Osteoarthropathy Associated with Weber-Christian Disease

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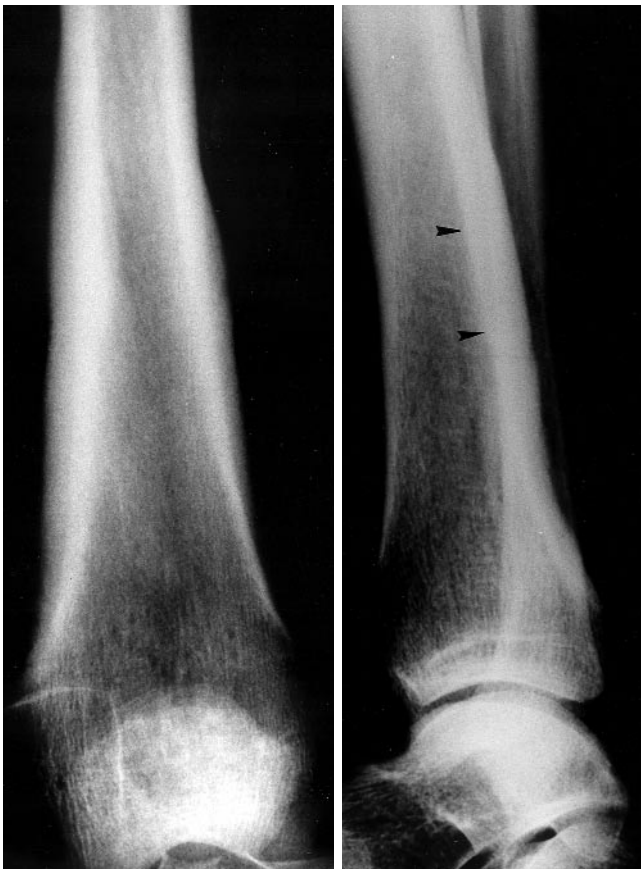
A 51-year-old man developed multiple, tender cutaneous nodules in both pretibial regions. He subsequently had a fever reaching 39°C. After a biopsy of the nodules, showing the typical findings of nodular panniculitis, and laboratory studies, he was diagnosed with Weber-Christian disease. The patient began treatment with oral prednisolone at a daily dose of 20 mg, and his symptoms were alleviated. Seven months later, the dose of prednisolone was gradually reduced to a daily dose of 10 mg, which controlled the patient's symptoms. However, 13 months after onset, the patient developed pain in the left knee and right ankle, and was referred to the orthopedic department. The patient had no history of pancreatic disease. Radiographically, the left distal femur and the right distal tibia showed cortical hyperostosis with periosteal reaction (Figures 1A and B). Computed tomographic (CT) scans of the left femur showed a markedly thickened cortex (Figure 2). Intramedullary spaces were inhomogeneously enhanced by contrast media. T1-weighted magnetic resonance (MR) images revealed an irregularly thickened cortex with hypointensity. In the bone



A B

Figure 1. A. An anteroposterior radiograph of the left distal femur shows cortical hyperostosis. B. A lateral radiograph of the right ankle shows cortical hyperostosis and irregularity in the posterior aspect of the tibia (arrowheads).

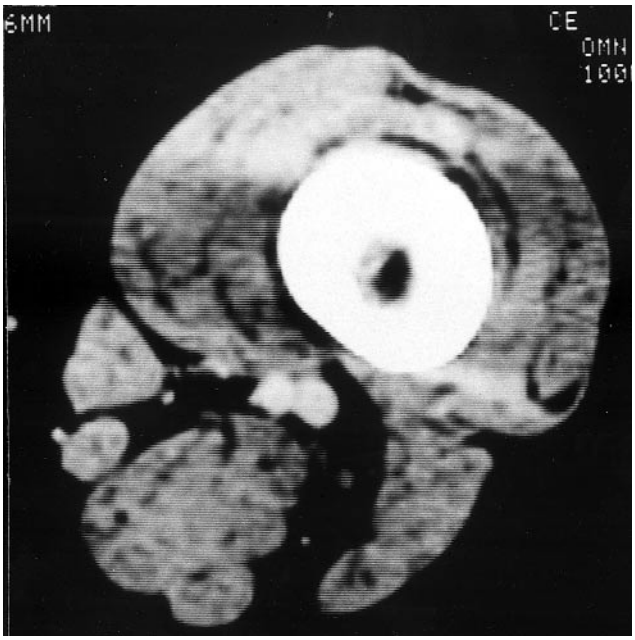


Figure 2. A contrast-enhanced CT scan of the left femur shows the markedly thickened cortex. The intramedullary space is inhomogeneously enhanced.

medulla, there were areas with a mixture of patchy hypointensity and hyperintensity compared to normal bone marrow (Figure 3A). T2-weighted MR images showed the bone medulla with inhomogeneous hyperintensity (Figure 3B). A ^{99m}technetium scintigraphic scan revealed an increased uptake of isotope in the right distal tibia, sternum, lumbar spine, both distal femora, ankles, knees, hips, hands elbows and shoulders (Figure 4). The patient subsequently underwent an open biopsy. Specimens from the left femoral bone marrow showed an admixture of fat necrosis and scattered chronic inflammatory cells (Figure 5). After biopsy, the patient was again treated with prednisolone at a daily dose of 20 mg, which alleviated the symptoms.

