FOLLICULAR ODONTOGENIC KERATOCYST MIMICKING DENTIGEROUS CYST-REPORT OF TWO CASES

S Hemavathy*, Swati Roy*

ABSTRACT

Odontogenic keratocyst (OKC) is one of the most aggressive odontogenic cysts owing to its relatively high recurrence rate and its tendency to invade adjacent tissue. Radiographically odontogenic keratocyst can be of different types – follicular, envelopmental, replacemental, extraneous and collateral. The biological behaviour, prognosis, recurrence and therapeutic approaches vary in different studies. Further research is necessary in order to establish relationship between markers of proliferation and aggressiveness with regard to the radiographic varieties in OKC. The present article reports 2 cases of follicular odontogenic keratocyst that was submitted to our department with the provisional diagnosis of dentigerous cyst.

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KEY WORDS: Odontogenic keratocyst; follicular odontogenic keratocyst.

* Department of Oral and Maxillofacial Pathology, Government Dental College and Research Institute, Fort, Bangalore, Karnataka, India.

INTRODUCTION:

The odontogenic keratocyst (OKC) was first described by Hans Philipsen in 1956, in a publication written in the Danish language, however it was only a few years later that this lesion became more widely recognised.1

OKC is a developmental odontogenic cyst with specific histopathological features and clinical behavior. Previously called primordial cyst, it may occur in place of a tooth where cystic degeneration of enamel organ epithelium may occur before the development of dental hard tissues. They account for about 3 to 11% of all odontogenic cysts.2

The histologic features are characterized by the presence of a thin band like parakeratinized or orthokeratinized stratified squamous epithelium, with a prominent basal layer composed of either columnar or cuboidal cells, and connective tissue wall that is usually free of inflammation.3

The OKC is one of the most aggressive odontogenic cysts owing to its relatively high recurrence rate and its tendency to invade adjacent tissue. The recurrence rate varies from approximately 20% to 62%. Some clinical and molecular studies showed that the parakeratinized and orthokeratinized OKCs were significantly different in molecular area as well as the recurrence rate; orthokeratinized OKCs (case 1) had a lower recurrence rate than the parakeratinized OKCs. (Case 2) However the relevance of the histology is not clear with respect to the behavior of both entities.3

Because of the aggressive growth pattern and neoplastic nature of the OKC, it is now designated by the World Health Organization (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.”4

Several theories of the development of the OKC have been presented. According to one theory, OKCs develop from the remains of the dental lamina. According to other theories, OKCs emanate from the basal cell layer of the oral mucosal epithelium.5

Thus radiographically odontogenic keratocyst can be of different varieties- follicular, envelopmental, replacemental, extraneous and collateral.6, 7

The follicular type of odontogenic keratocyst may be defined as a cyst with typical histology of an OKC which surrounds the crown of an unerupted tooth and is attached to the neck of the tooth. On radiography it mimics a dentigerous cyst.7, 8 Follicular OKCs are relatively uncommon. They account for 25 to 40% of all the OKCs.9 The OKC associated with an impacted tooth appears to arise at a younger age and grow more rapidly than a cyst unrelated to an impacted tooth. An impacted tooth and its dental follicle may affect the occurrence and proliferation of an adjacent OKC.10 On the other hand there are studies documenting no difference in the nature of follicular and extrafollicular OKC.

Hereby we are reporting two cases of follicular odontogenic...
keratocyst which came to our department with a provisional diagnosis of dentigerous cyst. The aim behind bringing about these two cases is to help the clinician in modifying the final treatment plan after the final diagnosis is achieved as OKCs (in contrast to dentigerous cysts) are potentially aggressive lesions with high recurrence rates so patients should be put up on regular follow up.

First case:

A 14-year-old male patient reported with swelling and pain in the maxillary anterior region since 1 year. The swelling had gradually increased in size. Intraoral examination revealed, obliteration of the right mucobuccal fold extending from 11 to 15. 13 was missing while 53 was retained. The other normal complement of teeth was present. Orthopantomogram revealed an impacted 13, coronally surrounded by a unicocular radiolucency. A provisional diagnosis of dentigerous cyst was made. Fluid was aspirated and a straw colored fluid recovered. The aspirated fluid was sent for biochemical investigation for protein level and was found to be less than 4.0 g per 100 ml, thus suggesting a keratinizing cyst. Surgical enucleation of the cyst was done. The enucleated specimen was send to our department in 10% formalin. The gross specimen revealed a cystic bag enveloping the crown of 13 and attached to the neck of the tooth (Figure 1a, 1b). A part of the lining was taken and subjected for processing. On microscopic examination, the specimen revealed a cystic lining of uniform thickness (6-8 cell layer thickness) with corrugated orthokeratinized stratified squamous epithelium with palisaded polarized basal layer of cell giving a picket fence appearance. Few areas of the lining epithelium exhibited characteristic parakeratinization. The connective tissue wall was made up of young to mature collagen fibers arranged radially to the epithelium and with few interspersed plump fibroblasts and mild inflammatory cell infiltrate (Figure 2a, 2b). Based on these finding diagnosis of odontogenic keratocyst was made.

