Catastrophic Bone Deformities Associated With Primary Hyperparathyroidism in a Middle-Aged Man

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In March 2008, a 43-year-old male presented with generalized leg aches and gait disturbances associated with multiple pathological fractures in both upper (Figure 1, left) and lower (Figure 1, right) extremities. His medical records documented that in March 2001, he presented with leg aches to another hospital. With the suspicion of malignancy, total spine magnetic resonance imaging had been performed, and lytic lesions in cervical, thoracic, and lumbar vertebrae were detected. A total-body skeletal scintigraphy had demonstrated widespread lytic lesions. He had had pathological fractures in the right femur, and humerus, and he had been operated for those fractures. The histopathological examination of bone biopsy suggested an increase in both osteoclastic and osteoblastic activities. Thereafter, he had been diagnosed with primary hyperparathyroidism (corrected serum calcium 18.5 [8.4–10.2] mg/dL, PTH 400 [8–76] pg/mL). A right inferior parathyroid adenoma (28 × 20 mm in diameter), probably unrecognized parathyroid carcinoma, had been excised. Early after the surgery, serum calcium levels were noted as 8.4 mg/dL. The patient was lost in the interval between 2001 and 2008.

His serum calcium, creatinine, PTH, and 25-hydroxyvitamin D levels were 15.6 mg/dL, 2.6 mg/dL, 1700 pg/mL, and 24.8 μg/L, respectively. Neck ultrasound showed multinodular goiter and multiple hypoechoic lesions adjacent to thyroid. Total parathyroidectomy and thyroidectomy performed. Histopathological and immunohistochemical examinations revealed a parathyroid carcinoma in 3 different loci, with an invasion to the adjacent perineural, vascular, and lymphatic tissues (Figure 2, right sections), and benign nodular goiter. He was discharged from the hospital with serum calcium and PTH levels of 7.6 mg/dL and 430 pg/mL, respectively, and rehospitalization as soon as possible was advised to perform neck dissection.

He disappeared and lost his follow-up once again until October 2010, when he presented with malaise, fatigue, gait disturbances, and marked renal insufficiency (calcium 16.5 mg/dL, PTH 1497 pg/mL, and serum creatinine 5.5 mg/dL). Radiography of the right femur demonstrated generalized brown tumors that were huge and incorporated in distal femur giving it the form bent bone, associated with osteitis fibrosa cystica (Figure 2, upper image). Neck ultrasonography showed an irregularly shaped parathyroid lesion (17 × 14 mm, Figure 2, lower left image), with a surrounding pathological lymph node (14 × 8 mm, Figure 2, middle image). He died of myocardial infarction when he was being prepared for neck exploration surgery and while he was awaiting for bureaucratic procedures that were started to import cinacalcet from other European countries.

Some of the radiologic findings in the last admission were calcifications in Waldeyer's ring, bilateral cervical lymph nodes 18 to 20 mm in diameter (neck computed tomography), atrophic kidneys with lobulated contours, indistinguishable cortex and medulla within the kidneys, opacities (probably cristaloids) within the collecting tubes of the urinary system (urinary system ultrasonography), mitral valve calcifications, myocardial hypertrophy (echocardiography), and 95% constriction within the left anterior descending artery (coronary angiography).

Although the clinical spectrum of primary hyperparathyroidism has changed during the last decades (1), still severe bone deformities may be observed in cases with severe and long-lasting hyperparathyroidism, especially in