Weber-Christian disease is characterized by relapsing febrile episodes and systemic, cytophagic histiocytic panniculitis. Patients with systemic panniculitis are known to manifest musculoskeletal symptoms. These include acute and chronic arthritis/periarthritis predominantly affecting the ankle and knee[1]. Radiographs of the affected bones may show diffuse osteolysis, endosteal scalloping, and mild cortical hyperostosis[2-4]. Radiological and histological studies in the current case showed the presence of inflammatory and necrotic processes of fat in the bone marrow. In particular, our MR images distinctly demonstrated intramedullary, diffuse areas with abnormal intensity, presumably concordant with fat inflammation

To our knowledge, multimodality imaging findings of osteoarthropathy in Weber-Christian disease have never been reported. These findings are helpful in distinguishing the osteoarticular lesions from chronic osteomyelitis, bone neoplasms and osteoarthropathy associated with other rheumatic diseases.

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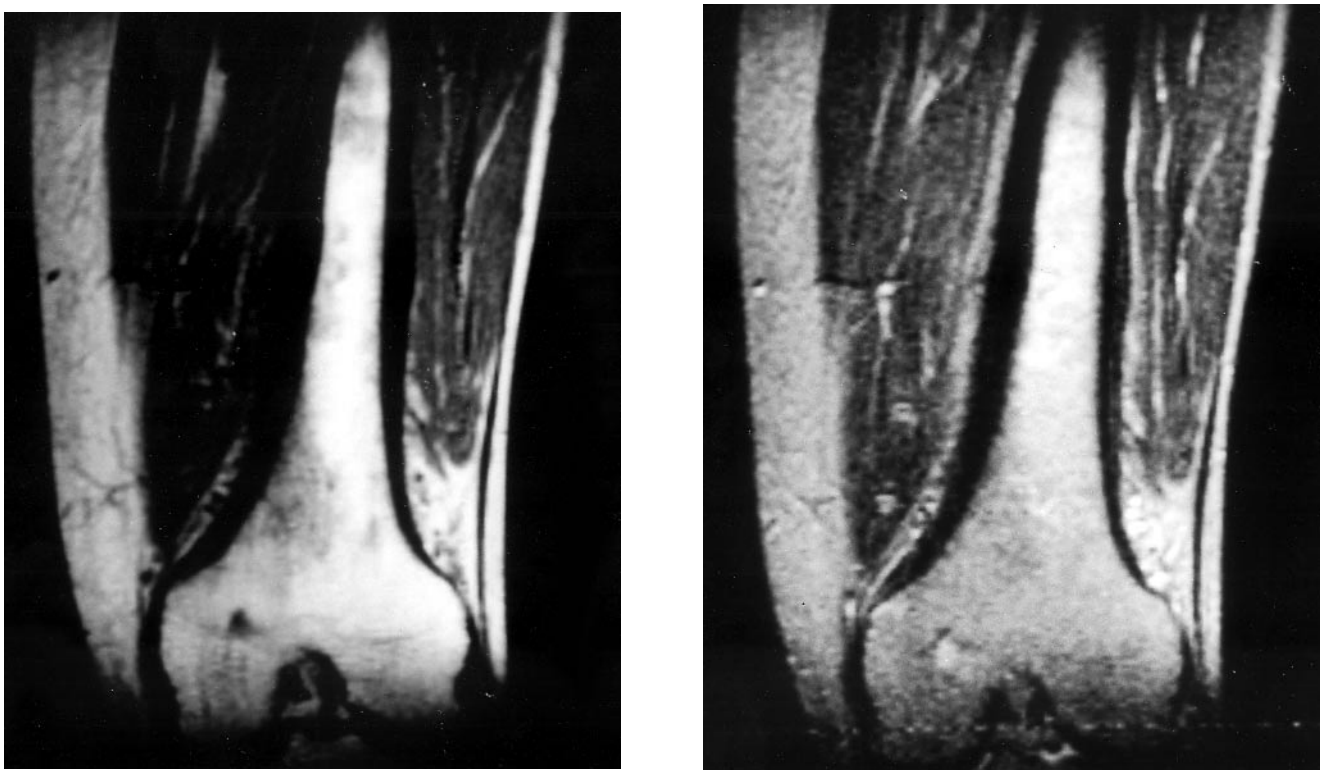

A B

Figure 3. A. A T1-weighted spin-echo MR image (TR/TE, 500/20) of the left femur shows the thickened cortex with hypointensity and intramedullary areas with a mixture of patchy hypointensity and hyperintensity. B. A T2-weighted image (TR/TE, 2000/70) shows areas with inhomogeneous hyperintensity in the bone marrow.

