Case Report

Ribbing's Disease: Radiographic-Scintigraphic Correlation and Comparative Analysis with Engelmann's Disease

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Engelmann's disease is an uncommon condition characterized radiographically by symmetric diaphyseal sclerosis involving predominantly the tubular bones. Ribbing described an entity with similar but less extensive or severe changes which he thought differed sufficiently to constitute a distinct process. We present two cases of Ribbing's disease and one of Engelmann's disease, emphasizing the differences between the two. Radiographic-scintigraphic correlation is provided to demonstrate the value of skeletal scintigraphy in cases of Ribbing's disease and progressive diaphyseal dysplasia.

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Engelmann's disease, or progressive diaphyseal dysplasia, is an uncommon condition characterized radiographically by symmetric diaphyseal sclerosis involving the tubular bones. The syndrome was first described by Cockayne (*1*) in 1920 and further defined by Camurati (*2*) in 1922. In 1929, Engelmann (*3*) reported a patient with diaphyseal sclerosis associated with abnormal gait, neurological disturbances, growth retardation and poor muscular development.

In 1949, Ribbing (*4*) described four siblings in their third decade with diaphyseal sclerosis of the femora and tibiae which was not always symmetric. He observed that his cases differed in several ways from the reported cases of Englemann's disease: (a) presentation occurred after puberty without gait or neurological abnormalities; (b) skeletal involvement was less extensive; and (c) histological findings consisted of only new bone formation, contrasting the bone formation and resorption seen in Engelmann's disease.

Thus, Ribbing concluded that his cases differed sufficiently to constitute a separate entity, which he termed "hereditary multiple diaphyseal sclerosis." Since that time, studies of several families with Engelmann's disease have demonstrated an autosomal dominant mode of inheritance (*5,6*). Contrariwise, a few authors have raised the question of autosomal recessive inheritance in Ribbing's disease (*5*) while others consider Ribbing's disease a "variant" or "adult" form of progressive diaphyseal dysplasia (*6*).

We present two cases of Ribbing's disease and one of Englemann's disease, emphasizing the differences between the two entities. Also, we present heretofore unreported radiographic-scintigraphic correlation in Ribbing's disease and suggest the potential value of bone scanning in this entity.

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CASE REPORTS

Case 1

A 27-yr-old black female, first seen at another institution in November, 1981, complained of a lump on her right leg. Several months prior to admission, the patient developed intermittent pain and discomfort over the pretibial region. The past medical history was unremarkable. The patient was first seen in January 1982 because of symptoms related to her right lower leg and abnormalities on radiographs at the outside institution. Physical examination revealed swelling and warmth over the anterior aspect of the right lower leg. The left lower extremity was normal.

Radiographs of the lower extremities demonstrated cortical thickening in the anterior tibiae, greater on the right (Fig. 1A).

A bone scan revealed increased activity in the mid-diaphyses of the tibiae (Fig. 1B). Serum calcium, electrolytes, uric acid, alkaline phosphatase and hematology profile were normal. There were no clinical or historical features to suggest a cause for the findings. The patient was given analgesics for symptomatic relief.

The patient returned in March 1984, with pain and tenderness in the left pretibial region. She was treated symptomatically and has not returned for followup visit.

Case 2

A 37-yr-old white female first became symptomatic in March, 1984 when she developed mild, intermittent right anterior tibial pain. Chemistry profile demonstrated an increased serum alkaline phosphatase which had been normal in 1983. Her most recent alkaline phosphatase remains elevated at 180 (N < 115 IU), the only laboratory abnormality. A VDRL was negative. In late 1984 and early 1985, the pain became more severe. Examination revealed tenderness and slight warmth over the anterior aspect of the right tibia.

Radiographs demonstrated mild cortical thickening in both tibiae (Fig. 2A). Bone scan on two occasions revealed increased diaphyseal uptake in both tibiae and femora (Fig. 2B), suggesting active disease while the radiographic findings were minimal.

Presently, the patient is being treated symptomatically. One of the patient's children complained of leg pain, but a bone scan was normal.

Case 3

A 29-yr-old white female described a 12-yr history of intermittent severe bilateral leg pain. She has also suffered from long-standing muscle weakness, fatigability and severe recurrent headaches. Her symptoms have subjectively worsened over time and at present she has difficulty climbing stairs. In addition, she has slight deafness in the right ear. Enlargement of the mandible and legs was evident at physical examination. Reflexes were brisk and symmetric. Laboratory studies were unremarkable, with normal ESR and biochemical indices of bone remodelling.

