Treatment of the Keratocystic Odontogenic Tumor (KCOT) in patients with Gorlin Goltz syndrome: a review of the literature with a case report

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KEYWORDS
Gorlin-Goltz Syndrome, Keratocyst, Keratocystic odontogenic tumor, Nevoid Basal Cell Carcinoma Syndrome.

ABSTRACT
Aim: This was to provide a literature review on the dental management of patients with Gorlin-Goltz syndrome and on the surgical treatment of Keratocystic odontogenic tumors (KCOT).

Materials and methods: Fifteen literature reviews published in English language within the last 10 years were selected through PubMed. Additionally, 2 textbooks and 21 clinical studies regarding the surgical approach to KCOTs were included. Data analysis focused on: prevalence rate of Gorlin-Goltz syndrome and related KCOTs; clinical diagnosis; first choice treatment for primary and secondary KCOTs; prognosis and recurrence rate of KCOTs after surgical removal. Through the report of a case, the diagnostic process of Gorlin-Goltz syndrome and the surgical treatment of the related KCOTs are described.

Results: Gorlin-Goltz syndrome is a rare disease with a prevalence of 1:57,000 to 1:256,000. In 75% of cases it is associated with single or multiple KCOTs. The diagnostic hypothesis arises from the observation of multiple/recurrent KCOTs or a single KCOT in patients under 20 years of age. Depending on the clinical presentation of the KCOT, two first choice surgical treatments are indicated: a) simple enucleation with surgical revision and application of a citotoxic agent on the site; b) "en-bloc" resection of the site with removal of at least 1 cm of marginal healthy tissue.

Conclusion: Despite the absence of specific data, several authors consider that KCOTs removal is a procedure with a high risk of recurrence, especially in cases associated with Gorlin-Goltz syndrome.

Introduction
The Gorlin-Goltz syndrome, or Nevoid Basal Cell Carcinoma Syndrome (NBCCS), is a rare form of ecto-meso-dermic polidipsisia. It presents an autosomal dominant transmission and has an incidence that varies between 1/56,000 a 1/256,000 people (1, 2, 3, 4, 5).

This syndrome is typically characterized by the occurrence of multiple keratocystic odontogenic tumors (previously defined as keratocysts), multiple basal cell carcinomas, calcifications of the falx cerebri, palmar and/or plantar pits (1, 6).

Each of these main signs may be present alone or be associated to the other ones. Furthermore, secondary signs can be observed asides from the main ones, which can involve the eyes, the nervous system, the endocrine and genital apparatus (4, 7).

From an oral point of view, the main sign of the Gorlin Goltz syndrome is the keratocystic odontogenic tumor (KCOT) (8, 9). The KCOT is a benign odontogenic neoplasm derived from the dental lamina. Its development is characterized by an aggressive expansion, an osteolytic activity, and the capacity to infiltrate adjacent soft tissues. In rare occasions, extensions into the skull base, the infratemporal fossa, and the orbit were also reported (4, 6, 7, 10, 11).

Because of this well documented aggressive clinical behavior, in 2005 the World Health Organization abandoned the previous designation of this lesion as an aggressive cyst (the odontogenic keratocyst - OKC), to classify it as an odontogenic tumor (KCOT) (8, 9).

Radiographically, KCOTs appear as round or ovoid radiolucent areas with a uni- or multi-locular configuration and variable dimensions. Margins are frequently irregular and corticated (2, 3, 4, 7).

Primary localisations include the posterior mandible and the mandibular ramus (2, 3, 4, 6, 7, 12). Maxillary lesions are less frequent and often located in the premaxilla (2).

Histologically, KCOTs are internally lined by a thin,
multi-layer, parakeratinized (96%) or orthokeratinized (4%), stratified squamous epithelium. This epithelium has five to eight cell layers, with no rete pegs. The basal layer is palisading with columnar or cuboidal cells.

Satellite cysts, cords or islands of epithelium may be seen in the fibrous connective tissue. The cystic lumen is usually filled with a cheesy material, but clear fluid may also be encountered (2, 3, 7).

From a diagnostic point of view, early stage lesions are typically silent. Their detection is usually fortuitous and concomitant to routine radiographic exams, such as panoramic radiographs. Advanced lesions, however, usually become clinically symptomatic inducing:

- a relevant expansion of the mandibular/maxillary bony profile, occasionally correlated with cortical erosion and soft tissues infiltration;
- sudden dental movements;
- paresthesia with or without pain component, secondary to compressions of the inferior alveolar nerve (7, 13, 14).

