Odontogenic Keratocyst in a 9-Month-Old Patient: A Case Report

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Odontogenic Keratocyst (OKC) is a benign, intraosseous, odontogenic cyst which originates from the basal cells of overlying epithelium or from the dental lamina remnants. Clinically, they are presented as asymptomatic swellings, although can sometimes be associated with pain. Growth of an OKC leads to expansion and destruction of bone as it infiltrates the tissue around it. It is commonly seen in males between the second and fourth decades of life. The aim of this study is to report on the clinicopathological characteristics of an odontogenic keratocyst in a 9-month-old female patient and posterior rehabilitation with a removable maxillary expander.

***Keywords**: Odontogenic keratocyst; odontogenic tumor; congenital; maxilla; oral cavity.*

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INTRODUCTION

Children and teenagers are less affected by the lesions of the maxillofacial complex[1]. Children have different ways of presenting with the diseases when compared to the adults. It may be explained by the different lifestyle, cultural habits, as well as physical, chemical and biological differences[2,3].

Among the cases reported in the literature, these age group can present different types of lesions, varying from benign to malignant[1,3]. Odontogenic tumours and cysts are one of the most frequently seen with the latter more common[1-3].

Odontogenic keratocyst (OKC) originates from the basal cells of overlying epithelium or from the dental lamina remnants and is classified as a benign intraosseous odontogenic cyst. This lesion demonstrates invasive and infiltrative behaviour and is commonly asymptomatic[4,5]. In addition, it shows a high recurrence rate, ranging from 0 to 62% which is commonly associated with the elected treatment[4,6].

Despite abundant research about OKC, just a little is known about the biological behaviour of OKC in children[3,6]. Therefore, the aim of this study is to carry out an analysis of the clinicopathological characteristics of the odontogenic keratocyst by reporting a new report and emphasizing the importance of posterior rehabilitation to provide the correct maxillary growth.

Case Report

A 9-month-old female patient was referred to the Oral Pathology and Surgery Centre of the João de Barros Barreto University Hospital, Belém, Pará, Brazil, for evaluation of a rapidly growing asymptomatic swelling in the left hand side of the anterior maxilla. It had reportedly increased in size in the past two months. Her parents stated that she was born with an abnormal elevation in the left alveolar ridge. The patient was delivered via Caesarean section

and no significant diagnosis was made during the antenatal period. The parents reported no significant event/condition and no trauma history of the affected site was observed.

Upon examination, there were no signs of any systemic illness or lymphadenopathy. Extra oral examination showed facial asymmetry due to the swelling, causing elevation of the left wing of the nose. The lesion was presented as a visible firm swelling on the left maxillary ridge, measuring 20 x 20 mm with a white speckled appearance. A CT scan was performed using general anesthesia and showed a hypodense, expansile mass in the left anterior alveolar ridge of the maxilla measuring 1.7 x 1.9 cm. Signs of thinning and rupture of the alveolar cortical bone was also evident (Figure 1A-B). These findings lead us towards the diagnosis of a cyst. The patient was then referred to the Oral and Maxillofacial Surgery department. Pre-operative exams were carried out and no systemic problems were noted that would prevent or complicate surgery.

Under general anesthesia, the patient was submitted to surgical excision of the lesion (Figure 2A). A creamy white suspension, characteristic of a parakeratinized squamous cell liquid, exuded upon puncture of the intracystic liquid (Figure 2B). An intrasulcular incision was then made to expose the lesion and the posterior aspect excised (Figure 2C). This was followed by enucleation of the lesion (Figure 2D). A fragment of cystic capsule, partially coated by a few layers of epithelium was analyzed microscopically. The basal layer of the epithelium consisted of columnar cells with palisade morphology and hyperchromatic nuclei. The most superficial layer of the epithelium was corrugated and consisted of keratin. The capsule was composed of loose connective tissue, ectasic blood vessels and moderate mononuclear inflammatory infiltrate. These findings lead us to the diagnosis of Odontogenic Keratocyst (Figure 2E-F). A panoramic radiograph was taken and revealed normal bone formation in the affected area after a 38-month follow-up, with no signs of recurrence (Figure 3A). The patient was referred to the Oral and Maxillofacial prosthetic and orthodontic department to undergo rehabilitation (Figure 3B). The group decided intervention was required in order to achieve successful maxillary growth. A removable palatal expansion appliance was created based on a cast model of the patient. The left maxillary incisor, lateral and canine were also replaced (Figure 3C-D). Satisfactory function and aesthetics was achieved after prosthetic/orthodontic treatment (Figure 3E). The patient is under follow-up after 67 months.