Radiographic survey revealed characteristic changes of Engelmann's disease (Figs. 3A–C) with a correspondingly abnormal distribution of uptake on bone scan (Figs. 3D–F).

Presently, symptomatic treatment with buffered aspirin is being undertaken. The patient has a 31-year-old sister with the same condition, but radiographs from two brothers were reported as negative. An 11-year-old daughter is presently asymptomatic.

DISCUSSION

Radiologically, Engelmann's disease is characterized by changes which begin in the diaphysis with a slight fusiform enlargement and progress towards the metaphyses. The epiphyses are spared. The long bones are usually affected symmetrically, with the tibiae, femora, fibulae, humeri, ulnae, and radii affected in decreasing order of frequency (*5*). The base of the skull is also commonly affected (*5*). Involvement of the mandible is less common but can occur in more severe instances. Bony involvement is usually quite extensive, even in patients who are asymptomatic or present after childhood.

In contrast, Ribbing's disease is characterized by less generalized involvement, usually of the tibiae and femora only, and sometimes with asymmetric involvement. Activity on bone scan would appear to correlate temporally with the clinical symptoms and complement the radiographic findings. In studying Engelmann's disease, Kumar et al., maintained that a normal scintigram indicates a "quiescent" or "mature" lesion while a normal radiograph with a positive scintigram indicates an early lesion (*7*). This is evidenced in Case 2 of Ribbing's disease, wherein minimal radiographic findings accompanied a bone scan with increased diaphyseal uptake. We believe these cases emphasize the role for isotope scanning in patients with lower extremity pain of obscure etiology or suspected Ribbing's disease, whether familial or sporadic.

Differential Diagnosis

Paget's disease, infantile cortical hyperostosis, syphillitic periostitis, hypervitaminosis A, hypertrophic osteoarthropathy, and venous stasis with periostitis can all be ruled out by clinical or laboratory tests and/or radiographic distribution. Chronic sclerosing osteomyelitis can be excluded by the bilaterality of involvement. Van Buchem's disease involves the epiphyses as well as diaphyses of tubular bones in addition to characteristic involvement of the mandible (*8*). Diaphyseal bone infarcts of sickle cell disease are excluded clinically. Engelmann's disease can be differentiated from craniodiaphyseal dysplasia, which is characterized by a combination of severe sclerosis involving the skull base and the cranial vault with cranial nerve compression, and involvement of bones of the hands and feet (*9*). Finally, stress reaction of the lower extremities is a diagnostic possibility (*10*), but can be excluded by history and more discrete isotope findings.

SUMMARY

We have reported two cases of Ribbing's disease and one of Engelmann's disease, contrasting the two entities. Severe cases of Engelmann's disease are readily recognized, but milder forms may pose a diagnostic dilemma. Case reports of "mild forms" may, in fact, represent Ribbing's disease and further study of this question is necessary. Our cases of Ribbing's disease illustrate the advantage of isotope bone scanning in assessing the degree of activity and distribution of the disease.

