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Orthokeratinized odontogenic cyst: case report of two cases with review of literature

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Abstract

An orthokeratinized odontogenic cyst (OOC) is a rare developmental jaw cyst, that has been considered as a variant of the keratocystic odontogenic tumour (KCOT), which was first described in 1927 by Schultz. We, report two cases of OOC along with review of literature. Of the two cases, the first one was noted in a 31 year old female, while the second one was seen in 48 year old male, both affecting the right posterior region of the mandible. Since, OOC has a less aggressive nature & no propensity to recur a conservative management with complete enucleation of the cystic lesion seems to be the treatment of choice compared to KCOT, where the ideal treatment involves more radical approach, through peripheral osteotomy, chemical curettage or block resection. Hence, now it has become essential to identify OOC as a distinct entity, as it is an odontogenic cyst with varied biological behaviour.

Keywords

odontogenic cyst; mandible; ameloblastoma

Introduction

An orthokeratinized odontogenic cyst (OOC) is a rare developmental jaw cyst that has been considered as a variant of the keratocystic odontogenic tumour (KCOT) & was first described in 1927 by Schultz. Wright in 1981 termed it as ''orthokeratinized variant of OKC,'' which showed minimal clinical aggressiveness [1]. Later on, in 1998 Li et al suggested a descriptive term ''orthokeratinized odontogenic cyst" which is the most accepted terminology till today [2].

In 2005, the World Health Organization, classified the odontogenic keratocyst as an odontogenic tumor derived from the odontogenic epithelium, so it is now essential to identify OOC as a distinct entity, separate from more aggressive KCOT so that the patient's receive appropriate treatment [3].

The purpose of this article is to represent the clinical, radiological and microscopic characteristics of OOC.

Case Presentations

Case Report 1: A 31 year old, female patient reported to the Department of Oral Pathology and Microbiology in our institute with the chief complaint of swelling & pain on chewing in right lower back tooth region since four months. Patient also gave history of pus discharge from the same area. For this,

patient went to a private clinic where she was prescribed antibiotics and analgesics which caused temporary regression of the lesion. Medical history, family history & habit history were non-contributory. The patient reported extraction of impacted48 two and half years back.

Extraoral examination revealed mild, diffuse swelling in right mandibular angle ramus region. Right submandibular lymph nodes were palpable and non- tender. (Figure.1a) Intraoral examination revealed no apparent swelling. (Figure. 1b) Pain on percussion w.r.t. 47 was noted.

Patient was subjected to radiological examination, both orthopantomogram (OPG) and Cone beam computed tomography (CBCT) was done.

OPG revealed a well-defined unilocular radiolucency extending from distal root of 47 up to the mid-portion of ramus. Involvement of anterior border of ramus was noted. (Figure. 2a)

CBCT examination revealed a radiolucent lesion in right ramus region. Lesion had caused slight expansion and thinning of buccal and lingual cortices. (Figure. 2b)

Based on history, clinical and radiological findings, a provisional diagnosis of odontogenic cyst was made with differential diagnosis of benign odontogenic tumour in consideration.

Prior to incisional biopsy the patient was subjected for routine blood investigations which were within the normal limits. Incisional biopsy was performed intraorally in the ramus region. But the incisional biopsy was small, insufficient, revealing only thin loose keratin strands & keratin flecks and hence was not conclusive of definitive diagnosis.

Patient was referred for excisional biopsy. Since the lesion was attached to the root of 47, RCT was done with 47 as a precautionary measure. Further enucleation of the lesion was done.

The macroscopic appearance of excised tissue was white in colour, soft in consistency with smooth surface. (Figure. 3)

On gross examination, as the tissue appeared white & was suggestive of a hyperkeratinisatized lesion.

Histopathological examination of Hematoxylin-eosin (H & E) stained slides revealed cystic lumen, lining epithelium and connective tissue capsule. Cystic lumen was filled with keratin in the form of keratin strands & flecks. It was lined by hyperorthokeratotic stratified squamous epithelium of 6-8 layer thickness with prominent stratum granulosum. Basal cell layer was cuboidal toflat. Epithelium & connective tissue interface was flat. The connective tissue capsule was fibro-cellular consisting of regularly arranged collagen fibre bundles. (Figure. 4a & 4b)

On the basis of histopathology, final diagnosis of an OOC was made.

