Clinical Images in Nephrology and Dialysis

Case Answer

A 44-year-old woman with ESKD treated with hemodialysis for >10 years presented with facial swelling, bone pain, and drifting of teeth leading to dysphagia and dysarthria (Figure 1). Past medical history includes chronic schizophrenia complicated by episodes of acute psychoses. Over the years, the patient had difficulty adhering to her treatment regimen. She regularly skipped hemodialysis sessions and was noncompliant with dietary and medical therapy (including phosphate binders), resulting in long-lasting uncontrolled hyperparathyroidism. Laboratory findings confirmed an extremely elevated parathyroid hormone (2150 ng/L, reference range 85–295 ng/L), a normal serum calcium (2.25 mmol/L, reference range 2.10–2.55 mmol/L), and an elevated phosphorus level (1.63 mmol/L, reference range 0.80–1.50 mmol/L). Alkaline phosphatase was extremely elevated (1820 U/L, reference range <125 U/L), consistent with a high-bone turnover state. Maxillofacial computed tomography scan showed hypertrophy of the maxilla and mandible with serpiginous soft tissue “tunneling” within the bone and loss of corticomedullary differentiation (Figure 2).

Leontiasis ossea or lion face, a rare type of osteodystrophy, is a descriptive term observed in a variety of bone diseases, including Paget disease, fibrous dysplasia, hyperparathyroidism, and condensing osteopathies (1,2). It is important to recognize because it may result in life-threatening upper airway obstruction and compressive cranial neuropathy. In recent decades, leontiasis ossea is rarely seen in CKD, most probably as a consequence of major advances in the treatment of secondary hyperparathyroidism (SHPT). SHPT describes a complex alteration in bone and mineral metabolism that commonly occurs in patients with CKD (3). Pathophysiology involves an overproduction of parathyroid hormone caused by multiple, inter-related disturbances as a result of decreased kidney function, including phosphate retention, hypocalcemia, vitamin D deficiency, and low levels of fibroblast growth factor 23 (3).

Bone disease represents only a small concern in light of the evidence that correlates SHPT with cardiovascular disease and an increased risk of morbidity and mortality (3). Treatment of leontiasis ossea usually entails parathyroidectomy along with appropriate replacement (calcium/vitamin D) or binding (phosphate) pharmacotherapy with close monitoring of serum calcium to avoid hungry bone syndrome (1,2). The presented patient highlights the importance of early diagnosis and treatment of SHPT. Perhaps the...
most difficult challenge is that of patient acceptance and adherence.

**Teaching Points**
- Leontiasis ossea is a rare craniofacial complication of renal osteodystrophy with distinctive clinical and radiographic findings
- Early diagnosis and treatment of secondary hyperparathyroidism is key in prevention

**Author Contributions**
- S. Dhont and L. Viaene conceptualized the study; L. Viaene and P. Evenepoel supervised the study; S. Dhont and L. Viaene wrote the original draft; and S. Dhont and L. Viaene reviewed and edited the manuscript.

**Disclosures**
- L. Viaene reports Amgen personal speaker’s fee. S. Dhont and P. Evenepoel have nothing to disclose.

**Funding**
- None.

**References**