Case reports

Surgical approach of brown maxillary tumor in Sagliker syndrome

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Abordaje quirúrgico de tumor pardo de maxilar en síndrome de Sagliker

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Abstract
Case presentation. This is a 20-year-old female patient with stage 5 chronic kidney disease on hemodialysis who noticed progressive enlargement of the palatal region in the last seven months, which generated dyslalia and difficulty in chewing with expansion of the maxillary bone, separation of the upper teeth and changes in the facial configuration and profile compatible with a brown tumor. Laboratory results confirmed hyperparathyroidism and hypocalcemia secondary to renal disease. Ultrasound findings were compatible with left-sided parathyroid hyperplasia. **Treatment.** Calcium carbonate and paricalcitol were administered during the two weeks prior to surgery. The treatment consisted of subtotal parathyroidectomy and surgical resection of the tumor in the maxilla and oral calcium and vitamin D3 supplements. **Outcome.** Calcium and phosphorus levels normalized and parathyroid hormone levels decreased. After a follow-up of more than two years, no recurrences were identified.

Keywords
Chronic kidney disease, hyperparathyroidism, maxillofacial surgery, renal osteodystrophy.

Resumen
Presentación del caso. Se trata de una paciente femenina de 20 años de edad con enfermedad renal crónica estadio 5 en tratamiento de hemodiálisis que notó aumento progresivo de la región palatina en los últimos siete meses, que le generaba dislalia y dificultad para la masticación con expansión del hueso maxilar, separación de las piezas dentales superiores y cambios en la configuración y perfil facial compatibles con un tumor pardo. Los resultados de laboratorio confirmaron un hiperparatiroidismo e hipocalcemia secundarios a la enfermedad renal. Los hallazgos ecográficos fueron compatibles con hiperplasia de paratiroides del lado izquierdo. **Intervención terapéutica.** Se administró carbonato de calcio y paricalcitol durante las dos semanas previas a la cirugía. El tratamiento consistió en una paratiroidectomía subtotal y la resección quirúrgica de la tumoración en el maxilar y los suplementos de calcio y vitamina D3 por vía oral. **Evolución clínica.** Se obtuvo normalización de los niveles de calcio y de fósforo y disminuyeron los niveles de la paratohormona. Después de un seguimiento de más de dos años, no se identificaron recurrencias.

Palabras clave
Insuficiencia renal crónica, hiperparatiroidismo, cirugía maxilofacial, osteodistrofia renal.

Introduction

Sagliker syndrome is the set of craniofacial and skeletal alterations produced by the elevated level of parathyroid hormone and the alteration in calcium metabolism[1], secondary to chronic renal failure[2,3]. It consists of renal osteodystrophy[4] that manifests itself as a disfigured appearance of the face, among the most common, protrusion of the maxilla and jaw, hyperplasia of the soft tissues of the palate, loss of nasal anatomy, dental and finger deformity and short stature[1].This syndrome was described by Sagliker *et al.* in 2004[1]. An incidence of 0,5 % of patients on haemodialysis has been reported and most frequently occurs in females aged 18 to 39 years[5].

Damage to renal function results in a decrease in the levels of 1,25-dihydroxycholecalciferol, which causes hypocalcemia and hyperphosphatemia due to the decrease in calcium absorption at the intestinal level and, together, an increase in phosphorus retention[6,7]. In response to these metabolic alterations, the cells of the parathyroid glands develop a process of adaptive hyperplasia that directly stimulates the increase in the release of parathyroid hormone[2]. The state of secondary hyperparathyroidism (SHPT) in Sagliker syndrome produces changes throughout the body, especially the long, vertebral and cranial bones, and contributes to the formation of brown tumors[8]. Brown tumors are also caused by the decrease in calcium levels in the body, which increase bone resorption by stimulation of osteoclasts induced by parathyroid hormone, and produce alterations that can be described as intraosseous tissue lesions, unique or multifocal, and are usually located in the maxillary bones[9].

This case presents the management and treatment of a patient with brown maxillary tumor with Sagliker syndrome who attends the oral and maxillofacial surgery service of the Rosales National Hospital in El Salvador.

Case presentation

This is a 20-year-old female patient with a history of eight years of having been diagnosed with stage 5[10] chronic kidney disease, on hemodialysis treatment for the last four years. She consulted the oral and maxillofacial surgery service of the Rosales National Hospital to notice a progressive increase in the palatine region in the last seven months that generated dyslalia and difficulty chewing with expansion of the maxillary bone. Also, it causes separation of the upper teeth and changes in the configuration and facial profile (Figure 1).

