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Case Report

Bilateral keratocystic odontogenic tumor of mandible – A unique pediatric lesion: Case report and review

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ABSTRACT

The keratocystic odontogenic tumor (KCOT) is the most common cystic neoplasm in the maxillofacial region. Multiple odontogenic keratocyst are usually associated with Nevoid Basal Cell Carcinoma (NBCC) syndrome. A variant of this neoplasm is the sporadic multiple KCOT in pediatric population. Presented here is a case of bilateral keratocystic odontogenic tumor in a fourteen year old. Multiple keratocysts in the pediatric population is rare, aggressive, and recurrent and the first sign of NBCC or the solo presentation of human homologue of the PTCH (Drosophila segment polarity gene Patched) gene mutation.

1. Introduction

The keratocystic odontogenic tumor (KCOT) is the most common cystic neoplasm in the maxillofacial region. Formerly known as odontogenic keratocyst (OKC), it constitutes about 3–21.5% of all odontogenic cysts. The peak incidence is second to fourth decades of life and usually shows a male predilection [1]. Majority of lesions occur in the mandible mainly in posterior body and ascending ramus region. Multiple odontogenic keratocyst are usually associated with Nevoid Basal Cell Carcinoma or Goltz Gorlin (NBCC) syndrome. Histopathological studies show that presence of parakeratinization, intramural epithelial remnants and satellite cysts are more among NBCCS associated OKC [2].

A variant of this neoplasm is the sporadic multiple KCOT in pediatric population. Multiple KCOTs have a propensity to recur. All clinicians need to be alert to clinicopathologic markers of recurrence namely younger age, parakeratinization, inflammation, daughter cysts, subepithelial split, sub epithelial hyalinization. Also, such a sporadic incidence may be the first sign Of Nevoid Basal Cell Carcinoma syndrome.

1.1. Case report

A 14-year-old male patient reported to the department with a 15-day-old right sided painful swelling. A 10 cm × 3 cm hard diffuse swelling was evident at right mandibular parasymphyseal region. No lymph nodes were palpable. Intraorally, an intraosseous lesion was seen in relation to teeth 31–85, 4 cm × 2 cm irregular with draining sinus in vestibule corresponding to 84 (Fig. 1). The teeth 41, 83, 84, 85 were mobile. Orthopantomogram showed radiolucency extending in the region of 31–85 with impacted 42, 43, 44, 45 (Fig. 2). An additional radiolucency was detected in left ramus in relation to impacted 38. Aspiration fluid from right parasymphyseal region was a serosanguinous yield and that of the ramus region was creamy semisolid type. Provisionally diagnosis was of multiple odontogenic keratocyst and dentigerous cyst was considered as a differential. Family history was noncontributory.

Excisional biopsy (with subsequent usage of Carnoy's solution) yielded from ramus region; a 4 cm × 2 cm × 3 cm enucleated cystic lining attached at the cervical portion of 38. Biopsy from the right parasymphyseal cyst was of dimension 1 cm × 1 cm × 1 cm (Fig. 3a–c). Marsupalization was carried out leaving the impacted teeth intact so as to allow them to erupt. A consultation with physician failed to add to the medical history.

Hematoxylin and eosin stained serial sections were evaluated histologically. Both the lesions showed cystic lining which was incomplete, parakeratinized stratified squamous epithelium showing folding. Keratin layer was thin and corrugated. Lumen showed almost nil keratin content. Basal cells were low columnar to cuboidal with palisaded nucleus (Fig. 4). Intraluminal epithelial plaques could be seen with basal cells hyper plastic, hyper chromatic, low cuboidal at places (Fig. 5). Mitosis could be seen both suprabasally and basally. Epithelial connective tissue

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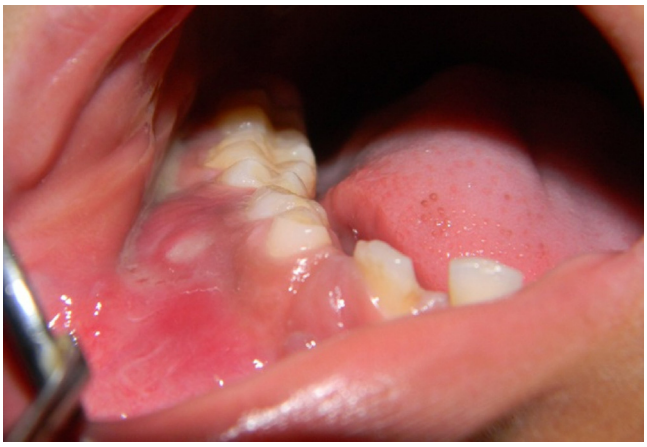

Fig. 1. Intraosseous swelling in region of 84 with draining sinus.

