Chapter 6
Multiple Histiocytic Fibromas with Extraskeletal Abnormalities (Jaffe-Campanacci's Syndrome)

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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 spots are usually seen. Occasionally, mental retardation, hypogonadism or cryptorchidism, 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 are quite different in the two lesions.

Pathology: 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.

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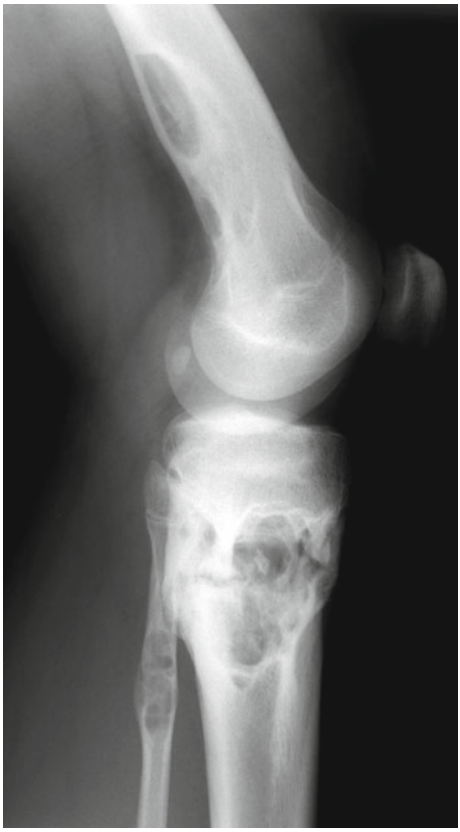
Radiograph of the knee, lateral view. Multiple histiocytic fibromas. The lesion of the tibia is broken. The one on the fibula appears centered, and not cortical. This pattern is frequent in thin bones



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