

原文題目(出處)：	Keratocystic odontogenic tumour: Reclassification of the odontogenic keratocyst from cyst to tumour. JCDA 2008;73:165-165h
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內文：

Abstract:



Purpose:

- to review the features and behaviour of the OKC (or KCOT);
- to analyze a series of histologically confirmed KCOT cases;
- to review and discuss the redesignation of KCOT and the implications for treatment



In a case series of 21 patients (27 KCOTs), recurrence rate was 29%, all recurrences occurred within 2 years after intervention.



WHO's reclassification of this lesion from cyst to tumour :

- underscores its aggressive nature
- motivate clinicians to manage the disease in a correspondingly aggressive manner



Most effective treatments are enucleation supplemented with Carnoy's solution, or marsupialization with later cystectomy

Introduction:



Odontogenic keratocyst (OKC) is now designated by the WHO as a keratocystic odontogenic tumour (KCOT) and is defined as "a benign uni- or multicystic, intraosseous tumour of odontogenic origin, with a characteristic lining of parakeratinized stratified squamous epithelium and potential for aggressive, infiltrative behaviour."

Case Series



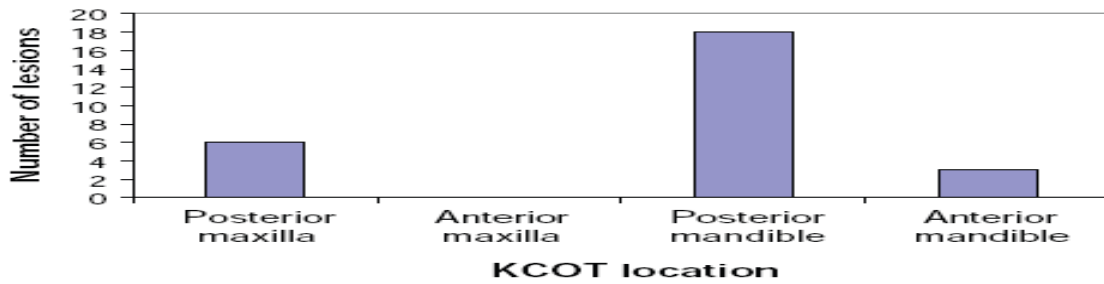
21 patient files on 27 histologically confirmed KCOTs were reviewed



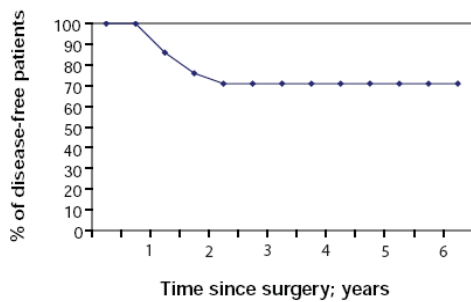
11 recurrences of lesions (5 elsewhere, 6 in author's clinic) and 16 de novo lesions



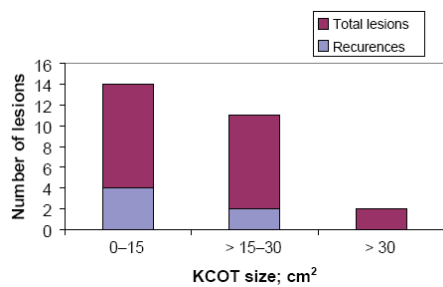
Site: posterior mandible – 18
 anterior mandible -- 3
 posterior maxilla – 6



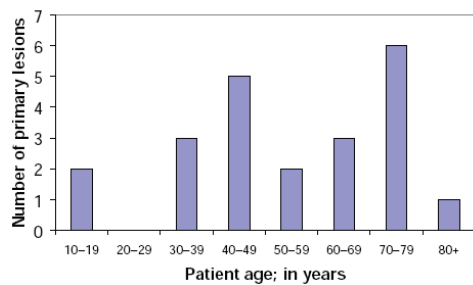
Treatment: enucleation and curettage -- 22
 resection -- 2
 marsupialization -- 3



- Follow up periods: 2 months to 7 years
- Overall recurrence rate: 29%
- All recurrences of lesions treated at author's clinic were within 2 years



- Average surface area of the lesions (radiographically) : 14 cm²
- Most lesions in 0-15 cm² range (greatest number and proportion of recurrences)



- No relation was found between age and number of primary lesions among patient group

Sample Cases

Patient 1 (born 1949, date of surgery: Dec. 16, 1999)

- Recurrence of a KCOT (treated elsewhere 10 years earlier), multilocularity and extent of soft tissue involvement
- Site: right mandible
- Size: 45 cm² radiographically

- ☞ Treatment: resection (complete removal of the right mandible from the condyle to the bone distal to tooth 44)
- ☞ Follow up 6 years: no recurrence