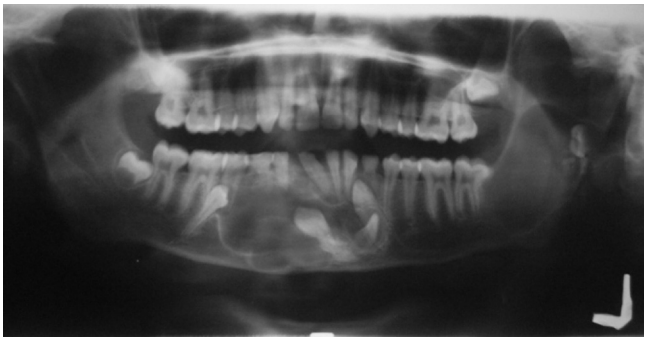

Fig. 2. Orthopantomogram parasymphyseal cyst is massive, crossing midline with several impacted permanent teeth, unilocular ramus cyst with scalloped margin and impacted 38.

interface was flat showing both sub-basal and supra-basal split (Fig. 6). At certain areas epithelium was effaced or atrophic in many areas and typical features were lost (Fig. 7). Capsule was composed of loosely arranged collagen bundles with generalized chronic inflammation in both the cysts. Early hyalinization could be seen subepithelially. Numerous daughter cysts could be detected (Fig. 8a and b). In the parasymphyseal cyst, parakeratinized stratified squamous gingival epithelium corresponding to the draining sinus could be seen. The ramus cyst may have shown the inflammation due to the preceding aspiration biopsy procedure.

Upon correlating clinico-pathologically, final diagnosis was of multiple sporadic Keratinizing Cystic Odontogenic Tumor: Juvenile, massive, envelopmental (right) and follicular (left).

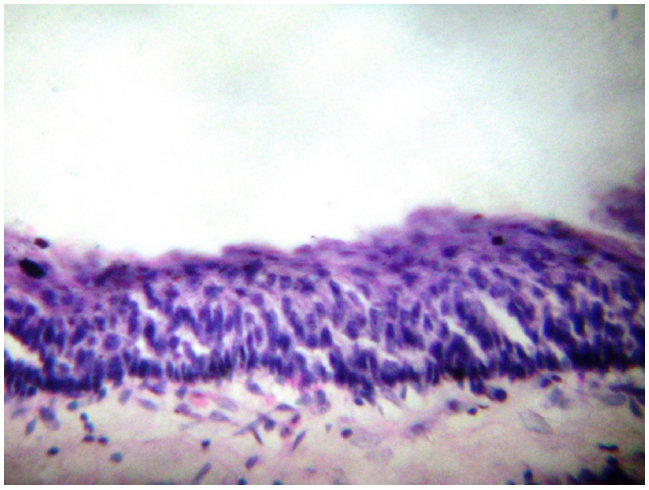

Fig. 4. Basal cells were low columnar to cuboidal with palisaded nucleus. Hematoxylin and eosin (450×).

2. Discussion

The term keratocystic odontogenic tumor (KCOT) has been defined as a benign uni or multicystic intraosseous tumor of odontogenic origin, with a characteristic lining of parakeratinized stratified squamous epithelium and potentially aggressive, infiltrative behavior. It may be solitary or multiple. The latter is usually the stigmata of the inherited Nevoid Basal Cell Carcinoma Syndrome (NBCCS) [3]. Owing to its clinical features, its potential for locally destructive behavior, high recurrence rate and tendency to multiplicity, the formerly cystic lesion was termed as a tumor [4].

There exists 43 cases of previously reported patients of multiple OKCs occurring in nonsyndromic patients out of which the pediatric population is involved only in 17 cases (Auluck et al., Parikh, Nirwan et al., Bartake et al., Meara et al.) (Table 1) (upper age limit being 15 years).

Reports say that sporadic KCOTs involve a 2 hit mechanism with allelic loss at9q22. The 2 hit mechanism is process by which a tumor suppressor gene is inactivated. The first hit leads to mutation in one allele which has no phenotypic effect although it can be dominantly inherited. The second hit refers to loss of heterozygosity. In case of KCOT it leads to dysregulation in cyclin D1 and p53 [15–17].

5%of cases of multiple KCOTs in children are indicative of multiple cysts without Goltz Gorlin syndrome [18]. Recurrence rate of such cysts is greater than solitary cyst by a value of 30%. This is of clinical importance and will influence the follow up of the case [6,7].

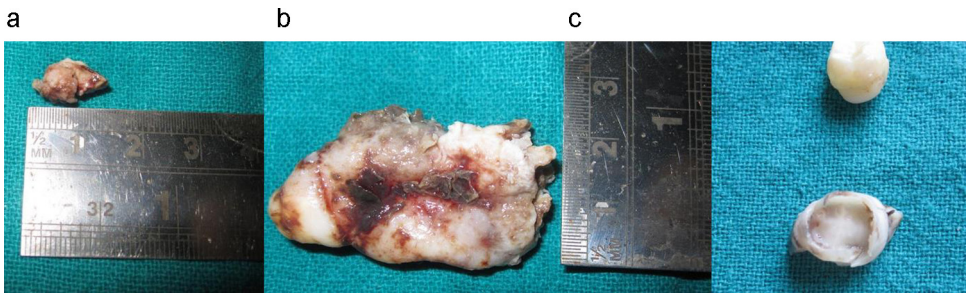

Fig. 3. Excised tissue from parasymphyseal region (a); from ramus region (b and c). 3c-Cusp imprint of impacted third molar crown cyst, dentigerous relation with tooth crown, root is underdeveloped.