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Gorlin-Goltz Syndrome: A Familial Case Report

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Abstract

Gorlin-Goltz Syndrome (GGS) also known as Nevoid Basal Cell Carcinoma Syndrome is a rare autosomal dominant disorder. Is characterized for presenting some clinical features such as palmar pits, fused eyebrows, multiple odontogenic keratocyst, which are considered some of the most common signs. In our case, a rare familial case highlights awareness when a member of the family is diagnosed with this condition, and the probability to find in one the first degree relative. Once again it is confirmed that is impossible to determine without the radiographical analysis such as Cone Beam Computed Tomography (CBCT), ortophantomography, magnetic resonance imaging, to corroborate with clinical aspects, advising that, this could be the best way for the sake of patients benefit. Many treatments less aggressive have been proposed along the years, with a lower percentage of success against the traditional way, specifically surgical enucleation/marsupialization combined with peripheral osteotomy, in order to approach the variable behavior of nevoid basal cell carcinoma syndrome. The early diagnosis is considered the key to well manage this kind of disease, due to its aggressiveness, potential to become malignant, and in some cases, with a silent destructive pattern.

Keywords: Gorlin goltz syndrome, Odontogenic keratocyst, Palmar pits, Nevoid basal cell carcinoma syndrome.

Abbreviations: GGS-Gorlin-Goltz Syndrome, CBCT-Cone Beam Computed Tomography

Introduction

Gorlin-Goltz syndrome also known as Nevoid Basal Cell Carcinoma Syndrome was described for the first time in 1894 from Jarisch and White [1]. Later in 1939, Straith described a familiar case in which multiple basocellular carcinomas and cysts appeared. In 1953, Gross presented a case suggesting additional signs such as synostosis of the first left rib and bilateral bifurcation of the sixth ribs. On the other hand, Bettley and Ward were the first to relate the presence of palmar and plantar pits with this syndrome. Nevertheless, it was not until 1960 when Robert James Gorlin and William Goltz established classical triad that characterizes the diagnosis of this syndrome (multiple basocellular epitheliomas, keratocysts in the jaws and bifid ribs [2]. Later this triad was modified by Rayner et al., who established that cysts had to appear simultaneously, either with calcifications of the falx cerebri, or with palmar and plantar pits, in order to arrive at a diagnosis [3]. The porpoise is discussing one familial case of Gorlin Goltz Syndrome and describing the characteristics and features this syndrome expresses.

Case Report

A patient of 18-year-old was referred to our hospital with a chief complaint of pain in the mandible accompanied with swelling in bilateral retromolar zone and symphysis area. Patient presented sensibility and mobility in lower dental pieces. Physical examination revealed fused eyebrows, mandibular prognatism, mild hypertelorism, bony bridging of sella turca, odontogenic keratocysts, punctat dyskeratotic pits on hand and face (**Figure 1 and Figure 2**), and hyperneumanization of paranasal sinuses.

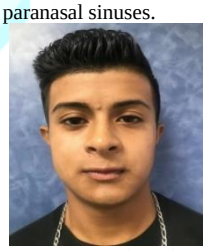

Figure 1: Frontal profile of the patient.

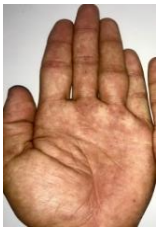

Figure 2: Palm pits.

The CBCT (**Figure 3**) and the orthopantomogram (**Figure 4**) was advised which revealed three cystic lesions in mandibular bone, one in the symphysis and one on each side of the mandibular angle, and two involving the maxillary canine region on both sides with the displaced permanent teeths. It also revealed dental agenesis of 1.8, 2.6, 2.7, 2.8, 3.8 and 4.8. The 1.3, 2.3, 3.3, and 3.7 were displaced owed to the presence of keratocysts. The odontogenic keratocyst typically shows a thin, friable wall, which if often difficult to enucleate from the bone in one piece. The cystic lumen may contain a clear liquid that is like a transudate of serum, on microscopic examination consists of keratinaceous debris.

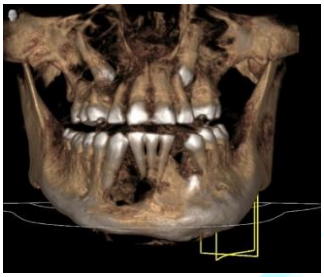

Figure 3: Cone beam computed tomography.

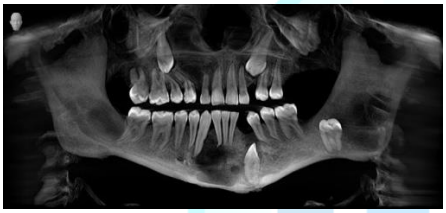

Figure 4: Orthopantomogram showing three cystic lesions in the mandible, and two involving the maxilla canine region.

