

CASE REPORT

Craniofacial approach in the treatment of invasive odontogenic keratocyst

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The odontogenic keratocyst (OKC) is an intraosseous cystic lesion of the bone covered by keratinized epithelium that originates from the dental lamina. OKC accounts for approximately 12% of cysts occurring in the maxillofacial region. When associated with Gorlin-Goltz syndrome (GGS), these cysts become more invasive, prone to relapse, and can affect several quadrants of the jaw bones simultaneously. The surgical approach chosen depends on the anatomical location and degree of invasion of the disease, but usually includes a superior coronal incision combined with an inferior transfacial approach in cases of extensive nasal involvement.

Keywords: odontogenic tumors; pathology, clinical; jaw cysts; craniofacial abnormalities.

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The odontogenic keratocyst (OKC) is an intraosseous cystic lesion of the bone covered by keratinized epithelium that arises from remnants of the dental lamina. It is the third most common odontogenic cyst and comprises approximately 12% of cysts occurring in the maxillofacial region¹.

This condition has been reported in both genders with a slight predominance in males and presents an overall peak around the second and third decades of life. It is insidious and painless, commonly diagnosed in routine radiographic examinations, showing a unilocular or multilocular radiolucent lesion with well-defined limits and a thin sclerotic halo. Although OKC clinical and radiographic characteristics are very suggestive, its diagnosis is only confirmed by histopathology¹.

Keratocysts are essential components of the Gorlin-Goltz syndrome (GGS), a rare autosomal dominant disorder with strong penetrance and highly variable expressivity. In addition to the presence of these cysts, GGS is characterized by greater predisposition to multiple basal cell carcinomas (BCCs), skeletal abnormalities, and neuro-ophthalmological disorders. It is a rare multisymptomatic disorder, with varying prevalence, caused by mutations of human patched gene (PTCH-1)².

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This study aimed to review the characteristics of the OKC and GGS, report a case of OKC, and describe the surgical approach used.

A 20-year-old female patient, with vitiligo and a family history of GGS (mother, uncle, two cousins, and three siblings) complained of nasal obstruction on the left side and diplopia. She had a previous medical history of seven surgeries to remove OKCs, without regular postoperative follow-up. She presented with an increase in volume in the middle hemi-third of the face (left) with ipsilateral proptosis and mobility of the adjacent dental elements. Computed tomography (CT) scans showed an extensive isodense mass affecting the entire left maxilla with invasion of the orbital floor, pterygoid process, greater wing of the sphenoid bone and nasal fossa, also compromising the nasal septum and the cribriform plate of the ethmoid bone (Figure 1). Although the lesion was benign, it presented large proportions and covered different areas, thereby an interdisciplinary (head and neck, neurological, and oral and maxillofacial surgery) approach was chosen. After careful evaluation of the CT scans, a bio-model (Figure 1D) was manufactured to plan the osteotomies for