Case Report 2: A 48 year old, male patient reported to the Department of Oral Pathology and Microbiology in our institute with the chief complaint of swelling in lower right side of the face, since 4 months. History revealed that the swelling was initially small in size and gradually achieved the present size, which was almost constant for past 2 months. Medical history, family history & habit history were non-contributory.

Extraoral examination revealed mild, diffuse swelling in right mandibular angle-ramus region.

(Figure. 5) Intraoral examination showed no apparent swelling.

On radiological examination, the OPG revealed a well-defined unilocular radiolucency involving the root apices 47 & 48. The mesial & distal roots of 48 showed widened PDL space. (Figure. 6)

Based on history, clinical and radiological findings, a provisional diagnosis of odontogenic cyst was made with differential diagnosis of benign odontogenic tumour in consideration.

The routine haematological investigations were within the normal limits. The patient was subjected to surgical excision.

On gross examination, the excised tissue appeared white to brown in colour, soft in consistency with smooth surface.

Histopathological examination of H & E stained slides revealed cystic lumen, lining epithelium and connective tissue capsule. Cystic lumen was filled with keratin flecks and lined by hyperorthokeratotic stratified squamous epithelium with prominent stratum granulosum. Basal cell layer was cuboidal to flat and epithelium - connective tissue interface was flat. (Figure.7a & 7b)

On the basis of histopathology, the case was reported as OOC.

Discussion

OOC is a rare developmental jaw cyst that occurs twice as frequently in the posterior region of mandible than the maxilla. It is predominantly seen in young adults, with a male to female ratio of 2:1 [4].

Dong Q et al in 2010 reviewed a total of 61 cases of OOC based on the observations of Vuhahula et al and Li et al [5,6]. They found that males are affected more frequently, with male to female ratio being 2.59:1. The age at time of diagnosis ranged from 13 to 75 years (average, 38.9 years), with a predilection for the third and the fourth decade. Mandible is more affected than maxilla and in mandible molar-ramus region is most common involved site compare to other areas of mandible [1,7].

MacDonald-Jankowski DS et al reviewed cases of Orthokeratinized odontogenic cyst in a Hong Kong community. In their study they reviewed histopathological files of Prince Philip Dental Hospital (PPDH), between 1988 and 2004 and found only five cases of OOC. Out of five two were male & three were female. The mean age of the patient's was 33.5 SD 13.06 years. Four lesions involved the mandible and only one involved the maxilla [8].OOC itself is a rare lesion and bilateral occurrence of OOC is said to be very rare with only few cases reported [9,10,11].

Radiographically, most of the cases appear as unilocular lesion with well-defined margins. Dong Q et al in their study of 61 cases, found radiographic data in 54 cases of OOC. Out of 54 cases, 47 cases showed unilocular radiolucency and 7 cases multilocular radiolucency [1]. Li et al in their study of 15 cases found that 14 cases were radiographically unilocular and only one case was multilocular [6].In our cases, one patient was male and the other was female belonging to the 3rd and 4th decade. In both cases posterior mandible was the site of involvement which is in accordance with previous literature. In our case, both the lesions showed unilocular radiolucency.

The lesions with similar clinical and radiographic presentation should be considered in the differential diagnosis such as dentigerous cyst, KCOT, ameloblastoma in particular Unicystic

ameloblastoma.

Histopathologically all cystic lesions show cystic lumen, lining epithelium and connective tissue wall. While making the diagnosis of any cystic lesion, content of cystic lumen, configuration of lining epithelium and cystic wall should be taken into consideration. Histopathologically KCOT shows more resemblance to OOC.

