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Visual Vignette

Osteitis Fibrosa Cystica With Complete Bony Destruction

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Case Presentation

A 60-year-old woman with a history of type 2 diabetes and end-stage renal disease on hemodialysis presented to the emergency room with a swollen right thumb and new-onset severe back pain. The patient denied any decrease in sensation of her lower extremities, bowel incontinence, or bladder incontinence. Physical examination showed an edematous right thumb without erythema and thoracic spine tenderness to palpation with limited thoracic spine flexion and extension due to pain. The patient had not missed any dialysis sessions. Laboratory studies showed a corrected calcium level of 8.6 mg/dL (8.5-10.5 mg/dL), parathyroid hormone level of >2500 pg/mL (12-88 pg/mL), alkaline phosphatase level of 1460 IU/L (42-121 IU/ L), and 25-hydroxyvitamin D level of 24.5 ng/mL (30.0-100.0 ng/mL). Radiography of the right hand showed complete bony destruction of the right first distal phalanx and multiple lytic lesions (Fig. 1). Magnetic resonance imaging of the spine demonstrated multilevel endplate erosions and a pathologic fracture of T12 (Fig. 2).

What is the diagnosis?

Answer

Osteitis fibrosa cystica. Osteitis fibrosa cystica is a bone disorder due to sustained primary, secondary, or tertiary hyperparathyroidism. Patients with concurrent chronic kidney disease and vitamin D deficiency are at particular risk.² It is characterized by increased osteoclastic activity leading to subperiosteal bone resorption and subsequent fibrous tissue replacement, sometimes resulting in formation of cystic spaces within the bone referred to as "brown tumors." Workup for both metabolic bone disease and malignancy was pursued. The patient underwent a right iliac bone marrow biopsy, which demonstrated medullary fibrosis with extensive bone remodeling and increased osteoblastic rimming and osteoclastic giant cells. The bone biopsy results, elevated parathyroid hormone level, lack of remarkable findings on flow cytometry, and clinical history led to the diagnosis of osteitis fibrosa cystica. Due to both the severity of the metabolic bone disease and concern that the patient had developed tertiary hyperparathyroidism, the decision was made for the patient to undergo 4-gland parathyroidectomy with autotransplantation in the forearm. The patient's dose of calcitriol was increased, and she was started on intermitted calcium gluconate infusions. Upon follow-up, her back pain is improved, thumb edema is decreased, and the 25-hydroxyvitamin D level remains within the normal range. The lesson learned from this case is that patients with chronic kidney disease are at risk for developing secondary and tertiary hyperparathyroidism, which puts them at risk for metabolic bone disease and pathologic fractures.

Disclosure

The authors have no conflicts of interest to disclose.

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Fig 1.



Fig 2.

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Statement of Patient Consent

Informed consent from the patient was obtained and permission to publish patient information without any identifiable features was granted.

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