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Uremic Leontiasis Ossea Secondary to Hyperparathyroidism in Three Patients with End Stage Renal Disease

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Abstract **Original Research Article**

Renal osteodystrophy is a common complication referring to the skeletal changes associated with the phosphocalcic disorders of chronic uremia in patients with end-stage kidney disease. Uremic leontiasis ossea (ULO) is a rare and severe form of renal osteodystrophy with characteristic overgrowth of the cranial vault and facial bones. Computed tomography (CT) findings are particularly characteristic and include serpiginous tunneling within the maxillofacial bones and cortical bone resorption. The recognition of its radiological appearance and abrupt management are essential to avoid its devastating esthetic and functional impairments. Today's better medical facilities have improved hemodialysis methods, and meticulous patient follow-up has decreased the incidence of leontiasis. In this paper, we report three rare cases of patients with end stage renal failure who presented with multiple face swellings and were diagnosed with ULO.

Keywords: Renal osteodystrophy, chronic uremia, Uremic leontiasis ossea (ULO), serpiginous tunneling.

INTRODUCTION

Leontiasis ossea also described in literature as lion face, refers to a number of conditions that affect a patient's facial and cranial bone structures. ULO is a severe and rare form of leontiasis ossea, and is directly related to the pathological alterations in calcium, phosphate and bone metabolism seen in patients with chronic kidney disease [1]. It typically affects the maxilla and mandible in a symmetrical manner which helps distinguish it from other forms of renal osteodystrophy [2]. The bone proliferation progressively leads to impairment in breathing and food intake, in addition to the evident esthetic impact.

Cranio-facial CT is the modality of choice in investigating ULO. The typical imaging features include bone swelling with tunneling pattern, along with diffuse cortical resorption and "salt and pepper" appearance of craniofacial bones [3].

During the last decade this uremic complication is less frequently reported in the literature. This is probably due to better dialysis and better medical control of secondary hyperparathyroidism [4].

We report three cases of patients who developed leontiasis ossea following a long-standing hyperparathyroidism and a poor adherence to dietary and medical treatment.

CASE REPORTS

Patient 1:

37-year-old female, with no family history of renal failure and related bone disease, who was diagnosed with end stage renal disease of undetermined etiology and has been on hemodialysis for over 10 years was referred to the radiology department for progressive painless facial swelling that progressively appeared and grew worse over the last months, with recent emergence of chewing difficulty. The patient also reported changes and enlargement in her fingers. In the clinical history, evaluation by the Nephrology Department recorded a year prior that the patient had hyperparathyroidism with the following results: PTH of 3102 pg/mL, normal range 12–88 pg/mL), hyperphosphatemia (7.4 mg/dL, normal range 2.3–4.7 mg/dL) and was normocalcemic (9 mg/dL, normal range 8.6–10.2mg/dL). A thyroid ultrasound was prescribed and found enlargement of the parathyroid glands. The patient refused the parathyroidectomy prescribed then.

Physical examination on current presentation demonstrated facial deformity with bilateral maxillary and mandibular swelling.

The non-contrast craniofacial CT showed important symmetric enlargement in the maxillary and mandibular bones with serpiginous lucencies in within. A "salt and pepper" appearance of the skull was also apparent on CT along with diffuse cortical resorption (Figure 1).

The patient agreed to the parathyroidectomy and the three month follow up revealed a normalization of the parathyroid panel and a stable appearance of the skeletal changes.

