Case Report

**Odontogenic Keratocyst of Maxilla Involving the Sinus – OKC to be a Cyst or a Tumour?**

Dr. Jyothi Mahadesh¹, Dr. Kokila², Dr. Laxmidevi B.L³
¹Prof. and H.O.D., ²Reader, ³Senior Lecturer, Department of Oral Pathology and Microbiology, Sri Siddhartha Dental College and Hospital, Tumkur, Karnataka.

**Abstract:**
OKC is an aggressive cystic lesion with high rate of recurrence comparable only to ameloblastoma. In majority of cases, it is located in mandibular posterior region. But it can also be found in the maxilla especially the canine region. We present a case of maxillary okc perforating the sinus and invading the infra-temporal fossa. The patient was operated under general anaesthesia, the lesion was removed and sent for histopathological examination which confirmed the diagnosis of para-keratinized OKC. In the recent past the para-keratinized OKC’s have been considered to be a benign tumour/keratocystic odontogenic tumour (KCOT) or true OKC while the ortho-keratinized OKC as a ortho keratinizing odontogenic cyst (OKOC). Can the clinical aggressive behaviour and infiltrating nature of the present case be construed to be a neoplastic behaviour is a matter of conjecture.

Key words: OKC, KCOT, OKOC

**Introduction:**
Odontogenic keratocyst (OKC) is a benign but locally aggressive developmental odontogenic cyst.¹ It is of great interest to dentists because of its high recurrence rate and its ability to grow to a large size before it manifests in the oral cavity. It is also a good mimic as it can have a variety of clinical appearances.² It is one of the common cysts in the oral cavity with a frequency of 5-11%.³ The term okc was first introduced by Philipsen in 1956. Although the
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The term only indicates the formation of keratin and is non-specific, as keratin formation may be seen even in radicular cysts and dentigerous cysts as a result of metaplasia, it is so well established in literature that it continues to be in use. Subsequently a lot of work on the molecular biology and genetics have been carried out by many researchers and based on which, it has been suggested to be considered as a benign tumour and hence be renamed as keratocystic odontogenic tumour (KCOT).

Case Report:

A 50 years old male patient reported to Sri Siddhartha College of Dental Sciences and Hospital with the chief complaint of swelling and pus discharge in the right side of the face. The swelling was reported to be present since two months.

Extra-orally the swelling extended antero-posteriorly from nose to cheek and superio-inferiorly from infra-orbital margin to upper lip (Fig. 1).

Fig 1: Extra oral clinical photograph showing swelling in the right maxilla.

Expansion of buccal plate was evidenced. The swelling was hard, non-fluctuant with diffuse margins. Intra-orally the swelling extended from right permanent lateral incisor to the second permanent molar. C T Scan showed enlarged sinus. The lesion was expansile and extensive. Posterior wall of the sinus was perforated with extension of the lesion into the infra-temporal fossa (Fig. 2). A clinical diagnosis was made as infected cyst of right maxilla. The left side sinus also showed
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generalised thickening as well as a small growth near the ostium and was thought to be due to inflammation.

Fig 2: Computed tomography showing hypodense lesion in the right maxilla with perforation of the sinus.

An incisional biopsy from the right affected sinus region was done and the tissue was submitted to the department of oral pathology. The gross appearance of the tissue was brownish in colour, soft in consistency and about 2”x3” in size.

The histopathology revealed a cystic epithelium overlying a connective tissue capsule. The epithelium was folded and showed uniform thickness of 5-10 layers except in one or two areas and the basal layer showed darkly stained tall columnar cells. The surface showed para-keratinization with corrugation (Fig.3).

Fig 3: Photomicrograph showing the cystic lining of uniform thickness, typical folding and parakeratinized surface corrugation and inflammation. (H&E,20x)

There was an artifactual separation of the epithelial lining and the capsule which may also suggest a weak junction between the lining and capsule (Fig.4).

Fig 4: Photomicrograph showing artifactual separation of cystic lining and the connective tissue capsule. (H&E,20x)
The capsule also showed chronic inflammatory cell infiltration. The diagnosis of inflammed para-keratinised OKC was made.

An exisional biopsy was done and the cystic lining, bone piece from right maxilla was sent to the department for histo-pathological examination. A diagnosis of para-keratinised OKC was confirmed. The patient was negative for basal cell nevoid syndrome.

Discussion:

OKCs of maxilla have diagnostic difficulties due to lack of specific clinical and radiographic characteristics.\(^1\) They are less common in maxilla than mandible with only 31.3% in maxilla.\(^3,4,7,8\) But when they do occur, they are more common in the canine region which was the case in our patient also. It’s radiographic image in such cases may be misinterpreted as radicular cyst or lateral periodontal cyst.\(^2\) OKC has been shown to have a bimodal age distribution with first peak in 2\(^{nd}\) and 3\(^{rd}\) decade and the second peak in the 5\(^{th}\) decade or older. It is said that the lesions in the second peak are more common in maxilla which corresponded with our case.\(^4\)

In our patient radiographically the lesion was radiolucent, which can be seen in dentigerous cyst, ameloblastoma or odontogenic keratocyst but there was no associated impacted tooth or root resorption, therefore the dentigerous cyst and ameloblastoma were less likely. Maxillary OKC tended to exhibit a unilocular, smooth, round border while mandibular ones had scalloped border.\(^5\) The occurrence of okc in maxilla is relatively rare and invasion of the maxillary sinus is unusual,\(^5\) which was seen in our case.

