

Unusual case of phosphaturic mesenchymal tumor

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A 40-year-old patient was referred to the Department of Endocrinology, Medical Center for Postgraduate Education, Warsaw, Poland, due to prolonged and profound hypophosphatemia, causing pain, cramps, and weakness of the proximal muscles. The patient was treated with 1 µg calcitriol, 1 µg alphadiol, 1.0 g calcium, and 1500 mg/d phosphorus. The regimen had no effect on serum phosphorus concentrations

and only a moderate effect on the clinical signs and symptoms.

Calcium and parathormone levels were normal, alkaline phosphatase slightly elevated (137 U/l; normal range, 40–129 U/l), and serum phosphorus very low (0.41 mmol/l; range, 0.81–1.45 mmol/l). A 24-hour urine collection showed high phosphorus excretion (66.5 mmol/24 h; range, 12.00–65.00 mmol/24 h).

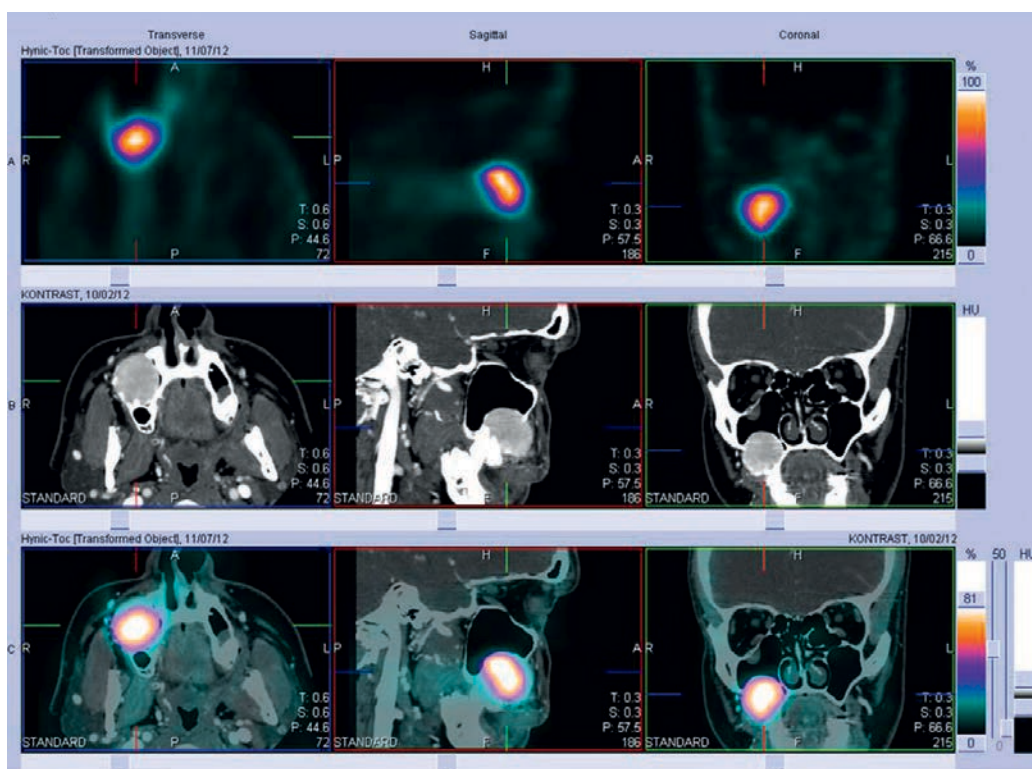


FIGURE Increased octreotide uptake in the right maxillary sinus shown on computed tomography (CT) scans and fusion of CT and somatostatin receptor scintigraphy scans

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We observed high levels of phosphaturic agent, fibroblast growth factor-23 (FGF-23; 260.4 RU/ml; range, 5–105 RU/ml). Therefore, tumor-induced hypophosphatemia was diagnosed and we started to search for the FGF-producing tumor.

Computed tomography (CT) scans did not show any abnormalities in the chest or abdomen. Abdominal magnetic resonance (MR) images were normal. Somatostatin receptor scintigraphy (SRS) showed increased octreotide uptake in the right maxillary sinus. On CT scans, an ovale hypodense tumor of 3 cm in diameter was found, and the CT/SRS scans confirmed labeled octreotide uptake in the tumor (FIGURE). Cytological examination of fine needle biopsy specimen suggested glomangiopericytoma, and the patient was referred to the Department of Cranio-Maxillofacial Surgery.

To increase phosphorus levels before surgery, we used intravenous phosphorus and (based on octreotide uptake in the tumor) somatostatin analogue. Serum phosphorus levels increased from 0.5 to 0.68 nmol/l and 0.75 nmol/l and reached the normal values within 10 days from tumor resection. Histology confirmed the diagnosis of glomangiopericytoma.

Tumor-induced osteomalacia is a rare condition associated with hypophosphatemia, myopathy, and systemic bone demineralization caused by renal phosphate wasting in the conditions of excessive FGF-23 production by neoplastic, most often benign, lesions. The tumors are usually very small and may develop in various sites of the body; therefore, their identification may be difficult.¹ In a recently described series of 39 patients with tumor-induced osteomalacia, Jiang et al.² reported the majority of lesions to be located in the lower extremities (56%) and in the head (31%).² Due to varied locations of FGF-producing tumors, CT and MR should be preceded by functional imaging – SRS or positron emission tomography.³ In our case, SRS not only showed the site of the lesion but also served as the basis for our decision to administer somatostatin analogue before surgery, which effectively increased the phosphorus level.

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