NEVOID BASAL CELL CARCINOMA SYNDROME: A CASE REPORT

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A síndrome dos múltiplos carcinomas basocelulares nevóides (síndrome de Gorlin-Goltz) é uma entidade composta principalmente de múltiplos carcinomas basocelulares, ceratocistos odontogênicos e anomalias esqueléticas. Aproximadamente 50% de todos os pacientes portadores da síndrome apresentam ceratocistos. Uma das características mais comuns dos ceratocistos é a maior agressividade e a alta taxa de recorrência quando comparados aos demais cistos odontogênicos, além de poderem ser a primeira manifestação da síndrome.

Os autores apresentam um caso da síndrome, com envolvimento extenso da mandíbula pelos ceratocistos.

Quando um paciente for diagnosticado como portador da síndrome deverá ser criteriosamente acompanhado, pois lesões que ainda podem não ter se manifestado podem surgir ao longo da vida.

Unitermos: *Carcinoma basocelular - síndrome Gorlin-Goltz. Síndrome basocelular - neoplasma. Ceratocistos odontogênicos.*

Keywords: *Nevoid basal cell - syndrome Gorlin-Goltz. Syndrome basal cell - neoplasms. Odontogenic keratocysts.*

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Introduction

Jarish (10) in 1894, was the first to report an entity with multiple basal cell nevi associated with neurologic problems and severe scoliosis. *Straith* (18) in 1939 described a family with multiple odontogenic cysts and some of them with basal cell carcinomas. In 1960, Gorlin and Goltz described a syndrome which bore their names, consisting of an association of basal cell carcinomas, odontogenic keratocysts and bifid ribs.

Actually the Gorlin and Goltz'syndrome has been described as an association of multiple basal cell carcinomas (nevoid or not), odontogenic keratocysts, skeletal anomalies (bifid ribs, enlargement of frontal bones), neurological problems, ocular hypertelorism and planto-palmar dysqueratosis (5, 6, 7, 8, 16, 17). It is more prevalent in females and 50% of the cases shows jaw involvement by odontogenic keratocysts (3, 17, 21, 22). The cysts are lined by a keratinized epithelium (6-8 layers thick) and a typical palisade cell layer (2, 12, 13). Keratocysts represent 3%-11% of all jaw and

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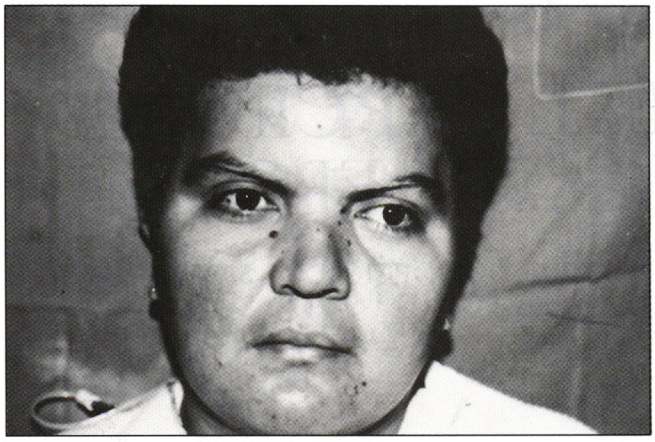

Figure 1 - Nevoid basal cell carcinomas, ocular hypertelorism, enlargement of frontal bones.

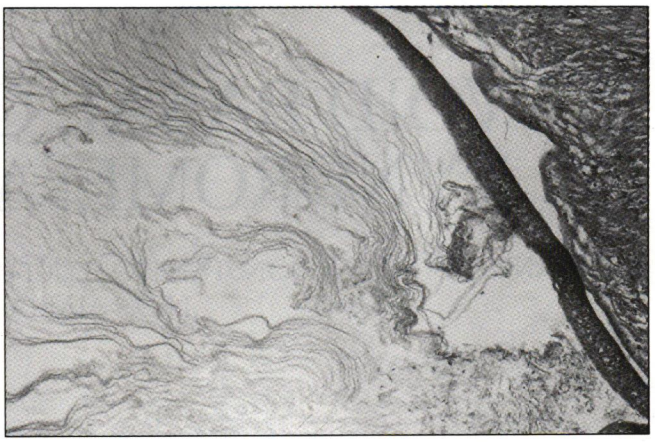

Figure 3 - Histological features of odontogenic keratocyst.