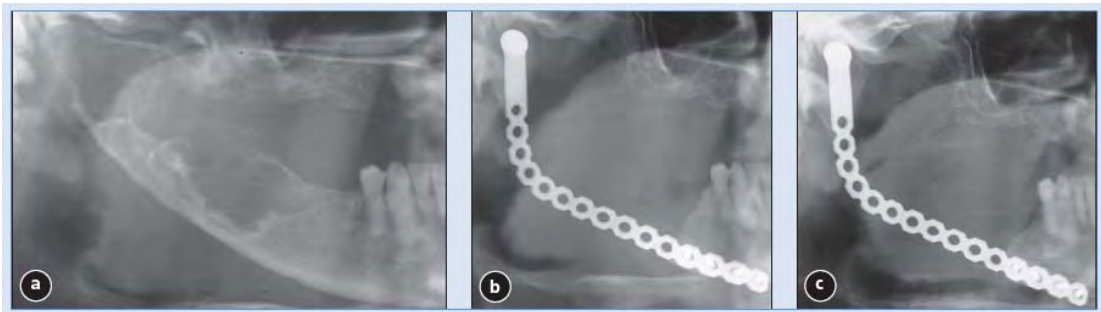


Figure 5: Partial panoramic radiograph taken (a) pre-operatively and (b) 6 days and (c) 6 years after resection.

☞ Patient 2 (born 1925, date of surgery: Nov. 22, 2001)

- ☞ primary KCOT
- ☞ Site: left mandibular ramus
- ☞ Size: 19 cm² radiographically
- ☞ Treatment: marsupialization
- ☞ Follow up 3.5 years: no recurrence

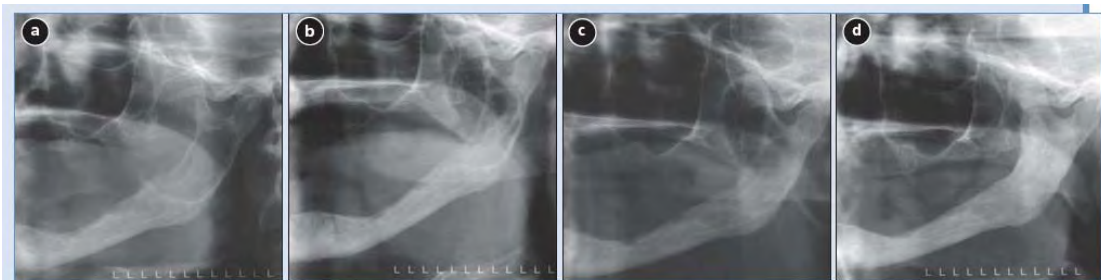


Figure 6: Partial panoramic radiograph taken (a) pre-operatively, (b) 9 days, (c) 3 months and (d) 3.5 years after marsupialization.

☞ Patient 3 (born 1949, date of surgery: Sept. 17, 1993)

- ☞ de novo KCOT
- ☞ Site: 18 cm² radiographically
- ☞ Size: anterior mandible
- ☞ Treatment: curettage
- ☞ Follow up 9 months: recurrence, this was curetted and followed up for 7 years



Figure 7: (a) Pre-operative radiographic appearance of the lesion. (b) Recurrence at 9 months after curettage; and (c) 16 months and (d) 7 years after curettage of the recurring tumour.

Clinical Features

- 👤 11% of all cysts of the jaws
- 👤 Occur most commonly in the mandible, especially in the posterior body and ramus regions
- 👤 Always occur within bone
- 👤 Patients may present with swelling, pain and discharge or may be asymptomatic.
- 👤 Local destruction and a tendency for multiplicity (NBCCS or Gorlin-Goltz syndrome)
- 👤 High recurrence rate, reportedly between 25% and 60% (NBCCS—82%)
- 👤 NBCCS : multiple KCOTs, nevoid basal cell carcinomas, bifid ribs, calcification of the falx cerebri, frontal bossing, multiple epidermoid cysts and medulloblastoma
- 👤 3 mechanisms for KCOT recurrence (Brannon, 1976):
 - 👤 incomplete removal of the cyst lining,
 - 👤 growth of a new KCOT from satellite cysts (or odontogenic rests left behind after surgery)
 - 👤 development of a new KCOT in an adjacent area
- 👤 Recurrence rates: follow-up times, surgical technique used, no. of cases
- 👤 Mostly recurred within 5–7 years → long-term follow-up

Common Treatment Modalities

- 👤 Morgan and colleagues : conservative or aggressive
 - 👤 Conservative treatment :“cyst-oriented”-- enucleation, with or without curettage, or marsupialization
 - Advantage: preservation of anatomical structures (including teeth)
 - 👤 Aggressive treatment: “neoplastic nature”-- peripheral ostectomy, chemical curettage with Carnoy’s solution or en bloc resection
 - Recommended for NBCCS cases, large KCOTs and recurrent lesions
- 👤 Site- and size-based approach to KCOT treatment planning
- 👤 Removal of the mucosa overlying the lesion has been recommended : clusters of epithelial islands and microcysts with the potential to cause recurrence