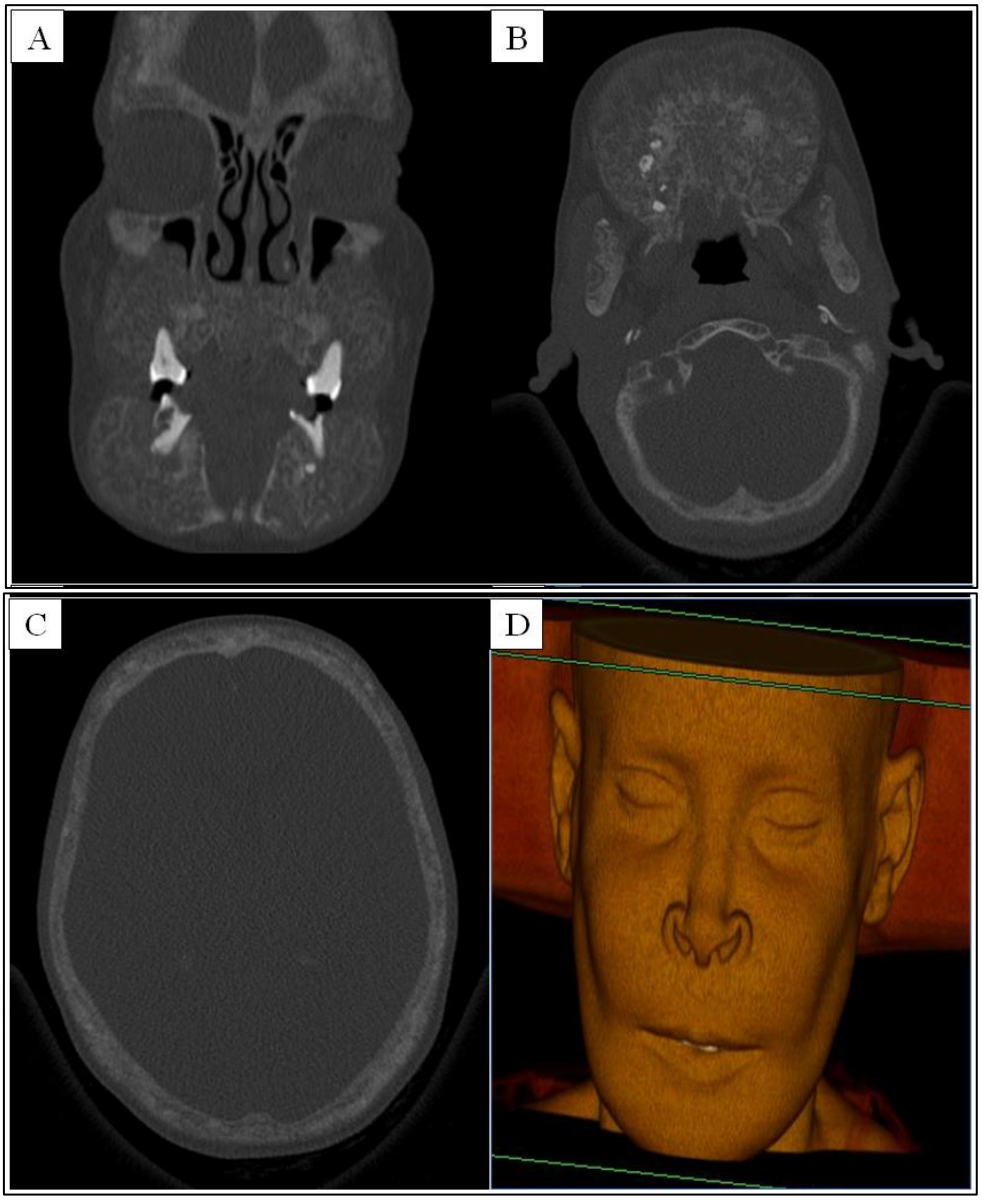

Figure 1: CT images of patient 1:

Coronal (A) and axial (B and C) CT images, bone reconstruction showing marked symmetric enlargement in the maxillary and mandibular bones with serpiginous lucencies in within, along with a "salt and pepper" appearance of the skull with diffuse cortical resorption. 3D soft tissue CT reconstruction (D) showing marked expansion of the maxilla and mandible with a "lion face" appearance.

Patient 2:

A 29-year-old man presented with progressive painless facial enlargement over the last five months. His past medical history included chronic renal failure of unknown etiology for the past ten years, under hemodialysis 3 times per week. The patient denied any history of dyspnea, or dysphagia or functional impairment. Physical examination found maxillary swelling with facial deformity.

The non-contrast craniofacial CT revealed bone remodeling with hyperostosis of facial and cranial bones, with a serpiginous "tunnelling" aspect of the maxillary bone, as well as an important widespread bone demineralization and diffuse lytic lesions with a "salt and pepper" appearance of the skull. It also revealed vascular parietal calcifications (Figure 2).

A thyroid ultrasound was requested and showed enlargement of all four parathyroid glands, that appeared hypo echoic with little vascularization on color flow Doppler (Figure 3). Laboratory investigations confirmed the underlying pathophysiologic mechanism in this case with the following results: PTH of 2072 pg/mL, normal range 12–88 pg/mL), hyperphosphatemia (5.9 mg/dL, normal range 2.3–4.7 mg/dL) and was normocalcemic (8.4 mg/dL, normal range 8.6– 10.2mg/dL).

The patient was initially put under calcimimetics and vitamin D analogs with no significant change in the laboratory results of the parathyroid panel and then underwent a parathyroidectomy. The histological findings confirmed parathyroid hyperplasia with no sign of malignancy. The patient's clinical condition remained stable and facial deterioration stopped at the two month follow up.

