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Keratocystic Odontogenic Tumor: A Comprehensive Review

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Abstract

Keratocystic odontogenic tumor (KCOT), previously termed odontogenic keratocyst (OKC), is a unique pathological entity characterized by aggressive behavior, a high recurrence rate, and association with nevoid basal cell carcinoma syndrome (NBCCS). Once classified as a cyst, KCOT has gained tumor status due to its neoplastic potential, distinctive histopathological features, and complex molecular profile. This article provides a detailed overview of KCOT, covering its classification, etiology, clinical presentation, radiographic features, histology, differential diagnosis, treatment options, and recurrence potential. Keratocystic Odontogenic Tumor (KCOT), previously known as odontogenic keratocyst (OKC), is a unique and enigmatic developmental cyst of odontogenic origin that has attracted considerable attention due to its aggressive clinical behavior, high recurrence rate, and controversial classification. Initially described as a cyst, KCOT was reclassified as a tumor by the World Health Organization (WHO) in 2005 owing to its neoplastic characteristics, only to be reclassified again in 2017 as a cystic lesion based on updated molecular and clinical evidence. KCOT is typically found in the posterior mandible of young adults and is often associated with the nevoid basal cell carcinoma syndrome (NBCCS or Gorlin-Goltz syndrome). Its pathogenesis is linked to mutations in the PTCH1 gene, part of the Hedgehog signaling pathway, which underscores its neoplastic potential in syndromic and some sporadic cases. Radiographically, KCOT often presents as a unilocular or multilocular radiolucency with well-defined margins, and histologically, it features a parakeratinized stratified squamous epithelial lining with a palisaded basal layer. Clinically, the lesion may remain asymptomatic or present with swelling, pain, and cortical expansion. Due to its high propensity for recurrence, especially when treated with simple enucleation, various management strategies ranging from conservative (e.g., decompression, marsupialization) to aggressive (e.g., resection, adjunctive therapies like Carnoy's solution or cryotherapy) have been employed.

This comprehensive review aims to explore the historical evolution, pathogenesis, clinical and radiographic features, diagnostic criteria, histopathological profile, treatment modalities, recurrence patterns, and future directions in the understanding and management of KCOT. By integrating the latest evidence from the literature, we seek to provide a nuanced and up-to-date perspective on this complex odontogenic lesion, highlighting the challenges in classification, diagnosis, and treatment planning, and underscoring the importance of long-term follow-up in clinical practice.

Keywords: Keratocystic odontogenic tumor (KCOT); Odontogenic keratocyst (OKC); Jaw cysts; Parakeratinized epithelium; PTCH1 gene mutation; Nevoid basal cell carcinoma syndrome; Gorlin-Goltz syndrome; Odontogenic tumors; Recurrence; Mandibular cysts; Enucleation; Marsupialization

Introduction

Keratocystic odontogenic tumor (KCOT) is a benign but locally aggressive developmental cyst derived from the dental lamina. The World Health Organization (WHO) reclassified odontogenic keratocyst as KCOT in 2005, owing to its tumor-like behavior [1]. Although it has been reclassified again in 2017 under "odontogenic cysts," the term KCOT is still widely used in research due to its clinical implications. KCOT presents a significant clinical challenge due to its aggressive growth, tendency for cortical perforation, and high recurrence rate following treatment [2].

The lesion was first described by Philipsen in 1956 as an odontogenic keratocyst (OKC). The aggressive nature and high recurrence rate prompted the WHO in 2005 to rename it as a keratocystic odontogenic tumor, placing it under odontogenic tumors [3]. However, in the 2017 WHO classification, it was moved back to the cyst category, mainly because molecular evidence of neoplastic transformation was considered insufficient. Nonetheless, understanding it as a potentially aggressive lesion is essential for clinical decision-making. Odontogenic lesions constitute a diverse group of jaw pathologies arising from the tooth-forming apparatus, among which the keratocystic odontogenic tumor (KCOT) stands out due to its distinct clinical and biological behavior [4]. First described by Philipsen in 1956 as the odontogenic keratocyst (OKC), this lesion was redefined by the World Health Organization (WHO) in 2005 as a benign neoplasm and renamed as KCOT in recognition of its aggressive growth, high mitotic activity, and recurrence potential. However, the 2017 WHO classification reversed this stance, reinstating the term "odontogenic keratocyst" and reclassifying the lesion as a cyst, reflecting ongoing debates about its true nature [5]. KCOT most commonly affects individuals in the second to fourth decades of life, with a predilection for the posterior mandible and a slight male predominance [6]. It can present as an incidental radiographic finding or manifest clinically with swelling, pain, tooth displacement, or jaw expansion. In syndromic cases, particularly in association with Gorlin-Goltz syndrome, multiple KCOTs may occur, often alongside other systemic features such as basal cell carcinomas,