DISCUSSION

Odontogenic keratocyst (OKC) originates from the basal cells of overlying epithelium or from the dental lamina remnants[4,5]. Philipsen first described this entity in 1956[6,7]. Histologically, they have aggressive characteristics and behavior. OKC's are associated with a mutation or inactivation of suppressor genes called the protein patched homolog (PTCH) gene[5,7].

It is commonly found in male patients between the ages of 20-40. OKC more frequently affects the third molar region in both the mandibular and maxillary arches[4,6]. The characteristics of the presented case report is not frequently seen since the patient is a 9- month old female. In addition, to the best of our knowledge, it is the first report of OKC in a newborn described in literature.

The lesions uncommonly show intraoral signs, except when inflamed. However, children present jaw bones in growing process and lesions tend to be more evident[1]. In early stages, a unilocular radiolucency is seen which later may develop into a multilocular lesion. This can lead to cortical thinning, as witnessed in our case[4,6]. In addition, large lesions can also cause thinning or erosion of the cortical bone, characteristics also observed in our case report[7].

Histologically, classic OKC shows a uniform layer of stratified squamous epithelium, 6-10 cells in thickness, which is corrugated and without rete ridge formation. The basal cell layer demonstrates a uniform and picket-fence-like characteristic, with ovoid, hyperchromatic nuclei often polarized away from the basement membrane[6,8].

Figure 1. TC aspects of the tumor. (A) Axial image showing a hypodense expansile mass in the left anterior alveolar ridge of the maxilla, with aspects of thinning and rupture of the alveolar cortical bone. (B) 3D view evidencing cortical bone thinning with rupture areas.