The aim of the present study is to conduct a literature review on the dental management of patients affected by the Gorlin-Goltz syndrome, with a specific focus on the surgical treatment of the keratocystic odontogenic tumor (KCOT). The most important aspects of diagnosis, treatment planning, surgical performance and patients monitoring will be exemplified through the report of a case.

Materials and methods

Studies included or considered for this review were identified through the PubMed database, using the following keywords: keratocystic odontogenic tumor; odontogenic keratocyst; Gorlin Goltz syndrome; nevoid basal cell carcinoma syndrome; basal cell naevus syndrome; multiple basal-cell carcinoma syndrome; multiple basal-cell naevi syndrome; nevoid basal-cell epithelioma.

Furthermore, the following limits were applied: articles published in english; articles published in the last 10 years; literature reviews; randomized controlled clinical trials (RCT).

After the exclusion of irrelevant results, 15 non-systematic literature reviews were selected out of 410 initial results. No RCTs were found during the search. In order to acquire additional data on the treatment of KCOTs, the search was integrated with 21 clinical studies cited in the selected papers.

The sources were analyzed according to the following queries.

1. Epidemiology
   a) Prevalence of the Gorlin Goltz syndrome in the general population.
   b) Incidence of KCOTs in patients affected by the Gorlin Goltz Syndrome.
   c) Prevalence of the Gorlin Goltz syndrome in patients affected by KCOTs.

2. Diagnosis
   a) Diagnostic criteria for the Gorlin Goltz syndrome.
   b) Major diagnostic criteria.
   c) Minor diagnostic criteria.

3. Treatment
   a) Surgical treatments described in the scientific literature.
   b) First choice alternatives.
   c) Indications for the different surgical approaches.

4. Prognosis
   a) Recurrence rates of KCOTs and their relation to the surgical technique adopted for removal.
   b) Factors that may influence the recurrence rate.
   c) Post-operative monitoring.

Results

1. Epidemiology
   a) Prevalence of the Gorlin Goltz syndrome in the general population: the Gorlin Goltz syndrome is a rare pathology with a prevalence that varies from 1: 57,000 to 1:256,000 according to geographic areas: the United Kingdom appears to have a higher rate of incidence (1:57,000) when compared to Australia (1:164,000) and Italy (1:256,000) (2). The male-to-female ratio is 1:1.2.
   b) Incidence of KCOTs in patients affected by the Gorlin Goltz Syndrome: approximately 75% of patients affected by the Gorlin Goltz syndrome develop single or multiple KCOTs (15).
   c) Prevalence of the Gorlin Goltz syndrome in patients affected by KCOTs: approximately 5% of patients that present a KCOT are affected by the Gorlin-Goltz syndrome (15).

2. Diagnosis
   a) Diagnostic criteria for the Gorlin Goltz syndrome: the diagnosis of the Gorlin Goltz syndrome is primarily based on clinical findings, which can be divided in major criteria and minor criteria of pathology (1, 3, 4). Specifically, a diagnostic hypothesis is formulated in the presence of two major criteria and a minor one, or in the presence of one major criteria and three minor ones (4). With respect to the oral diagnosis, a hypothesis of Gorlin Goltz syndrome can be formulated in the presence of a single KCOT lesion in patients younger than 20 years, or multiple/recurring KCOTs (2). On average, the syndrome is first diagnosed around 13 years of age, and nevoid basal cell carcinomas usually appear around 20 years of age (1).
b) Major diagnostic criteria:
1. one or more basal cell carcinomas (BCC) in patients under 20 years, and multiple (more than two) BCCs in patients older than 20 years;
2. odontogenic keratocysts of the jaws proven by histopathology;
3. palmar or plantar pits (3 or more);
4. bilamellar calcification of the falx cerebri;
5. bifid, fused or markedly splayed ribs;
6. first degree relatives with NBCCS (2, 16).

c) Minor diagnostic criteria:
1. macrocephaly determined after adjustment for height;
2. congenital malformation: cleft lip or palate, frontal bossing, “coarse face”, moderate or severe hypertelorism;
3. other skeletal abnormalities: Sprengel's deformity, marked pectus deformity, marked syndactyly of the digits;
4. Radiological abnormalities: bridging of the sella turcica, vertebral anomalies such as hemivertebrae, fusion or elongation of the vertebral bodies, modeling defects of the hands and feet, or flame-shaped lucencies of the hands or feet;
5. ovarian fibroma;
6. medulloblastoma (2, 16).