The physical evaluation showed short stature (1,46 m), growth of the scapular bone in recent years of renal failure, abnormalities and shortening of the distal phalanges (Figure 1). Moreover, in the intraoral evaluation, a tumor was observed in the maxilla that affected the hard palate and the separation of the teeth was identified in the frontal dental region, which confirmed the alteration in the growth of the maxilla.

Laboratory test results confirmed hyperparathyroidism, hypocalcemia, hyperphosphatemia, and elevation of alkaline phosphatase (Table 1).

Figure 1. a. Intraoral clinical image of the palate. b. Frontal intraoral clinical imaging. c. Facial images with convex facial profile characteristics. d. Upward shortening and curvature of distal phalanges. e. Tendency to scapular growth in the sagittal direction

The CT scan showed hypodense images with osteolytic pattern that compromised the hard palate towards the bony part of the maxillary sinuses, the frontal bone, the sphenoid bone and a portion of the bilateral temporal bones, identifying irregularities in bone trabeculation at the aforementioned sites (Figure 2).

Ultrasound findings were described as compatible with left-sided parathyroid hyperplasia; moreover, parathyroid gammagraphy with Technetium 99-sestamibi (Tc99m) Technetium-99m-sestamibi parathyroid gammagraphy (Tc99m) combined with a monophoton emission computed tomography of the neck confirmed glandular tissue with excess cell activity in static images of the neck and mediastinum at 15 minutes and two hours (Figure 2).

Treatment

The case was analyzed with a multidisciplinary team (maxillofacial surgery, nephrology and endocrinology) for the choice of medical treatment, from the preoperative moment to the surgical management. Calcium carbonate 3600 mg daily orally and paricalcitol 15 µg were intravenously administered in three doses during the two weeks prior to surgery. The surgical treatment consisted of a subtotal parathyroidectomy without thymectomy, with the complete removal of three parathyroid glands and approximately two-thirds of the left lower parathyroid gland (Figure 3). Then, a surgical resection of the tumor in the maxilla was proceeded (Figure 3) and a sample of the palatine region was obtained, which measured approximately 4,5 × 4,3 × 1 cm. Finally, it was sent for histopathological study (Figure 3). In the immediate postoperative period, calcium gluconate 10 % was administered, one ampoule every eight hours intravenously In the immediate postoperative period and in the postoperative period, treatment with vitamin D3 0,25 µg daily and oral calcium supplement 3600 mg daily orally was initiated.

The histopathological study of the tumor of the maxillary palatine region reported the finding of moderately pleomorphic spindle cells intermingled with multinucleated giant cells of the osteoclast type, hemosiderin deposits and abundant osteoid of benign appearance of diffuse distribution, compatible with a brown maxillary tumor.

It was maintained in control and follow-up with the multidisciplinary team to monitor the evolution and continue the treatment of kidney disease.

Table 1. Comparison of laboratory results in the pre- and postoperative periods

Serology	Preoperative	Postoperative	Range
Parathyroid hormone	2870,5	650	12-88 pg/mL
Calcium	8,44	8,99	8,5-10,5 mg/dL
Phosphorus	6,74	4,40	3-5 mg/dL
Alkaline phosphatase	1807	322	30–125 UI/L

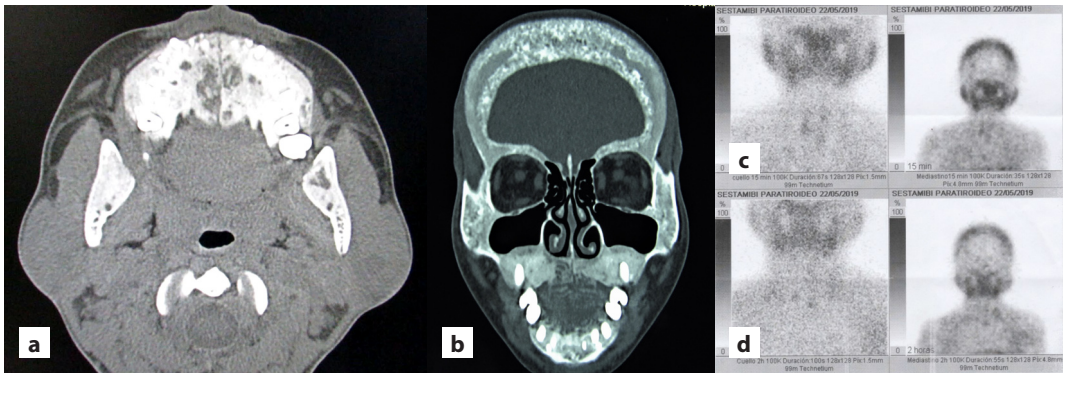

Figure 2. a. Tomographic image of axial cut with hypodense areas in palatine bones of the maxilla. b. Coronal tomographic image with salt and pepper-like lesions on the cranial bones. c. Parathyroid gammagraphy at 15 minutes at the level of the neck and mediastinum with diffuse uptake of radiotracers in the thyroid tissue. d. Parathyroid gammagraphy at two hours with capture of a small zone in the area corresponding to the lower pole of the left thyroid lobe

