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NON-SYNDROMIC MULTIPLE ODONTOGENIC KERATOCYST IN A19 YEAR OLD PATIENT

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ABSTRACT

Odontogenic keratocyst (OKC)is the second most commonly occurring developmental cyst in the jaw region next to the dentigerous cyst, accounting for about 11% of the total odontogenic cysts. Approximately 8% of multiple OKC's are associated with syndromes like Basal Cell Nevoid Syndrome (BCNS), Noonan syndrome or other syndromes. Around 5% of OKC's are non-syndromic without any symptoms. OKC's usually arise from remnants of dental lamina and epithelium of other developmental cysts. Genetic mutations involving PATCH gene are also considered in the pathogenesis of OKC. Such genetic alterations often lead to BCNS The cause of multiple OKC's without associated systemic manifestations is ill-defined. In this article, we present a case report of non-syndromic multiple OKC in a 19-year-old male patient.

KEYWORDS: *Basal Cell Nevoid Syndrome (BCNS), Non-Syndromic Multiple Odontogenic Keratocyst*

INTRODUCTION

Odontogenic keratocyst (OKC) is the cyst arising from the cell rests of dental lamina[1]. It can occur anywhere in the jaw, but commonly seen in the posterior part of the mandible. Radiographically, most OKCs are unilocular or multilocular. Histopathologically, it comprises of fragile cystic wall, corrugated epithelium and palisaded basal layer of epithelium.

CASE REPORT

A 19yearold male patient reported to the Department of Oral Medicine and Radiology, with a chief complaint of pain and swelling along lower left back tooth region for one week. History of present illness revealed severe pain which was intermittent in duration and pain aggravated on chewing. A patient also complained history of swelling along the lower third of face near chin region on the left side for one week. Past medical history was nothing relevant. No systemic manifestations were revealed.

A patient had a fall 8 years back. On intraoral examination, a Fixed Partial Denture(FPD) in relation to 11,21 and 22 was seen. Mild asymmetry was seen on left side of face near the angle of the mandible. The lesion extended from body of the mandible to ramus of the mandible. Grade I mobility was seen in relation to involved region in relation to 32,31,41 and 42. OPG revealed multilocular radiolucency in relation to impacted 43. The lesion extended surrounding impacted 43, extending inferiorly to the border of mandible crossing midline with involving 32,31,41,42,43,44,45 and 46. Another unilocular radiolucency is seen in relation to impacted 38. A circumscribed radiolucency was noticed in relation to impacted 18. Lesion involved inferior mandibular canal.