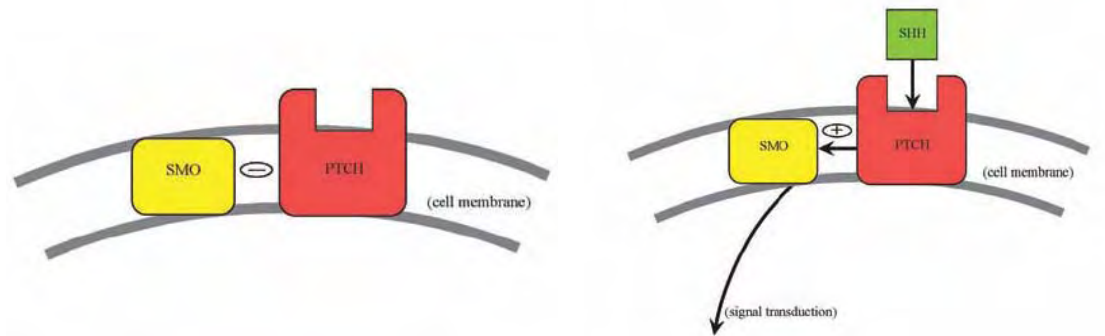
Recurrence

- 👤 Aggressive treatment → relatively low recurrence rates
- 👤 Conservative treatment → more recurrences
- 👤 The most effective treatment option: enucleation of the KCOT and subsequent application of Carnoy’s solution
- 👤 Alternatively, marsupialization followed by cystectomy is likewise effective

KCOT: The Neoplasm

- 👤 Factors that influenced WHO to reclassify KCOT:
 - 👤 Behaviour: KCOT is locally destructive and highly recurrent

- Histopathology: basal layer of the KCOT budding into connective tissue, and mitotic figures are frequently found in the suprabasal layers
- Genetics: PTCH (a tumour suppressor gene, occurs on chromosome 9q22.3-q31) PTCH binding to SMO(oncogene) inhibits growth-signal transduction. SHH binding to PTCH releases this inhibition.



The pathogenesis of NBCCS and sporadic KCOTs involves a “2-hit mechanism”, with allelic loss at 9q22.

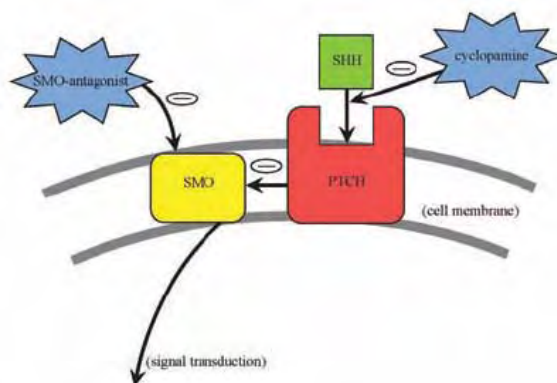
- 1st hit: mutation in one allele, although it can be dominantly inherited, has no phenotypic effect
- 2nd hit: “loss of heterozygosity” (LOH). In KCOTs, leads to the dysregulation of the oncoproteins cyclin D1 and p53.

Implications and the Future of KCOT Treatment

The most appropriate action would be enucleation of the KCOT plus use of Carnoy’s solution or marsupialization followed by enucleation

Lesion size and associated recurrence are inconclusive (Forssell and others found that lesion size does not affect recurrence rate)

Taipale and colleagues, cyclopamine, a plant-based steroidal alkaloid, inhibits the cellular response to the SHH signal



- Zhang and others postulate that antagonists of SHH signalling factors could effectively treat KCOTs (intracystic injection of an SMO protein-antagonist)

Conclusion

- ☞ The aggressive nature of KCOT warrants an aggressive treatment strategy
- ☞ As researches continue, treatment may become molecular in nature→reduce or eliminate the need for aggressive methods

題號	題目
1	Which of the following statements is wrong? (A) OKC may have scalloped outline radiographically (B) The most common location of an OKC is the posterior body of the mandible (C) OKC showed cortical border radiographically (D) The presence of internal keratin increase the radiopacity
答案(D)	出處：Oral Radiology-Principles and Interpretation (5 th edition, pg 394)
題號	題目
2	Which of the following statements about NBCCS is wrong? (A) Prevalence of Gorlin syndrome is estimated to be 1 in 50,000 (B) It is caused by mutations in PTCH, a tumor suppressor gene (C) Multiple basal carcinoma (D) Odontogenic keratocyst
答案(A)	出處：Oral & Maxillofacial Pathology (2 nd edition, pg 598)