Multiple odontogenic keratocysts are seen in 85% of cases, nearly all of which develop between the ages of 7 and 40 years. The peak age of development is between 12 and 25 years. Odontogenic keratocysts develop three times more frequently in the mandible than in the maxilla. The growth rate and aggressiveness of odontogenic keratocysts associated with basal cell nevus syndrome are no different than those associated with isolated odontogenic keratocysts. However, the emergence of odontogenic keratocysts from odontogenic [4].

Microscopically, the thin fibrous wall is essentially devoid of any inflammatory infiltrate. The epithelial lining is composed of a uniform layer of stratified squamous epithelium, usually six to eight cells in thickness. The epithelium and connective tissue interface is usually flat, and rete ridge formation it's inconspicuous (**Figure 5**) [5].

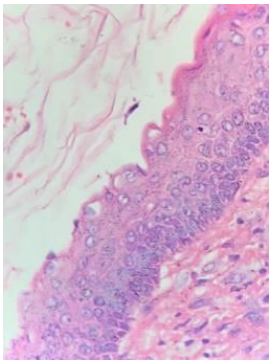

Figure 5: Odontogenic Keratocyst: The epithelial lining is from 6 to 8 thick.

The patient was asked about the other family members and revealed that his mother, sister, and brother had similar characteristic lesions. His mother was diagnosed at the age of 27, her principal complain was pain in the lower jaw, specifically in the mental zone (**Figure 6**). The orthopantomogram showed four dark radiolucent lesions compatible with odontogenic keratocyst. Biopsy was performed to confirm the disease (**Figure 7**).

Figure 6: Frontal profile of the mother.

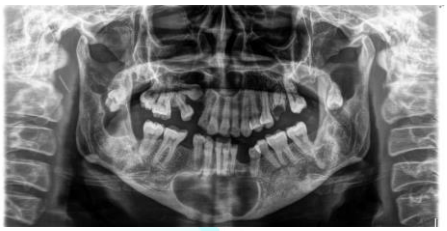

Figure 7: Ortophantomogram showing four cystic lesions, located in the intermaxillary suture zone, both mandibular ramus and symphysis.

The patients underwent surgical treatment for elimination of the jaw and maxillary cysts with general anesthetic. At first, we approached the maxillary with a periodontium flap discovering the cysts, we continued with an enucleation with peripheral osteotomy of left cyst involving the upper left canine and then with the enucleation with peripheral osteotomy of the right cyst involving the upper right canine repositioning the periodontium flap afterwards. Then we proceed with the lower jaw starting with a periodontium flap of the left cyst located in the body of the jaw, the symphysis cyst and the right angle of the jaw cyst perferming an enucleation with peripheral osteotomy finishing with the repositioning of the periodontium flap. The monetary capability of the patient and the lack of supplies of the hospital made it impossible to use any bone substitute nonetheless the result of the surgery was good (**Figure 8 to Figure 15**).

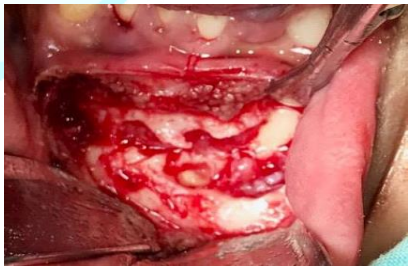

Figure 8: Odontogenic Keratocyst, symphysis area.

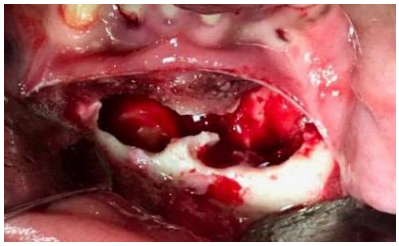

Figure 9: Removed odontogenic keratocyte in symphysis area.

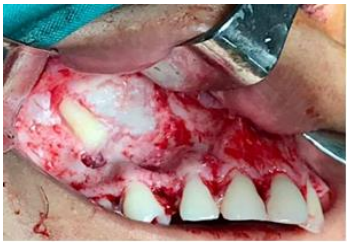

Figure 10: Odontogenic Keratocyst surrounding upper right canine.

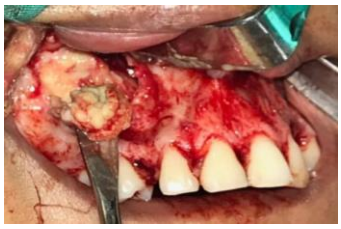

Figure 11: Exposing Odontogenic Keratocyst.

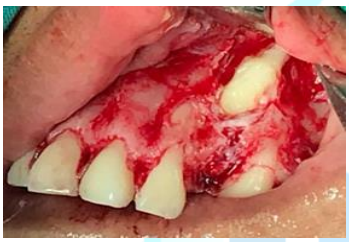

Figure 12: Odontogenic Keratocyst surrounding upper left canine.

Figure 13: Removed odontogenic keratocyst surrounding the left canine.

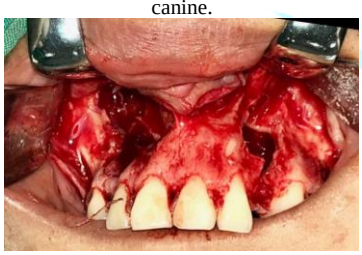

Figure 14: Removed left and right odontogenic keratocysts.