Outcome

The patient satisfactorily progressed in the immediate postoperative period; however, on the fifth postoperative day, she presented an area of necrosis in the mucosa of the palate in which hyaluronic acid was applied in 0,2 % bioadhesive gel every six hours and hygiene measures were included in the intervened area, which favorably developed. Moreover, control laboratory tests were performed, which reported normal levels of calcium, phosphorus and alkaline phosphatase; as well as decreased paratohormone levels (Table 1).

After two weeks post surgery, a granulation tissue with the secondary healing was presented on the palate, which favored the feeding ability and improved the quality of life of the patient. Also, the clinical evolution was monitored by controls every four months for more than two years. No recurrences were identified during this period (Figure 4).

Clinical diagnosis

A diagnosis of brown maxillary tumor was established in a patient with Sagliker syndrome since the clinical, radiographic, biochemical and histopathological findings.

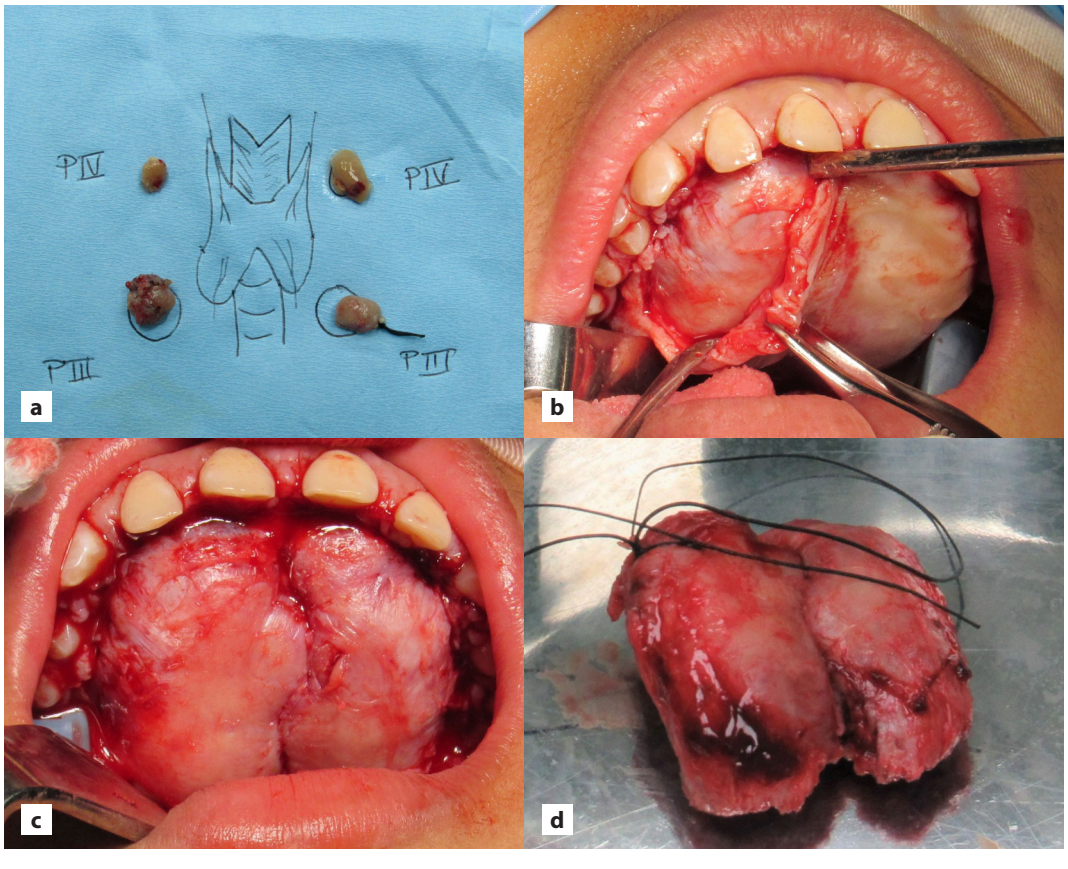

Figure 3. Transoperative imaging. a. Subtotal parathyroidectomy. b-c. Complete surgical resection of tumor. d. Sample obtained from the palatine bone of the maxilla

