

The relevance of dental surgeon on Gorlin-Goltz syndrome

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SUMMARY

Multiple odontogenic keratocysts could be linked to different conditions. Then, to achieve the correct diagnosis whether their presence is associated to a syndrome, some criteria must be followed. The present study aims to report a case of a 21 years-old male patient whose was referred with several radiolucent lesion on the maxilla-mandibular complex. The lesions were biopsied and the diagnosis of the histological exam hypothesized as odontogenic keratocyst. After complete evaluation, others abnormalities were also found such as calcification of falx cerebri, palmar and plantar pits, and multiple basal cell on feet. The patient was diagnosed with Gorlin-Goltz syndrome and was referred to other medical specialties to adequate follow-up. Dental surgeon represents an important role on correct diagnosis of the Gorlin-Goltz syndrome and could avoid further complications.

Keywords: keratocysts, Gorlin-Goltz Syndrome, diagnosis.

INTRODUCTION

Gorlin-Goltz Syndrome (GGS), first described in 1894 by Jarish & White, is a condition characterized by the presence of odontogenic keratocysts, multiple basal cell nevi, and skeletal anomalies such as bifid ribs and calcification of falx cerebri. Palmar and plantar pits are usually associated to this syndrome (1, 2) In a large number of cases, odontogenic keratocyst represents the first sign of GGS and can be detected at the first decade of life. In addition of skeletal manifestations already mentioned, ophthalmic, neurological, endocrine, and genital anomalies can also be found in such patients (3) The early diagnosis of this syndrome by the dentist could prevent further complications (4).

CASE REPORT

A 21 years-old man referred to the Diagnosis and Surgery Department of the Dental School of Araraquara presenting facial asymmetry and malocclusion (Figure 1). The radiographic exam revealed different radiolucent lesions associated to impacted

teeth involving maxillary and mandible regions (Figure 2). Biopsy of the lesions was performed and histopathological results indicated to odontogenic keratocysts (Figure 3).

Due to the presence of multiple odontogenic keratocysts, patient evaluation was conducted to discover other abnormalities that could be leading to Gorlin-Goltz syndrome. Cervical radiograph showed spina bifida of the cervical vertebrae, CT scan revealed calcification of falx cerebri and others intracranial structures (Figure 4), and teeth agenesis and mandibular hyperplasia can be seen at the orthopantomography (Figure 2). Moreover, plantar pits and multiple basal cell nevi on feet were found and posteriorly confirmed with histological exam (Figure 5).

Under general anesthesia, the impacted teeth were removed and its associated lesions were enucleated. After collected all the clinical, radiographic, and histological findings, the final diagnosis was for Gorlin-Goltz Syndrome. Disorders involving the ocular, cardio-vascular, genito-urinary, auditory, respiratory, and gastro-enteric system were not found. However, even after two years, the patient remains under follow-up by a multidisciplinary medical team.

DISCUSSION

The multiple radiolucent lesions found in this case, posteriorly diagnosed as odontogenic kerato-

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Fig. 1. Clinical aspect: facial asymmetry and malocclusion



Fig. 2. Radiographic exam revealing radiolucent lesions associated to impacted teeth and agenesis of tooth 38. Size of keratocysts: 3.0×1.7×0.8 cm (18); 3.3×2.1×0.7 cm (28); 2.7×1.5×0.5 cm (48).

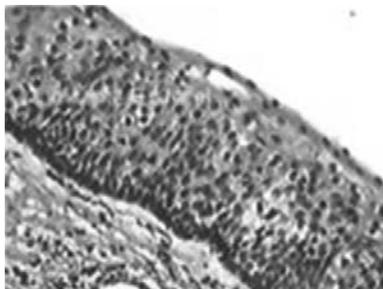


Fig. 3. Histopathological results of maxillomandibular keratocysts



Fig. 4. Cervical radiograph: spina bifida of the cervical vertebrae; CT scan: calcification of falx cerebri; 3D reconstruction.



Fig. 3. Plantar pits and multiple basal cell nevi on feet. Histological exam result: diffuse hyperpigmentation into the basal layer of the epidermis; incipient intradermal nevus.

cyst, may be the first sign and a good evidence of Gorlin-Goltz syndrome (5). However, those lesions can be present in others syndromes (6). According to MacDonalds-Jankowski (2011), the odontogenic keratocysts are commonly located at molar region of the mandible, asymptomatic, and can evolve to facial swelling (7). The patient reported in this case presented the same characteristics cited above.

As well as showing the cystic lesions and impacted teeth, the imaging exams also showed calcifi-

cation of the falx cerebri and bifid cervical process. Kimonis et al. (1997) (8) reported that 31% of patients can present vertebral anomalies while Shanley et al. (1994) (9) revealed that ectopic calcification of falx cerebri can occur in 92% (8, 9). Moreover, de Ortega et al. (2008) described the calcification of falx cerebri as one of the most common sign in syndromic patients (2).

The patient in this case was presenting palmar and plantar pits as well as multiple basal nevi. Tit-

inchi et al. (2013) (10) evaluated 15 individuals that were diagnosed as Gorlin-Goltz syndrome along 40 years in the South African population. Their results revealed 13% of patients presenting multiple basal cell nevi and a quarter showing palmar-plantar pits (10). However, other authors reported that more than 80% of patients can present palmar-plantar pits (8, 9). Although basal cell carcinoma is found in 80% of affected individuals (8), no one was found in this case. Nevertheless, the patient was referred to dermatologist to skin lesions follow-up.

The establishment to the correct diagnosis of Gorlin-Goltz syndrome two major criteria or two minor and one major criteria must be present (8). Therefore, the patient presented odontogenic keratocyst, palmar and plantar pits, and calcification of falx cerebri as major criteria. Besides that, presented also vertebral anomalies as minor criteria. Furthermore, Reyes Macias et al. (2002)

affirmed that, when the Gorlin-Goltz Syndrome is diagnosed, the recurrence rate of keratocysts is 63% while is 37% when they are not associated to the syndrome (11).

CONCLUSIONS

In conclusion, the Gorlin-Goltz syndrome is a condition that requires great attention of oral medicine and oral maxillofacial surgery professionals. Besides, involves also others medical specialists (12). All these professionals are important not only for the correct and early diagnoses but also for the adequate follow-up. As soon as the diagnosis is achieved the prognosis will be better controlled.

CONFLICT OF INTERESTS

None.

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Received: 17 01 2017
Accepted for publishing: 27 12 2017