

Short Case Report

A non-syndromic case of maxillo-mandibular keratocysts

P.M. Brechard^{1,2,*}, G. Hervé³, V. Descroix^{1,4}, G. Lescaille^{1,4}, A. Guyon^{1,4}

¹ Département de Chirurgie Orale Service Odontologie [Department of Oral Surgery and Odontology], Hôpital Pitié-Salpêtrière-Paris, Paris, France

² Service d'Anatomo-Pathologie [Pathology Department], Hôpital Pitié-Salpêtrière-Paris, Paris, France

³ UFR Odontologie [UFR Odontology], Université Paris Descartes, Paris, France

⁴ UFR Odontologie [UFR Odontology], Université Paris Diderot, Paris, France

(Received: 23 September 2019, accepted: 27 September 2019)

Keywords:
odontogenic
keratocyst /
non-syndromic

Abstract – Introduction: Odontogenic keratocysts (OKCs) are frequent, aggressive lesions with a strong tendency to recur, particularly in their para-keratinized majority form. Although they are mainly non-syndromic, these lesions are found in a large majority of patients with Gorlin syndrome. Thus, multiple forms are almost always associated with this syndrome and require investigation to prevent the risk of various cancers. Non-syndromic multiple forms are exceptional. **Observation:** A 20-year-old patient presented with dual localization of maxillary and left mandibular OKC at consultation. Under general anesthesia, excision of the lesions and extraction of the impacted wisdom teeth 28 and 38 were performed. The patient showed no clinical sign of Gorlin syndrome. **Discussion:** The OKC or epidermoid cyst is derived from the dental lamina or its remnants and from the basal part of the oral epithelium and represents between 10 and 20% of all cystic lesions in the maxillae. Its peak of incidence is between the second and fourth decade (or earlier in case of association with basal cell necrosis). OKC occurs mainly in the mandible and preferentially at the Ramus, where its frequency can reach 70% depending on the series. **Conclusion:** The management of OKC by oral surgeons must be conducted in a multidisciplinary setting in close collaboration with dermatologists, geneticists, and anatomic pathologists. Due to the strong recidivating character of OKCs, patient monitoring is essential.

Introduction

Odontogenic keratocysts (OKCs) were reclassified as part of the new 2017 WHO classification, as OKCs since 2005 and are no longer classified as benign odontogenic tumors considering their pathogenic cystic processes; therefore, they cannot be considered tumors. Furthermore, it has been demonstrated that keratocyst occurrence in the framework of Gorlin-Goltz syndrome and basal cell nevomatosis is all the more likely because they often affect multiple locations and can manifest early in childhood. Multiple keratocysts have also been reported in certain syndromes, such as oral-facial-digital syndrome, Noonan syndrome, Ehlers-Danlos syndrome, and Simpson-Golabi-Behmel syndrome. Non-syndromic cases are very rare, with only four cases having been reported in the literature [1].

Observation

The present case involves a 20-year-old patient who had no prior medical or surgical history and was referred by a dentist

for the management of two osseous lesions that were discovered by chance following an orthopantomogram. The orthopantomogram showed a radioclear image of the left mandibular angle, as well as a radiopaque image of the left maxillary sinus.

No mandibular or sinusoidal symptomatology was mentioned during the interview. The clinical extraoral examination was unremarkable, and no asymmetry or neurosensitivity disorders affecting either the middle or lower face were found. The intraoral examination revealed that tooth 38 was missing from the arch, and there was a light bluish discoloration caused by transparency through the mucous membrane of the retromolar trigone without any drainage or fistula. Dental vitality testing was positive for non-mobile teeth 36 and 37. The maxillary examination found that tooth 28 was missing from the dental arch without any other abnormalities. Dental sensitivity tests were also positive for the teeth in sector 2.

Dental occlusion was normal. There was no dermal staining.

The sinus scan revealed a mixed lesion that extended throughout the left maxillary sinus, which was defined as a vast radioclear lesion with tooth 28 lodged in the middle meatus area. Furthermore, the scan found a perforation of the sinus cortices on the lateral and posterior walls (Fig. 1).

* Correspondance: pmbrechard@gmail.com



Source : Dr Alice Guyon, 2019.

Fig. 1. Top: Facial scan centered on the included tooth 28. Bottom: Facial scan centered on the included tooth 38.