3. Surgical treatment of KCOTs
a) Surgical treatments described in the scientific literature
Different treatment strategies were suggested for the treatment of KCOTs, including:
1. simple/conservative enucleation (7);
2. enucleation combined with an aggressive curettage of the residual bone walls (25);
3. decompression via marsupialization, followed by enucleation and curettage at a second surgical time (26);
4. enucleation and curettage followed by the application of the Carnoy solution (27);
5. enucleation and curettage followed by osteotomy of the residual bone walls, guided by cellular stains like methylene blue (28);
6. en bloc resection of the lesion with asportation of at least 1 cm of peripheral healthy tissue, through a marginal or segmental resection (7).

b) First choice alternatives
There are no prospective controlled trials that compare the long-term efficacy of the different approaches. The selection of a specific technique can only be based on retrospective data and surgical experience. Multiple authors consider as first choice treatment: a) the conservative enucleation combined with an aggressive curettage or osteotomy of the residual bony walls that faced the lesion, and the additional application of a cytotoxic agent, such as the Carnoy solution; b) the en bloc resection of the area affected by the lesion, that includes at least 1 cm of macroscopically healthy peripheral tissue (17, 18).

c) Indications for the different surgical approaches
There are no specific indications with respect to the degree of surgical invasiveness that is required in the treatment of a KCOT. Some authors agree that:
1. the conservative enucleation with adjuvant mechanical and chemical treatment of the residual bony walls is primarily indicated in localized mandibular lesions with no interruption of the cortical bony walls. In such conditions, an aggressive curettage combined with the application of the Carnoy solution seems to be efficacious in the removal of residual epithelial cells that can induce/promote a recurrence of the lesion;
2. the en bloc resection is primarily indicated in case of recurring lesions that arise after an initial, more conservative treatment. A resective approach is also indicated in primary extensive lesions of the upper jaw, that are adjacent or in direct communication with the paranasal sinuses (7, 12, 19).

The reason why a more aggressive approach is usually adopted in the maxilla, is related to its local anatomy. Specifically, the higher content of spongious bone offers a scarce resistance to the expansion of the tumour mass. Furthermore, the upper jaw is in close proximity to noble anatomical structures like the infratemporal fossa and the skull base (7, 12, 19).

4. Prognosis
a) Recurrence rates of KCOTs and their relation to the surgical technique adopted for removal: Zhao et al. published results with a 3 to 29 year follow-up, of 255 KCOTs treated with enucleation with curettage (n=163), enucleation with curettage and application of the Carnoy solution (n=29), masupialization and second stage enucleation (n=11), and en bloc resection (n=52). A mean recurrence rate of 12,16% was observed. Specifically, 17,79% in patients treated with enucleation with curettage, 6,70% (p<0,05) in patients treated with enucleation with curettage and application of the Carnoy solution, and 0% (p<0,01) in patients treated with marsupialization or en bloc resection (18). However, there are no long term studies that compare in a standardized way the recurrence rate of homogenous groups of patients treated with different surgical approaches. Long term follow-ups are essential to compare the different approaches, as the development of recurrent lesions was described to arise even 25 years after the first surgical treatment. However, the majority of recurrences are generally observed within 5 years from the original treatment (20).
b) Factors that may influence the recurrence rate: the analyzed studies support that:
1. the recurrence rate of KCOTs is always high;
2. the recurrence rate is higher in the presence of multilocular lesions, fistulae that originate from the primary lesion, perforations of the cortical bone walls, and supra-infections;
3. patients affected by the Gorlin Goltz syndrome present a higher rate of recurrence for KCOTs;
4. the recurrence rate does not seem to be related to the position nor the dimension of the lesion;
5. the primary cause of recurrence seems to be the permanence of islets of epithelial cells within the marginal peripheral connective tissue, that were not adequately removed during the surgical treatment of the primary lesion (13, 18, 21, 22).

c) Post-operative monitoring: as recurrent lesions can appear even after many years from the treatment of a KCOT, postoperative monitoring should be conducted for at least 10 years after surgery (19, 23, 24).

Case report
A 22 year old, systemically healthy woman, was referred to our oral surgery unit because multiple radiolucent lesions, involving the upper and lower jaws, were observed in a routine panoramic radiograph. During the previous 4 months, the patient was referring episodes of transient paresthesia of the lower left lip.