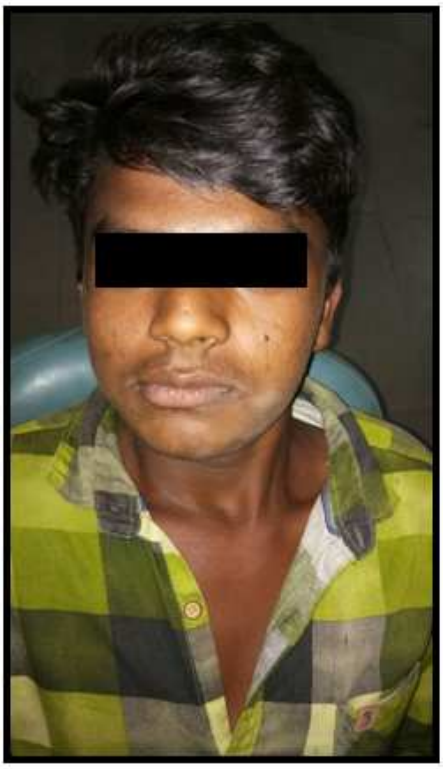

Figure 1: Extra Oral Swelling on the Left Side and Also Facial Asymmetry

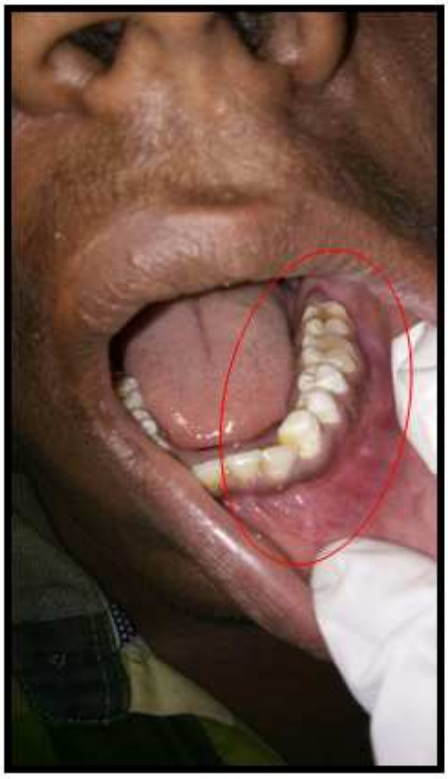

Figure 2: Intra Oral Cortical Expansion on the Left Side

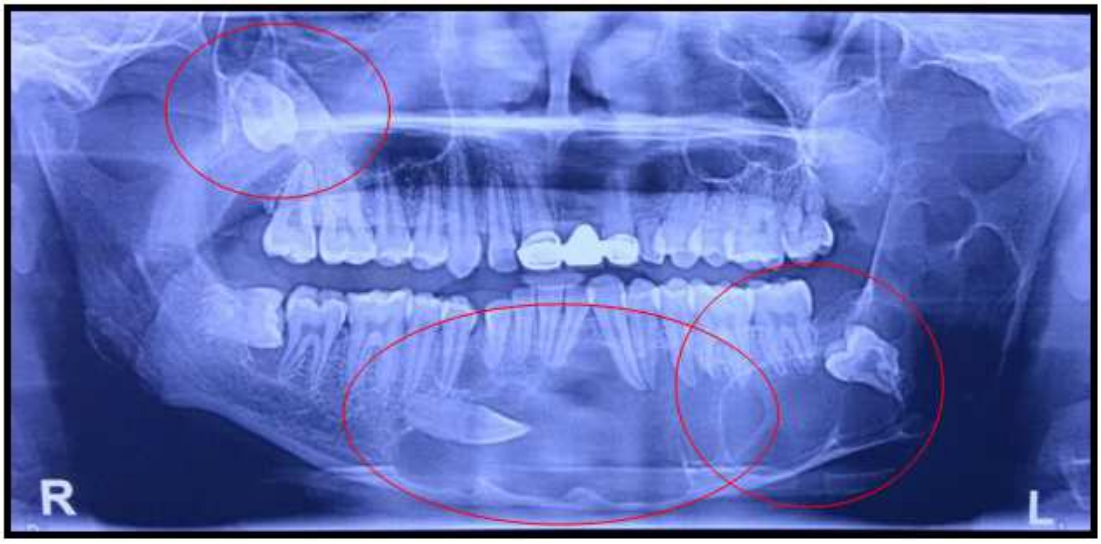

Figure 3: Maxilla – Radiolucency Associated with Right Third Molar Mandible – Radiolucency Associated with Impacted Canine and Third Molar

A provisional diagnosis of the dentigerous cyst was given due to the association of impacted teeth. Incisional biopsy was performed at the body of the mandible and also at the ramus of the mandible. All the lesions under a light microscope showed odontogenic cystic epithelium with surface corrugations. The epithelium was fragile with detachment from the underlying connective tissue in certain areas. Basal cells of the epithelium showed palisading. Hyperparakeratosis was also prominently observed. Similar findings were noted in another lesion involving. As there are no external findings such as pigmented nevoids a final diagnosis of non-syndromic multiple odontogenic keratocyst was made. Complete excision of the lesion was done.

