CASE REPORT

Keratocystic odontogenic tumor

Anjani Kumar Jha1, Nonitha S2, Tejavathi Nagaraj3, Hema Keswani2, Sarvesh Vijay2

1Department of Dentistry, Narayan Medical College & Hospital, Jamuhar, Rohtas, Bihar, India, 2Department of Oral and Maxillofacial Pathology, Sri Rajiv Gandhi College of Dental Sciences and Hospital, Bengaluru, Karnataka, India, 3Department of Oral Medicine and Radiology, Sri Rajiv Gandhi College of Dental Sciences and Hospital, Bengaluru, Karnataka, India

Abstract

Odontogenic cysts are considered as non-neoplastic benign lesions. Odontogenic keratocyst is a cystic lesion of tooth origin with an intrusive clinical behavior involving an elevated recurrence rate. It has been renamed to keratocystic odontogenic tumor (KCOT), as it explains its tumor nature which is distinguished by stratified squamous epithelium with parakeratinization and a prospective for destruction, impregnating behavior, and for the possibility for malignant transformation in the wall of the lesion. Histologic examination is necessary for diagnosis as the clinician find it difficult to manage the lesion since the true nature of the lesion is not revealed. This article is an attempt to extend an outline of a diverse strand of KCOT.

Keywords:
Benign tumor, odontogenic tumor, PTCH, satellite cyst

Introduction

Originally recounted by Philipsen in 1956, the odontogenic keratocyst (OKC) is now nominated by the World Health Organization (WHO) as a keratocystic odontogenic tumor (KCOT) and is defined as “a benign uni- or multi-cystic, intraosseous tumor of odontogenic origin, with a characteristic lining of parakeratinized stratified squamous epithelium and potential for aggressive, infiltrative behavior.”[1] WHO “endorse the title KCOT, as it imitates its tumor nature.” KCOT possibly contemplate as a benign tumor rather than a traditional cyst depending on its clinical behavior (Toller 1967).[2]

Ahlfors et al. proposed that “if OKC was identified as an actual, benign cystic epithelial tumor, the query of advanced treatment modalities has to be answered.”[3] Harring et al. designated this cystic lesion by declaring that “After more than 30 years of research, the aspects of KCOT in terms of histogenesis, pathogenesis, histology, high recurrence rate, and malignant possibility are still being puzzled.”[4] The WHO reclassified OKC as a tumor.

• Behavior: As proposed before, the KCOT is regionally catastrophic and eminently recurrent in nature.
• Histopathology: KCOT is characterized by budding of basal layer into connective tissue and mitotic figures in the suprabasal layers.[5]
• Genetics: PTCH (“patched”), a tumor suppressor gene intricate in both nevoid basal cell carcinomas (NBCCS) and sporadic KCOTs, usually diverse a receptor complex with oncogene SMO (“smoothened”) for the SHH (“sonic hedgehog”) ligand. PTCH attaching to SMO obstructs growth-signal transduction. The bond between SHH and PTCH releases this inhibition. If the normal functioning of PTCH is off-track, the proliferation-stimulating effects of SMO are allowed to prevail.[6]

KCOT is a benign growth of odontogenic origin with a probable destructive and penetrating behavior. It is most predilected in mandible and manifests a unilocular, round, oval, scalloped radiolucent area, while huge lesions may show multilocular areas.[7] A prime feature of KCOT is its proclivity to spread in an anterior to posterior direction.[2] The aggressive behavior of KCOT to the extent that they impale cortical bone and involved surrounding soft tissues have been reported. According to (Browne, 1971), he showed that after the removal of KCOT the recurrence rate with satellite cysts was 23.7% and those without satellite cysts was 24.4%.[2]

Possible reasons for recurrence are:
1. Insufficient eviction of the native cyst’s lining.
2. Spread of a new lesion from tiny satellite cysts of odontogenic epithelial rests left behind after the surgical treatment.
3. Evolvement of an unrelated KCOT in an adjoining radius of the jaws, which is interpreted as a reappearance.[8]

Case Report

A 32-year-male came with a swelling on the right side of the face since 8 months. On extra-oral assessment, facial asymmetry was
observed toward the right side of mandible [Figure 1]. Intra-oral swelling was present on the right lower vestibular area measuring around 1.5 cm × 3 cm with obliteration. On palpation, the swelling was soft to firm in consistency. Provisional diagnosis was given as KCOT with differential diagnosis of ameloblastoma.

