The Keratocystic Odontogenic Tumor

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KEYWORDS

• Keratocystic odontogenic tumor • Odontogenic keratocyst • Enucleation • Radiolucency

KEY POINTS

- In 2005, the World Health Organization renamed the lesion previously known as an odontogenic keratocyst as the keratocystic odontogenic tumor.
- The clinical features associated with the keratocystic odontogenic tumor show it to be a unilocular or multilocular radiolucency, occurring most frequently in the posterior mandible.
- These tumors are normally diagnosed histologically from a sample of the lining.
- With simple enucleation, it seems that the recurrence rate may be from 25% to 60%.

In 2005, the World Health Organization renamed the lesion previously known as an odontogenic keratocyst as the keratocystic odontogenic tumor (KOT or KCOT).[1,2] The term odontogenic keratocyst was first used by Philipson in 1956[3] and its clinical and histologic features were confirmed by Browne in 1970 and 1971.[4,5] At that time, it was believed to be a benign, but potentially aggressive and recurrent, odontogenic cyst, and probably represented the lesion previously termed a primordial cyst.[6] Although most of these cysts were lined by parakeratinized epithelium, a few were orthokeratinized. Over the years, it has generally been agreed that the orthokeratinized versions have a lower incidence of recurrence than the parakeratinized version. As initially described, it was believed that the primitive nature of the epithelium may have a premalignant potential,[7] but this is now believed not to be true, and the incidence of malignant transformation is probably extremely low,[8] if it exists at all.

However, since its designation, some have believed that although it was designated as an odontogenic cyst, the lesion behaved more like a tumor.[9–11] The reasons for this belief include its clinical behavior, with a high recurrence rate after simple enucleation, the histologic appearance, and, more recently, the presence of tumor markers within the cyst. These markers consist of specifically proliferating cell nuclear antigen (PCNA), Ki67, BCE 2 sequence of the enzyme dihydrolipoyl acetyltransferase, matrix metalloproteinase (MMP) 2 and 9, and p53.[12–14] This combination of features led to the 2005 reclassification of this lesion, although a PubMed search of articles published since 2005 found that the lesion is still mostly referred to as an odontogenic keratocyst.[15] Even the term KCOT refers only to the parakeratinized version of the odontogenic keratocyst, and this leaves the orthokeratinized version of the cyst without a new designation. Until further reclassification, these orthokeratinized cysts are grouped with other benign odontogenic cysts.

CAUSE

It is generally believed that these lesions originate from remnants of the dental lamina in the same way as the primordial cyst.[6] However, a tooth is generally not missing and, therefore, they are believed to originate from additional remnants of the lamina not involved in tooth formation. Alternatively, in some cases they may arise from the oral mucosa, particularly in the retromolar region, because daughter cysts are found between the oral mucosa and the cyst in the retromolar

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area.[16] Therefore, there may be 2 possible sites of origin of this lesion (**Fig. 1**).

CLINICAL FEATURES

The clinical features associated with the KCOT show it to be a unilocular or multilocular radiolucency, occurring most frequently in the posterior mandible (the same site as the primordial cyst). It may or may not be associated with a missing tooth (usually not) (**Fig. 2**). Expansion of the buccal and lingual plates occurs late with this lesion (in contrast to the ameloblastoma), because it primarily tends to invade the marrow. However, it does cause some expansion of the lingual plate and can cause lingual plate perforation (**Fig. 3**). Inferior alveolar nerve involvement occurs late. Clinically, the lesion has a high recurrence potential if purely enucleated. Reports in the literature vary, but can show a recurrence rate of from 25% to 60% after local enucleation.[17–21] The reasons for this recurrence rate are believed to be 3-fold:

- They have a thin lining, which is friable, and portions are easily left behind.
- Daughter cysts occur beyond the visible margin of the lesion.
- Some of these lesions may originate from the oral mucosa and daughter cysts are seen between the oral mucosa and the cyst itself. Unless these lesions are removed, recurrence is likely (**Fig. 4**).

The basal cell nevus syndrome (also called Gorlin syndrome or Gorlin-Goltz syndrome) is a genetic condition with an autosomal-dominant inheritance pattern that includes a triad of KCOTs of the jaws,

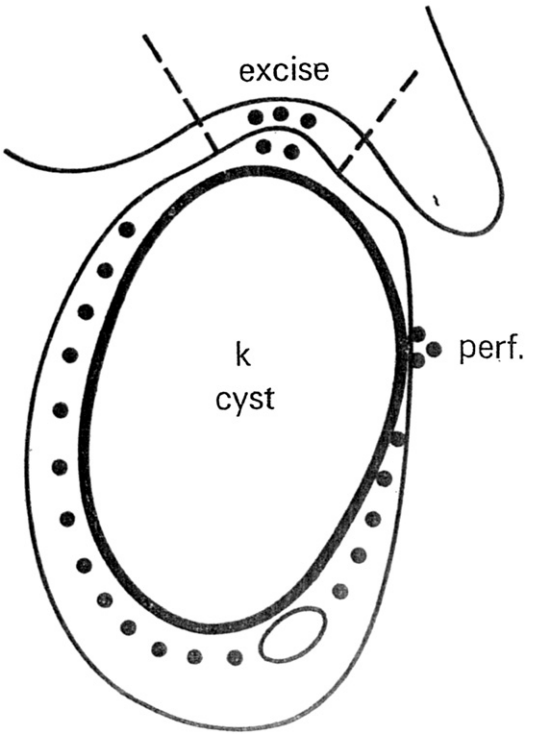

Fig. 1. KCOT with daughter cyst beyond the lining of the cyst and daughter cysts between the cyst and the alveolar mucosa. The area of alveolar mucosa that should be excised with the lesion is indicated. There is also a lingual perforation with associated daughter cysts. (*Adapted from* Bradley PF, Fisher AD. The cryosurgery of bone, an experimental and clinical assessment. Br J Oral Surg 1975;13:122; with permission.)

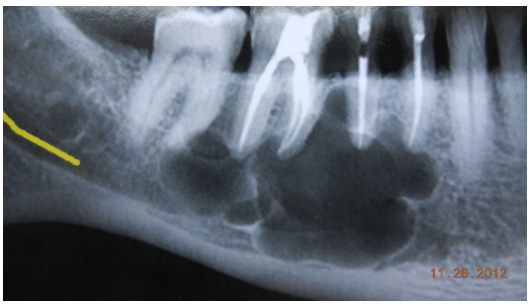

Fig. 2. A multilocular, multicystic KCOT of the right mandible not associated with a missing tooth. The complexity of the lesion contributes to difficulty in total removal.

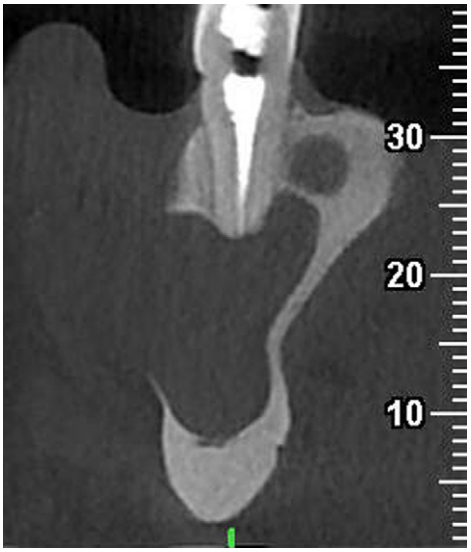

Fig. 3. A coronal cone-beam computed tomography scan showing a multilocular, multicystic KCOT, with lingual perforation and the inferior alveolar nerve embedded in the base of the lesion.