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Case Report

Uremic leontiasis ossea, a rare presentation of severe renal osteodystrophy secondary to hyperparathyroidism

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ABSTRACT

Renal osteodystrophy is a common complication of end-stage renal failure patients. It's most severe osseous complication is characterized by massive thickening of the cranial vault and facial bones, called uremic leontiasis ossea (ULO), with only few cases reported in the literature. A case of a 47-year-old female patient with ULO is presented. Physical examination showed enlargement of the jaws, which hinders proper ventilation and feeding. The computed tomography examination showed marked osseous proliferation in the jaws causing severe bony expansion and loss of normal bony architecture in the skull and the skull base. The most relevant clinical, histopathological and laboratory findings are discussed. The uremic leontiasis ossea causes significant aesthetic and functional changes. Correct diagnosis and management of the factors responsible for the development of bone lesions due to altered bone metabolism are key factors. The maxillofacial surgeon must have the proper knowledge of patient's medical condition and bone maturation status to address an adequate surgical strategy.

1. Introduction

Chronic renal failure (CRF) is a multifactorial syndrome characterised by progressive and irreversible loss of renal mass and function [1]. CRF is associated with several complications influenced by aetiology, residual renal function, response to treatment, and individual variation [2]. In patients with end-stage renal failure, a common complication is renal osteodystrophy (RO), which is a descriptive term for the skeletal complications that results from pathologic alterations in calcium, phosphate, and bone metabolism [3]. Findings of RO caused by secondary hyperparathyroidism (SH) in cranial bones are frequent and include osteomalacia, osteosclerosis, and erosion of the cortical bone, brown tumours, and resorption of the lamina dura [4].

The most severe osseous complication is characterized by massive thickening of the cranial vault and facial bones, called uremic leontiasis ossea (ULO), with only few cases reported in the literature [4,5]. The term leontiasis ossea is a descriptive term applied to such hyperostotic changes in the facial bones that can lead to bilateral expansion of the malar processes, thus reducing the nasomaxillary angle [4,5].

The progressive enlargement of the facial bones and the facial deformation can lead to encroachment upon the orbital, oral and nasal cavity with its accessory sinuses, exophthalmos, optic nerve compression and potential airway obstruction [4,5].

The differential diagnosis between leontiasis ossea and other conditions with similar clinical appearances is made by clinical and laboratory findings [4]. Management of this condition includes reduction of phosphate levels, treatment of hyperparathyroidism and surgical contouring of the enlarged facial bones [4].

In the current article, a rare case of uremic leontiasis ossea with previous history of end-stage CRF in haemodialysis who developed secondary hyperparathyroidism is reported. The most relevant clinical features and laboratory findings are discussed, highlighting the complex and interdisciplinary manage these patients must have.

2. Case presentation

A 47-year-old female with CRF on hemodialysis for 6 years is referred from the endocrinology service to Maxillofacial Surgery Department with a chief complaint of severe maxillary and mandibular enlargement.

In the clinical history, evaluation by Nephrology and Endocrinology Department recorded three years ago reported that the patient had hyperparathyroidism (PTH 2500 pg/mL, normal range 12–88 pg/mL), hyperphosphatemia (5.5 mg/dL, normal

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range 2.3–4.7 mg/dL) and was normocalcemic (8.4 mg/dL, normal range 8.6–10.2). Parathyroid ultrasound showed increased thyroid size, presenting a complex node in the right thyroid lobule, and complex cysts and calcified nodule with benign appearance in the left thyroid nodule. Bone scintigraphy showed abnormalities with craniofacial predominance, compatible with bone changes.

Patient did not attend to follow-up and the parathyroidectomy surgery was postponed. Two years later, the patient reappeared. At that time, a soft and mobile 1 centimetre size nodule was found in the right thyroid lobule during physical examination. Laboratory screening was made, showing PTH levels over normal range values (3825 pg/mL, normal range 12–88 pg/mL). Hyperthyroidism was diagnosed.

Parathyroid scintigraphy was requested and suggested a parathyroid hyperplasia and showed a nodule in the right thyroid gland with radioisotope hyper-uptake. A total thyroidectomy and 3½ parathyroidectomy was performed, plus auto transplantation of half of the left superior parathyroid gland.

When the patient was admitted at the maxillofacial service, she complaint about significant enlargement of both, maxillary and mandibular bones, with subsequent dyspnea, malocclusion and dysarthria. The physical examination revealed maxillary and mandibular bone tumors, loss of nasal commissure, tooth mobility, and a tumor of the hard palate that compromised proper swallowing and adequate ventilation (Figs. 1 and 2).

