CLINICAL SNAPSHOT

12 Vertebral-Body Fractures and Simultaneous Skin Changes

A 57-year-old woman had developed a pruritic, brownish-red, maculopapular rash on her legs at age 40 (positive Darier's sign) that persisted thereafter, with variable intensity. A physician had suspected urticaria pigmentosa at the time, but no treatment was recommended. At age 51, she had sustained a femoral neck fracture and four vertebral body fractures, without any antecedent trauma and without any known risk factors. Osteoporosis was diagnosed and treated with strontium ranelate. At age 57, the patient had fractures of all vertebral bodies from T6 to L5 and a threefold elevation of the serum tryptase level. Laboratory findings, skin and bone marrow biopsy fulfilled the criteria for systemic mastocytosis (major: multifocal mast-cell infiltrates, minor: fusiform mast cells, expression of CD25 and CD2, KIT mutation, serum tryptase level). The patient was initially treated with zoledronate 4 mg every six months. At one year, no further compression fractures were seen.

Systemic mastocytosis can cause cytokine-mediated stimulation of osteoclasts, leading to osteoporosis. Osteoporosis of unclear origin (in a young patient without any known risk factors) should be thoroughly evaluated, and systemic mastocytosis should be considered in the differential diagnosis.

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The authors state that they have no conflict of interest.

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a) Lateral lumbar spine x-ray: there are osteoporotic fractures of all vertebral bodies from T10 to L5 (superior endplate impression fractures, concave fractures of both endplates, wedge fractures). Bone cement has been introduced into T11, T12, and L1 by vertebroplasty.

b) Red-brownish maculopapular efflorescences along the thighs. Skin changes were augmented by rubbing (positive Darier's sign). These findings fit urticaria pigmentosa. The scars were related to a fracture of the femoral neck 6 years earlier.