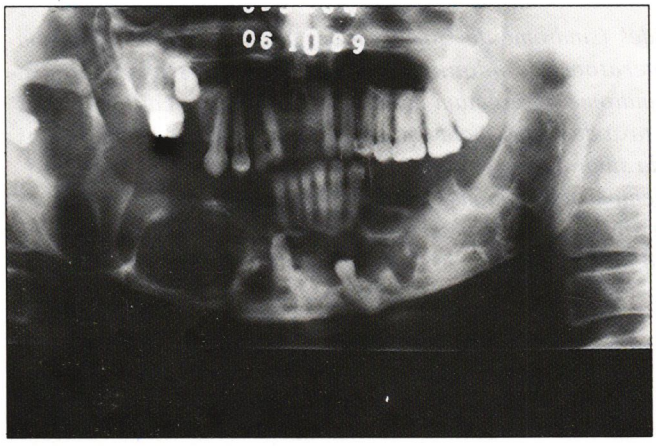

Figure 2 - Odontogenic keratocysts.

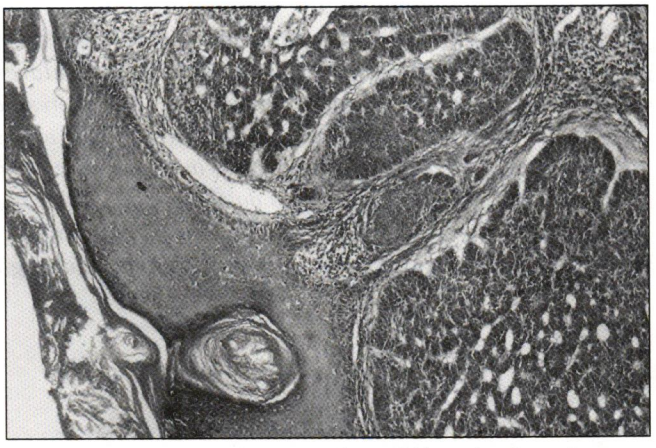

Figure 4 - Histological features of nevoid basal cell carcinoma.

maxillar cysts and the dental lamina rests are the possible origin of these cysts (2, 8, 12, 16). Several studies have show differences in the clinical and histological features of the solitary keratocysts and those presented in the Gorlin and Goltz'syndrome (3, 22). The studies revealed that the keratocyst presented in the syndrome has a more agressive behaviour than the solitary, thus requiring a more radical surgical treatment (9, 11, 14, 19, 20).

Case report

A 26 year old white female was first examined in the Stomatology Department, A. C. Camargo Hospital, in September 1989. The patient reported a painless enlargement of the right mandible for six months, following a dental extraction performed elsewhere.

The external examination revealed and increase in volume with 4cm in the largest diameter, in the right side of the mandible. The lesion had a hard consistence and was covered by normal skin. In the perinasal skin there were 6 nodular pigmented lesions measuring less than 0,5cm in diameter each. The patient also presented enlargement of frontal region and ocular hypertelorism (figure 1). The oral exam revealed the disapearance of the mucosal sulcus on the right side of lower jaw and a fistula in the fibromucous area of the bicuspid region where the tooth was previously removed. Purulent secretion was present at the site of the fistula. The radiological examination revealed an extensive involvement of the entire mandible by radiolucent cystic lesions, with light expansion of the cortical bone of the right retromolar and sigmoid notch. It was also evident two dental inclusions in the anterior region and one close to the right condyle (figure 2).

With these findings, the initial diagnosis was multiple odontogenic keratocysts. In view of the extensive jaw involvement the surgical treatment was performed in two sessions. In October 1989 the lesions were biopsed and after the histopathological diagnoses the cysts of the right side of mandible were carefully enucleated. Five months later (March 1990) the left side mandibular cysts were removed. This planning was necessary to avoid jaw fracture. The pathological exam confirmed the clinical diagnoses of multiple keratocysts (figure 3).

The cutaneous lesions were surgically removed in May 1990 and the histopathological exam revealed nevoid basal cell carcinomas (figure 4).