The craniofacial computer tomography (CT) showed extensive bone involvement that compromised the frontal bone, skull base, craniofacial bones, and specially the jaws, the zygomatic and the nasal bones. In the jaws, the most affected portion was the hard palate, characterized by bone expansion with thinning and loss of cortical, with an alternated pattern of osteolysis and osteosclerosis, resembling a tabby appearance. Osteolysis of the right mandibular condyle was also present (Figs. 3 and 4).

An incisional biopsy of palatal bone tissue was performed, showing multiple immature bone trabeculae with multinucleated giant cells and augmented vascularity. Histopathological diagnosis is consistent with a hyperparathyroidism brown tumor (Fig. 5).

Despite the parathyroidectomy, PTH levels remained high (1907 pg/mL, normal range 12–88 pg/mL). Subsequently a scintigraphy was performed revealing an ectopic gland located at the posterior portion of sternum-manubrium.

Four months later, a new parathyroidectomy was performed to remove the residual parathyroid glandular tissue. PTH values 5 days after surgery decreased to normal ranges (63.8 pg/mL, normal range 12–88 pg/mL). After successful surgical treatment, administration of calcium, vitamin D, folic acid and iron supplementation is indicated.

During the follow-up, the maxillary bone continues to change. A second incisional biopsy would be required to assess the bone maturation and schedule the facial bones remodelling.

3. Discussion

When the glomerular filtration rate decreases below 25% of normal, phosphate excretion is impaired [3]. Hyperphosphatemia leads to hypocalcemia, because phosphate renal retention decreases renal synthesis of calcitriol (1,25-dihydroxyvitamin D3), the active form of vitamin D3 [3]. This inadequate activation of vitamin D leads to a decreased intestinal absorption of calcium [2].

Parathyroid hormone (PTH) is produced and secreted by the parathyroid glands, whose activities are controlled by free (ionized) serum calcium levels [6]. Hyperphosphatemia and hypocalcemia increased parathyroid activity (secondary hyperparathyroidism), with subsequent hypercalcemia [5]. Hypocalcemia and hyperphosphatemia mark the beginning of the biochemical sequence that culminates in renal bone disease [3,5].

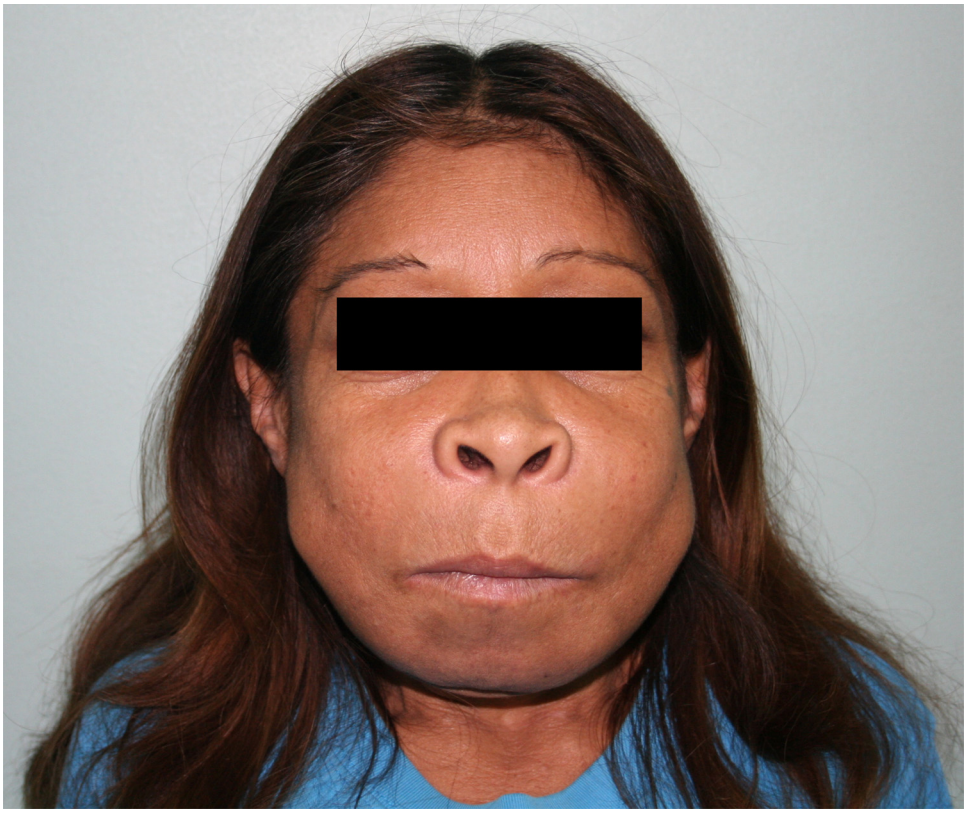

Fig. 1. Lion-like expression caused by maxillary and mandible deformation.

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