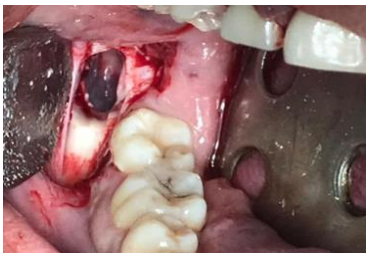

Figure 15: Retromolar zone cavity.

Patient was followed up four months, where he presented bone regeneration, stability of dental pieces, no paresthesia, and lack of swelling and better breath smell **(Figure 16)**.

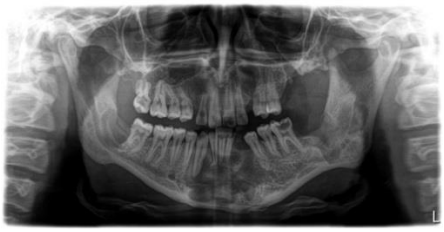

Figure 16: Four Months followed up panoramic radiography.

Do to the genetical component and aggressive behavior of Gorlin Goltz Syndrome, the decision to advise a control orthopantogram for his sister, revealed one cyst in left base of the mandible **(Figure 17)**.

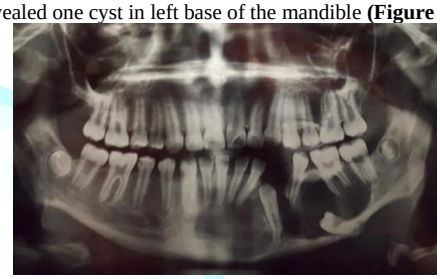

Figure 17: Sister's panoramic x-ray shows and important growing of a cystic lesion in the left base of mandible responsible for 3.3 and 3.5 dental pieces displacement.

Discussion

Odontogenic Keratocyst is considered by the WHO as a benign and multicystic, intraosseous tumor of odontogenic origin with a characteristic location of parakeratinized squamous epithelium with defined as a potentially aggressive, infiltrating behavior [6].

In differential diagnoses are to be mentioned:

- Follicular cyst
- Ameloblastoma
- Ameloblastic fibroma
- Odontogenic Myxofribroma
- Plasmocytoma (multiple myeloma)
- Aneurysmal bone cyst

To confirm the diagnosis of odontogenic keratocyst, a histological examination as a trial biopsy or examination of the entire primary removed cyst bladder [6].

The Gorlin-Goltz syndrome is an autosomal dominant inherited syndrome manifested by multiple defects involving the skin, nervous system, eyes, endocrine system, and bones. It is also known as basal cell nevus syndrome, multiple basal cell carcinoma syndrome, Gorlin syndrome, or hereditary cutaneomandibular polyonocosis, multiple nevoid basal cell epithelioma-jaw cysts, or bifid rib syndrome [7].

The incidence of this syndrome is estimated to be 1 in 50,000 to 150,000in the general population bur perceived incidence may vary by region. There is no racial/ sex predilection associated with Gorlin syndrome [8].

The tumor suppressor gene called Patched (PTCH), located in the 9q22.3 chromosome, has been identified as the cause of Gorlin-Goltz syndrome. Data suggest that a product of this gene acts as tumor suppressor and Gorlin- Goltz syndrome's typical malformative patterns suggest that the main function is to control the growth and development of normal tissues [1,9].

The diagnostic criteria for nevoid basal cell carcinoma was established by Evans et al., and modified by Kimonis et al. in 1973. According to them diagnosis of Gorlin-Goltz syndrome can be established when two major or one major and two minors are present which are described below [7].

In our patient showed three major criteria's, three odontogenic keratocyst in jaw and two in maxilla, palmar pits and a first degree relative. Minor criteria were such as fused eyebrows, prognathism, hyperpneumanization of paranasal sinuses and bony bridging of sella turca. In the mandible, 43% Odontogenic keratocysts (OKCs) occurs in the molar ramus region, followed by 18% in the incisor-canine area. In the maxilla, 14% OKCs were found to occur in the incisor-canine area, followed by molar tuberosity with 12%, 7% in the mandibular premolar region and 3% in the maxillary premolar region [11].

Treatment

There are two methods of treating odontogenic keratocysts: Conservative or aggressive. In the conservative method, simple enucleation with or without curettage and marsupialization are suggested. Aggressive methods include peripheral ostectomy, chemical curettage with Carnoy's solution and resection [3]. Cryosurgery using liquid nitrogen is indicated in the large complex mandibular lesions if there is a risk of damage to vital structures with conventional treatment methods [10]. The liquid nitrogen treatment showed very good results [1].

Conclusion

Gorlin Goltz syndrome is an autosomal dominant mode of inheritance, in this case from the mother to her children. Major and minor criteria must be present when identifying odontogenic keratocyst for diagnoses this syndrome. Radiographic and genealogical analysis is essential to establish an accurate and early diagnosis.

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