Scientific Research Publishing

Open Journal of Stomatology, 2023, 13, 167-173
https://www.scirp.org/journal/ojst
ISSN Online: 2160-8717
ISSN Print: 2160-8709

Leontiasis Ossea about a Case Treated at Chu Owendo

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How to cite this paper: Makungu, A.P., Atsame, G., Roukaya, M., Ogoula, S.N. and Kenguej, G. (2023) Leontiasis Ossea about a Case Treated at Chu Owendo. *Open Journal of Stomatology*, **13**, 167-173.
https://doi.org/10.4236/ojst.2023.135014

Received: April 4, 2023
Accepted: May 16, 2023
Published: May 19, 2023

Abstract

Introduction: Leontiasis ossea is the most severe bone complication of the maxillae of chronic renal failure. The objective of this work is to determine the clinical, paraclinical and therapeutic aspects of this condition. **Observation:** A 42-year-old patient with chronic anuric terminal renal failure dialysis for 12 years had consulted for multiple maxillo-mandibular swelling evolving for 2 years deforming the face with discomfort to chewing. The biological assessment was: PTH: 5252 pg/mL, Ca^{2+}: 2.86 mmol/L, free T3: 4.16 pmol/L, free T4: 6.55 pmol/L, TSH: 3.13 U/mL, serum protein: 61 g/L, Creatinine: 835 µmol/L. The maxillofacial CT noted heterogeneous diffuse thickening of the cranial vault and bones of the facial massif evoking leontiasis ossea with right mandibular brown tumor. The biopsy diagnosed fibrous gingival epulis. The patient had undergone modeling surgery, under general anesthesia after dialysis, of the right mandibular tumor whose histological result was Leontiasis ossea. **Conclusion:** Treatment is modeling surgery under hemostatic control.

Keywords

Leontiasis Ossea, Hyperparathyroidism, CKD, Bone Hypertrophy

1. Introduction

Leontiasis ossea is an enlargement of the bone tissue composing the face and is accompanied by a leonine appearance [1]. It is the most severe bone complication of the maxillae of chronic renal failure. There are two forms of Leontiasis ossea, one that touches the outer shell of the bone; it is the periosteal form with a bone with a bumpy and irregular surface; an osteal form where hyperostosis results in a regular and smooth appearance of the bony surface. Disease of un-

known etiology begins during the first or second decade of life and its evolution is very slow, so the survival time can reach several decades. The initial lesion is located on the ascending branch of the maxilla at its union with the forehead. Then the hypertrophy reaches the external face of the maxilla, the malar bone, the zygomatic arch, fills the canine fossa and increases the volume of the entire upper maxilla. From then on, the nose has lost its saddle, the nasolabial folds are filled, the vestibular region prominent: a real muzzle is thus constituted giving the facies a leonine appearance. The mandible is also thickened, creating a prognathism that is accompanied by tooth loss. The slow evolution is burdened with complications: obstruction of the cavities of the face (sinuses and nasal cavities, frontal sinuses), the ducts that deliver passage to the cranial nerves and holes in the base of the skull [2]. In patients with end-stage renal disease, a common complication is renal osteodystrophy whose findings caused by secondary hyperparathyroidism (SHPT) in cranial bones are common and include osteomalacia, osteosclerosis and erosion of the cortical bone, brown tumors and resorption of the lamina dura. The most severe bone complication is characterized by a massive thickening of the cranial vault and facial bones, called Uremic Leontiasis Ossea (ULO), with only a few cases reported in the literature [3].

The objective of this work is to determine the clinical, para-clinical and therapeutic aspects of this condition.

2. Observation

A 42 years old patient with chronic end-stage renal failure anuric who had been dialyzed for 12 years had consulted for a maxillo-mandibular tumefaction evolving for 2 years deforming the face with discomfort to the mastication (**Figure 1** and **Figure 2**). Exofacial clinical examination noted bilateral genian swelling resulting in facial asymmetry without sensory-motor disturbance. In endobucal, a hard vestibular swelling was objectified forming part of the maxillary and mandibular bones with healthy mucosa and filling the vestibule without obstruction of the upper airways (**Figure 3** and **Figure 4**).

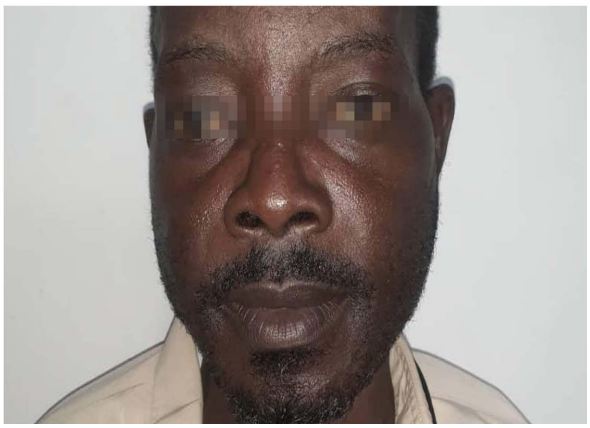

Figure 1. Asymmetry of the face with chubby appearance in 2 years of evolution.