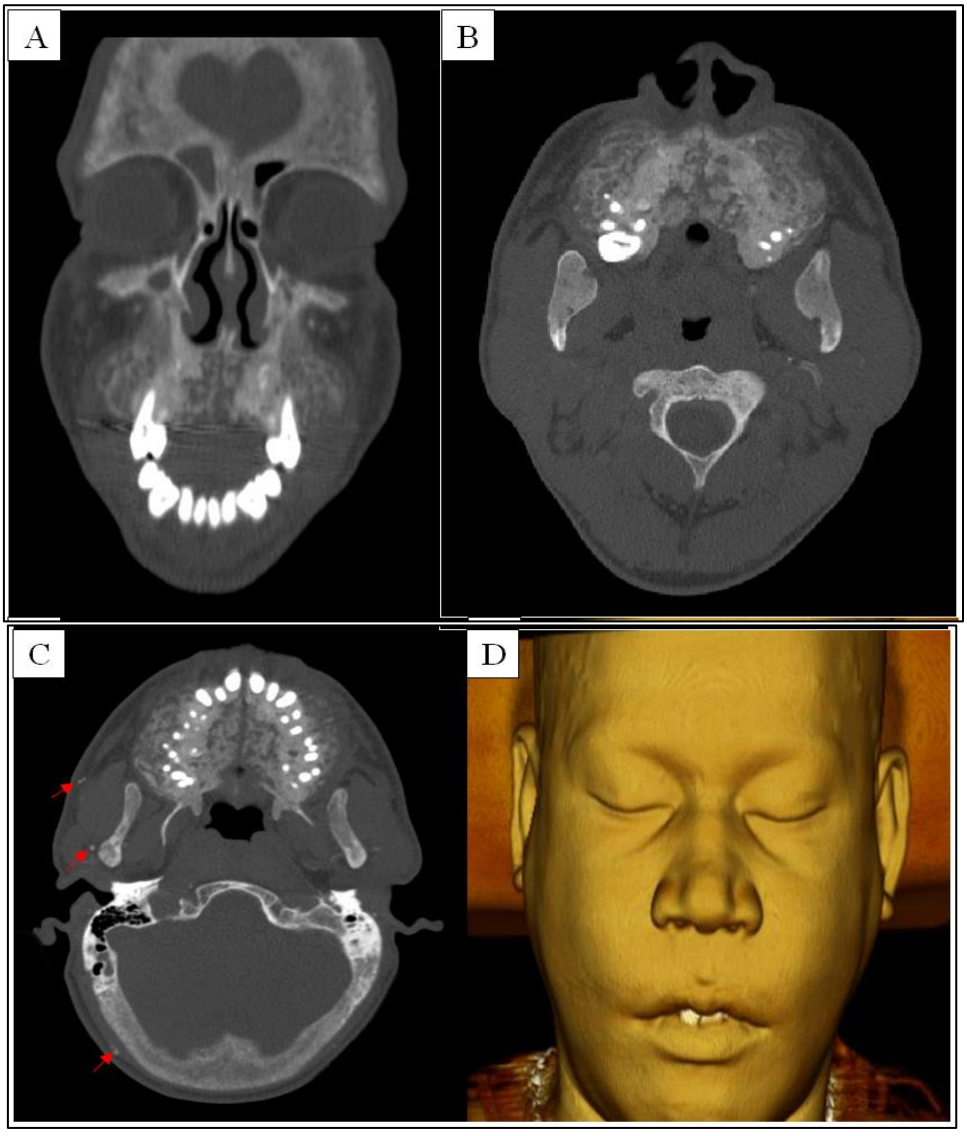

Figure 2: CT images of patient 2:

Coronal (A) and axial (B and C) CT images, bone reconstruction showing marked symmetric enlargement in the maxillary bone with typical serpiginous lucencies in within, along with a "salt and pepper" appearance of the skull with diffuse cortical resorption. Note the parietal vascular calcification (arrow). 3D soft tissue CT reconstruction (D) showing marked expansion of the maxilla with a "lion face" appearance.

