

Case 1 [2010]



A 45-year-old-woman presented with a long-standing history of left lower leg pain distributed from the midportion of the lower leg to the foot and ankle. She had no history of relevant trauma or constitutional symptoms suggesting systemic illness. The patient experienced little relief of her leg pain with 4 weeks of rest. A physical examination showed no swelling, redness, or warmth. She had normal laboratory findings. No leukocytosis was evident and ESR was 16 mm/hour. C-reactive protein was 0.02 mg/dL (normal range, 0–0.5 mg/dL), and serum alkaline phosphatase (ALP) was 44 IU/L .

Your Diagnosis?

Diagnosis: Ribbing Disease

1. X rays



Radiographs of both lower legs were obtained when the patient's left lower leg pain worsened

Cortical thickening and increased intramedullary bone density were observed. Left worse than right

There was suggestion of early subtle changes in the right midtibial diaphysis.

D/d

Ribbing disease

Camurati-Engelmann disease

Intramedullary osteosclerosis

Erdheim-Chester disease

Chronic multifocal sclerosing osteomyelitis

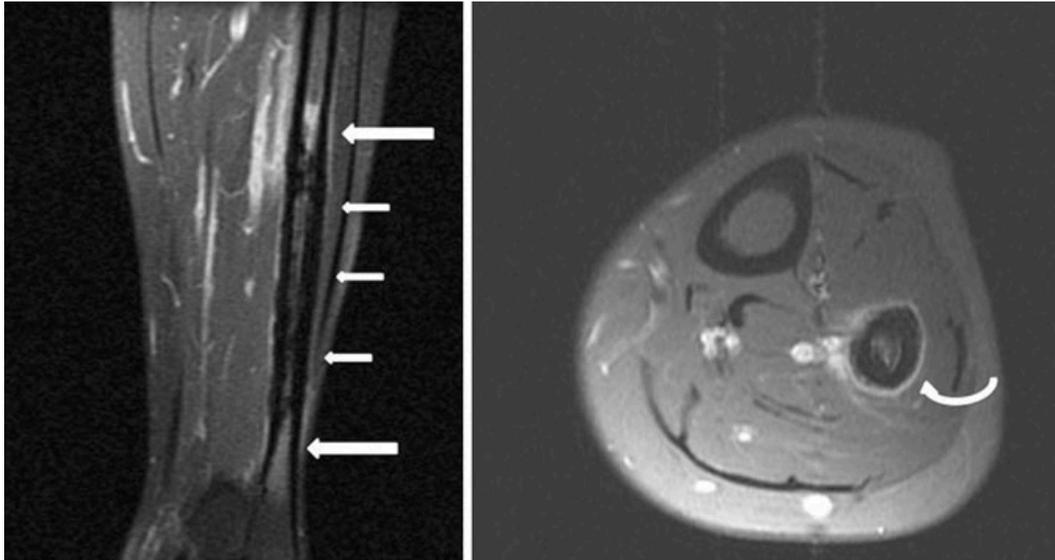
Hardcastle syndrome

2. MRI

Cortical thickening (small arrows) and bone marrow edema (large arrows) of the diaphysis of the left fibula are seen on this coronal MRI.

An axial gadolinium enhanced fat-suppressed image shows periosteal reaction (curved arrow) of the left fibula.