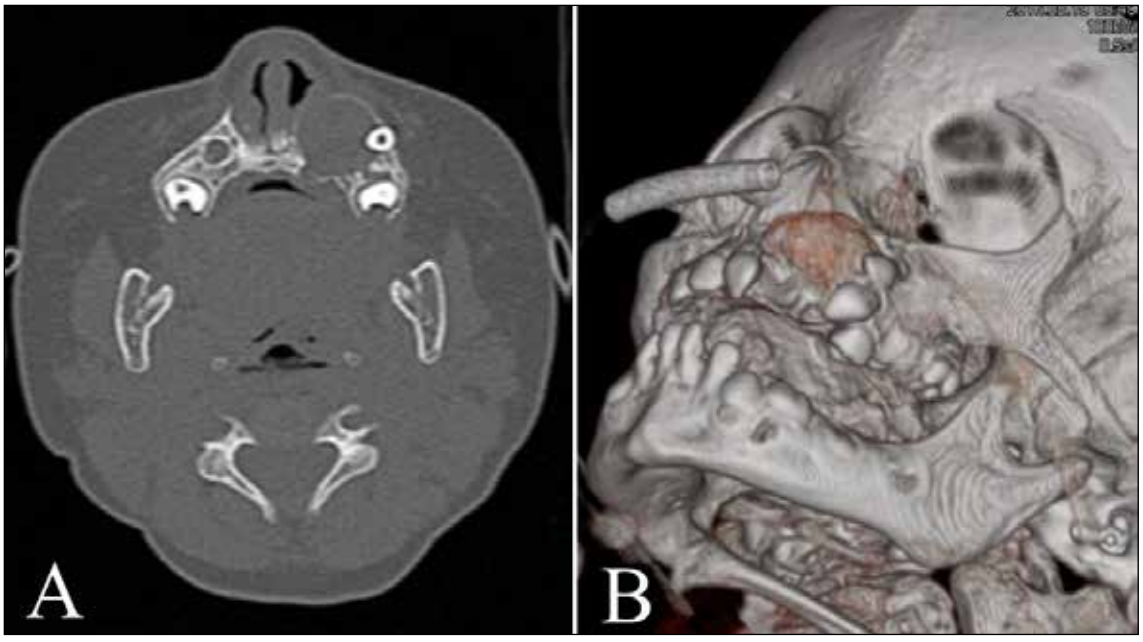


Figure 2. Clinical aspects, transurgical procedures and histological aspects. (A) Lesion showing a whitish to normal colored areas, located on the left side of the maxillary ridge. (B) Puncture of the cyst demonstrating a creamy white suspension liquid. (C) Cystic cavity after enucleation. (D) Lesion fragment measuring 20x20 mm. (E) Basal layer of the epithelium show columnar cells with palisade morphology and hyperchromatic nuclei. The capsule of the lesion is formed of loose connective tissue, with ectasic blood vessels and moderate mononuclear inflammatory infiltrate. (F) The most superficial layer of the epithelium with corrugated keratin.