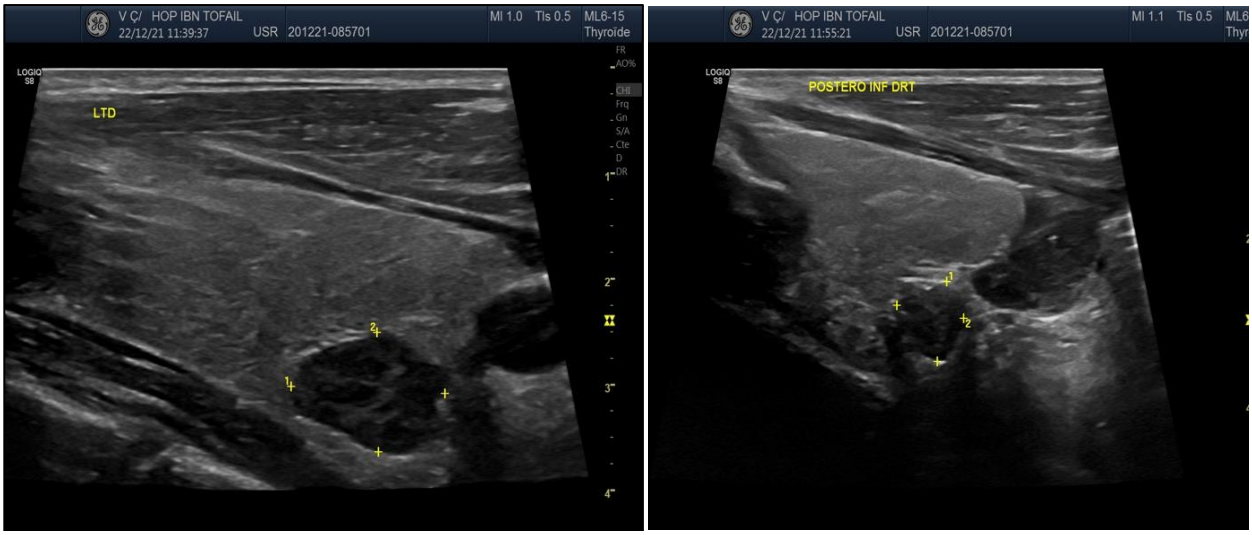

Figure 3: Ultrasound images of patient 2:

Ultrasound images of the thyroid and parathyroid showing bilateral hypoechoic heterogenous lesions in contact with the posterior side of the thyroid, separated from the latter by a thin hyperechoic line, later confirmed to be parathyroid hyperplasia.

Patient 3:

A 30-year-old man with a long history of renal failure under poor observance of hemodialysis presented with progressive painless facial enlargement over a period of three months. His past medical history included chronic renal failure of unknown etiology for the past ten years, under hemodialysis 3 times per week. The patient denied any history of dyspnea, or dysphagia or functional impairment. Physical examination found maxillary swelling with facial deformity.

The non-contrast craniofacial CT revealed bone remodeling with hyperostosis of facial and cranial bones, with a serpiginous "tunnelling" aspect of the maxillary bone, as well as an important widespread bone demineralization and diffuse lytic lesions with a "salt and pepper" appearance of the skull. It also revealed vascular parietal calcifications (Figure 2).

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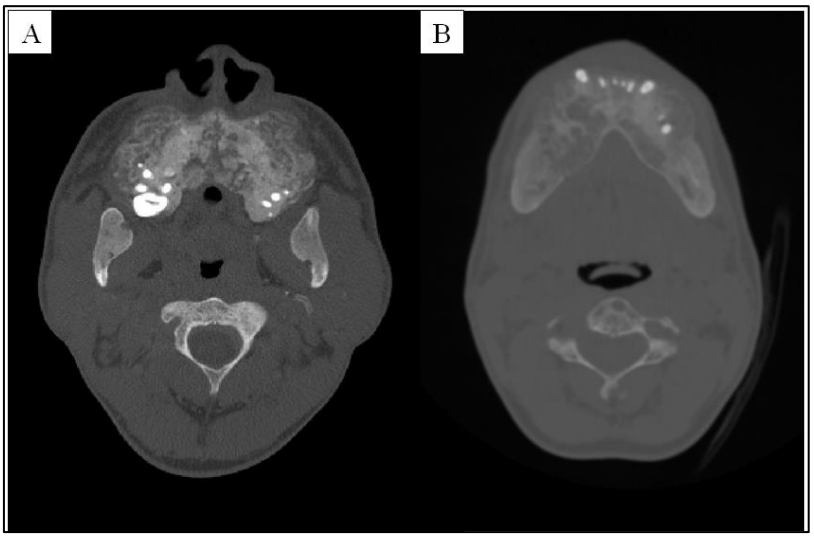