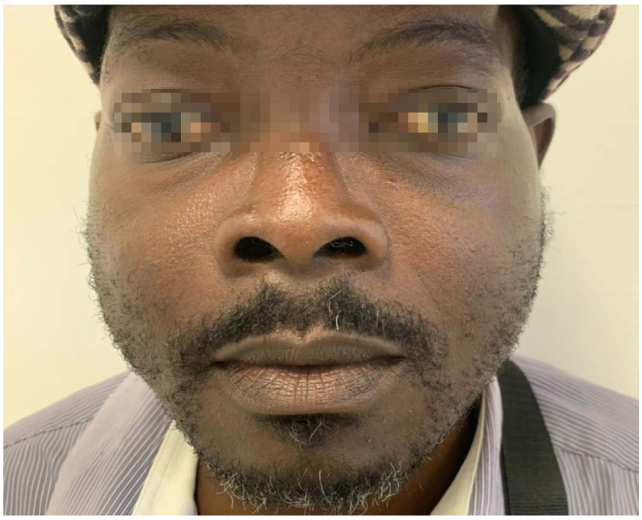

Figure 2. Asymmetry of the face with chubby appearance in 2 years of evolution.

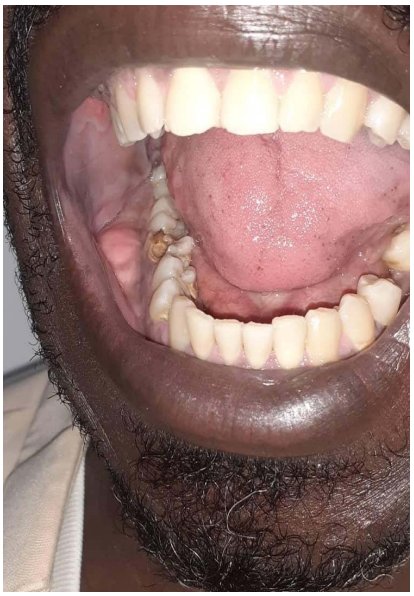

Figure 3. Mandibular vestibular swelling.

Paraclinical examination revealed on maxillofacial CT a heterogeneous diffuse thickening of the cranial vault and bones of the facial massif, a thickening of the two mandibular ascending branches with lytic lesions breaking the bony cortices. Leontiasis ossea with right mandibular brown tumor was suspected (**Figure 5**). The laboratory assessment carried out confirmed the disruption of parathyroid hormone, phosphocalcic function and renal function (**Table 1**). The analysis of the biopsy performed concluded that there was gingival fibrous epulis with bone metaplasia ranges. The indication for an intervention under general anesthesia was posed. The surgery performed was a modeling surgery with monobloc excision of the right mandibular tumor. This gesture induced a significant bleeding requiring the application of hemostatic bone wax on the site of excision

Figure 4. Maxillary vestibular swelling.

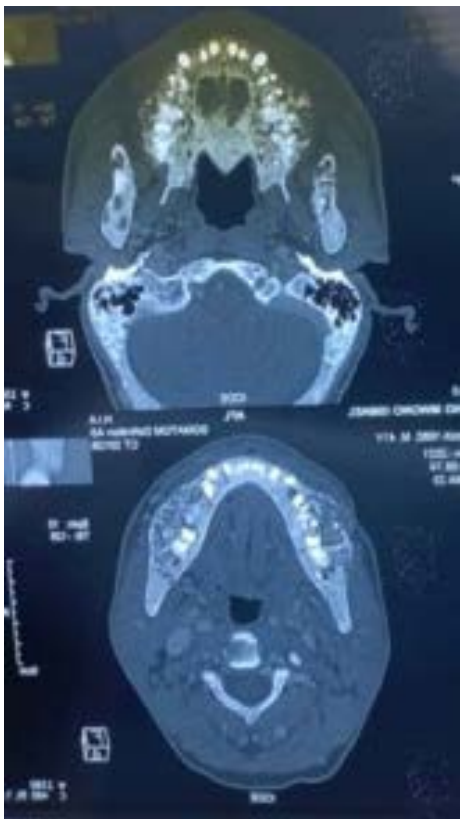

Figure 5. CT axial section, bone hypertrophy of the cranial arch of the maxilla and mandible with mandibular brown tumor.

which had made it possible to obtain hemostasis. This procedure could not be done without a per and post-operative dialysis session, and a transfusion of a bag of red blood cells per and postoperative. The post-operative follow-up was considered simple. The patient was allowed to leave on Day 15 with continued care

in the nephrology and endocrinology unit. It was reviewed on postoperative Day 60 and there was no sign of a progressive recovery of the tumor.

The histological result of the analysis of the surgical specimen showed a proliferation limited by a fibrous capsule, a neoplasm consisting of a well vascularized fibroblast proliferation with anastomosed osteoid fragments surrounded by a border of osteoblasts without malignancy. The diagnosis of leontiasis ossea was made (Figure 6 and Figure 7).

Table 1. Laboratory tests were performed to confirm diagnosis.