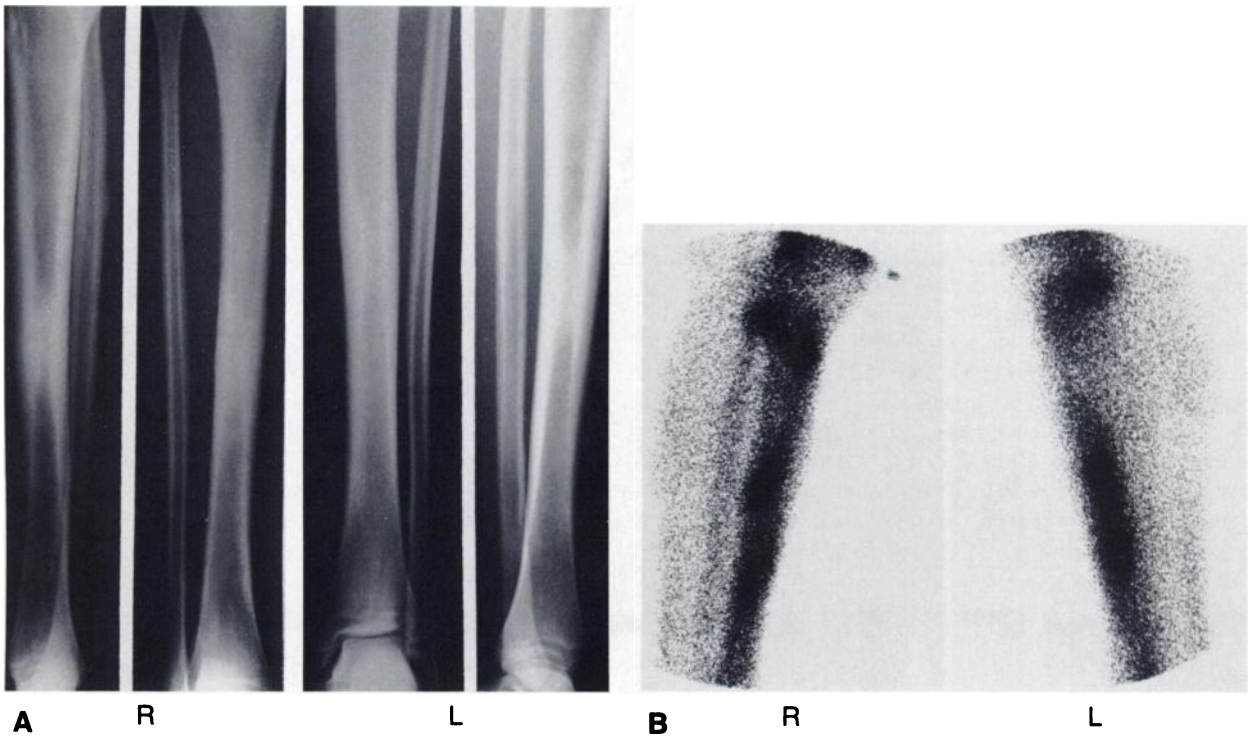

FIGURE 1
A: Cortical thickening in the mid-diaphyses of the tibiae, greater on the right. B: Bone scan. Symmetric, increased uptake in the mid-diaphyses of the tibiae.

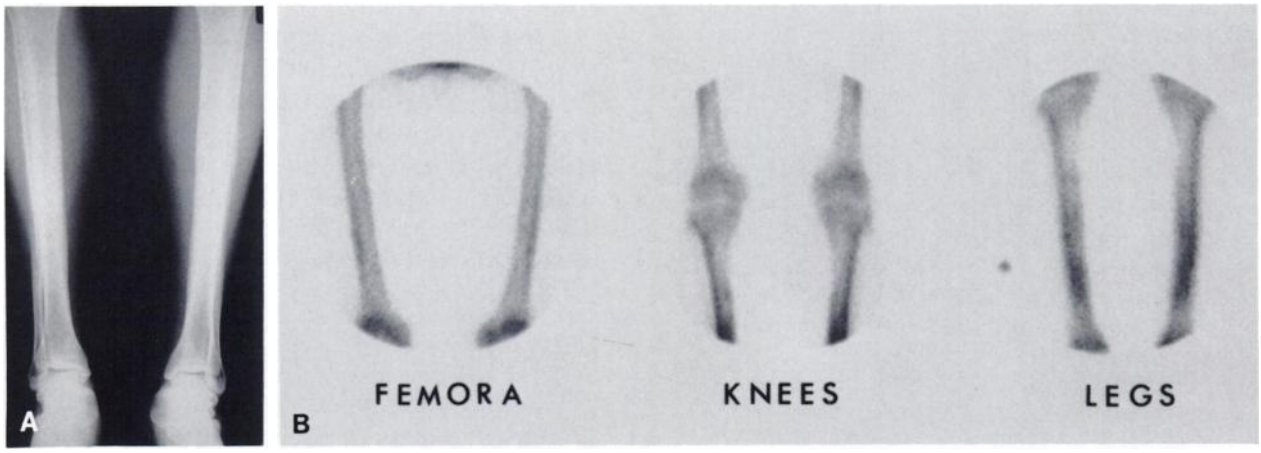

FIGURE 2
A: Minimal cortical thickening in both tibial diaphyses. B: Bone scan: Symmetrically increased diaphyseal uptake in the tibiae and, to a lesser extent, the femora.