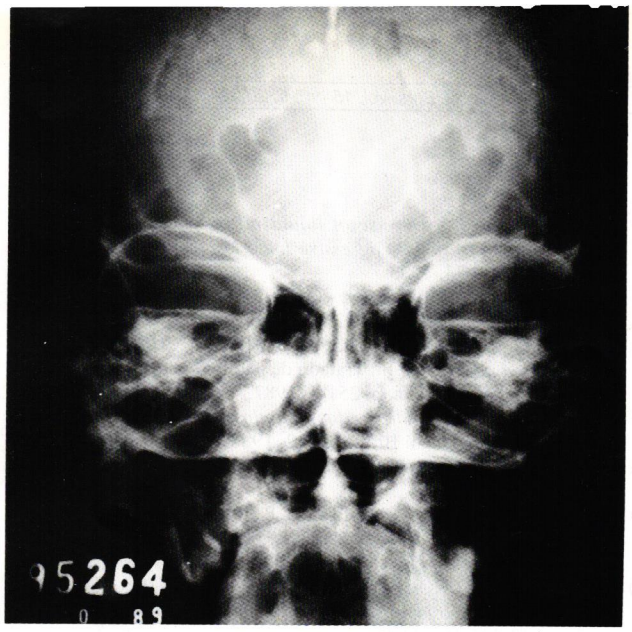

Figure 5 - Calcification of cerebri falx.

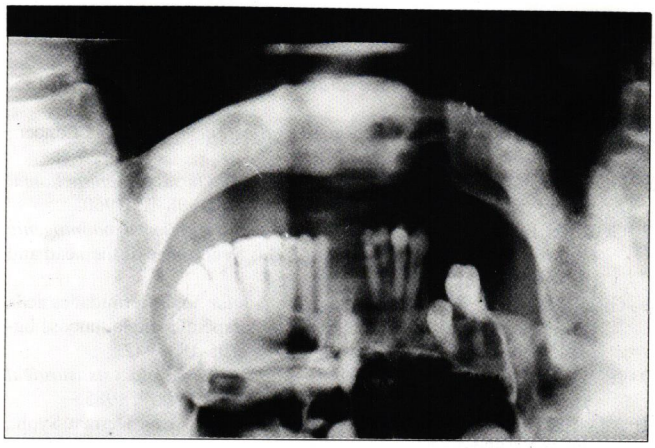

Figure 6 - Follow up three years after the first mandibular surgery.

Additional radiological study revealed calcification of the cerebri falx (figure 5).

Based in the clinical, radiological and histopathological findings, the case has all criteria to be classified as a Gorlin Goltz'syndrome.

Radiographic post operative control showed continued osseous neoformation, filling the cystic cavities of mandible. The patient is living without any evidence of recurrence until January 1995.

Discussion

The Gorlin and Goltz'syndrome is an hereditary entity caused by a dominant autossomic gene of reduced penetration and variable expression (5, 6, 7, 8, 16, 17). Therefore the cromossomal alteration involved in this syndrome are still not clear. The most important features are nevoid basal cell carcinomas of the skin, multiple keratocysts and skeletal anomalies (5, 6, 7, 8, 16). No component of the syndrome is presented in all cases, so there is variable expressivity of the several features (5). As odontogenic keratocyst in the most common oral manifestation and often is the first signal of this syndrome, it is likely that the dentist may be the first to detect the condition (15, 23).

The odontogenic keratocyst present in the syndrome have a different behaviour than other solitary keratocysts, with a recurrence rate of 12%-65% (1, 2, 4, 12). *Brannom* (1) described a higher incidence of satelite cysts and epithelial rests in the syndrome keratocysts than in solitary ones, and sugested that this evidence may reflect a greate proliferative potential in cysts from syndrome patients.

To decrease the rate of recurrence it is essential to perform a careful removal of the cysts as proposed by several authors (9, 11, 14, 19, 20). It has also been suggested additional treatment of the bone cavity with Carnoy's solution, or the use of cryotherapy in the bone cavity resulting from the cyst removal.

For the correct diagnosis and treatment of this entity it is essential a close cooperation between dentist and the oncologist. If the dentist suspect of multiple keratocysts in a given patient, the nevoid basal cell lesions should be sought by the oncologist and vice-versa.

The earlier diagnosis of Gorlin Goltz'syndrome surely is better for patient's treatment and prognosis, because could prevent great deformities and mutilations (17).

Summary

Nevoid basal cell carcinoma syndrome (Gorlin Goltz' syndrome) is a entity composed mainly of multiple basal cell carcinomas, odontogenic keratocysts and skeletal anomalies. Aproximatelly 50% of all syndrome patients have odontogenic keratocysts. The most common feature of keratocysts is the agressivity and high rate of recurrence, compared with other types of odontogenic cysts, and may be the first signal of the syndrome.

A case of the syndrome with extensive jaw involvement by odontogenic keratocysts is presented.

After the definitive diagnosis of the syndrome it is fundamental that be established a continuous follow up, because new lesions arise in long time of life.

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