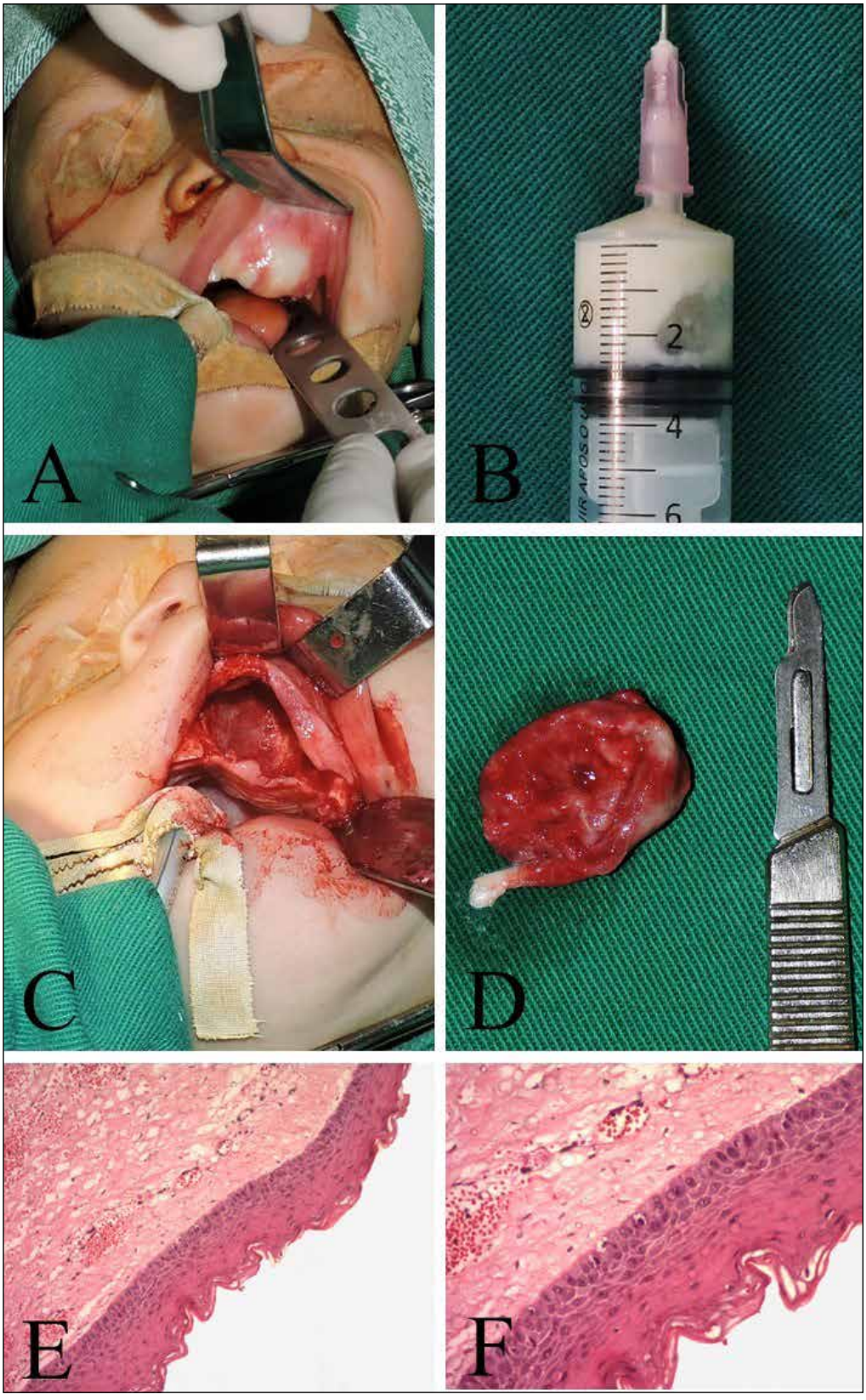
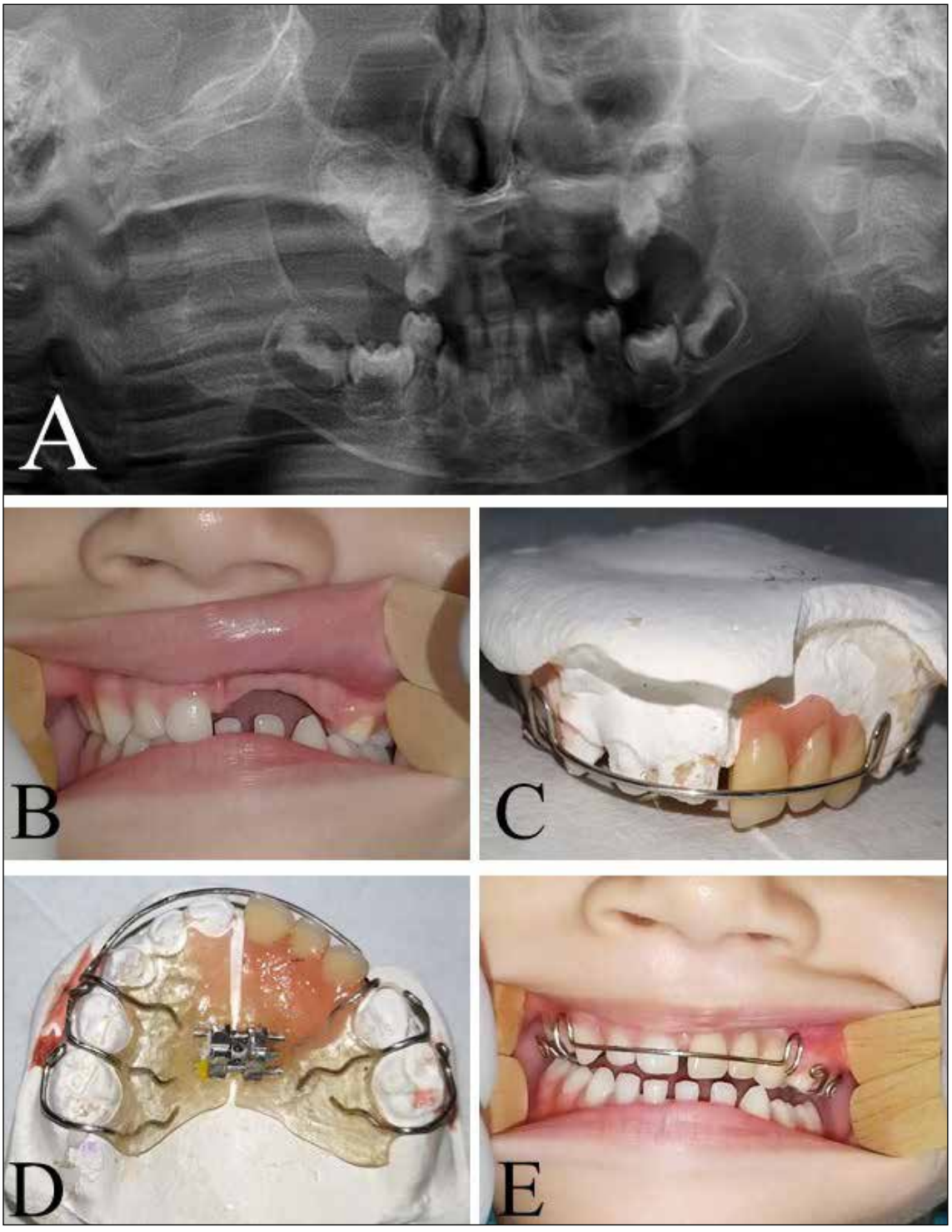