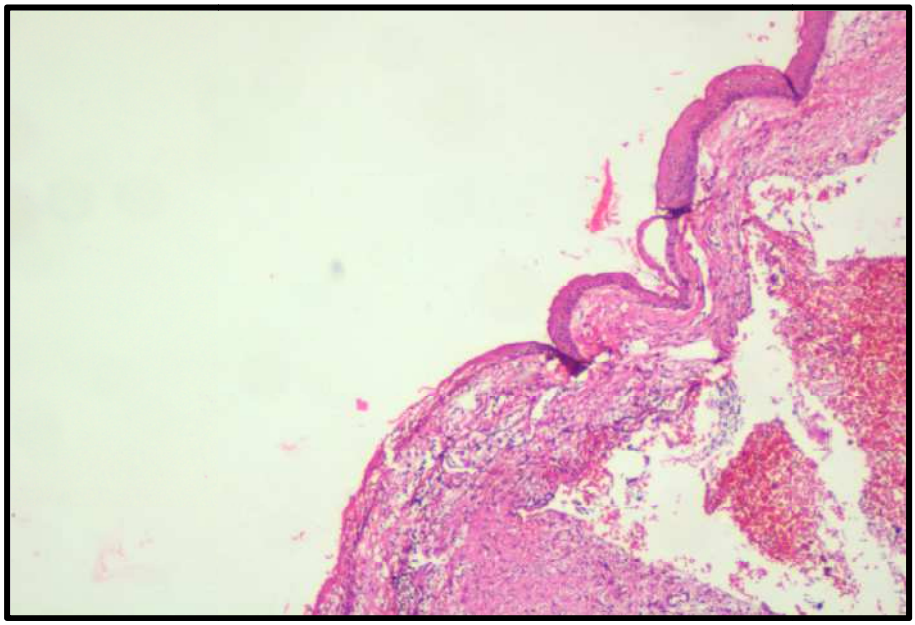

Figure 4: H&E of Mandibular Lesion – Showing Odontogenic Cystic Epithelium and Underlying Connective Tissue Stroma (10x)

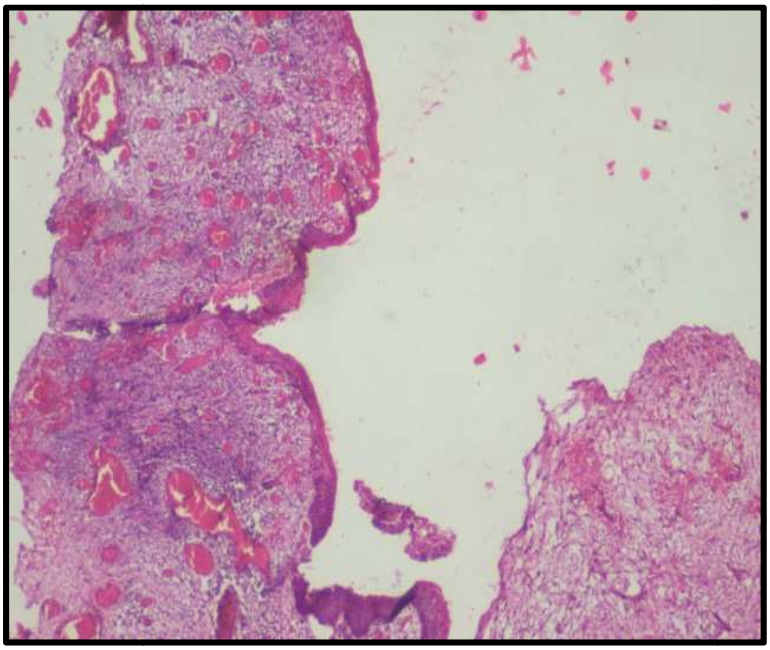

Figure 5: H&E of Mandibular Lesion – Showing Thin, Fragile Cystic Epithelium and Underlying Connective Tissue Stroma (5x)