A scan of the mandibular lesion revealed a 4-cm multilobed radioclear lesion along the major axis, which was homogenous and well delineated with a peripheral border of osteocondensation, with a close connection to the cemento-enamel junction of tooth 38, which was affected by the lesion. The inferior alveolar nerve seemed to be repelled at the lower boundary of the lesion. The vestibular and basilar cortices were not affected; however, the lingual cortex was perforated by the lesion at the level of the mylohyoid muscle insertion (Fig. 1).

The intervention was performed under general anesthesia owing to the patient's wish as well as because of the risk of intraoperative mandibular fracture. The intervention took place under general anesthesia with nasotracheal intubation on the right in the dorsal recumbent position.

During the intervention, the intravenous administration of broad-spectrum antibiotics, namely Augmentin 80 mg/kg, was used for antibiotic prophylaxis. Anti-inflammatory steroids were also administered, namely Solupred 1 mg/kg, to limit postoperative edema of the floor of the mouth. The intervention began with the mandible, the area most likely to experience complications, mainly fractures. The preoperative scan showed that the basilar edge was weakened by the extension of the OKC.

After the administration of adrenaline lidocaine, an incision was made with blade 15 along the rising branch and up to the vestibulo-distal region of tooth 37 with permanent bone contact. A full-thickness flap was repositioned in the vestibular and lingual tissues, thereby exposing the rising branch up to approximately 1 cm from the mandibular crown. This was followed by an initial superior osteotomy using the tungsten carbide round bur under irrigation with frozen saline. The cyst was carefully removed from the walls using sinus curettes without rupturing the cyst and respecting the lower alveolar nerve. Tooth 38 was extracted without an

additional alveolectomy. A detersion of the cavity was performed using a 10 vol. hydrogen peroxide and green betadine mixture and then rinsed thoroughly using a saline solution. The sutures consisted of a single layer of vicryl 3.0 separated into many individual stitches. There was no perioperative sign of fracture (Fig. 2).

After infiltration into the sector 2 vestibule with adrenaline lidocaine, an incision was made with blade 15 in the vestibule at a distance from the necks of tooth 23 to tooth 27. Removal of a full-thickness flap exposing almost the entire left side of the mandible. This specific approach was chosen to increase the extraosseous exposure of the cyst on the lateral part of the sinus using the tungsten carbide round bur at a distance from the apex of tooth 23. The cyst was removed from the walls without rupturing it, and tooth 28 was extracted at the same time. Next, a detersion of the cavity was performed using the same mixture used previously. Finally, the site was closed using a vicryl 3.0 continuous suture (Fig. 2). The surgical specimens were sent for analysis to the pathology laboratory.

The postoperative prescription consisted of a broad-spectrum antibiotic therapy based on amoxicillin/clavulanic acid at 3 g/d for 7 days. Tier I and II analgesics were prescribed: paracetamol/codeine 500/30 mg/cp, as well as prednisolone corticosteroid therapy at 1 mg/kg/day for 4 days. Finally, nasal washes with sterile saline and an anti-inflammatory mometasone spray were given. A strict soft diet for 6 weeks was also prescribed.

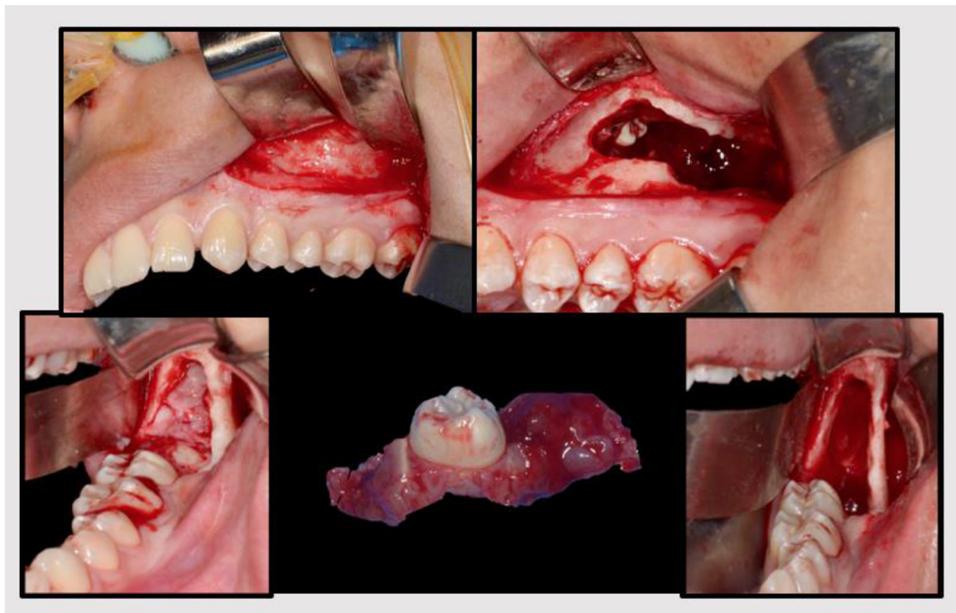
The follow-up was established as follows: a postoperative appointment after 7 days, after 14 days, and then finally after 30 days.