The review of literature revealed that two third of OOCs are commonly associated with impacted teeth most often unerupted mandibular 3rd molar and are generally thought to be dentigerous cyst based on clinical and radiologic findings [1,4]. The dentigerous cyst is also a developmental odontogenic cyst with similar clinical findings as that of OOC, but dentigerous cyst can cause severe root resorption while OOC does not. Radiologically it is associated with impacted tooth with attachment at CEJ [12].

Vuhahula E et al., stated that "OOCs associated with impacted teeth may represent true dentigerous cysts with orthokeratinization, due to the pleuripotential characteristic of odontogenic cystic epithelium, which is capable of inducing proliferation of orthokeratinized epithelium on the surface of a DC" [5].

Although, KCOT and ameloblastoma are similar to OOC, they also show predilection for posterior region of mandible but both are more aggressive lesion than OOC and frequently cause root resorption which is not seen in case of OOC. Moreover OOC are sporadic & do not have syndromic variant as seen in KCOT which is associated with basal cell nevus syndrome [12].

In our cases on the basis of clinical and radiological findings KCOT and ameloblastoma were considered in the differential diagnosis, but were ruled out on the basis of histopathological findings.

Histologically, the OOC shows a cystic cavity filled with keratin flecks and strands. Cystic cavity is lined by an orthokeratinized epithelium of varying thickness with thick keratinization, displaying a well-developed stratum granulosum and a well-defined flat or low cuboidal basal cell layer. Basal cells show no tendency to palisade, or nuclear polarization and hyperchromatism, which are characteristics of the odontogenic keratocystic tumor. A hypocellular spinous cell layer is usually made up of polyhedral to flattened cells with eosinophilic cytoplasm [1,4,12, 13].

Immunochemistry: T. Aragaki et al studied the immunohistochemical profiling of the keratin expression in KCOT & OOC as shown in Table 1. Their results indicated that KCOT and OOC expressed unique sets of keratin subtypes, suggesting that each constitutes a distinct entity defined by the keratin profile. On the basis of their finding keratin profile of OOC is almost identical to that of the epidermis and keratin profile of KCOT is similar to that of the dental lamina (which has intrinsic growth potential). [14].

Compared with KCOTs, expression level of Ki-67 and p63 are significantly lower in OOCs, suggesting a lower proliferative activity [1, 15].As the epithelial cell proliferation and differentiation pattern varies in OOC & OKC, it may be a possible reason for the different biological behaviour. Due to the less aggressive nature and no propensity to recur, a conservative management with complete enucleation of the cystic lesion seems to be the treatment of choice in OOC compared to KCOT, where the ideal treatment involves more radical approach, through peripheral osteotomy, chemical curettage or block resection [6, 7, 8]. Hence, definitive diagnosis and categorization of the cystic lesion would aid in the management and evasion of recurrence in a patient.

Conclusion

Orthokeratinized odontogenic cyst is an independent clinical and pathological entity with biological behaviour, management & prognosis different from that of keratocystic odontogenic tumour (KCOT). Although it is a rare developmental jaw cyst which is commonly associated with impacted third molar and histopathologically similar to KCOT, it should be considered in the differential diagnosis of well-defined radiolucent lesion involving the mandibular molar ramus region.