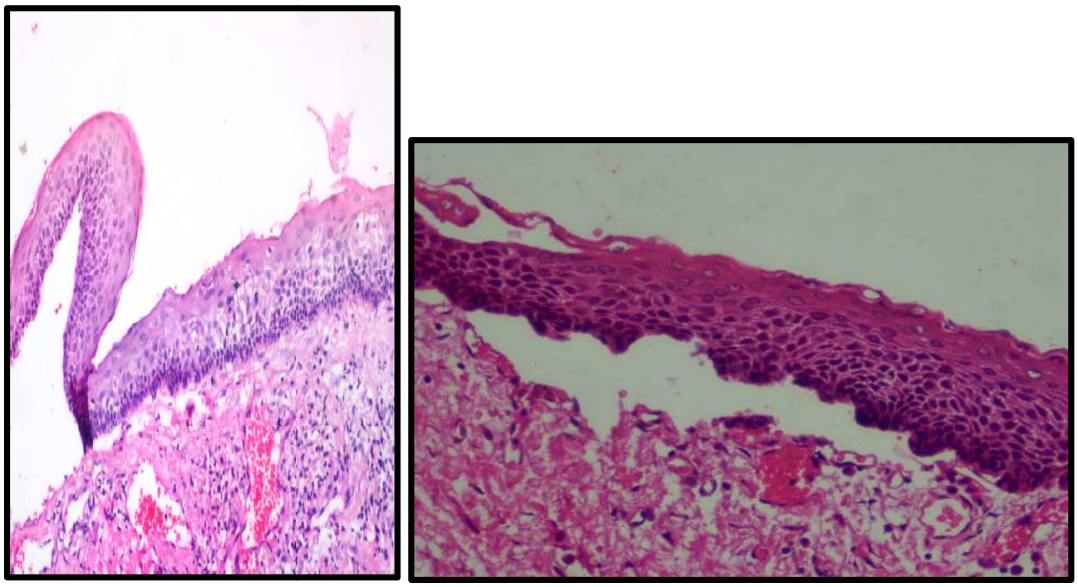

Figure 6: H&E of Maxillary Lesion – Showing Odontogenic Cystic Epithelium with Surface Corrugations that is Fragile and Separated from Underlying Connective Tissue Stroma

DISCUSSIONS

OKC is an epithelial odontogenic cyst which is generally non- inflammatory and developed from remnants of dental lamina. The main site of OKC occurrence is posterior mandible. It manifests as a unilocular radiolucent lesion. Sometimes impacted teeth may be involved which is considered a variant of OKC, called as 'Follicular OKC'[2]. OKC is also known to be genetic in origin with PTCH gene mutations which encode cell fate and patterning in many tissues including teeth. Other mutations of tumor suppressor genes are also involved in the development of OKC like p53, L-myc and so on[3].

However, the molecular pathogenesis of OKC is vast and is out of the scope of this article. Genetic mutations are mostly associated with NBCCS. Some of the syndromes where the possibility of occurrence of OKC is seen are Noonan syndrome, Marfan syndrome, Ehler – Dalnos syndrome, and Gardner syndrome. Multiple OKCs are commonly known to occur in syndromic cases. An occurrence of multiple OKC's without any other systemic manifestations is regarded as non-syndromic multiple OKC. According to the study conducted by Brannon 5.8 % of the cases out of 312 cases were non-syndromic multiple OKC's[4].

Major and minor criteria for diagnosing NBCCS is presented in table 1[5].

The pathogenesis of non – syndromic OKC and its diagnostic criteria are ill-defined.

Table 1

Major Criteria
>2BCC, or 1 BCC if <20 years
Multiple OKC of Jaws
Falxcerebri calcification
Bifid, fused or markedly splayed ribs
Palmar/plantar pits
First degree relative with NBCCS
Minor Criteria
Macrocephaly
Medulloblastoma
Congenital malformations
Radiologic abnormalities
Ovarian fibroma

Two theories are suggested in the pathogenesis of non – syndromic multiple OKC's, the first theory emphasizes the partial expression of mutated genes without systemic manifestations. Auluck et al suggested that the genetically mutated PTCH affects only locally susceptible cells in sporadic OKC's which are non-syndromic multiple OKC's[6]. Whereas in syndromic OKC's the genetic mutation occurs in precursor cells of the various tissues. Another hypothesis of occurrence of multiple OKC's without systemic manifestations is the hyperplastic potential of the dental epithelial remnants. The present case showed no systemic manifestations and no family history of multiple OKC's. The occurrence of multiple OKC's in the present case may be due to the hyperplastic potential of dental epithelial remnants with multiple focal proliferations. As such in the present case, no impacted teeth are found associated with the OKC's.

Non-syndromic OKC's are known to occur in younger age groups than syndromic OKC's. The present case also is seen in 19 years of male patient without systemic manifestations. The patient was regularly followed up for recurrences.

CONCLUSIONS

OKC is a developmental odontogenic tumor occurring in young adults in 2nd to 3rd decades of life with a male predilection. It occurs as a painless asymptomatic swelling unless secondarily infected. OKC is associated with syndromes like BCNS, non-syndromic OKC's are also present which are still an enigma to pathologists. They also have been known to cause recurrences. This case report adds to the present literature of non-syndromic multiple OKC's and may provide a clue to study this rare entity.

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