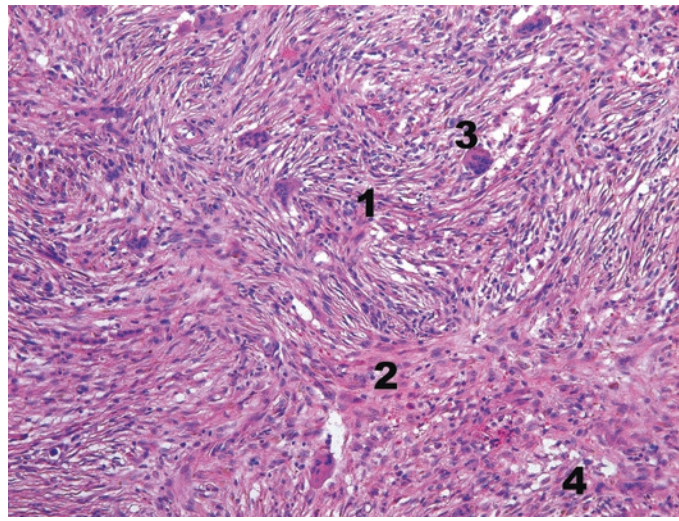
Course and Staging: Tumor growth stops at skeletal maturity and very frequently even before. The lesion then tends to ossify slowly. The stage is initially 1 or 2 and then constantly becomes 1.

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 in a storiform pattern
• Differential diagnosis	Chondromyxoid fibroma (at imaging) Fibrous dysplasia



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



(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)

11.1 Multiple Histiocytic Fibromas with Extraskeletal Abnormalities (Jaffe–Campanacci's Syndrome)

It is a very rare condition possibly linked to neurofibromatosis. Multiple large histiocytic fibromas extend to the long bones of one or both lower limbs or to the four limbs with prevalence in one side of the body and even including the pelvis. Café au lait skin spots are usually seen. Occasionally, mental retardation, hypogonadism or criptorchidism, ocular and cardiovascular anomalies, and other skin alterations suggestive of neurofibromatosis are associated.

More frequently symptomatic, as compared to the usual histiocytic fibroma, they cause slight

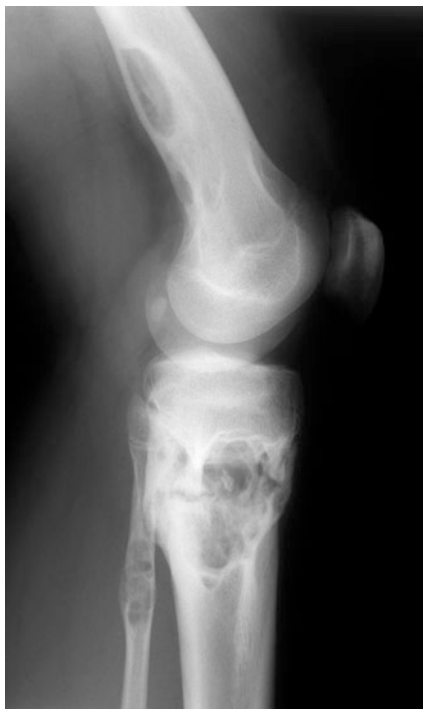
expansion of the bone, stress or pathologic fractures, and sometimes deformity or limb length discrepancy.

Imaging: Lesions are rather extensive and, from the metaphysis, tend to involve the diaphysis. Osteolyses are mostly intracortical or eccentric. The cortex may be very thin or absent. Differential diagnosis includes multifocal fibrous dysplasia, but imaging and histopathology is quite different in the two lesions.

Histopathology: Same as conventional histiocytic fibroma.

Course: Similar to conventional histiocytic fibroma. The lesions do not expand after skeletal maturity and then tend to be replaced by sclerotic bone.

Treatment: Aimed to prevent pathologic fractures and address deformities.



Radiograph of the knee, lateral view. Multiple histiocytic fibromas. The lesion of the tibia is broken. The one on the fibula appears centered and not cortical. This pattern is frequent in thin bones

Selected Bibliography

Betsy M, Kupersmith LM, Springfield DS. Metaphyseal fibrous defects. J Am Acad Orthop Surg. 2004;12:89–95.

Jee WH, Choe BY, Kang HS, et al. Nonossifying fibroma: characteristics at MR imaging with pathologic correlation. Radiology. 1998;209:197–202.

Levine SM, Lambiase RE, Petchprapa CN. Cortical lesions of the tibia: characteristic appearances at conventional radiography. Radiographics. 2003;23:157–77.

Mankin HJ, Trahan CA, Fondren G, Mankin CJ. Nonossifying fibroma, fibrous cortical defect and Jaffe-Campanacci syndrome: a biologic and clinical review. Chir Organi Mov. 2009;93(1):1–7. Review.

Mirra JM, Gold RH, Rand F. Disseminated nonossifying fibromas in association with café-au-lait spots (Jaffe-Campanacci syndrome). Clin Orthop Relat Res. 1982;(168):192–205.

Vanel D, Ruggieri P, Ferrari S, Picci P, Gambarotti M, Staals E, Alberghini M. The incidental skeletal lesion: ignore or explore? Cancer Imaging (2009);9(Spec No A):S38–43. Review.