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The Clinical Picture

The shrinking woman



FIGURE 1. Severe kyphoscoliosis and barrel chest deformity on lateral view of chest.

A 45-YEAR-OLD WOMAN who has been undergoing hemodialysis for 20 years presents with diffuse bone pain, pruritic skin, muscle weakness, and disability. About 8 years ago, she was diagnosed with uremic hyperparathyroidism and underwent two parathyroidectomy procedures and eight sessions of percutaneous alcohol ablation of the parathyroid gland.

On physical examination, she appears to have shrunk by 20 cm over the past 8 years, measured as the vertical height from head to foot, or by 13 cm of total body length when the curvature of the thoracic spine is taken into account. Radiography clearly shows severe deformity of the spine and limbs (**FIGURE 1** and **FIGURE 2**).

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FIGURE 2. Bowing deformity of the right arm (left) and the right leg (right).

Laboratory testing reveals a serum calcium concentration of 11.9 mg/dL (reference range 8.2–10.2), inorganic phosphorus 6.6 mg/dL (reference range 2.3–4.7), albumin 3.9 g/dL (reference range 3.5–5.0), and parathyroid hormone 1,747 pg/mL (reference range 10–69).

A technetium-99m sestamibi radionuclide scan shows bilateral parathyroid hyperplasia with no ectopic parathyroid adenomas. Although a surgeon she consulted 1 month ago declined to perform another parathyroidectomy for technical reasons, another surgeon agreed to do it at this time. Parathyroidectomy was successfully performed, after which the parathyroid hormone level decreased drastically, to 85 pg/mL.



FIGURE 3. “Rugger-jersey” spine (arrows) on lateral view of the thoracic spine.

To our surprise, her total body length increased by 6 to 7 cm after surgery, with partial straightening of the back and legs noted about 4 to 5 weeks after surgery. Furthermore, she was able to walk a short distance just a few weeks after surgery. Unfortunately, she died from sepsis the next year.

■ SEVERE UREMIC HYPERPARATHYROIDISM: A CLINICAL DILEMMA

The clinical appearance of reduced body length and diffuse bony deformity leading to “shrinking” as a consequence of prolonged severe uremic hyperparathyroidism has only rarely been reported.¹ However, it is not uncommon for surgeons to decide against parathyroidectomy because of concerns of extensive subcutaneous fibrosis and recurrent laryngeal nerve damage associated with previous operations. The result is that the patient’s uremic hyperparathyroidism goes untreated, increasing the risk of long-term complications, as in this patient.

■ TYPICAL RADIOGRAPHIC FEATURES

This patient had all the cardinal radiographic manifestations of uremic hyperparathyroidism—a “salt-and-pepper” appearance of the skull, which is a sign of trabecular bone re-

sorption; “rugger-jersey” spine, or widespread osteosclerosis below the end plates of each vertebral body (thus giving the appearance of the stripes on a rugby jersey) (FIGURE 3); and twisted upper and lower limbs with scattered phalangeal subperiosteal erosions due to exaggerated bone resorption and vascular calcification. Although the gold standard for diagnosing uremic osteodystrophy remains bone biopsy, these characteristic radiographic manifestations of extensive bone resorption with simultaneous biomarkers referring to high bone turnover are usually diagnostic. In fact, they may predict the duration and severity of the disease.^{2,3}

Why the body length increases after parathyroidectomy is not yet known, but plausible mechanisms are the effects of a recovery of muscle strength and the vigorous bony remineralization that strengthens weight-bearing bones after resolution of uremic hyperparathyroidism.

■ THE DANGERS OF DELAYED TREATMENT

Delaying parathyroidectomy may induce prolonged and severe uremic hyperparathyroidism, as in this patient. Nevertheless, despite the delay, surgery was able to partially ameliorate the symptoms of hyperparathyroidism and improve the extreme bone deformity. However, the patient’s informed consent, a detailed preoperative evaluation, and exclusion of ectopic parathyroid adenomas are imperative before surgical treatment. ■

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