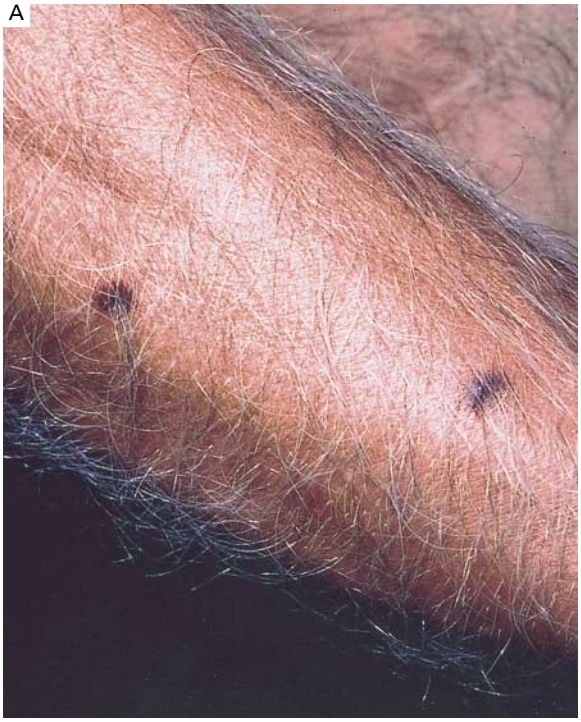
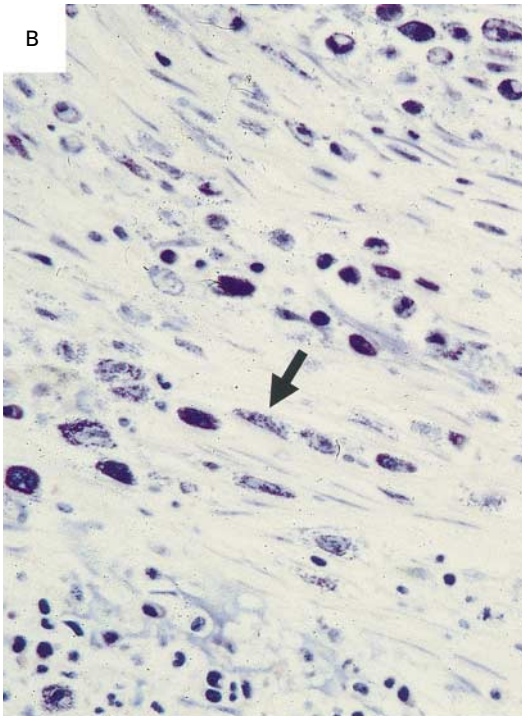
Images in Clinical Medicine

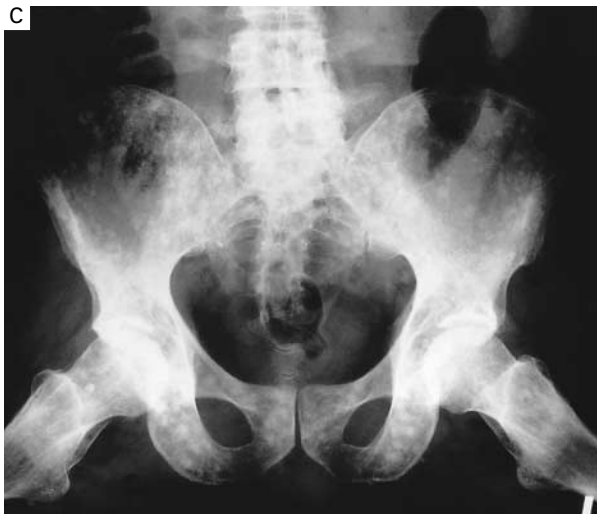
A



B



C



Systemic Mast-Cell Disease (Mastocytosis)

A 71-year-old man presented with hepatosplenomegaly and multiple macular, hyperpigmented, nonpruritic skin lesions on the trunk, legs, and arms (Panel A). The hemoglobin level was 9.1 g per deciliter, the platelet count was 112,000 per cubic millimeter, and the white-cell count was 36,200 per cubic millimeter, with monocytosis, lymphopenia, 16 percent myelocytes, and 3 percent blasts. A bone marrow biopsy revealed findings consistent with the presence of chronic myelomonocytic leukemia, fibrosis, osteosclerosis, and increased numbers of mastocytes, but not of blasts. There were numerous mastocytes, including spindle-cell forms (arrow in Panel B; Giemsa stain, ×40), and multiple extracellular mastocyte granules. The finding of increased plasma tryptase levels (141 μg per liter; normal, <13.5) and the results of a skin biopsy confirmed the diagnosis of systemic mastocytosis. Multiple osteoblastic lesions were visible on a radiograph of the pelvis (Panel C). Treatment with interferon alfa (3 million to 5 million U three times a week subcutaneously), prednisone (0.5 to 1 mg per kilogram of body weight per day orally), and epoetin (10,000 U once or twice per week subcutaneously) caused a decrease in tryptase values (80 μg per liter), a reduction in transfusion needs, and normalization of the leukocyte count, with stable thrombocytopenia (platelet count, 70,000 to 100,000 per cubic millimeter). Seven months later, the patient died of acute myeloid leukemia.

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