

Decompression Followed by Targeted Secondary Enucleation of an Extensive Mandibular Odontogenic Keratocyst in Gorlin-Goltz Syndrome: A Case Report with Differential Radiologic Response of Two Lesion Compartments

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Publication Date: 2026/10/22

Abstract: The odontogenic keratocyst (OKC) is a benign but locally aggressive intraosseous lesion with a high recurrence rate and a well-established association with Gorlin–Goltz syndrome. In young patients with Gorlin–Goltz syndrome and extensive mandibular involvement, radical resection can cause severe functional and aesthetic morbidity, prompting interest in conservative, bone-sparing alternatives. Previous reports have described decompression and staged enucleation as effective strategies for reducing lesion volume and enabling less morbid definitive surgery, yet evidence in syndromic cases with bilateral or multilocular mandibular disease remains limited.

This case report describes the outcome of a two-stage conservative approach in an 18-year-old man with Gorlin–Goltz syndrome and extensive multilocular mandibular OKCs at high risk of pathological fracture. The scope includes clinical, radiographic, and histopathological assessment, decompression with a custom stent, and secondary enucleation with peripheral ostectomy. The novelty lies in achieving complete resolution of one compartment and marked volumetric reduction of the other, enabling nerve-sparing surgery without neurovascular deficit or fracture, thereby supporting decompression as a viable first-line option in selected syndromic patients.

Keywords: Odontogenic Keratocyst; Gorlin-Goltz Syndrome; Nevroid Basal Cell Carcinoma Syndrome; Decompression; Mandible; Bone Regeneration.

How to Cite: Sarra Azzez; Hela Zouaghi; Chaima Haj Hassine; Chiraz Baccouche (2026) Decompression Followed by Targeted Secondary Enucleation of an Extensive Mandibular Odontogenic Keratocyst in Gorlin-Goltz Syndrome: A Case Report with Differential Radiologic Response of Two Lesion Compartments. *International Journal of Innovative Science and Research Technology*, 11(10), 1300-1305. <https://doi.org/10.38124/ijisrt/26oct560>

I. INTRODUCTION

The odontogenic keratocyst (OKC) is a benign but locally aggressive intraosseous lesion with a well-known tendency to recur. Since Philipsen's characterization in 1956 [1] and Pindborg and Hansen's subsequent clinical and radiographic description in 1963 [2], OKC has prompted debate about its biological nature. The WHO reclassified it as a keratocystic odontogenic tumor in 2005, reinstated the term OKC in 2017 [3], and has maintained this designation in the 2022 classification [4].

OKCs predominantly affect young adults in the posterior mandible and are histologically defined by a thin, parakeratinized epithelium with a palisaded basal layer. Recurrence rates vary widely (15% to over 60%) by treatment modality and have been attributed to capsule fragility, cortical perforation, and satellite microcysts [5,6].

Multiple OKCs are a hallmark of Gorlin–Goltz syndrome, in which lesions present earlier, contain more satellite cysts, and recur more frequently [5]. Optimal management of extensive mandibular OKC remains debated,

with options ranging from radical resection to conservative decompression or marsupialization, often with adjuncts such as Carnoy's solution or peripheral ostectomy [5]. In young or syndromic patients, radical surgery carries substantial morbidity, motivating two-stage strategies that reduce lesion volume before selective enucleation [7,8]. We report a Gorlin-Goltz patient with a two-compartment mandibular OKC in whom decompression completely resolved one compartment and enabled safe, targeted enucleation of the residual ramus lesion, highlighting this approach's potential and limitations.

II. CASE REPORT

An 18-year-old man presented to the Department of Oral Surgery at Taher Sfar Teaching Hospital with a 3-day history of rapidly progressing swelling and severe tenderness over the right lower jaw, localized discomfort, and difficulty chewing, consistent with left lower buccal space cellulitis. His past medical and family history was unremarkable for systemic metabolic disease.

The acute infection was managed with amoxicillin 1 g 3 times/day for 7 days, paracetamol 1 g 3 times/day for 4 days, and an antiseptic mouth rinse.

Extraoral examination showed facial asymmetry with soft-tissue edema over the left mandibular angle and cheek (Fig.1). Intraoral examination showed cortical expansion along the right posterior mandibular alveolar ridge and ascending ramus. Dermatological examination showed multiple discrete punctate depressions on the palmar surfaces of both hands (palmar pits) (Fig. 2).



Fig 1 Initial Acute Presentation



Fig 2 Palmar Pits

Panoramic radiography and high-resolution computed tomography (CT) showed multiple extensive, well-demarcated multilocular radiolucent lesions of the mandible and the maxilla (Fig.3). The two left mandibular lesions extended from the distal root of the first molar (tooth 36) to the left mandibular angle, and the entire right ascending ramus up to the coronoid process.

Multiplanar reformations of the CT scan showed marked thinning, expansion, and focal dehiscence of the buccal and lingual cortical plates, creating a critical risk of pathological fracture. The lesion was intimately related to the inferior alveolar canal, displacing the nerve bundle towards the inferior cortical border (Fig. 4).