At the observation of the panoramic radiograph, four radiolucent lesions were found in conjunction with fully impacted upper and lower third molars. Furthermore, a fifth lesion was observed in contact with the upper right canine and first bicuspid. All lesions presented...
Radiographic features compatible with a diagnosis of KCOT (Figure 1).

At the intraoral examination, the patient presented normally erupted teeth, except for all the third molars. All erupted teeth presented a normal response to pulp vitality tests (Figure 2).

At the general extra-oral examination, palmar pits were observed in both hands (Figure 3). In the light of those radiographic and clinical findings, a diagnostic hypothesis of Gorlin Goltz syndrome was formulated.

A computed tomography of the upper and lower jaw was requested, in order to better assess the location and extension of all radiolucent lesions. The radiographic field of view was extended to the base of the skull, in order to assess the presence of calcifications of the falx cerebri, which were not found.

Under local anesthesia, an incisional biopsy was performed on the lesion in the right inferior retromolar area. The histopathological diagnosis confirmed the presence of a KCOT (Figure 4). Thus, the surgical treatment of all radiolucent lesions was planned in conjunction with the extraction of the impacted third molars, under general anesthesia.

The preoperative patient preparation included: a) professional oral hygiene 2 weeks prior to surgery; b) local antisepsis with 0.2% chlorhexidine mouthwashes starting 3 days before surgery; c) antibiotic prophylaxis with 2 g of oral amoxicillin and clavulanate, administrated one hour before surgery.

After nasal-tracheal intubation, general anesthesia induction, and local infiltration of mepivacaine 2% + epinephrine 1:100,000, the surgical enucleation of the five lesions was performed through a conservative approach, in conjunction with the extraction of the impacted third molars (1.8, 2.8, 3.8, 4.8) and the right upper canine (1.3). In order to thoroughly remove any lesion remnant from the surrounding bone walls,
Figure 5 Conservative removal of the osteolytic lesion located in the right retromolar region

Figure 6 Avulsion of tooth 4.8 in deep bone impaction, contiguous to the osteolytic lesion

Figure 7 Excision of the osteolytic lesion localized in the posterior upper maxilla, in communication with the maxillary sinus
Figure 8 Excision of the osteolytic lesion located close to teeth 1.3 and 1.4

Figure 9 Avulsion of tooth 2.8 in bone impaction

Figure 10 Removal of the osteolytic lesion annexed to tooth 2.8
adjunctive mechanical and chemical treatment was conducted via aggressive curettage with manual curettes and application of the Carnoy solution.

In the lower jaw (Figure 5, 6), both lesions were in direct contact with the right and left inferior alveolar neurovascular bundles. Care was taken to preserve their integrity both during lesions enucleation, third molars extractions, and adjunctive treatments delivery. Specifically, during the application of the Carnoy solution, the bundles were protected with sterile gauzes or momentarily displaced, in order to avoid any damage.

In the upper jaw (Figure 7, 8, 9, 10), bilateral oro-antral communications were found after the extraction of the third molars and the associated osteolytic lesions. Thus, a Rehrmann’s advancement flap with the additional support of the isolated buccal fat pad was conducted bilaterally, to close the surgical access through the advancement of a thick buccal flap, and reach a first intention wound healing.

Both in the upper and lower jaws, wound closure was performed with non-resorbable 4/0 monofilament sutures. Simple interrupted and horizontal internal mattress sutures were applied (Figure 11, 12). All surgical procedures were performed by the last author (M.C.).

All the removed pathologic material was submitted for histopathologic examination to the Unit of Human Pathology, Department of Health Sciences, San Paolo Hospital, University of Milan, Italy. The histopathological diagnosis of all lesions further confirmed the presence of KCOTs.

The following post-surgical instructions were prescribed:

1. application of ice packs over the surgical site for 6-8 hours after surgery;
2. oral antibiotics (amoxicillin+clavulanate - 2 g per day) for 6 days post-operatively;
3. post-surgical non-steroidal analgesic therapy for 2 to 3 days after surgery;
4. cold and liquid diet for the first two days after surgery and soft diet until suture removal;
5. mouth rinses with chlorhexidine digluconate 0.2%, twice a day for 15 days, combined with normal oral hygiene manoeuvres on the remaining dentition.