Figures

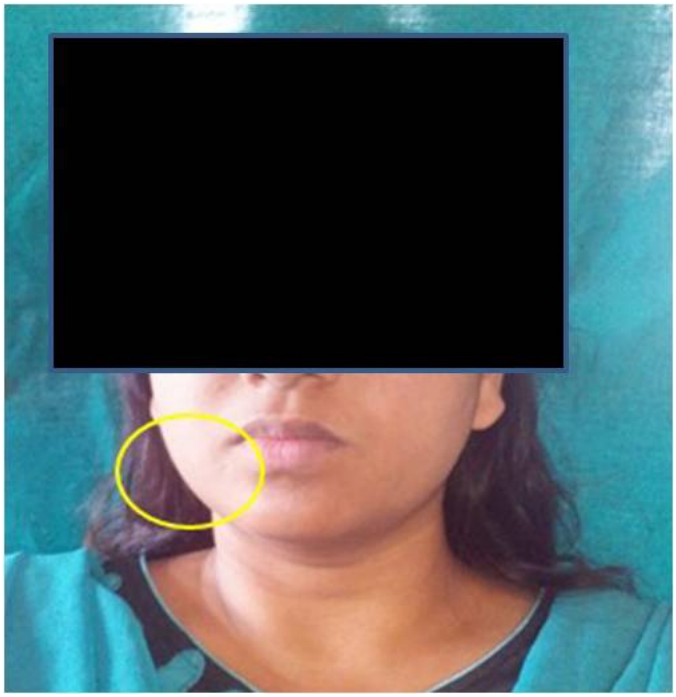

Figure 1a: Mild diffuse swelling in right mandibular angle ramus region.

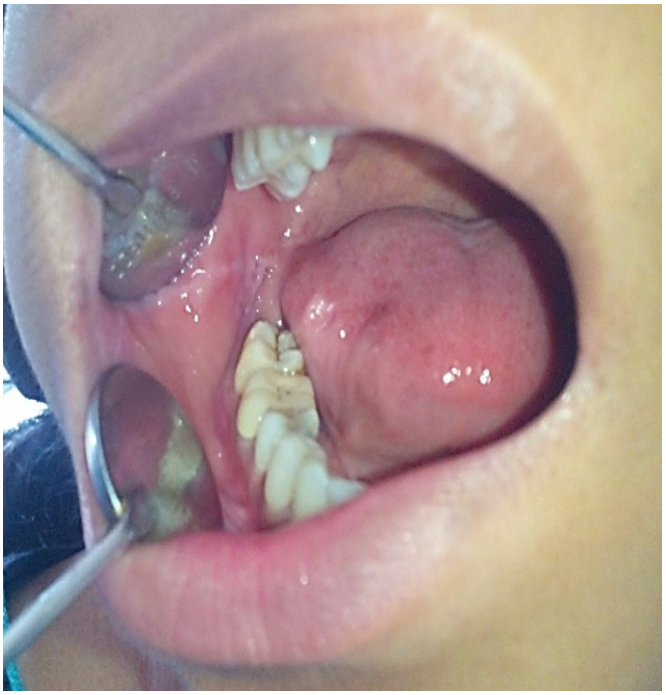

Figure 1b: No obvious swelling seen.

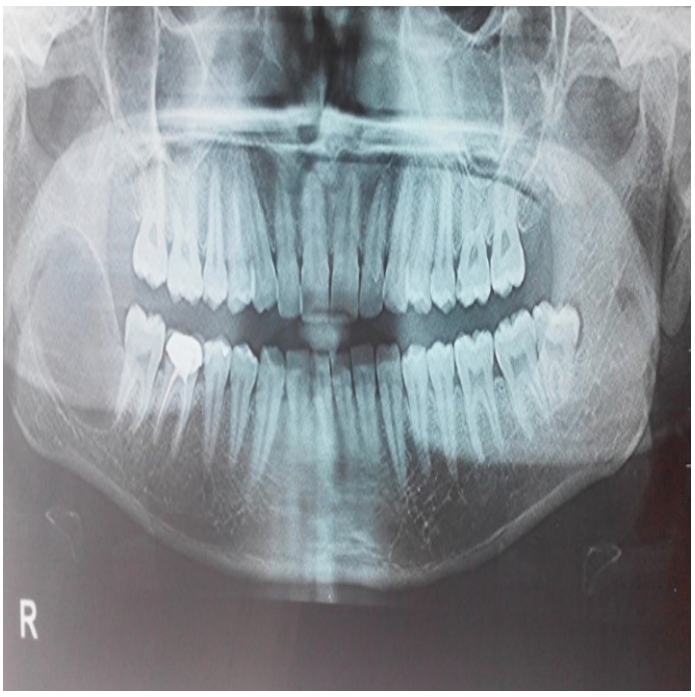

Figure 2a: OPG showing well defined unilocular radiolucency seen in right ramus region.