Fig 3 Preoperative Imaging Showing Extensive Multiple Multilocular Radiolucent Lesions Involving the Mandible and the Maxilla

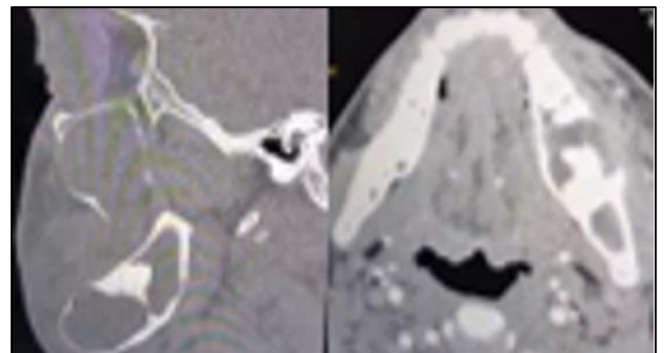


Fig 4 Computed Tomography Scan: Sagittal and Axial Views Showing the Two Left Mandibular

An incisional biopsy under local anesthesia showed a uniform parakeratinised stratified squamous epithelial lining 5–8 cell layers thick, a palisaded columnar basal layer, absence of rete ridges, and a corrugated keratinized surface, confirming the diagnosis of odontogenic keratocyst.

On the basis of the clinical findings, imaging, and biopsy, a diagnosis of Gorlin-Goltz syndrome was retained.

Because of the extent of the lesion, the severe cortical destruction, the high risk of fracture, and the patient's age, primary enucleation or radical resection was considered inappropriate owing to the risk of permanent nerve damage and disfigurement, and a two-stage plan based on decompression was selected. Under local anesthesia, the left posterior mandible was approached. Tooth 37 and impacted

tooth 38 were extracted, and a bony fenestration of approximately 15 to 20 mm was created. Thick keratinaceous debris was gently evacuated. A customized decompression stent, fabricated from a modified surgical drainage cannula, was placed in the bony opening and sutured to the wound margins with non-absorbable sutures to maintain continuous communication between the cavity and the oral environment (Fig.5).



Fig 5 Intraoperative View of the Decompression Cannula in Situ.

The patient and his caregivers were instructed to irrigate the cavity through the cannula with sterile normal saline two to five times daily. Healing was uneventful, with a well-epithelialized mucosal decompression port surrounded by healthy oral mucosa after cannula retrieval (Fig.6).



Fig 6 Removal of the Customized Drainage Stent, Showing an Epithelialized Mucosal Decompression Port/Fistula with Healthy Surrounding Soft Tissue

Serial clinical and radiographic monitoring was performed during decompression. At 10 months, panoramic radiography and CBCT showed complete radiological and clinical resolution, with bone fill and disappearance of the radiolucent cavity; no secondary enucleation of the anterior compartment; and marked volumetric contraction with centripetal bone deposition and thickening of the buccal and lingual cortical walls of the Posterior ascending ramus compartment (Fig.7).



Fig 7 10-Month Post-Decompression Radiograph Demonstrating Resolution of the Anterior Lesion and Marked Bone Apposition in the Ramus.

Because the posterior lesion had substantially shrunk and moved away from the mandibular canal, Phase 2 was performed; the residual cystic tissue of the right ascending ramus was enucleated, with adjunctive peripheral osteotomy to remove potential daughter cysts at the bony margins. Postoperative healing was uneventful. Sensation in the

inferior alveolar nerve distribution remained normal, mandibular continuity was preserved without pathological fracture, and facial symmetry and function were satisfactory. At the one-year follow-up, the panoramic X-ray showed complete reossification of the mandibular lesion (Fig. 8).



Fig 8 Final Postoperative Radiograph Following Secondary Enucleation (at 12 Months Follow-Up)

Table 1 Timeline

Time Point	Event
Day 0	Presentation with left lower buccal space cellulitis; CBCT; incisional biopsy confirming OKC; palmar pits identified; Gorlin-Goltz syndrome diagnosed
Phase 1 [Janvier 2024]	Decompression under local anesthesia; teeth 37 and 38 extracted; fenestration; custom stent; saline irrigation two to five times daily
10 months	Panoramic radiography and CBCT: anterior compartment resolved; posterior compartment reduced with cortical thickening
Phase 2 [December 2024]	Enucleation with peripheral ostectomy of the residual ramus lesion; nerve preserved
Last follow-up [February 2026]	Complete reossification

III. DISCUSSION

In this patient with Gorlin-Goltz syndrome, decompression of an extensive mandibular OKC was followed at 10 months by complete radiological resolution of the anterior compartment and volumetric reduction with centripetal cortical thickening of the posterior compartment, allowing nerve-sparing secondary enucleation. Staged protocols combining decompression with secondary enucleation have been described for large mandibular OKCs [9], and long-term outcomes of decompression-based management have been reported in young patients with NBCCS [7,10]. Decompression before enucleation has been associated with a lower recurrence rate [8]. What distinguishes the present case is the differential response of two anatomically distinct compartments within the same syndromic patient under identical decompression conditions.