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bifid ribs, and skeletal anomalies [7]. The lesion's behaviorcharacterized by satellite cysts, daughter cysts, and potential cortical perforation often complicates complete removal and contributes to a recurrence rate reported to be as high as 60% in some studies. From a histopathological standpoint, KCOT is distinguished by a thin, uniform epithelial lining of parakeratinized stratified squamous epithelium, a palisaded basal cell layer, and a corrugated surface. The lesion often lacks significant inflammatory infiltrate unless secondarily infected. Molecular studies have implicated aberrations in the Hedgehog signaling pathway, especially PTCH1 mutations, further supporting the view of KCOT as more than a simple cyst. These findings have spurred interest in targeted therapies and non-surgical treatment avenues, although surgical excision remains the mainstay of management [8].

Given the lesion's clinical variability, histological uniqueness, and therapeutic challenges, KCOT continues to be a subject of intense research and discussion. This review seeks to consolidate the current understanding of KCOT, tracing its historical classification changes, examining the etiopathogenesis, detailing diagnostic approaches, and critically appraising the spectrum of treatment modalities and their outcomes. Emphasis is placed on the importance of individualized treatment planning, considering lesion size, location, patient age, and recurrence risk, along with the need for vigilant long-term surveillance to prevent recurrence and complications.

Etiopathogenesis

KCOT arises from the remnants of the dental lamina (rests of Serres). The proliferative activity of the epithelial lining is responsible for its growth.

PTCH1 gene mutations, associated with both syndromic and sporadic cases.

Sonic Hedgehog (SHH) pathway, dysregulation leads to increased epithelial proliferation.

Ki-67 and p53, Overexpression of these markers suggests high proliferative potential.

KCOT is a key feature of Nevoid Basal Cell Carcinoma Syndrome (Gorlin-Goltz syndrome), characterized by,

- Multiple KCOTs
- Basal cell carcinomas
- Bifid ribs
- Intracranial calcifications

Clinical features

- Age, typically presents in the 2nd to 4th decades of life.
- Gender, slight male predominance.
- Location, most common in the posterior mandible, particularly the angle and ramus region.
- Often asymptomatic
- Swelling
- Pain or discomfort
- Tooth displacement
- Rarely, pathologic fracture or drainage
- Unilocular in smaller lesions
- Multilocular in larger, more aggressive lesions
- Tendency to grow anteroposteriorly within the medullary bone
- Minimal buccolingual expansion initially
- Frequently associated with impacted teeth
- Panoramic Radiograph (OPG)
- Cone Beam CT (CBCT), Reveals cortical perforation and soft tissue involvement

Histopathology

KCOT has a characteristic histological appearance,

- Lumen filled with keratinaceous debris
- Epithelial lining, 6-10 cell layers thick, parakeratinized stratified squamous epithelium
- Corrugated surface of parakeratin
- Palisaded, hyperchromatic basal cell layer
- Lack of rete ridges
- Satellite (daughter) cysts in the fibrous wall

Differential diagnosis

- Dentigerous cyst
- Ameloblastoma
- Radicular cyst
- Lateral periodontal cyst
- Residual cyst

Treatment and Management

- Conservative Approaches
- Enucleation and curettage
- Useful for large lesions to reduce size before enucleation
- Considered for recurrent or multilocular lesions
- Peripheral ostectomy and chemical cauterization
- Use of Carnoy's solution to fix residual cyst lining

KCOT is notorious for its high recurrence rate, ranging from 10% to 30%, and even higher in syndromic cases.

Reasons for recurrence

- Incomplete removal
- Presence of satellite cysts
- Thin, friable lining difficult to enucleate entirely
- Long-term follow-up for 5–10 years is essential
- Periodic radiographs and clinical evaluation

Conclusion

Keratocystic odontogenic tumor, though benign, exhibits neoplasm-like behavior with a high rate of recurrence. Proper diagnosis, complete removal, and long-term follow-up are crucial. While the terminology may shift between "tumor" and "cyst," the clinical approach should

remain cautious and guided by the lesion's biological behavior and radiographic features.

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