In the CT Scan the lesion appeared to be aggressive and destructive as seen by the large size, irregular borders, perforation of the sinus and extension of the lesion towards the infra temporal fossa and
bone loss. It is very rare for the maxillary OKC’s to show perforation of the sinus and seems that less than 1% of cases showing this feature.\textsuperscript{6} Buccal cortical plate expansion was noticed which is seen in only one third cases of maxillary cysts.\textsuperscript{4}

In radicular and dentigerous cysts the expansion is mainly due to the pressure exerted by cystic fluid leading to the buccal and lingual cortical plate expansion which is called as the pushing type of growth. Whereas in OKC the extension is more in antero-posterior direction and the pressure of the fluid is quite low and grows by extension rather than by expansion. The extension here is due to reasons like finger-like projections from the cyst wall into the marrow spaces, and enlarges slowly but relentlessly along the path of least resistance.\textsuperscript{3} So, not much of cortical expansion is seen in the initial stages, and by the time it shows clinical swelling the lesion would have been quite huge.

Histopathologically our case showed parakeratinized lining and a capsule which is slightly inflammed with chronic inflammatory cell infiltration. A diagnosis of inflammed para-keratinised OKC was made. Para-keratinization was mentioned in the report as this lining is considered by a few authors as true OKC or a tumour because of its aggressiveness, high recurrence and their tendency to be more associated with the Gorlin-Goltz Syndrome.\textsuperscript{3}

Cawson et al in 2004 have considered OKCs as keratinising cysts and have divided it into para-keratinised and ortho-keratinised linings. They have called para-keratinised cysts as odontogenic keratocysts or keratocystic odontogenic tumor (OKC/KCOT). Ortho-keratinised cysts have been called as orthokeratinised odontogenic cysts (OKOC).\textsuperscript{3} Philipsen and Riechert have suggested that OKC should be considered as a benign tumour and hence be called as KCOT or keratocystic odontogenic
tumour. Shear has countered this argument by saying that even if it is a neoplasm, it is convenient to call it as OKC as many neoplasms do not necessarily have a suffix-oma.4

This debate was started by shear(2003) that OKC should be called as keratocystoma which led to Philipsen and Riechert suggesting keratinising cystic odontogenic tumour in 2004 and then Philipsen suggesting keratocystic odontogenic tumor in 2005. Shear suggested that the term keratocyst was so widely used by clinicians and pathologists that the term should be continued even if it is agreed to be a neoplasm. The neoplastic nature of the lesion when debated in 2006 at The International Association of Oral Pathologists, it was concluded that only the molecular findings were not sufficiently definitive to support the thesis that the lesion was a benign neoplasm. Based on the suggestions given by Toller in 1967 who termed them as benign neoplasm because of their intrinsic growth potential, Alfors et al in 1984 called it as benign cystic neoplasm, which was also considered by shear in 2003.4

Therefore to avoid any confusion we mentioned the diagnosis as inflammed para-keratinized OKC.

The defects in tumor suppressor genes, infiltrative growth pattern, recurrence and high proliferative activity of the epithelium along with rarity of squamous cell carcinoma developing within OKC are thought to be some of the evidences that OKC may be a neoplasm.1,3

Although the genetic theory may be convincing that OKC may be a neoplasm, neoplasms are defined on the basis of their relentless growth and not on their molecular genetics. The OKCs may show relentless growth but respond well to marsupialisation provided all the locules are opened to the surface. This is not a feature of a neoplasm.3 Carolina Cavalieri Gomes et al in their review of the molecular pathogenesis of the okc have
concluded that genetic alterations, epigenetic alterations, miRNA expression need more translational research as advances in one area may improve the overall knowledge related to different tumours. Therefore the debate whether OKC are neoplastic or not still continues. Further studies are required to conclusively call it either as a cyst or a neoplasm.

**Conclusion:**

In conclusion, the present case report emphasizes that any cystic lesion should be evaluated cautiously. Though okc’s are rare in maxilla especially in the 5th decade when radicular cyst would have been a more probable diagnosis, biopsy and CT Scan should be done to evaluate the exact lesion and it’s extension. Because a parakeratinized okc can be associated with Gorlin-Goltz Syndrome, the clinician should properly evaluate the presence of features of the syndrome which was found negative in our case. This case also validates that para-keratinized lesions are aggressive as shown by its generous bone involvement and erosion of the sinus wall. Considering the lesion’s extension towards the infra-temporal fossa, this type of infiltrative growth and not expansion is contingent to be a neoplastic behaviour can be subject to further discussion. Whether the lesion shows recurrence or not will be confirmed by regular follow ups. The current dilemma of the various terminologies of okc is also discussed.

**References:**

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Correspondence:

Dr. Jyothi Mahadesh,
Department of Oral Pathology and Microbiology,
Sri Siddhartha Dental College and Hospital, Tumkur, India.
Email: jyothi.desh@gmail.com