Figure 3. Patient's follow-up and rehabilitation. (A) Panoramic radiograph revealing a normal bone formation in the affected area after 38-months follow-up. (B) Clinical aspect before rehabilitation. The patient had the left maxillary central, lateral and canine missing. (C) Front view of the removable palatal expander in the plaster model before the fit stage. (D) Palatal view of the removable palatal expander. (E) Final aesthetical and functional result.



Inflammation can also be found in the connective tissues and often results in the loss of keratinisation, differing thickness of the epithelium, and loss of the uniform basal cell layer[8,9]. Immunohistochemical analysis can be a useful tool to confirm the diagnosis. Several cytokeratins (more specifically cytokeratin 10) can present a significant marker for differentiating odontogenic cysts. Moreover, several studies confirmed the significance of p53, Ki-67 and other markers used in the diagnosis[5].

OKC can have a wide variety of mutations, such as PTCH, CDKN2A, TP53, MCC, CADMI, and FHIT. PTCH is the most common and widely described in nevoid basal cell carcinoma syndrome (NBCCS) or Gorlin-Goltz syndrome, which is characterized by multiple OKCs in the jaw[6]. In addition to this, patients commonly present with basal cell carcinomas, palmar and/or plantar pits and ectopic calcifications of the falx cerebri[4-6]. In the presented case report, the child did not report any of these characteristics but is under regular follow-up with a geneticist and oral pathologist as the patient is young and can demonstrate them along time.

Treatment varies from conservative to aggressive methods[4,6]. Conservative treatments consist of marsupialization, decompression and enucleation. Aggressive approaches are based on ostectomy, resection, or use of adjunctive therapies like Carnoy's solution and liquid nitrogen[4]. Finkelstein and collaborators described that recurrence rate is decreased in aggressive treatments[9]. However, these techniques present many disadvantages, such as significant morbidity associated with the loss of the jaws continuity or facial disfigurement. In addition, when Carnoy's solution and liquid nitrogen are used as adjuvant therapy, they can cause toxicity to the adjacent tissue, leading to necrosis of the bone[10]. For these reasons, along with the fact that the case report was a child in the osseous growing period, we decided to take a conservative approach and no recurrence was observed. In addition, it is crucial for correct maxillary development to perform rehabilitation of the patients with a multidisciplinary team, including prosthetic and orthodontic professionals, aiming a better maxillary growth[3,6].

In conclusion, the diagnosis of lesions affecting younger patients is challenging due to their rarity, especially in newborn patients. After diagnosis, a multidisciplinary team approach is essential to evaluate the presence of any syndromes and the possibility of recurrence. In addition, it is necessary to monitor the patient's rehabilitation, leading to a better social and functional development.

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None.

Conflict of Interest

The authors state that they have no potential conflict of interest.

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