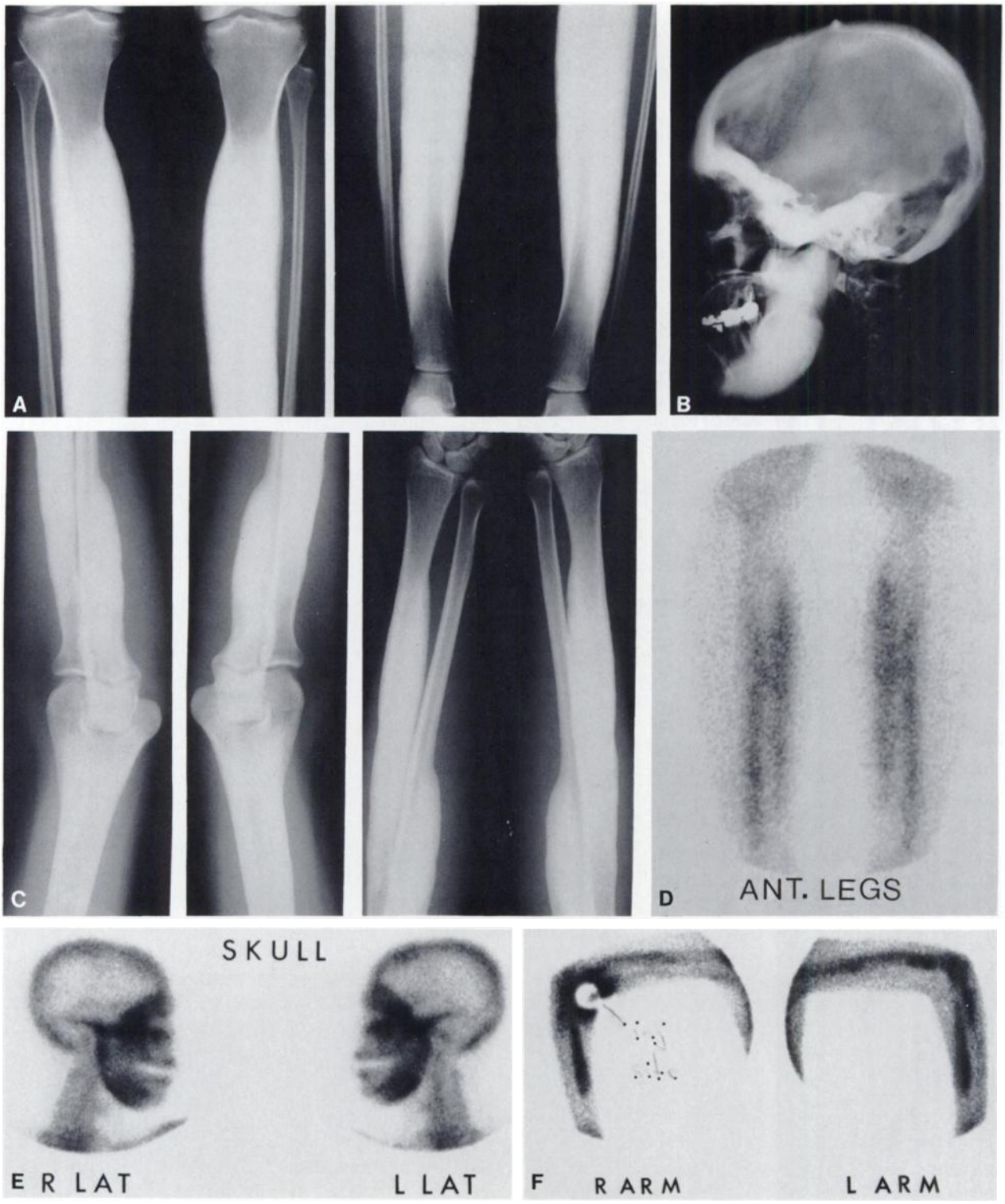

FIGURE 3
A: Legs. Marked, symmetric diaphyseal cortical thickening, sparing proximal and distal epiphyses. B: Skull. Sclerosis at base of skull with atypical involvement of the mandible. C: Forearms and elbows. Diaphyseal sclerosis sparing epiphyses and correlating with sites of increased uptake on isotope bone scan. D: Bone scan. Symmetric, increased diaphyseal uptake in the tibiae and fibulae. E: Bone scan (500k, 10% window). Increased activity in the mandible and skull base. F: Bone scan. Proximal ulnae and mid-radii demonstrate increased uptake, correlating with the radiographic sites of involvement. The humeri are involved as well.

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