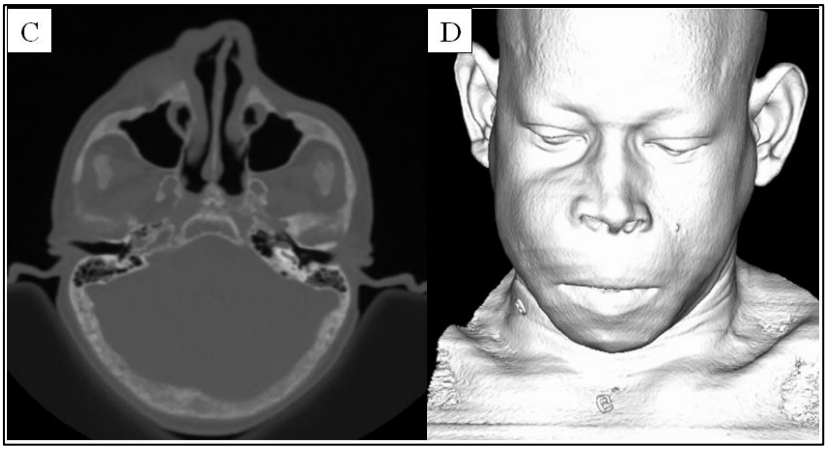

Figure 4: CT images of patient 3:

Axial (A, B and C) CT images, bone reconstruction showing marked enlargement in the maxillary and mandibular bones with serpiginous lucencies in within, along with a "salt and pepper" appearance of the skull with diffuse cortical resorption. 3D soft tissue CT reconstruction (D) showing a "lion face" appearance.

DISCUSSION

Uremic bone leontiasis is an uncommon, chronic condition caused by secondary hyperparathyroidism in patients with end-stage renal disease [5]. Poorly managed chronic renal failure results in decreased calcium resorption and phosphate excretion, which activates the parathyroid gland. In addition, decreased activation of vitamin D in the kidney leads to increased PTH secretion. Excess PTH leads to an unbalanced enhancement of osteoclast and osteoblast activity, leading to an increase in non-mineralized bone contributing to ULO; the same pathogenesis is seen in renal osteodystrophy [5, 6]. The pathophysiology is unclear, and in particular the enormous remodeling of facial bones versus the rest of the skeleton. ULO preferentially affects young people with poor access to health care and poor management of chronic renal failure [7].

Its usual clinical presentation includes progressive, painless, massive jaw enlargement, widening of the nostrils, flattening of the nasal bridge, and increased interdental spacing. If left untreated, bone remodeling can lead to progressive disfigurement, visual impairment, deafness, dysphagia, airway obstruction, and even compressive myelopathy [5].

Classic findings, on CT imaging, seen in patients with ULO are hypertrophy and hyperostosis of maxillary and mandibular bones with diffuse serpiginous lucent "tunneling" or channeling in the marrow space. This marbled appearance with alternating bands of hyperdensity and hypodensity throughout the bony matrix has been speculated to be due to necrosis and cystic degeneration representing the hypodense band and opaque osteosclerosis representing the hyperdense band [3, 6]. Thinned cortical bone with indistinct corticomedullary differentiation and possible obliteration of the maxillary sinuses were also frequently described. CT findings found in both our patients fit this description as well. On MRI, the expanded bone marrow appears as intermediate signal intensity on T1-weighted images and low-to-intermediate signal intensity on T2- weighted images [7, 8].

Bone biopsy is not particularly relevant in diagnosing ULO, as it shares similar histologic findings with other causes of craniofacial overgrowth and can often be interpreted as fibrous dysplasia [7, 9].

Early recognition of uremic bone leontiasis both clinically and radiologically, is essential to prevent the progression to disfigurement that may result from sustained and untreated secondary hyperparathyroidism. An appropriate clinico-radiological examination can provide a sure diagnosis without resorting to invasive procedures.

Therapeutic management is limited. The treatment primarily resides on the management of hyperparathyroidism by correcting the calcium and vitamin D levels and decreasing the phosphate level with either parathyroidectomy or the administration of calcitriol in conjunction with phosphate binding agents and a phosphate-controlled diet [8].

Parathyroidectomy ceases further facial deterioration although, it may be complicated by hungry bone syndrome as reported in several cases in literature. Surgical correction of facial bones is controversial and facial deterioration can stabilize or improve mildly after parathyroidectomy, which highlights the importance of early recognition to prompt management and prevent severe disfigurement. This emphasizes the importance of recognizing the distinctive phenotype of this rare and serious complication [10].

CONCLUSION

ULO has specific imaging features including extensive jaw proliferation and very typical internal serpiginous tunnels. Recognition of its radiological appearance and abrupt management are paramount to prevent its devastating esthetical and functional consequences. Biological testing during hemodialysis is essential, as abnormal parathyroid panel results often precede skeletal changes, allowing a preventive approach.

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