Nonsyndromic Multiple Odontogenic Keratocysts: A Case Report

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Abstract: Multiple Odontogenic keratocysts (OKCs) are one of the most frequent features of nevoid basal cell carcinoma syndrome (NBCCS). NBCCS is a genetic disorder transmitted by an autosomal dominant gene with variable expressivity, which is important to recognize when a patient has multiple OKCs. It is linked with mutation in the PTCH gene. Partial expression of the gene may result in occurrence of only multiple OKC. We hereby report a case of multiple OKCs in a non-syndromic patient and highlight the general practitioner the importance of diagnosing the disease and enforcing a strict long-term follow-up whenever such a case is identified. [Parikh Neelampari et al NJIRM 2012; 3(4) : 142-145]

Key words: Multiple odontogenic keratocysts; Non-syndromic; Gorlin Goltz syndrome

Introduction: Usually, multiple odontogenic keratocysts (OKCs) occur as a component of Nevoid Basal Cell Carcinoma Syndrome (NBCCS) with concomitant cutaneous, skeletal, ophthalmic and neurologic abnormalities. Gorlin and Goltz first described the spectrum of features associated with this syndrome in 1960; hence, it is also called Gorlin-Goltz syndrome.¹

On basis of analysis of clinical features of 312 acceptable cases of odontogenic keratocysts, R. B. Brannon² found that 5.8 per cent were from patients with multiple keratocysts but with no other features of the syndrome. Though it is very rare, more cases have been reported by Auluck A,³ AR. Bartake,⁴ Rudagi B M,⁵ Amar A. Sholapurkar.⁶ However, a case is reported here so as to add to the growing number of such cases in the literature.

Case report: A 12 year-old male patient reported to Karnavati School of Dentistry, Gandhinagar with a complaint of swelling in lower anterior portion of the face since 1 year. The swelling was small initially which gradually increased to the present dimension which was progressive. The patient stated that he had pain in this region before 3 months. Intra-oro, there was a marked bone expansion of the labial plate in mandibular anterior region with obliteration of the vestibule. Overlying mucosa was normal in color. In associated soft tissues there was no ulceration or fistula formation. On palpation, the swelling was bony hard in consistency but no tenderness.

A panoramic radiograph (Figure: 1) revealed presence of two radioluencies with corticated border in relation with mandibular left and right impacted canines.

Provisional diagnosis of dentigerous cyst or odontogenic keratocyst was made. The patient’s chest and skull radiographs were unremarkable. Dermatology consultation did not reveal any cutaneous abnormality. Hematologic investigations were within normal limits. Enucleation of the cystic lesions was performed under local anesthesia and tissue samples (Figure: 2) were sent for histopathologic examination. The tissues were then processed, sectioned and stained with Hematoxylin and Eosin stain. The histopathological report revealed that the cystic lining of both lesions was parakeratinized stratified squamous epithelium of uniform thickness of 5-6 cell layers. The epithelium lining was corrugated with absence of rete pegs and palisaded basal cell layer giving an appearance of tombstone or picket fence. The connective tissue wall was composed of an inflammation-free fibrous connective tissue. (Figure: 3 and 4) Presence of few epithelial remnants (Figure: 5) and satellite cyst was noted. (Figure: 6).
All these features, correlated with the clinical and radiographic findings, established the diagnosis of Odontogenic Keratocyst for both cystic lesions.

**Discussion:** Multiple OKCs usually occur as a component of NBCCS or Gorlin-Goltz syndrome, Orofacial digital syndrome, Ehler-Danlos syndrome, Noonan syndrome. Our patient reported only odontogenic keratocysts and had no features suggestive of these syndromes, such as basal cell carcinoma, skeletal or orofacial defects, stunted growth, bleeding diathesis, hyperextensible skin and hypermobile joints or other congenital anomalies.

NBCCS is characterized by multiple OKCs, nevoid basal cell carcinomas of the skin, bifid ribs, calcification of the falx cerebri, and other features. However, except for odontogenic keratocysts other features were not present in our case.

OKCs occur in a wide age range, with a peak incidence in the second and third decades of life. Patients with multiple OKCs with or without NBCCS are generally younger than those with single OKCs.

The mandible: maxilla ratio was 2:1, with the mandibular third molar area and ramus being the most common sites. These features coincide with the case presented in our study.

Radiographically, OKCs present as a well-defined radiolucent lesions with smooth, usually corticated margins and may be either multilocular or unilocular. There is involvement of an unerupted tooth in 25% to 40% of cases. Our case complied with these findings, with the detected radiolucencies being unilocular in relation with unerupted mandibular right and left canine, having well corticated margins.

Histologically, OKCs show the presence of a thin band-like parakeratinized or orthokeratinized stratified squamous epithelium, with a prominent basal layer of columnar or cuboidal cells, and an inflammation-free connective tissue wall. Microscopic examination in our case revealed corrugated parakeratinized stratified squamous epithelial lining with absence of rete pegs and
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In children who have permanent teeth that have yet to be erupted, conservative management should be considered first because an aggressive operation can have an adverse effect on teeth development, the eruption process, and the development of the involved jaw. It has been reported that marsupialization followed by enucleation results in the lowest recurrence rate among the conservative treatment. 14 OKCs associated with NBCCS are more aggressive and have higher recurrence rates than those associated without syndrome.15

The PTCH (patched) gene, a tumor suppressor gene mapping to 9q22.3, is known to be involved in NBCCS, KCOT, PTCH binds to and inhibits the oncogene SMO (smoothened). However, in the presence of SHH (sonic hedgehog), SMO is released from the PTCH-SMO transmembrane receptor complex by the binding of SHH to PTCH. In recent studies, the hypothesis that suppression of the SHH signaling pathway can be an effective treatment for KCOT has been postulated. 16

Conclusion: In any patient with multiple OKCs, the possibility of NBCCS must be considered. As multiple OKCs might be the first & only manifestation of NBCCS, the dentist may be the first to detect it and refer the patient to a clinical geneticist for counselling and careful follow-up of such cases.

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References:
4. AR. Bartake, NG. Shreekanth , S. Prabhu , K. Gopalkrishnan Non- Syndromic Recurrent Multiple Odontogenic Keratocysts: A Case