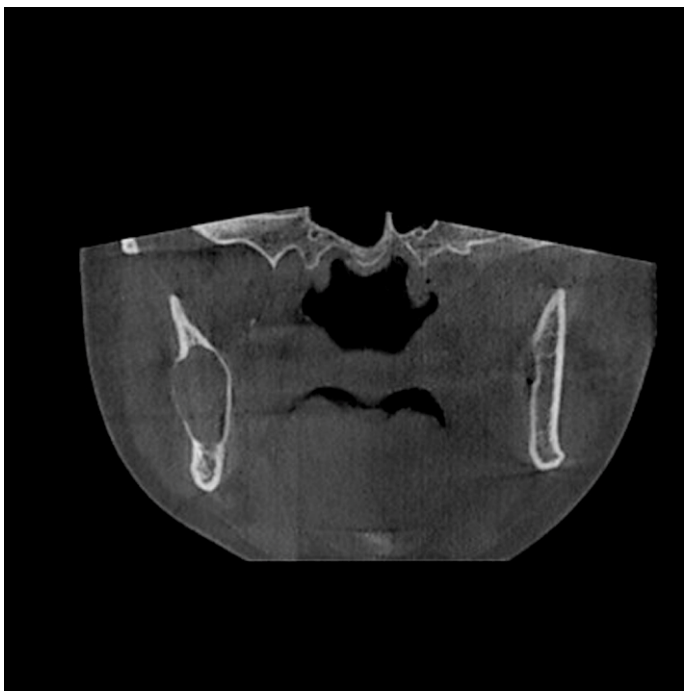

Figure 2b: CBCT showing unilocular radiolucency with expansion of buccal & lingual cortical plates noted.

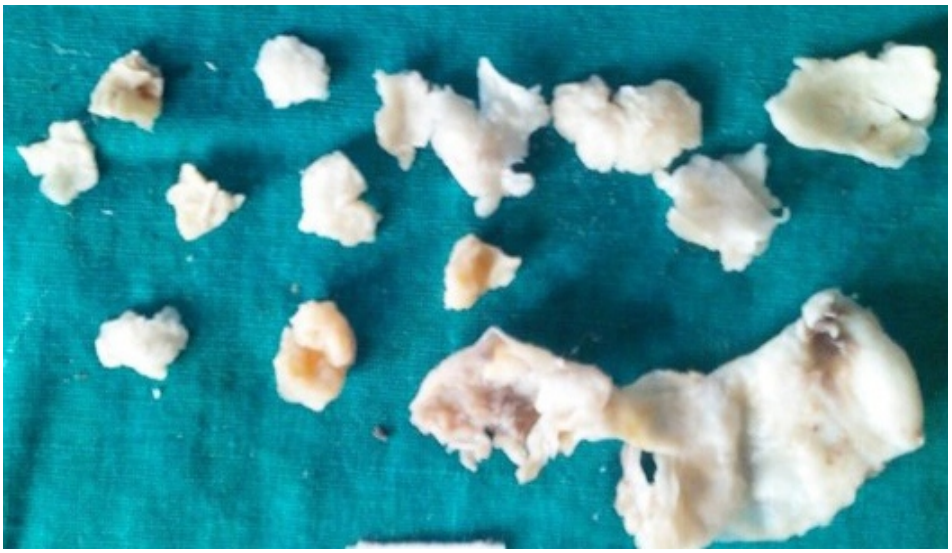

Figure 3: Gross specimen: white in colour with smooth surface.

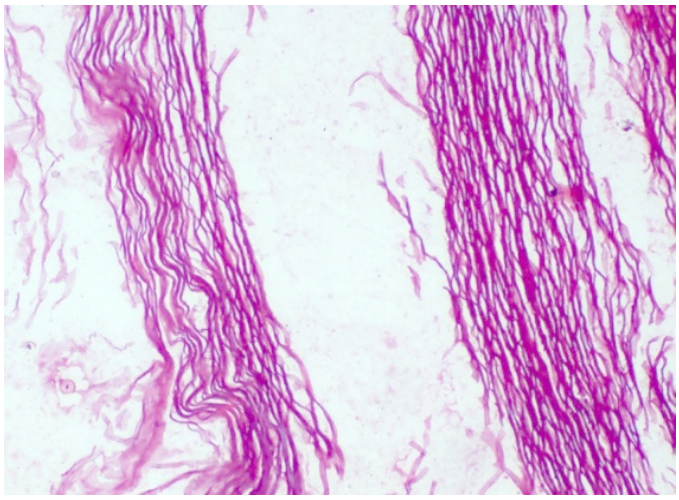

Figure 4a: Abundance of keratin flecks within cystic lumen (Hematoxylin-eosin stain X100).