Furthermore, the patient was instructed to sneeze with her mouth open and not to blow her nose for the first 15 post-operative days, to avoid dangerous pressure peaks within the maxillary sinuses, which could hamper the healing of the closed oro-antral communications.

Sutures were removed 7 days after surgery. No short-term complications were observed and the surgical sites healed uneventfully.

Clinical follow-ups were conducted at 1, 6, 12, and 24 months after surgery.

Panoramic radiographs were acquired immediately after surgery, 12 and 24 months afterwards, to inquire whether an adequate re-ossification of the residual cavities was occurring during healing. At the last radiograph (24 months) the degree of radiopacity observed in the post-surgical sites was comparable with the adjacent healthy bone (Figure 13, 14).

Discussion

In the last decades, multiple alternatives were described in the surgical treatment of the KCOT, which differ in terms of aggressiveness, efficacy, and efficiency (7, 19, 25, 26, 27, 28).

Ghali et al. reported how the sole conservative enucleation with no adjunctive treatments is characterized by a high rate of recurrence, which varies between 9 and 62.5% (19). Today, most authors support the adoption of more invasive solutions, which were associated to significantly lower rates of recurrence (7, 19).

Zhao et al. reported a recurrence rate of 7% when treating KCOTs with a conservative enucleation combined with an aggressive manual curettage of the residual bony walls that faced the lesion. This approach seems to be more efficacious in the removal of those remnants of pathologic tissue from which a secondary lesion can arise (18).

In order to further increase the capability to eliminate residual pathologic cells, and thus to lower the recurrence rates, some authors added further adjunctive treatments to the conservative enucleation with manual curettage (27, 28). Specifically, Voorsmit, Stoelinga and Van Haelst introduced in 1981 a surgical protocol that implements three adjunctive procedures to the enucleation with manual curettage:

1. application of the Carnoy solution on the surface of the residual bone walls that faced the lesion. This was originally composed of absolute alcohol, chloroform, acetic acid and iron chloride;
2. removal of areas of buccal mucosa that directly faced the lesion, in case of erosion of the buccal cortex;
3. electrocauterization of areas of lingual mucosa that directly faced the lesion, in case of erosion of the lingual cortex.

This approach was validated through a retrospective study conducted on 82 KCOTs with a mean follow-up of 11.8 years (from 2 to 25 years, STD 7.2 years) in maintained patients, and of 4.1 years (from 1 to 10 years, STD 3.1 years) in patients that were lost during follow-up. Three cases of recurrence were observed in 38 KCOTs treated with the suggested protocol, whether 6 cases of recurrence were observed in the 44 patients that received a simple conservative enucleation (20). However, chloroform has today been removed from the composition of the Carnoy
solution, as it was observed to express a carcinogenic activity (29).

An additional adjunctive treatment following conservative enucleation with manual curettage is represented by the ostectomy of the residual bony walls that faced the lesion, performed with rotary instruments (7, 25, 28).

This approach provides a higher surgical aggressiveness than the conventional manual curettage, but is still more conservative than a radical resection. It is primarily indicated in cases at higher risk of recurrence, were the integrity of the bony process affected by the lesion needs to be preserved (25). Specifically, in 1991 Williams proposed to guide the ostectomy procedure by applying methylene blue on the residual bony walls, in order to highlight areas of peri-lesional tissue that still contain residual epithelial cells (28). Madras et al. observed a significant reduction of the recurrence rate when comparing this protocol to the conservative enucleation (18% vs. 30% of recurrences). However, the recurrence rate was higher if compared to a conservative enucleation with curettage and application of the Carnoy solution (9% of recurrences). A mild reduction was observed when combining the two approaches, performing first the enucleation with curettage and the cellular stain guided ostectomy, and subsequently the application of the Carnoy solution (8% of recurrences) (9). However, no data are available with respect to the depth at which methylene blue is able to penetrate the peripheral bone and the extent to which the ostectomy procedure should be performed in order to guarantee an efficacious removal of all epithelial remnants of the lesion (19).

An alternative approach to the conservative enucleation with adjunctive measures, is the initial decompression of the lesion via masupialization, followed by enucleation and curettage in a second surgical time (7, 25). The procedure aims to first achieve an initial reduction of the lesion extension, and then to enucleate the residual mass with a less invasive approach. Furthermore, the marsupialization seems to induce an inflammatory reaction that thickens the perilesional connective tissue, thus facilitating the
surgical manipulation of the lesion during the second stage enucleation (25).