Reviews conducted	Value	Norms
PTH	5252 pg/mL	14 - 65 pg/mL
Calcemia	2.86 mmol/L	2.25 - 2.62 mmol/L
Free T3	4. 16 pmol/L	2.0 - 7.0 pmol/L
Free T4	6.55 pmol/L	12 - 30 pmol/L
TSH	3.13 µU/mL	0.5 - 5 µU/mL
Serum protein	61 g/L	60 - 83 g/L
Creatinine	835 µmol/L	53 - 97.2 µmol/L
Urea	22.4 mmol/L	2.1 - 8.5 mmol/L
Phosphoremia	1.63 mmol/L	1.12 - 1.45 mmol/L

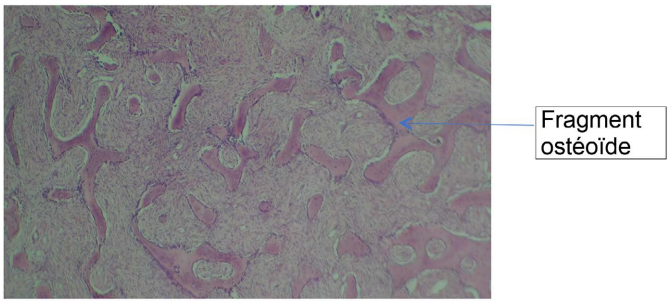

Figure 6. Fibroblastic proliferation with osteoid fragments.

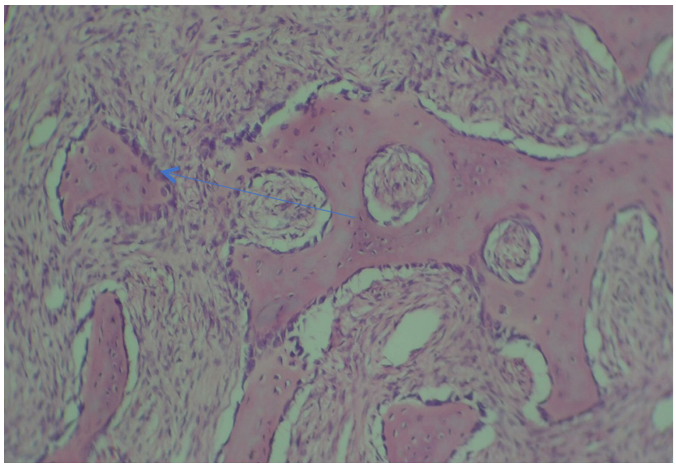

Figure 7. Border of osteoblastic cells bordering the osteoid fragments (blue arrow).

3. Discussion

This condition is manifested by the appearance of swelling of the maxilla, but is predominant in the upper maxilla. These hyperostotic changes in the facial bones can lead to a bilateral expansion of malar processes, thereby reducing the nasomaxillary angle. Progressive enlargement of facial bones and facial deformity can lead to encroachment on the orbital cavity with exophthalmos and compression of the optical nerve, oral and nasal with its accessory sinuses, and potential airway obstruction [4] [5]. The diagnosis of leontiasis ossea according to Donoso-Hofer is based on paraclinical results. Thus, the decrease in the glomerular filtration rate below 25% of normal due to renal failure leads to hyperphosphatemia responsible for hypocalcemia which will increase parathyroid activity (secondary hyperparathyroidism), followed by hypercalcemia. Hypocalcemia and hyperphosphatemia directly stimulate the production of PTH by the parathyroid glands. Excessive secretion of PTH after old secondary hyperparathyroidism leads to tertiary hyperparathyroidism, which is characterized by the development of autonomic hypersecretion of PTH and then hypercalcemia. Thus, markedly elevated parathyroid hormone levels mean secondary hyperparathyroidism and chronic renal failure [6] [7]. Computed tomography is the diagnostic and prognostic guidance tool for assessing leontiasis ossea [8]. For Yeon-Hee, this imagery can be complemented or supplemented by scintigraphy. According to Donoso-Hofer, the bone biopsy that would make the diagnosis does not seem particularly useful for distinguishing ossea leontiasis from fibrous dysplasia and Paget's disease. Only the histological analysis of the surgical specimen whose most relevant histopathological feature is the presence of immature bone, which therefore implies the presence of giant cells, makes the diagnosis. The primary treatment is parathyroidectomy according to Duan, it reduces the parathyroid mass and the number of cells and thus normalizes the serum calcium concentration, it is coupled with the medical intervention specifically targeting hypercalcemia and high PTH levels. Facial modeling surgery will be done by the maxillofacial surgeon. Thus, for Donoso-Hofer, the maxillofacial surgeon must know the state of bone maturation to decide on the appropriate surgical strategy before the onset of life-threatening upper airway obstruction and compressive cranial neuropathy.

4. Conclusion

It is a condition of the young adult with Chronic Renal End-Stage Disease. PTH, serum calcium, phosphorus, and urea are elevated. CT is essential to evoke the diagnosis. Biopsy analysis is not very contributive. It is the analysis of the surgical specimen that confirms the diagnosis. Modeling surgery in case of bone maturation is necessary.

Conflicts of Interest

Informed consent was obtained from the patient.

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