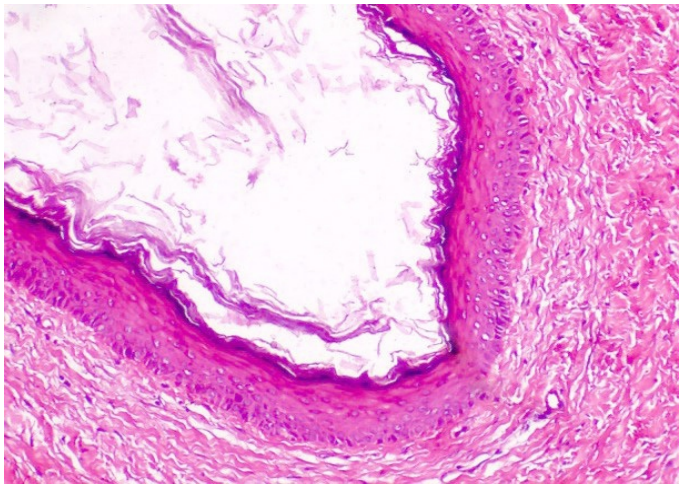

Figure 4b: Hyperorthokeratotic stratified epithelium with prominent granular layer (Hematoxylin-eosin stain X100).

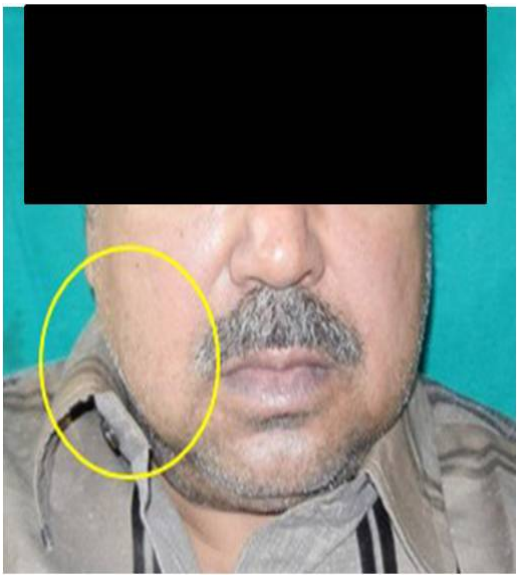

Figure 5: Mild, diffuse swelling seen in right angle ramus region.

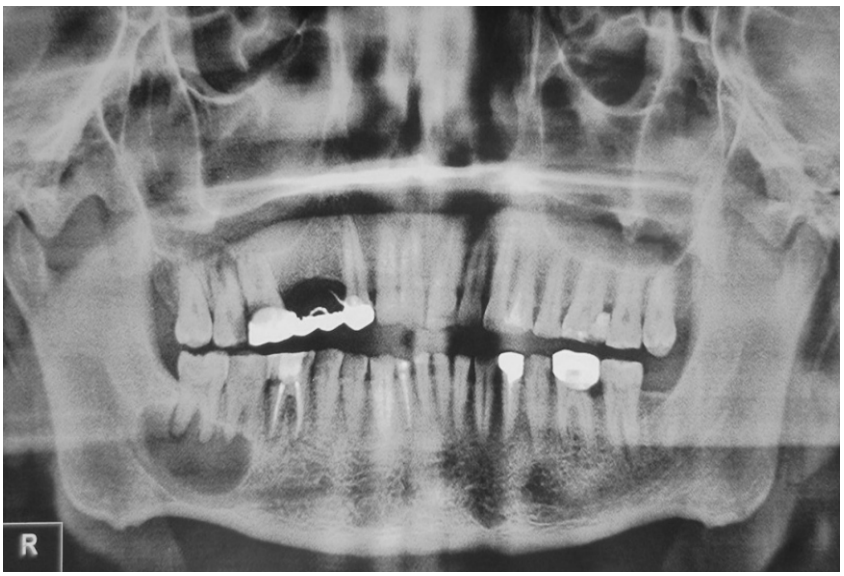

Figure 6: OPG showing unilocular radiolucency associated with 47 and 48, extending from mesial of 47 to 3mm posterior to distal root of 48.