?Chronic infection

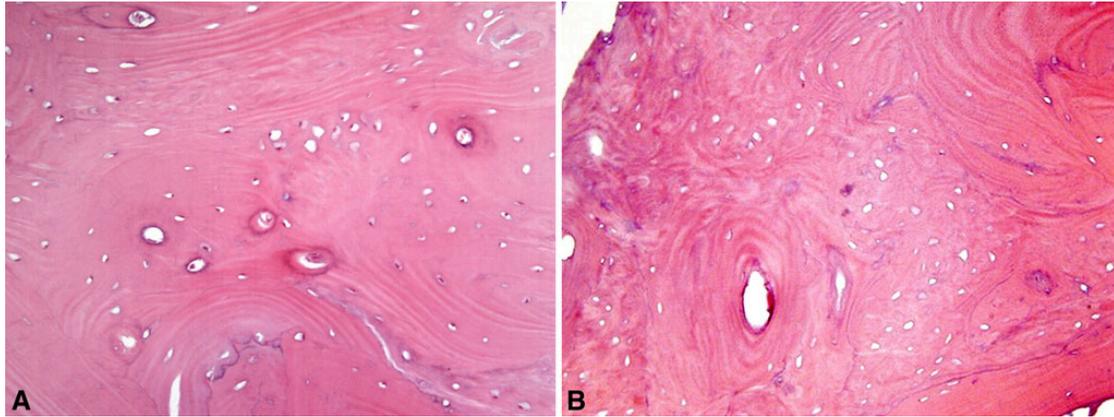


3. Bone Scan: showed hot uptake of the left tibia, , and a new subtle uptake on the right tibia.



4. Biopsy:

The permanent histologic section showed thickened trabecular and a variably sized Haversian system at the sclerotic bony lesion. Many empty lacunae, which could suggest dead bone fragments.



Discussion

Clinical and radiologic features of our patient showed asymmetrical bilateral diaphyseal involvement of long bones with surrounding bone marrow edema. Those features combined with the absence of inflammatory signs confirmed laboratory findings, and analysis of the biopsy specimen suggested Ribbing disease. Here involvement is unilateral or when bilateral, symmetrical. Only long bones are involved.

We considered the primary differential considerations to be Camurati-Engelmann disease, intramedullary osteosclerosis, Erdheim-Chester disease, chronic multifocal sclerosing osteomyelitis, and Hardcastle syndrome.

Camurati-Engelmann disease, an autosomal dominant disorder, manifests as pain, progressive leg weakness, elongated extremities, and gait disturbances, and is characterized by a symmetric and bilateral distribution with an onset during childhood. It may be associated with gait and neurologic abnormalities, which are absent with Ribbing disease. Long bones and skull may be involved.

Intramedullary osteosclerosis is an abnormal intramedullary new bone formation located mainly in the shaft of a long bone, especially the tibia, in adults. It is a rare condition and is more common in females. The presence or absence of associated reactive marrow edema can be used to distinguish intramedullary sclerosis from Ribbing disease. Whereas intramedullary sclerosis is characterized chiefly by

increased bone density, Ribbing disease usually shows edema and enhancement on T2-weighted and postgadolinium T1-weighted MR images, respectively, in the adjacent marrow.

Erdheim-Chester disease is a rare condition characterized by an abnormal increase in histiocytes, and usually occurs during middle age. It has some histiocytic disorder-favoring signs like exophthalmos, diabetes insipidus, and fever. In addition, biopsy specimens of Erdheim-Chester disease lesions differ from those of Ribbing disease.

The laboratory findings of chronic multifocal sclerosing osteomyelitis are characterized by elevated ESR and leukocyte count, but laboratory findings for our patient were within normal.

Hardcastle syndrome is characterized by diaphyseal medullary stenosis of long bones surrounding cortical thickening. However, contrary to Ribbing disease, Hardcastle syndrome manifests as a history of pathologic fracture after minor trauma, radiographs showing multiple permeative cortical radiolucencies and longitudinal metaphyseal striations, and malignant transformation between the second and fifth decades of life.

Ribbing disease is a rare condition of unknown etiology reflected by a sclerosing dysplasia of the diaphyses of long bones. It was first described by Ribbing in 1949. The tibia and femur usually are involved, but it also affects the fibula, radius, and ulna. The disease is asymmetric or asynchronous bilateral, and is suspected to show an autosomal recessive trait. It occurs after puberty. The typical presenting symptom is bone pain, which may be associated with swelling. Furthermore, the pain appears to be resistant to medication, and to be associated with physical activity. It can resolve spontaneously with time or progress.

On MR images, the intramedullary and bone marrow edema are characteristic of Ribbing disease. Histologically, the most commonly described findings are increased cortical thickness and Haversian canal narrowing. And empty lacunae.

This patient was treated with intramedullary reaming. When the patient returned to our clinic 2 months later, her left lower leg pain had subsided. We presume that

decompression resulted in pain relief and that obliteration of the medullary canal caused recurrence of the pain.

Reference: AJR 167: 689 [1997]