Panoramic radiograph revealed radiolucency with respect to 45 extending till angle and condylar region of the mandible. The margins were sclerotic [Figure 2]. An excisional biopsy specimen was consigned for histopathological evaluation [Figure 3].

**Histopathological findings**

The H and E stained soft tissue section shows cystic, corrugated parakeratinized stratified squamous epithelium of 6–8 layer thickness with characteristic basal cell palisaded arrangement. The capsular connective tissue is mainly composed of fibrous tissue. Separation of epithelium from connective tissue is seen at the focal area. Scanty inflammatory infiltrate is also seen in the capsular tissue wall [Figures 4 and 5]. The presence of satellite cysts was noted in few areas [Figure 6].

Based on clinical, radiographical and histopathological findings, the final diagnosis was concluded as KCOT.

**Discussion**

Philipsen in 1956 was the first person to give the description of OKC histologically for all the cysts that showed keratinization. Pindborg and Hansen 1963 concluded the histological criteria, which were later confirmed by Browne in the year 1970 and 1971. KCOTs incorporate almost 11% of all cystic lesion of the jaws. They appear frequently in the lower jaw region, specifically in the posterior body and ramus.[2] The case reported here also shows the same site predilection. KCOT as a matter of course generally effect bone, but a few cases with peripheral KCOT have also been published in the English literature. Patients may complain of swelling, pain, and discharge or may be asymptomatic in nature[9] whereas our patient reported with asymptomatic swelling. KCOT has many differentiating clinical and histologic features which includes: (i) Prospect for provincially devastative manner; (ii)
comparatively more recurrence rate, and (iii) titled as a undeviating finding in the NBCCS syndrome, or Gorlin syndrome. It is most frequently reported in the second, third, and fourth decades of life at the posterior lower jaw region in case of male patients. The features of the reported case are also in concurrence with the previous reported cases. The radiographic finding of KCOT includes unilocular or multilocular lesion. Tiny unilocular cystic lesions can be startled with periapical, dentigerous, lateral periodontal cysts or gingival cysts, and huge unilocular KCOT can resemble ameloblastoma. A unilocular KCOT appears as a radiolucent lesion with well-defined margins. Resorption of root, eviction of erupted tooth or migration of impacted erupted teeth may be obvious.\(^{26}\)

On microscopic assessment, KCOT shows invariable parakeratinized epithelium with corrugation, thick cells showing a flat basal surface lining called the fibrous wall. The cystic chamber is bounded with a slim coating of connective tissue roofed by stratified squamous epithelium which is either orthokeratinized or parakeratinized in nature.\(^{5,8}\)

Morgan et al., 2005 classified surgical regimen for KCOT as moderate or assertive. The moderate management is "cyst-oriented" and thus comprises enucleation, with or without curettage, or marsupialization. The convenience is maintenance of anatomy. Assertive treatment approaches the "tumor nature" of the KCOT and constitutes peripheral ostectomy, chemical curettage with Carnoy’s solution or en bloc resection. As per literature review, the recurrence rate is proportionately low with assertive management, whereas more moderate method gravitate to outcome high recurrence rate.\(^{10}\)

**Clinical significance**

KCOTs have a more reversion rate, observed ranging from 25% to 60%.\(^{5}\) The Angle of mandible and Ramus are the areas in the mandible where vision is compromised at the operative field because of its anatomical site, which results in the incomplete removal of tumor, which causes subsequent recurrences.\(^{9}\) Hence, the proper diagnosis and management of KCOT are necessary.

**References**


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