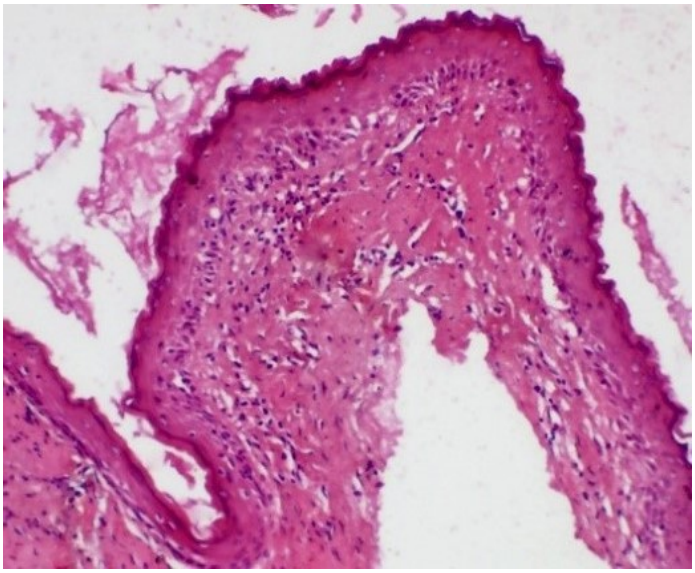

Figure 7a: Cystic lumen filled with keratin, Othokeratotic stratified epithelium with prominent stratum granulosum (Hematoxylin- eosin stain X100).

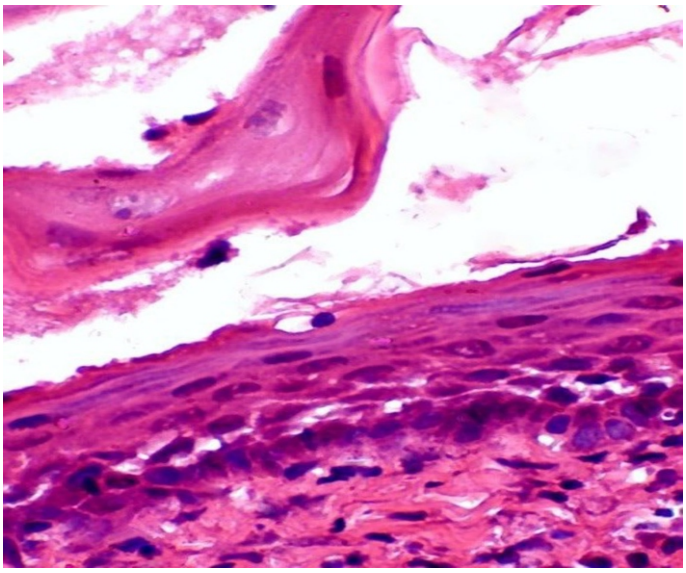

Figure 7b: Hyperothokeratotic stratified squamous epithelium with flat cuboidal basal layer (Hematoxylin-eosin stain X400).

Table

Table 1: Keratin profile in OOC, KCOT & their localization within epithelium

Keratin profile	Expression within epithelium	OOC	KCOT
K4	Superficial layer	- *	+*
K13	Superficial layer	-	+
K17	Basal & suprabasal	-	+
K19	Basal & suprabasal	-	+
K1	Suprabasal layer	+	-
K10	Suprabasal layer	+	-
LOR	Superficial layer	+	-

* positive : - (+), negative: - (-), K:-Keratin, LOR:-Loricrin

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