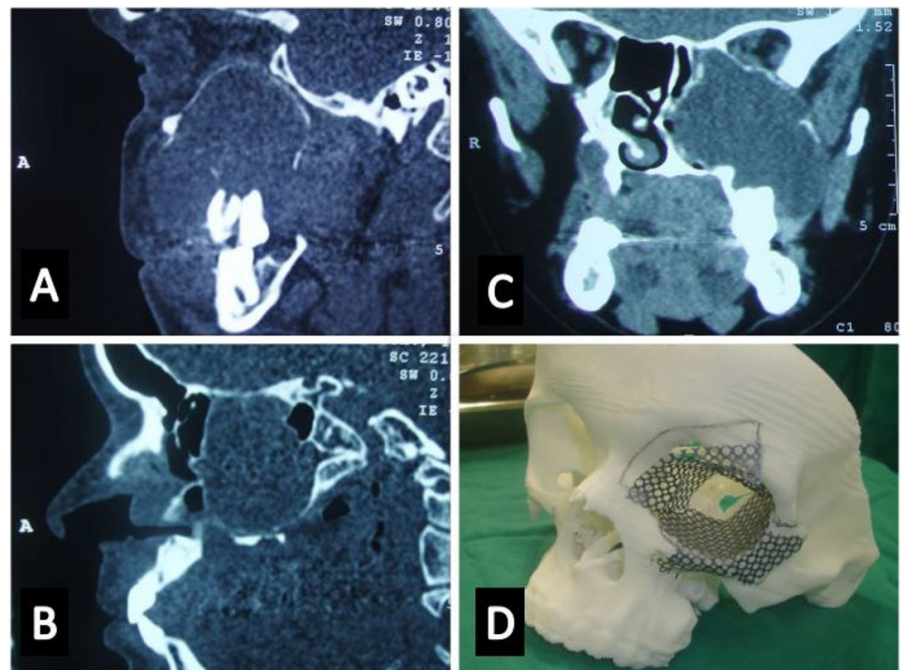


Figure 1. Image exams and bio-models. **A** and **B** - Sagittal sections of computed tomography (CT) scan showing a well-defined invasive isodense mass in the middle hemi-third of the face. **C** - Coronal incision with a lesion causing bone erosion and invading the orbit, nasal cavity, ethmoid sinus, and oral soft tissues. **D** - Bio-model with titanium mesh adapted to the orbital walls for installation in the patient immediately after tumor excision.

resection and immediate reconstruction with titanium mesh, thus providing support to the eyeball after removal of the orbital floor. To this end, a 1.5 mm titanium mesh was modeled on the prototype. The patient received the

necessary clarifications about her condition, signed an informed consent form, and underwent surgical treatment through a craniofacial approach. This approach consisted of a combination of the Weber-Ferguson-Diefenbach and coronal (or bitemporal) incisions. The first access (Figure 2A) enabled excision of the left maxilla that involved almost the entire lesion, followed by installation of the periorbital titanium mesh. Osteotomies were performed using drills, reciprocating saws, and chisels. The neurosurgical approach (Figures 2B, C, and D) consisted of a coronal incision with manufacturing of a pericranial flap, bitemporal craniectomy, and extradural dissection of the

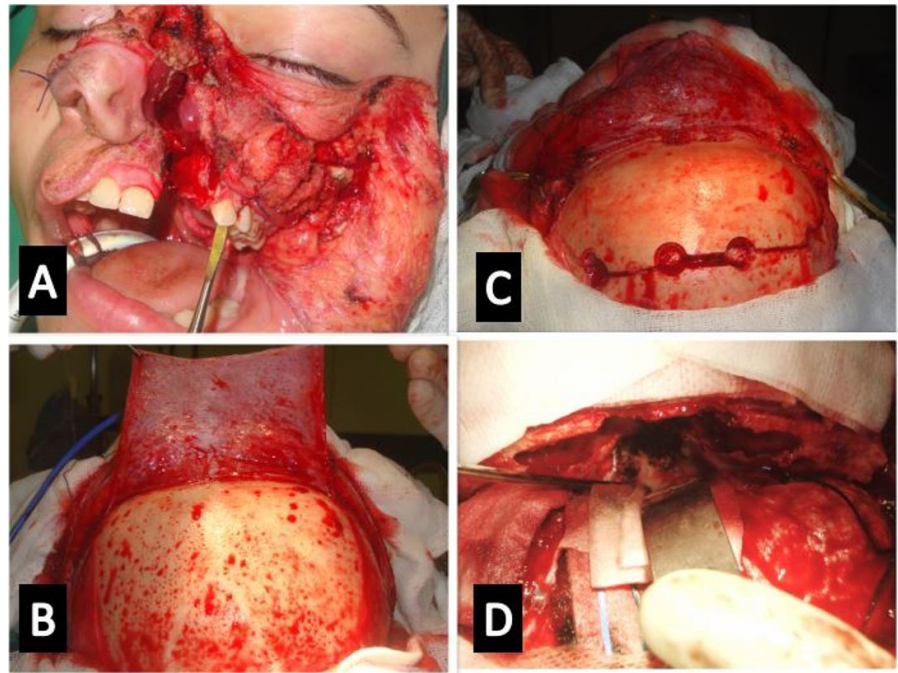


Figure 2. Trans-operative. **A** - Access by Weber-Ferguson-Diefenbach access. **B** - Coronal (bitemporal) soft tissue incision and folding of the pericranium pedicled flap. **C** - Bitemporal craniectomy. **D** - Extradural peeling of the dura mater of the anterior cranial fossa.

anterior skull base in order to view of the crista galli and ethmoid bone, with subsequent excision of the compromised part. After that, the skull base was lined with a pedicled flap, Omnex® surgical sealant (3 mL) was applied to prevent cerebrospinal fluid (CSF) leak, and the dura mater was anchored. Next, cranial reconstruction was performed using bone cement (methacrylate) and four titanium miniplates and screws (Figure 3C). After hemostasis, a subgaleal drain was placed and the soft tissues were closed. At the end of the procedure, the dead space was buffered with lubricated gauze, a custom-made filling prosthesis was installed to occlude the oro-antral communication, and compression dressing was applied. There were no complications during the surgery, and the patient was subsequently sent to the ICU. Postoperatively, she developed CSF rhinorrhea (with spontaneous resolution) and meningitis (properly treated with antibiotic therapy). After three years of follow-up, the patient was diagnosed with a new small lesion in the right maxilla, which was