The biological rationale for decompression is to reduce intraluminal hydrostatic pressure by connecting the cystic lumen to the oral cavity [7]. This pressure release, together with reduced osmolality and locally produced bone-resorbing cytokines such as interleukin-1 α , interleukin-6, and tumor necrosis factor- α acting through the RANKL pathway, is thought to favor osteoblastic apposition and centripetal bone regeneration over osteoclastic resorption [7,9]. Decompression has also been reported to alter the epithelial lining, reducing cytokeratin-10 expression and converting the parakeratinised epithelium into a thicker, non-keratinized

lining supported by denser connective tissue, potentially facilitating subsequent enucleation [7].

Complete radiological resolution of the anterior compartment within 10 months after decompression alone was unexpected, as decompression is generally considered a preparatory step before cystectomy, although complete resolution has been reported, particularly in younger patients [9]. Young patients may have heightened osteogenic capacity and faster bone remodeling, which may contribute to this outcome [7]. Nonetheless, radiological bone fill does not exclude persistent microscopic satellite cysts or epithelial rests in adjacent marrow spaces, particularly in syndromic OKC, and decompression alone therefore carries a risk of late recurrence. Complete resolution should thus be regarded as a radiological observation rather than proof of cure, and continued surveillance of the anterior compartment is warranted. Delayed, targeted enucleation with peripheral ostectomy of persistent compartments, as performed here, combines bone preservation with removal of residual epithelium. Chemical adjuncts such as Carnoy's solution were not used, given the proximity of the inferior alveolar nerve, although a network meta-analysis ranked enucleation with peripheral ostectomy and 5-fluorouracil highest for recurrence reduction, albeit on very low-quality evidence [5].

Treatment selection for large mandibular OKC requires balancing recurrence risk against procedural morbidity. Radical resection offers the lowest recurrence rate but carries severe, disfiguring morbidity and high risk of neurovascular

deficit [7], whereas enucleation alone and marsupialization alone present intermediate recurrence rates with varying degrees of morbidity. Decompression followed by secondary enucleation offers a low recurrence rate with minimal to low, bone-sparing morbidity and preservation of neurovascular structures [11]. Patients with Gorlin–Goltz syndrome are predisposed to multiple and recurrent OKCs, so repeated radical resections would accumulate morbidity in the form of disfigurement, loss of dentition, and functional and psychological impairment [7,10,12]. In this patient, decompression as the initial step preserved mandibular continuity and the inferior alveolar neurovascular bundle, avoided free-tissue transfer or grafting, and restricted secondary surgery to the residual ramus lesion, with cortical thickening reducing the risk of pathological fracture. Bone-regenerative adjuncts such as guided tissue regeneration have also been combined with conservative surgery for mandibular OKC [13], though none was used here.

Custom drains and cannulas for OKC decompression have been described as practical technical solutions [11]; our device was fabricated from a modified surgical drainage cannula and sutured to the wound margins. The principal limitation of decompression is its dependence on adherence, as the patient and caregivers must irrigate the cavity several times a day for months, and inadequate hygiene may cause infection, stent obstruction, or premature closure of the window [7,12]. Because Gorlin–Goltz syndrome is a multisystem disorder, oral and maxillofacial management should be integrated into lifelong multidisciplinary surveillance involving maxillofacial surgeons, dermatologists (for basal cell carcinoma), geneticists, and neurologists (for intracranial lesions) [7]. No standardised protocol defines the optimal duration of decompression or the objective criteria for its completion. Three-dimensional CBCT volumetric analysis has been used to quantify decompression effects on jaw cystic lesions and may help track reduction rates and timing of secondary enucleation [14]. Standardised volumetric monitoring criteria are needed to guide clinical decision-making.

IV. LIMITATIONS

This is a single case with a short reported interval without genetic confirmation of the syndrome. The outcome depended on patient adherence and cannot be generalized. Without longer surveillance, recurrence of the resolved anterior compartment and the enucleated ramus lesion cannot be excluded.

V. CONCLUSION

In this patient with Gorlin–Goltz syndrome, staged management with decompression followed by targeted secondary enucleation with peripheral ostectomy achieved complete radiological resolution of one compartment, volumetric reduction and cortical thickening in the other, and nerve-sparing removal of the residual lesion without pathological fracture or sensory deficit at the reported follow-up. This single case suggests that decompression may be a bone-sparing first step in selected syndromic patients with

extensive mandibular OKC; longer follow-up and larger series are needed to establish recurrence rates and optimal protocols.

➤ Declarations

- *Informed Consent:*

The patient provided written consent for publication of clinical details and images.

- *Conflicts of Interest:*

The authors declare no conflicts of interest.

- *Funding:*

This study received no specific funding.

- *Acknowledgments:*

None.

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