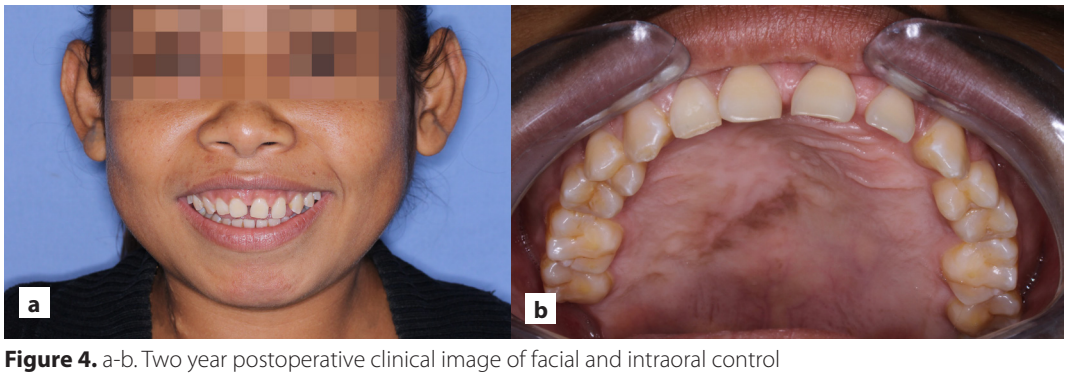

Figure 4. a-b. Two year postoperative clinical image of facial and intraoral control

Discussion

Brown tumors are intraosseous lesions that are produced by the action of osteoclasts[3]. These cells cause a replacement of bone tissue by fibrous tissue, multinucleated giant cells and hemosiderin deposits, mainly in the maxillary bones[9,11]. This is because the maxilla has a rapid rate of bone resorption and apposition. Some authors report an incidence of 1,5 % in patients with SHPT[12].

Clinical, biochemical, imaging and histopathological findings are the main tools to establish the diagnosis of brown tumors in SHPT in individuals with Sagliker syndrome[5,9]. Serum levels of calcium, phosphorus, alkaline phosphatase and parathyroid hormone in the initial assessment are of importance for proper management[13]. Likewise, imaging studies allow to identify bone alterations not only in the maxilla but also in other anatomical areas[13]. Salt-and-pepper-like lesions in the cranial bones are common to be observed in Sagliker syndrome[8].

The clinical manifestations detected in the patient are related to those described by other authors, including the characteristics of convex facial profile and malocclusions due to the protrusion of the maxillary angle in relation to the mandibular angle[14], the short stature, the growth of the scapular bone, the shortening with upward curvature of the distal phalanges, the deformity of the knees, scapulae and certain gait disturbances[1,15]. Furthermore, some authors mention other findings that are less frequent, such as moderate to severe neurological alterations, including headaches and auditory abnormalities[15].

Parathyroid gammagraphy with Technetium 99-sentamibi (Tc99m) combined with a monophotonic emission computed tomography of the neck are used as auxiliary methods of diagnostic imaging[5,9]. These allow to identify the increased cellular function of the parathyroid glands and are a guide tool for the surgeon in the surgical approach of the parathyroid glands[5].

The definitive diagnosis of brown tumor does not depend only on the histopathological result, since it presents histological characteristics similar to other giant cell tumors; therefore, this case required a combination of clinical, biochemical and imaging findings that corroborated the SHPT, and the identification of the specific signs and symptoms that determined the presence of Sagliker syndrome[12].

For many authors, the conservative therapeutic approach should encompass SHPT control and vitamin D administration to improve serum calcium levels; consequently, this improves parathyroid hormone levels and do not generate deformities in cases of brown tumors in the maxillary area and allows a gradual regression of the tumor[16-18]. Patients with very severe SHPT may not respond to high doses of vitamin D, and patients with large tumors in the maxillary or intraoral area require evaluation for parathyroidectomy to control hormonal imbalance[2,12,19,20].

The choice of treatment of brown tumors in the maxillary or oral area in SHPT depends on the location, size, generated functional alterations, the presence of facial deformations or the invasion of important anatomical structures in the maxillary or intraoral area[9,12].

Surgical treatment was decided in addition to parathyroidectomy because it invaded the nearby anatomical structures with airway involvement and swallowing with a continuous and accelerated expansion of the tumor due to the pathophysiology of the SHPT base disease with Sagliker syndrome.

Ethical aspects

The presentation of this case has the informed consent of the patient and the approval of the Research Ethics and Research Coordination Committee of the Rosales National Hospital was received.

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