Visual Vignette

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Case presentation: A 63-year-old male presented with generalized bone pain and moderate to severe right hip pain of 4 months' duration. His daily activities were partially restricted due to pain. There was no history of preceding trauma, fever, arthritis, hearing loss, or proximal muscle weakness. There was a history of mild anorexia with a 3-kg weight loss over the past 6 months. Examination was unremarkable except for marked tenderness over the right hip. There was no focal neurological deficit or proximal myopathy. Blood biochemistry tests revealed elevated alkaline phosphatase at 655 U/L (normal: 40-125), high calcium of 14 mg/dL (normal: 8.3-10.4), decreased phosphate of 2.3 mg/dL (normal: 2.5-4.6), 25-OH vitamin D 28 ng/mL (normal: 30-75), and markedly increased parathyroid hormone (PTH) of 1,013 pg/mL (normal: 8-50). His prostate-specific antigen level was 2 ng/mL (normal: <4). Ultrasound abdomen did not reveal renal calculi. Evaluation of bone mineral density showed osteoporosis of the spine (T Score –3.2). We performed a pelvic X-ray and technetium-medronic acid bone scan (Fig. 1 *A* and *B*), as well as a sestamibi scan (Fig. 2). **What is the diagnosis?**

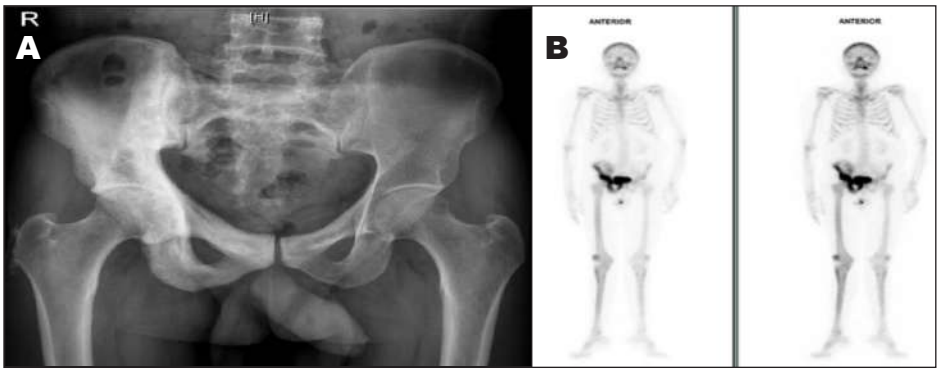

Fig. 1

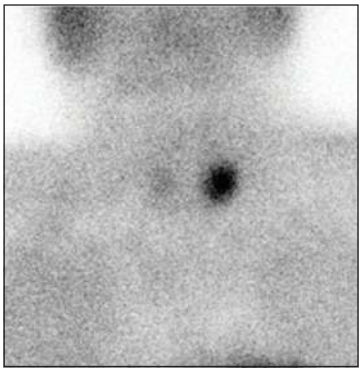

Fig. 2

Answer: Monostotic Paget disease of bone with primary hyperparathyroidism. Biochemically, he had PTH-dependent hypercalcemia along with elevated alkaline phosphatase and vitamin D insufficiency. The pelvic X-ray showed sclerotic and lytic changes in the right hip (Fig. 1 *A*), and the bone scan revealed a localized increased uptake in the right ilium, ischium, and acetabulum (Fig. 1 *B*) with decreased renal uptake. A right iliac bone biopsy showed features of Paget disease including osteoblastic rimming, giant osteoclasts, and a characteristic mosaic pattern. There was also moderate cellular marrow fibrosis. The technetium sestamibi scan (Fig. 2) showed a left inferior parathyroid adenoma. Prior to surgery, the patient was hydrated and treated with parenteral bisphosphonates. After parathyroidectomy, his calcium levels normalized and his bone pain was greatly reduced within 2 weeks.

The co-existence of Paget disease and primary hyperparathyroidism is quite rare; fewer than 100 cases have been reported in the literature. Both disorders have overlapping clinical, biochemical, and pathologic features like bone pains, elevated bone turnover markers, marrow fibrosis, and increased vascularity, making it difficult to simultaneously diagnose the 2 disorders. The present case had clinical and bone biopsy features suggestive of Paget disease, with serum chemistries and imaging diagnostic of primary hyperparathyroidism. Elevated PTH is seen in 12 to 18% of patients with Paget disease; in most of such cases, it is ascribed to secondary hyperparathyroidism caused by vitamin D deficiency, low calcium intake, and/or increased requirement of calcium during the bone formation phase of Paget disease (1).

Primary hyperparathyroidism is a systemic metabolic disease, whereas Paget disease usually involves just a few skeletal sites (2). The present case had more or less localized involvement, but the postoperative resolution of his symptoms implicates that his pagetic symptoms were aggravated by hypercalcemia. Although the occurrence of primary hyperparathyroidism in Paget disease is rare, it is prudent to periodically monitor blood calcium for early detection of primary parathyroid disease and plan timely interventions to reduce the morbidity associated with this condition.

DISCLOSURE

The authors have no multiplicity of interest to disclose.

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