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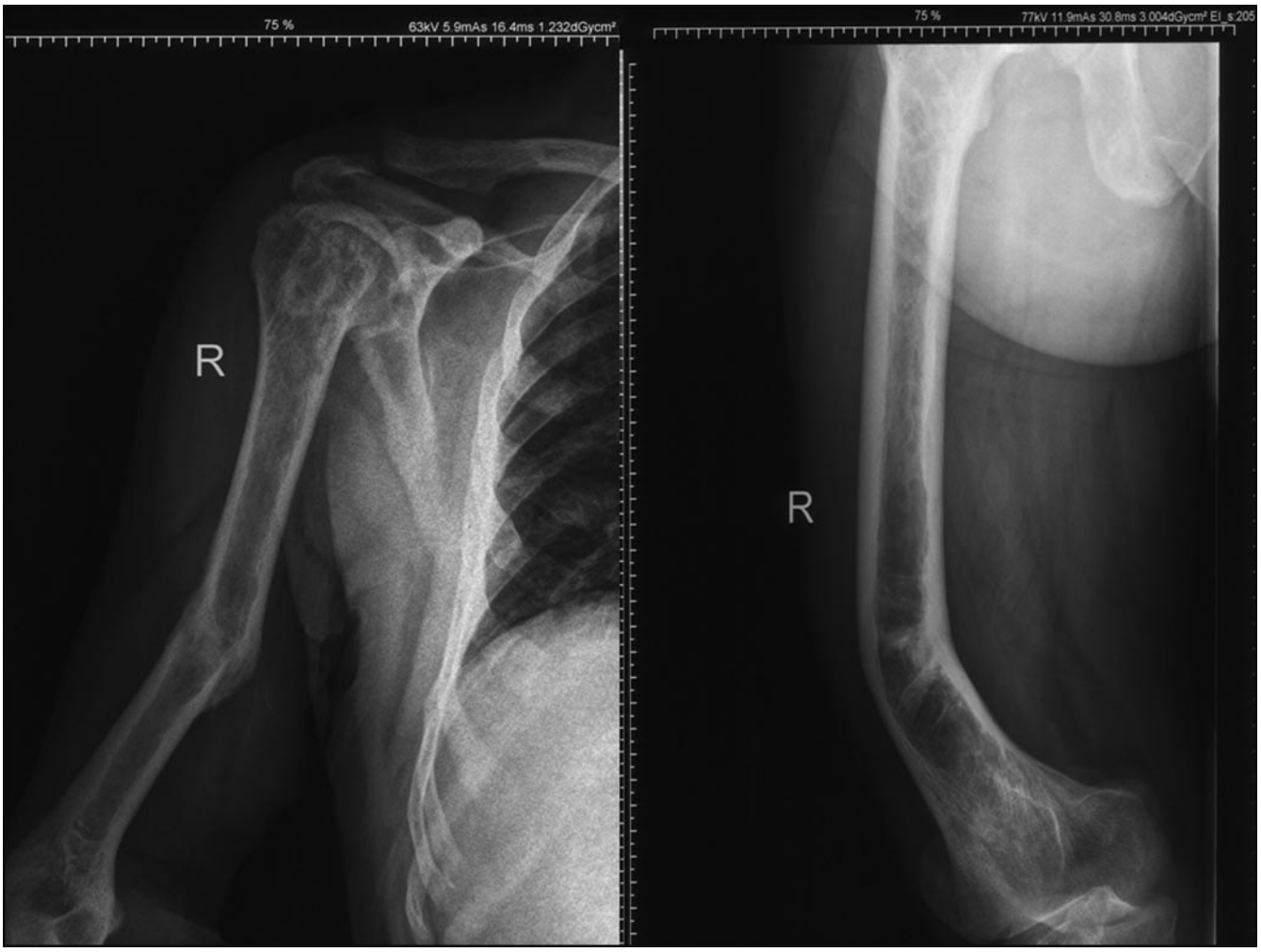


Figure 1. Multiple pathological fractures that give the shape of bent bone in the right midhumerus (left side) and in the right mid-distal femur (right side).

severe hypercalcemia that may be associated with parathyroid carcinoma (2–5).