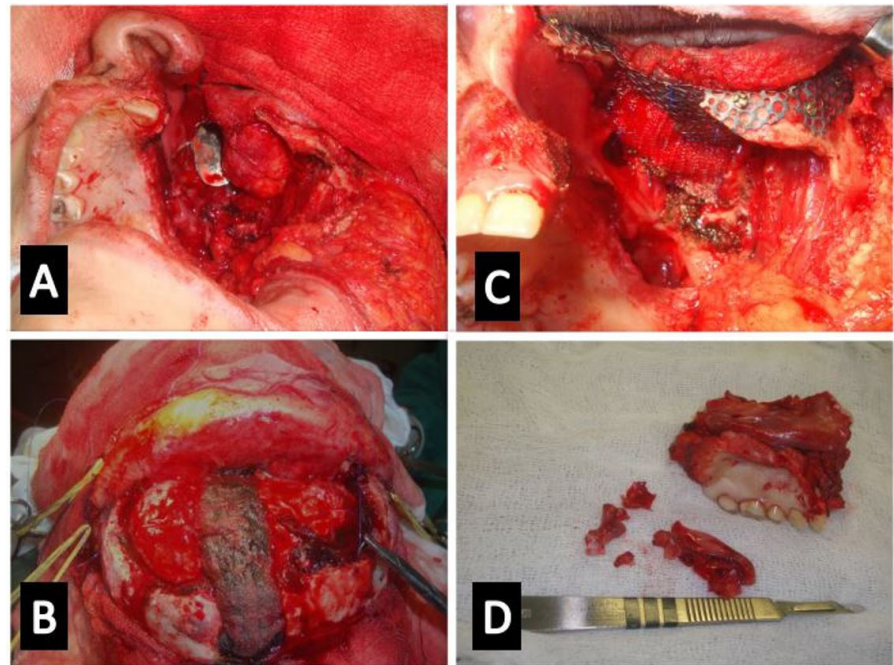


Figure 3. Trans-operative. **A** - Immediate appearance after excision of the left maxilla; **B** - Hemostasis. Dura mater anchorage points on the left and dura mater leaflet of the superior sagittal sinus covered by Surgicel® Fibrillar™ medially can be observed. **C** - Installation of periorbital titanium mesh. **D** - Left maxilla after surgical resection.

early treated. Currently, she has been under regular follow-up for five years, since the craniofacial approach, and has shown no signs of recurrence in the operated area. The suspicion of OKC was confirmed by histopathology.

There has been great controversy in the world scientific literature regarding the classification of odontogenic cysts either as cysts or tumors. Odontogenic cyst was accepted as a neoplastic lesion in the World Health Organization (WHO) 2005 classification, designated as keratocystic odontogenic tumor (KCOT); however, the latest WHO classification (2017) redesignated KCOT in the cyst category³.

One factor that seems to increase the aggressiveness of odontogenic cysts is their association with GGS. They become more invasive, susceptible to relapse, and can affect several quadrants of the jaw bones simultaneously. In addition, odontogenic cyst is related to mutations in the tumor suppressor PTCH-1 gene, which is also responsible for the development of basal cell carcinoma².

There are several techniques for the treatment of odontogenic cyst, namely, enucleation with curettage, enucleation with osteotomy, and bone resection with a safety margin. There are also other conservative techniques, such as marsupialization, cyst decompression followed by enucleation, and cryotherapy. In this case report, resection with a safety margin using a

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craniofacial approach was chosen, as this technique has been suggested as efficient in the literature⁴. Moreover, anterior craniofacial resection is the most common indication for nasossinus tumors⁵.

The surgical approach chosen depends on the anatomical location and degree of invasion of the disease, but usually involves a superior coronal incision combined with an inferior transfacial approach⁴. The case reported here used a combination of the Weber-Ferguson-Diefenbach and coronal (bitemporal) incisions.

OKC is an asymptomatic disease with aggressive potential that affects especially the jaw bones. Resection of the lesion, particularly when associated with GGS, is essential for treatment success and recurrence reduction. The craniofacial approach is presented as a viable alternative for maxillary invasive lesions at an advanced stage.

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