This approach was described to be very efficacious, with recurrence rates that are only matched by the en bloc resection (18). However, Bhargava et al. objected that, in the light of the recent WHO classification that has redefined the keratocyst as an odontogenic tumor (KCOT) (8,9), a masupialization based approach does not seem to be appropriate (7).

A different, more aggressive approach, is the en bloc resection of the lesion with asportation of at least 1 cm of peripheral healthy tissue. The resection can maintain the integrity of the bony process (marginal resection) or interrupt it abruptly (segmental resection) (7, 17, 18, 19, 25).

Even though this approach was associated with a 0% recurrence rate (14, 18, 23, 30, 31, 32), the higher morbidity limits its indications in non malignant lesions such as the KCOT, which does not express any real metastatic potential. Consequently, the en bloc resection does not represent a first choice alternative in most cases of primary KCOT (7). In those cases where a segmental resection or a broad marginal resection are performed, it is mandatory to sustain the residual bone structure with osteosynthesis plates and to apply an intermaxillary fixation for the first post-operative period (19, 32).

In the light of those considerations, the authors of the present paper believe that: the first choice alternative for the treatment of primary KCOTs is represented by the conservative enucleation with aggressive manual curettage and application of the Carnoy solution; the en bloc resection is mainly indicated in case of: 1) secondary lesions that recur after treatment with a
more conservative approach; 2) extended lesions that eroded the cortical bone walls of the mandible/maxilla and invaded the surrounding soft tissues to an extent that does not allow to apply a conservative enucleation with curettage and application of the Carnoy solution; 3) vast lesions located in the posterior maxilla, that extended to the paranasal sinuses, and/or reached the proximity of the skull base, the infra-temporal fossa, the pterigo-palatine fossa, or the orbits (7, 19).

The only case in which a resective approach must be applied even in the treatment of primary and confined KCOTs, is the observation of a carcinogenic evolution of the mass during the preliminary histopathological examination performed on an incisional biopsy (25).

The marsupialization based approach has very limited indications, such as the presence of extended primary lesions that are in close contact with noble anatomical structures. In these cases, a preliminary reduction of the tumor mass can allow a more conservative enucleation in a second stage procedure.

In the reported clinical case, the diagnostic hypothesis of the Gorlin Goltz syndrome arose from the observation of multiple KCOTs and was confirmed, accordingly to the published diagnostic criteria, by the co-presence of palmar pits in both hands (16).

After acquiring all preliminary data from a computed tomography and the histological analysis of an incisional biopsy, a treatment approach based on conservative enucleation with aggressive manual curettage and application of the Carnoy solution was selected. This was in accordance with data from the literature, as none of the lesions were associated with cortical erosion. As it was reported that patients affected by the Gorlin Goltz syndrome exhibit a higher rate of KCOT recurrence, the patient was included in a recall regimen where a panoramic radiograph is acquired every year.

Additionally, as it was reported that the etiology of the Gorlin Goltz syndrome has a genetic component, associated in 80% of cases with a mutation of the PTCH1 gene, all close relatives of the patient were screened for radiolucent lesions of the jaws and other signs of the syndrome. None of them resulted to be affected.

At a 2 year radiographic follow-up, a regular reossification of all surgical sites was observed in the panoramic radiograph, with no signs of recurrence.

Conclusions

The Gorlin Goltz syndrome is a rare disease, with a prevalence of 1:57,000 to 1:256,000 individuals. In 75% of cases, Gorlin Goltz patients are affected by KCOTs. Multiple surgical approaches were suggested for the treatment of these lesions, including a simple conservative enucleation with or without adjunctive mechanical (manual curettage and/or peripheral ostectomy) or chemical (application of the Carnoy solution) measures, and the en bloc resection of the affected area, via a marginal or segmentary resection. Even though prospective datas are lacking, most authors agree that:

- KCOTs are always associated with a high risk of recurrence;
- patients affected by the Gorlin Goltz syndrome present a higher rate of recurrence for KCOTs;
- at present, the enucleation with aggressive curettage and application of the Carnoy solution represents the most conservative approach that is capable to provide satisfactory clinical outcomes. The simple conservative enucleation with no adjuvant measures does not represent a reliable alternative, as it was associated to a high rate of recurrence;
- even though the en bloc resection is the most efficacious approach to prevent KCOTs recurrences, in most cases of primary lesion it is associated with an excessive amount of surgical invasiveness, thus being not indicated.

References