Second Case:

A 25 year-old male patient reported with swelling and pain in the mandibular left posterior region since 1 week. Intraoral examination revealed obliteration of the right mucobuccal fold extending from 45 to 47. 48 was missing. Aspirate was taken and a straw colored fluid
recovered. The protein level of the fluid was less than 4 mg per 100 ml. Orthopantomogram revealed an impacted 48, coronally surrounded by a unilocular radiolucency. A provisional diagnosis of dentigerous cyst was made. The size of the lesion was 3cm x 2cm. Surgical enucleation of the cyst was done but in this case the cystic lining could not be enucleated in toto and was fragmented. The excised specimen was send to our department in 10% formalin. The gross specimen revealed fragmented cystic bag enveloping the crown of 48 and was attached to the neck of the tooth (Figure 3). A part of the lining was taken and subjected for processing. On microscopic examination the lesion exhibited the same features as that of the first case except that the epithelium did not exhibit orthokeratinization. It was completely parakeratinized (Figure 4a, 4b). So the case was diagnosed as follicular odontogenic keratocyst.

**DISCUSSION:**

The OKC is one of the most aggressive odontogenic cysts owing to its relatively high recurrence rate and its tendency to invade adjacent tissue. In 1967, Toller suggested that the OKC may best be regarded as a benign neoplasm rather than a conventional cyst based on its clinical behaviour. In 1984, Ahlfors and others suggested that “if the OKC were recognized as a true, benign cystic epithelial neoplasm, the question of modified treatment schedules would be raised.” In the years since, published reports have influenced WHO to reclassify the lesion as a tumor. Several factors form the basis of this decision.

- **Behaviour:** As described earlier, the OKC is locally destructive and highly recurrent.

- **Histopathology:** Studies such as that by Ahlfors and others show the basal layer of the OKC (KCOT) budding into connective tissue. In addition, WHO notes that mitotic figures are frequently found in the suprabasal layers.

- **Genetics:** PTCH (“patched”), a tumour suppressor gene involved in both NBCCS and sporadic KCOTs, occurs on chromosome 9q22.3-3q1.36-40 Normally, PTCH forms a receptor complex with the oncogene SMO (“smoothened”) for the SHH (“sonic hedgehog”) ligand. PTCH binding to SMO inhibits growth-signal transduction. SHH binding to PTCH releases this inhibition. If normal functioning of PTCH is lost, the proliferation-stimulating effects of SMO are permitted to predominate.4

OKCs are usually discovered during the course of a routine radiographic examination and demonstrate a well-defined unilocular or multilocular radiolucency with smooth and often corticated margins which may simulate that of a dentigerous, radicular, residual or a lateral periodontal cyst. Root resorption is seldom a feature.7

An odontogenic keratocyst may envelop an adjacent impacted tooth (envelopmental keratocyst). Occasionally the lining of a cyst in a true dentigerous relationship may be identical to that of an odontogenic keratocyst. Altini et al termed this follicular primordial cyst (follicular keratocyst) and postulated that follicular keratocysts might arise following eruption of a tooth into a pre-existing keratocyst cavity in the same way that a tooth erupts into the oral cavity.7,8,11,12 Follicular odontogenic cysts were first described by Browne in the year 1970. He pointed out the in such cyst the epithelium immediately around the neck of the teeth was not keratinized and showed inflammatory changes in the underlying capsule.12

In a study done by Altini and Cohen in the year 1987, they were able to support their hypothesis in a series of experiment
done on velvet monkeys, recipient sites were prepared by drilling holes in the alveolar bone and Autogenous palatal mucosa was placed in them. In one of the animal killed after 52 weeks, the follicle of an erupting premolar tooth collided with one of the cyst, the lining of which became incorporated into the follicle, partly replacing the follicular reduced enamel epithelium and forming an integral part of this follicle of the erupting tooth.11

In an immunohistochemical study done by Kim DK et al in 2003, it was revealed that the staining pattern and intensity for Ki-67 was same for both the follicular (associated with impacted tooth) and extrafollicular variant of OKC. Thereby suggesting that the aggressiveness of the follicular OKC is similar to the extrafollicular one and should be attended with the same therapeutic approach in order to prevent recurrence.9

In the cases reported to our department, we also saw that the gross specimen resembled a dentigerous cyst. But on histopathological examination the cystic lining resembled that of an OKC. Thus co relating the radiographic, gross and the histopathological view, we arrived to a diagnosis of follicular odontogenic keratocyst.

Very few cases of follicular OKCs have been reported so far. This makes our case a rare occurrence.

CONCLUSION:

By reporting these two cases of follicular OKC, we would like to state two points. Firstly the final diagnosis of any odontogenic cyst requires a proper clinical, radiographic and histopathological coordination. The cases described above are good example of the same.

Secondly, the most common impacted tooth in jaws is third molar followed by maxillary canine so the most common cyst associated with these impacted teeth is dentigerous cyst, encircling the neck of impacted tooth. Nevertheless according to these reported cases in young adults other than dentigerous cyst, follicular type of OKC should be considered because the clinical behavior and prognosis is much different for these two entities. However according to the present reported cases can we still consider KCOT a cyst is a research question. Further research should carry on for such reported cases with proper follow up of patients to note the prognosis of follicular type of OKC.

REFERENCES:


Correspondence:
Dr. S Hemavathy
Professor and Head,
Department of Oral and Maxillofacial Pathology
Government Dental College and Research Institute
Fort, Bangalore-560002, Karnataka, India.
E-mail: hemas21@rediffmail.com