

Removal of Multiple Keratocystic Odontogenic Tumors in a Nonsyndromic Patient

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Abstract

Multiple keratocystic odontogenic tumors are one of the key features of Gorlin–Goltz syndrome. A 15-year-old nonsyndromic female child presented with multiple keratocystic odontogenic tumors. The presence of the tumors was observed in immunological examinations. The images led to the suspicion of Gorlin–Goltz syndrome which was discarded after analyzing the patient’s medical history and complementary examinations. Le Fort I osteotomy was opted to access the maxillary tumors favoring visibility and allowing simultaneous bilateral accesses. A sagittal vestibular incision in the lower rim was performed to access the mandibular lesions. After 3 months, the patient underwent a bilateral myotomy to reduce the volume of the masseter muscles. The occurrence of nonsyndromic multiple keratocystic odontogenic tumors is rare. Clinicians facing this situation shall seek for other known features of the Gorlin–Goltz syndrome and follow up closely these patients for the possibility of occurrence of basal cell carcinoma.

Keywords: Basal cell nevus syndrome, carcinoma, Gorlin–Goltz syndrome, keratocyst, odontogenic

INTRODUCTION

Keratocystic odontogenic tumor stems from the odontogenic organs mostly localized in the mandible, particularly in the posterior body and ascending ramus.^[1]

When these lesions are multiple, the Gorlin–Goltz syndrome is usually associated.^[2] Few are the reported cases of nonsyndromic patients with multiple keratocystic odontogenic tumors.^[1,3,4]

This paper describes the case of nonsyndromic multiple keratocystic odontogenic tumors in a young woman, her treatment, and follow-up.

PROCEDURE

A 15-year-old female child attended the Mato Grosso Cancer Hospital accompanied by her mother reporting enlargement on both sides of the face and pain in the region of the upper third molars with 3 months of evolution. In the anamnesis, the patient denied any systemic diseases.

On extraoral examination, there was a mild swelling on both mandibular angles [Figure 1]. Cutaneous abnormalities were absent.

In intraoral examination, mucosa had normal color and there was not buccal and lingual cortical expansion of the mandibular bone [Figure 2].

Panoramic radiograph revealed radiolucent cystic-like lesions in the mandible body. The radiolucencies circumscribed the dental roots apex of anterior teeth and the region of the right second premolar, which had root dilaceration. Dental germs of the third molars were also present [Figure 3].

The multiple radiolucent images with approximately 3 cm of diameter and a rim with space of more than 3 mm circumscribing the crowns of the intraosseous lower third molars led to the suspecting of Gorlin–Goltz syndrome. Thus, complementary examinations were solicited.

Computed tomography, in axial cut, showed cystic-like images surrounding the dental germs of the upper third molars

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invading the maxillary sinuses, lesions with cystic aspect in mandible body, and bone fenestration [Figure 4].

Chest radiograph showed normal lungs and preserved cardiac area, with no signs of bifid ribs. Skull radiograph also showed normal aspect. Ultrasound examination of the ovaries and hematologic examinations were within normal limits.

Le Fort I osteotomy was opted as a way to access all maxillary lesions to favor the visibility and allow simultaneous bilateral accesses [Figure 5]. To access the mandibular lesions, a sagittal vestibular incision in the lower rim was performed and the lesions were then removed [Figure 6].

No reconstruction was necessary. The sites were filled with clot after curettage and cryotherapy. Healing occurred naturally.

Histopathological analysis showed cystic cavity limited by layer of regularly parakeratinized stratified squamous epithelium and fibrous connective tissue [Figure 7], leading to the diagnosis of keratocystic odontogenic tumors.

As her main complaint was the enlargement on both mandible angles, diagnosed as muscle hyperplasia, the treatment chosen was bilateral myotomy of the masseter muscles. Two incisions behind mandibular angles were realized in both sides to partly remove the masseter muscle hyperplasia. That intervention was performed 3 months after the removal of the keratocystic odontogenic tumors.

The patient is kept under clinical and radiographic follow-up for 6 years without recurrence of the tumors [Figures 8 and 9] and without manifestation of other lesions.

DISCUSSION

The nevoid basal cell carcinoma syndrome, also called Gorlin–Goltz syndrome, is a rare autosomal dominant inherited disorder and is characterized by a wide range of developmental abnormalities and predisposition to neoplasms.^[2] This syndrome was reported in 1894 by Jarisch and White, but the developmental malformation and basal cell epitheliomas were associated by Binkley and Johnson in 1951 and Howell and Caro in 1959.^[5] In 1960, Gorlin and Goltz characterized the Gorlin–Goltz syndrome with the presence the three alterations: basal cell carcinomas, keratocystic odontogenic tumors, and bifid ribs.^[5] Some other clinical characteristics include malformation in structures of the nervous system as the corpus callosum,^[6] calcification of the falx cerebri, palmar and plantar epidermal pits, spine and rib anomalies, relative macrocephaly, facial milia, frontal bossing, ocular malformation, medulloblastomas, cleft lip and/or palate, and developmental malformations.^[5]



Figure 1: Extraoral photograph showing the face with mild swelling on both mandibular angles



Figure 2: Intraoral examination showing normal color mucosa and no buccal and lingual cortical expansion



Figure 3: Panoramic radiograph revealing unilocular pericoronal radiolucencies in the lower third molars and apical area of anterior teeth

