Keratocystic odontogenic tumor with ossification and calcification: A case report with unusual histological findings

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ABSTRACT

The keratocystic odontogenic tumor (KCOT), formerly known as odontogenic keratocyst, is a benign developmental odontogenic tumor with many distinguishing clinical and histologic features. Hard tissue deposits, which usually take the form of dystrophic calcifications, cartilaginous tissue, or dentinoid, are uncommon findings in the connective tissue capsule of the KCOT. We report a case of a 33-year-old female with KCOT showing osseous tissue and calcified deposits close to its epithelial lining, which is an extremely rare occurrence. A brief review on the reported prevalence of hard tissue deposits in KCOTs and possible mechanisms that has been implicated in mineralization and bone formation has been discussed.

Key words: Calcifications, keratocystic odontogenic tumor, ossifications

CASE REPORT

A 33-year-old female reported with a complaint of an inability to open her mouth and pain in the left side of the lower jaw for 15 days. Extraoral examination revealed tenderness on palpation over the ramus of the left side of the mandible. Intraoral examination also revealed restricted mouth opening without any facial asymmetry [Figure 1]. Radiological investigations were significant as the orthopantomogram revealed a large radiolucency with sclerotic margins associated with the left mandibular first molar extending up to the condyle [Figure 2]. A provisional diagnosis of KCOT was made, and the lesion was excised under general anesthesia.

The gross specimen consisted of multiple gray-colored soft tissue bits that appeared like cystic linings; they were firm in consistency with largest bit measuring 1.9 cm × 0.9 cm × 0.2 cm in dimension [Figure 3].
Microscopic examination revealed a squamous parakeratinized corrugated epithelial lining which was approximately 8–10 cell layers in thickness showing basal cell palisading and hyperplasia, reverse polarity, and no evidence of rete ridges [Figure 4]. The surrounding connective tissue capsule was fibrocellular with blood vessels and dense infiltrate of chronic inflammatory cells in some areas. Areas of bone and dystrophic calcification were seen close to the epithelial lining [Figures 5-7]. The overall microscopic features were suggestive of KCOT with ossification and calcification.

DISCUSSION

The KCOT is a well-known pathologic lesion of the jaws that arises from the remnants of the dental lamina. KCOTs may also arise from the extension of basal cells from the overlying oral epithelium. The histologic features of KCOT are highly specific. These features include stratified squamous parakeratinized corrugated epithelial lining devoid of rete ridges. There is a well-defined, palisaded basal layer consisting of columnar cells with intensely basophilic nuclei that tend to be oriented away from the basement membrane. Cystic cavities often show desquamated keratin.

The fibrous tissue capsule is characteristically thin although infected cyst or presence of satellite cysts may show thickenings of capsule. Hyalinization of fibrous tissue is seen in 10% of the cases. However, hard tissue deposits, which usually take the form of dystrophic calcifications, cartilaginous tissue, or dentinoid, are uncommon findings in the connective tissue capsule of the primary KCOT.

Ng and Siar noted scattered irregular eosinophilic masses with evidence of tubule formation and calcospherite type of mineralization. The masses stained positive for collagen (Massons trichrome and Van Gieson) but were negative for amyloid (Congo red). Similar histological findings were reported by Shetty and Srilatha and Naveen et al. who suggested that inductive changes with
A case of KCOT with mural cartilaginous metaplasia was reported by Fornatora et al. who found foci of cartilaginous tissue in areas immediately subjacent to the epithelial lining.\(^9\) Histologic findings of the present lesion showed the presence of bone which was also noted adjacent to the epithelial lining.

The pathologic mechanism of bone formation so close to the epithelium despite the presence of inflammation is unknown. Epithelial elements of odontogenic tumors actively express bone sialoprotein (BSP) which may play an important role in tumor formation and differentiation with respect to pathological calcification. BSP is synthesized and secreted by bone, dentin, and cementum-forming cells and has been implicated in \textit{de novo} bone formation and mineralization.\(^{10}\) Studies by Malaval et al. demonstrated that BSP (gene) knockout in mice displayed thinner cortical bones than the wild type mice models and suggested that BSP deficiency impairs bone growth and mineralization, with dramatically reduced bone formation.\(^{11}\) Expression of the BSP gene in other odontogenic tumors such as ameloblastomas is consistent with the expression of BSP by the enamel epithelium and also with the expression of BSP by neoplastic tissues, which could play a possible role in tumorigenesis.\(^{12}\) Osseous metaplasia could be another possible explanation for the presence of bone in the fibrous tissue wall of KCOTs.

Areas of dystrophic calcifications were also seen in the present case. These are caused by degeneration as a result of necrobiosis or a foreign body reaction. A high incidence of crystalline calcium phosphates, hydroxyapatite and whitlockite, and inorganic phosphates that is found in the aspirated fluid of the KCOTs may be responsible for an increased frequency of calcified deposits in the walls of these lesions.\(^4\)

The significance of calcification and osseous tissue formation in the biologic behavior of KCOTs is not clear. Few studies have shown that primary nonrecurrent KCOTs showed a slightly higher prevalence for dystrophic calcifications than recurrent KCOTs.\(^1\)

**CONCLUSION**

A case of KCOT with bone formation and dystrophic calcifications is presented. Expression of BSP by the odontogenic epithelial component is a favorable explanation for the presence of osseous tissue in the fibrous tissue wall of KCOTs, which is an extremely rare occurrence. The significance of the presence of osseous tissue in the biologic behavior and prognosis of KCOT still need to be investigated.

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Conflicts of interest
There are no conflicts of interest.

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