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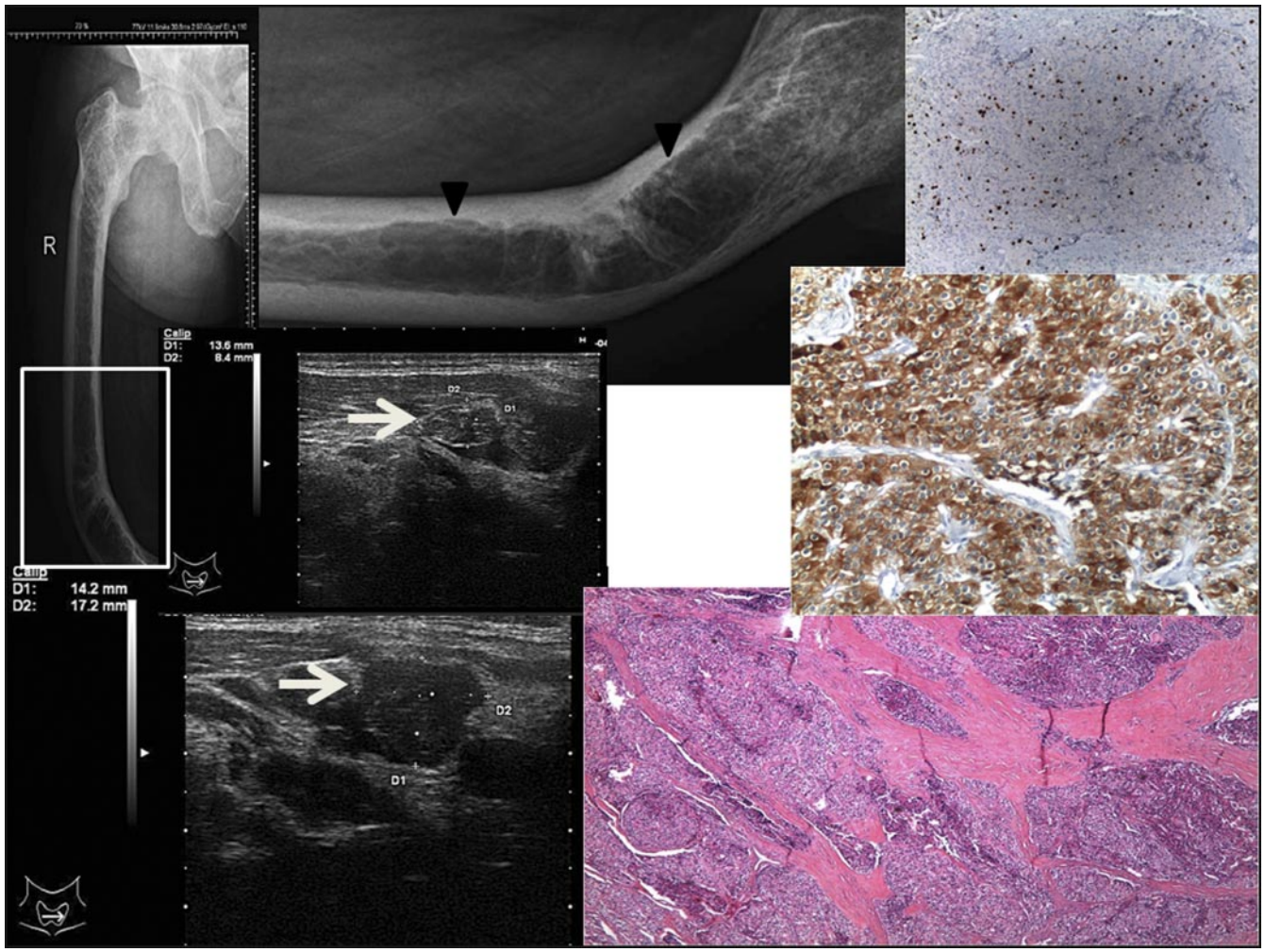


Figure 2. Left, Generalized brown tumors that are huge and incorporated in distal femur, associated with osteitis fibrosa cystica (top), caused by long-lasting severe hypercalcemia due to uncured parathyroid carcinoma (bottom), and the surrounding pathological lymph node (middle). Right, Parathyroid tumor with sharply outlined fibrous bands dividing tumor in to lobules (hematoxylin and eosin, ×40 (bottom), PTH positivity in tumor cells (streptavidin-biotin peroxidase, ×200) (middle), and high number of tumor cells with immunoreactivity for Ki-67 (streptavidin-biotin peroxidase, ×200) (top).

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