The postoperative sequelae were uncomplicated; the postoperative facial edema had almost disappeared by the 14-day follow-up. In addition, there was a slight hyposensitivity of the territory of the left lower alveolar nerve, which was clearly regressing at the time of the 30-day follow-up (Fig. 3).

Histological results of the pathology analysis found two OKCs. The remote follow-up appointment revealed that the patient no longer suffers from hyposensitivity in the area around the left lower alveolar nerve.

Discussion

OKs or squamous cysts are derived from the dental lamina or its vestiges and from the basal foundation of the oral epithelium [2]. It represents between 10% and 20% of all cystic lesions of the maxilla [3]. This makes it the third most frequent odontogenic cyst after root and dental cysts, according to the WHO classification. Its peak incidence is between the ages of 10 and 30 years (or earlier if associated with basal cell nevomatosis). It primarily affects the mandible and usually affects the ramus with a frequency of up to 70% depending on the course. The maxilla (either anterior or posterior depending on the course) comes in second position.



Source : Dr Alice Guyon, 2019.

Fig. 2. Top: Intraoperative maxillary. Bottom: Clinical view of mandibular keratocyst and the part in contact with tooth 38.



Source : Dr Alice Guyon, 2019.

Fig. 3. Clinical and radiographic follow-up at 6 weeks postoperatively.

In addition, OKCs are characterized by their local aggressive potential with the invasion of neighboring structures, particularly bone and their high recurrence rate after treatment (up to 60% depending on the course of

treatment), which led to its classification as an odontogenic tumor between 2005 and 2007. However, since 2017, keratocystic tumors have again been classified as odontogenic cysts due to their pathogenic cystic process and therefore cannot be classified as tumors [2]. These lesions have strong similarities with other lesions, particularly when the lesion represses a dental element, as was the case here for the two lesions, for which the diagnosis of follicular cyst is part of the differential diagnosis. The polylobate form, although more frequent for ameloblastoma, is also described for OKC. Although frequent in their unique form, multiple OKs are readily associated with syndromes, including Gorlin-Goltz syndrome. It is an inherited disease with autosomal dominant transmission caused by the mutation of the *PTCH14* gene, characterized by, among other elements, multiple OKCs and primordial cysts, dysmorphic syndrome (facial dysmorphism with hypertelorism, spina bifida, brachymetacarpia), mental retardation, and multiple nevi that can degenerate. In this syndrome, OKCs appear early in childhood and can be both numerous and voluminous. Therefore, some authors recommend that a genetic investigation be carried out to identify Gorlin-Goltz syndrome in patients with OKCs before the age of 20, which is when the OKCs are more likely to be numerous and/or when certain elements of the examination or interview point to developmental abnormalities, considering the predisposition to develop different cancers [4]. The genetic diagnostic examination must be considered in the light of arguments in favor of a syndrome. In this patient's case, the general clinical examination did not reveal anything remarkable, and no other disorders other than OKC were present.

Aggressiveness and a higher rate of recurrence are related to the “parakeratinized” nature of these cysts (compared to orthokeratinized cysts). Location seems to play an important role; the recurrence rate is 75% in the mandibular molar region and 31.8% in the mandibular region. The transformation of keratocysts into ameloblastomas is rare, but they are nevertheless more frequent in Gorlin syndrome keratocysts. Very rare cases of degeneration have been described [5].

In this patient, given the dual factors of the location and parakeratotic nature of OKCs, increased surveillance was implemented in relation to the significant risk of recurrences, ranging from 10% to 40% according to the authors [6].

Conclusion

OKCs are commonly managed by oral surgeons, who should consider the possible association of OKCs to a syndromic form of the disorder, which should be evaluated in the event of multiple lesions. Thus, OKC management must be performed in a multidisciplinary setting in close collaboration with dermatologists, geneticists, and pathologists. Due to the high recurrence rate of OKCs, patient follow-up is essential.

Conflicts of interests: The authors declare that they have no conflicts of interest in relation to the publication of this article.

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