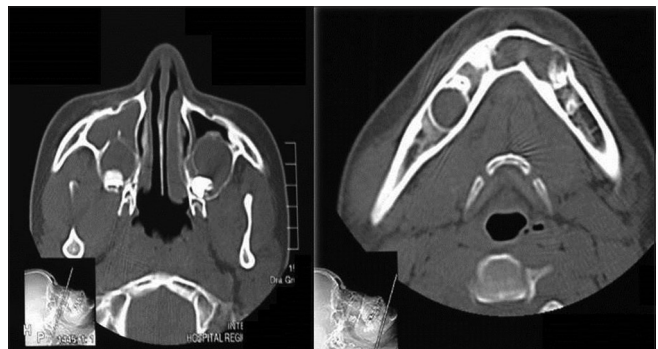


Figure 4: Computerized tomography scan showing two well-limited areas with increase of soft tissue in the maxillary sinuses on both sides

In rare cases, patients may present multiple keratocystic odontogenic tumors without being a syndromic patient; in other cases, the tumors are the primary manifestation of the Gorlin–Goltz syndrome^[3] or even a partial expression of the syndrome.^[4]

Despite the absence of basal cell carcinomas in patients with Gorlin–Goltz syndrome was reported previously,^[7,8] other cases of patients with several keratocystic odontogenic tumors were not diagnosed with the syndrome^[1,3] as in the present case. The patient did not have cutaneous lesions or skeletal anomalies to characterize Gorlin–Goltz syndrome. Nevertheless, she is being kept under close control for the possible manifestation of new keratocystic odontogenic tumors or even other late syndromic signs.

The lesions in the maxillary sinuses observed in computed tomography were accessed by means of Le Fort I osteotomy surgery as a way to decrease the removal of bone tissue and the damage to nerves and blood vessels^[9] and to promote a direct line of vision and improved exposure^[10] to the tumors.

The bilateral myotomy of masseter muscles was successful on solving the patient's main complaint.

CONCLUSION

The occurrence of nonsyndromic multiple keratocystic odontogenic tumors is rare, so clinicians facing this situation shall seek for the other known features of the Gorlin–Goltz syndrome. Even with the absence of the syndromic characteristics, it is prudent to follow-up closely these patients for the possibility of occurrence in the future of new keratocystic odontogenic tumors or even the basal cell carcinoma, also associated with the syndrome.

The approach adopted in this case for the multiple tumors removal and the reduction of masseter volume was successful and produced very few side effects.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other

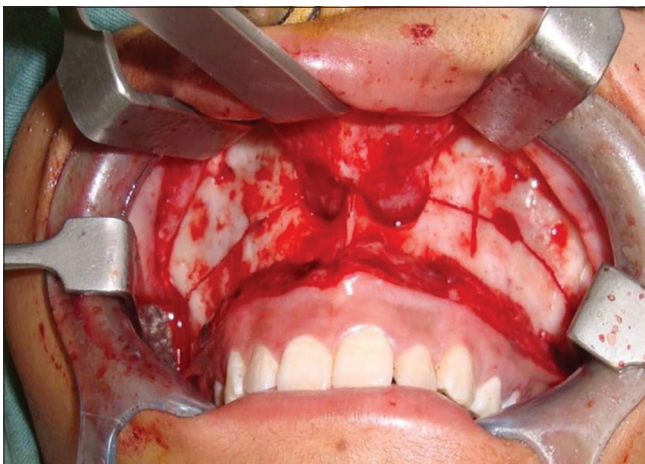


Figure 5: Intraoperative Le Fort I osteotomy

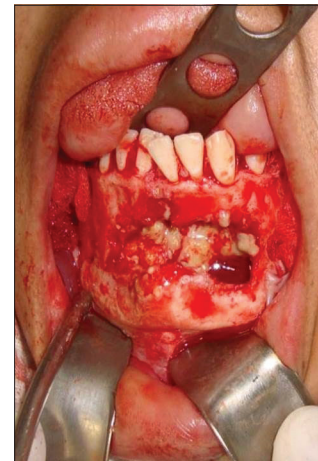


Figure 6: Intraoperative sagittal vestibular incision

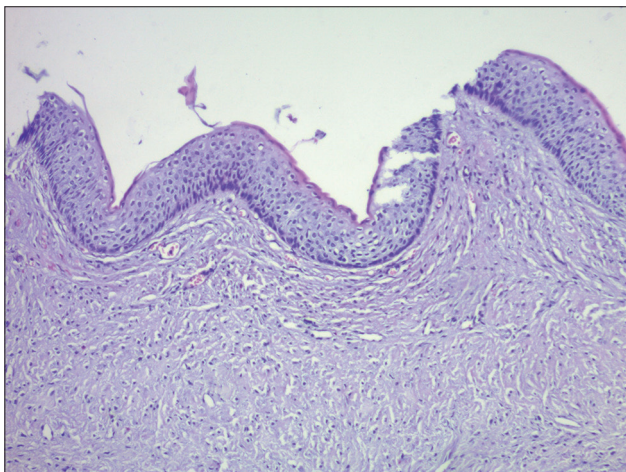


Figure 7: Histopathological analysis showing cystic cavity limited by layer of regularly parakeratinized stratified squamous epithelium and fibrous connective tissue



Figure 8: Six-year follow-up with no signs of recurrence



Figure 9: Panoramic radiograph showing no signs of recurrence after six years

clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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