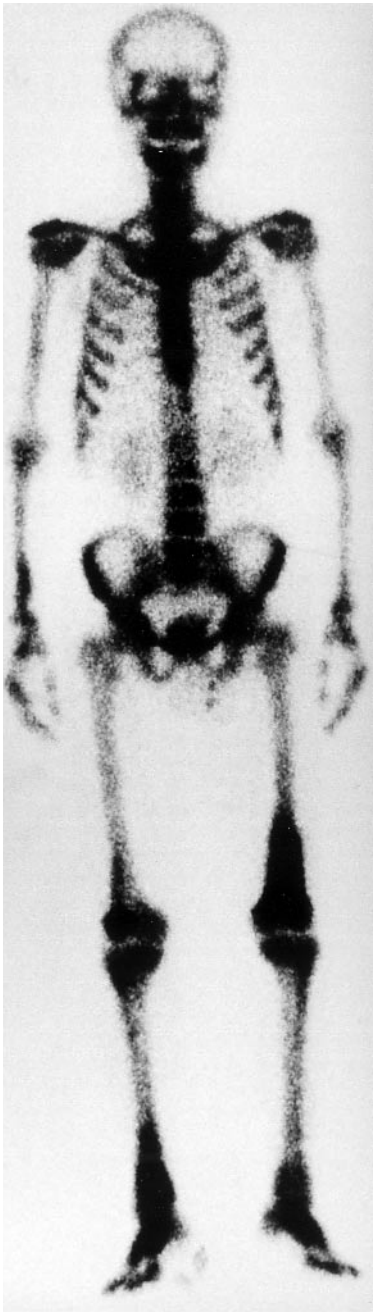

Figure 4. A ^{99m}technetium scintigraphic scan shows an increased uptake of isotope in the right distal tibia, sternum, lumbar spine, both the distal femora, ankles, knees, hips, hands, elbows, and shoulders.

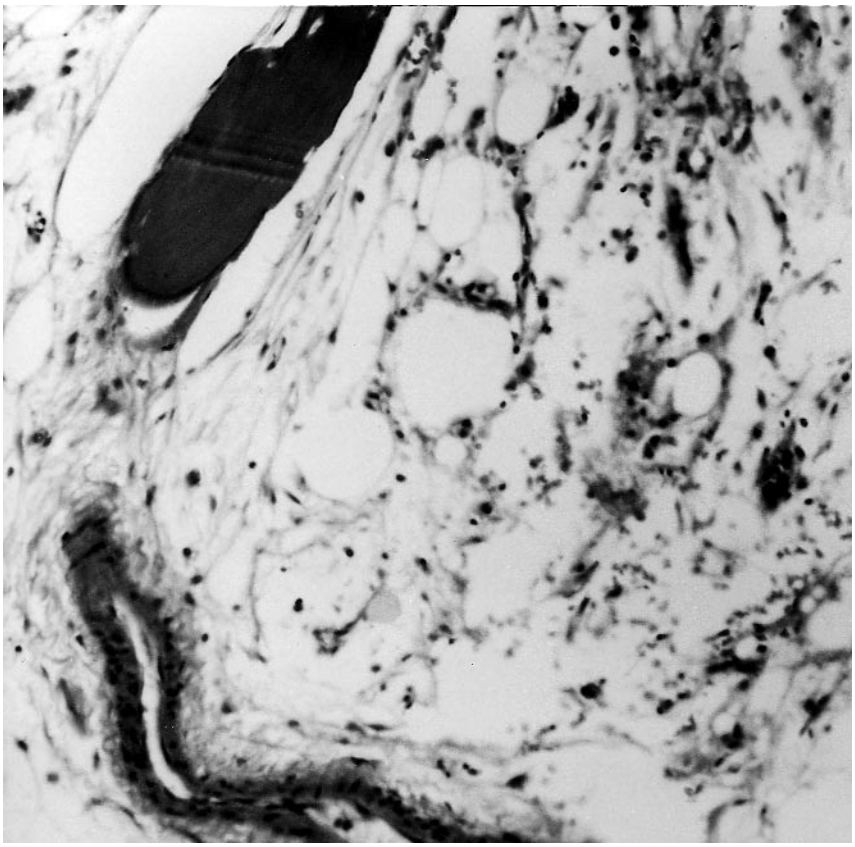

Figure 5. Histologically, the bone marrow in the femur comprised fat necrosis and inflammatory granulation tissues (hematoxylin and eosin, original magnification × 200).