

Multiple Odontogenic Keratocyst In A Non-Syndromic Patient

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Abstract: Odontogenic keratocysts are very well documented in the literature. Multiple odontogenic keratocysts (OKCs) are one of the most frequent features of nevoid basal cell carcinoma syndrome (NBCCS). Rarely do these multiple OKCs occur in non syndromic patients. Such a case of multiple odontogenic keratocysts unassociated with any syndrome is reported here, so as to add to the growing number of such cases in the literature. Vigilance should be kept in such cases so as to monitor the occurrence of syndrome later on in life.

Keywords: Multiple Ododontogenic keratocyst, non syndromic

I. Introduction

Odontogenic keratocysts (OKCs) are the most common form of cystic lesions affecting the maxillofacial region.¹ They are clinically aggressive lesions which are thought to arise from the dental lamina or its remnants². The OKC was first described in 1876,³ and named by Phillipson in 1956.⁴ It is one of the most aggressive odontogenic cysts of the oral cavity. OKC is known for its rapid growth⁵ and its tendency to invade the adjacent tissues including bone.^{3,7-9} It has a high recurrence rate⁹⁻¹¹ and is associated many a times with nevoid basal cell carcinoma syndrome (NBCCS).¹² Usually, multiple OKCs occur as a component of 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.¹³ Multiple OKCs have been known to occur in non-Syndromic cases, though it is very rare.¹⁴ These multiple lesions may be the first manifestation of the NBCCS or otherwise it may be because of the multifocal nature of OKCs.^{15,16} We discuss the possibility that the current case is a partial expression of NBCCS and briefly review the current trends in treatment of recurrent OKCs associated with this syndrome.

II. Case Report

A 25 year-old male patient reported to K M Shah Dental College, Vadodara, with a chief complaint of swelling in lower left and right side of the face since 2 months (Fig 1). The swelling was small initially which gradually increased to the present dimension which was progressive. Patient also complained of pain in the lower posterior region bilaterally.



Figure 1: Extraoral picture of patient showing asymmetrical face

Intra-orally, there was a marked bone expansion of the buccal cortical plate in mandibular left region with obliteration of the vestibule (Fig 2). In associated soft tissues there was no ulceration or fistula formation. On palpation, the swelling was bony hard in consistency but no tenderness. No swelling or tenderness in relation to maxillary posterior region on both sides.



Figure 2: Intraoral picture showing expansion of the cortical plate with obliteration of buccal vestibule on the right side

A panoramic radiograph revealed Multiple unilocular radiolucent lesions in right mandibular body, symphysis and left parasymphysis region. Internally, lesions were homogeneously radiolucent. They were well defined with corticated border. Scalloped margins were seen in the lesion of symphysis and left parasymphysis region. No evidence of root resorption were seen. Loss of PDL space and lamina dura in coronal third of root were evident. Mandibular canal seemed to be widened on both the sides. (Fig 3).



Figure 3: Panoramic Image showing multiple radiolucencies in mandible

For Histopathological diagnosis, we received 4 bits of tissues from posterior radiolucency, one bit was associated with the teeth. (Fig 4) Measuring collectively 5 cm X 6 cm X 7 cm. color was brownish black with soft consistency and irregular margins. Surface was smooth. The specimen received from the anterior radiolucency was measuring collectively 8 cm X 5 cm X 7 cm. color was brownish black with soft consistency and irregular margins. Surface was irregular and borders were corrugated.



Figure 4 Showing specimens received for histopathology

Histology sections stained with H & E showed cystic lumen lined by parakeratinized stratified squamous epithelium showing tall columnar basal cells with hyperchromatism, cellular atypia, altered nuclear and cytoplasmic ratio and reverse polarity. Presence of flat epithelial connective tissue interface. Epithelium is separated from the connective tissue. Underlying connective tissue stroma is composed of loosely arranged collagen fiber bundles and fibroblasts. There is presence of satellite cysts and daughter cysts in the connective tissue stroma. Presence of keratin flecks in the lumen. (Fig 5-A) (Fig 5-B),



Figure 5-A Showing histopathological sections at 4X magnification.

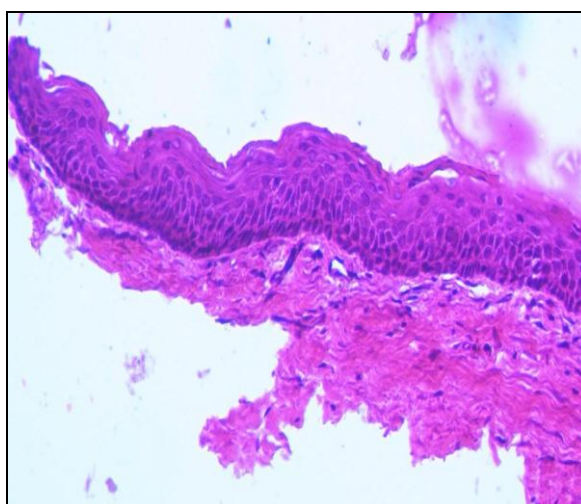


Figure 5-B Showing histopathological sections at 10 X magnification.

III. Discussion

OKC was first identified by Philipsen in 1956 and was defined by the WHO as a cyst “characterized by a thin fibrous capsule and a lining of keratinized stratified squamous epithelium, usually about five to eight cells in thickness and generally without rete pegs.” An OKC may envelope an adjacent impacted tooth (envelopental keratocyst). Very occasionally, the lining of a cyst in a true dentigerous relationship may be identical to that of an OKC.^{1,17}

The maxillofacial region is more prone to cystic lesions than any other part of the body and OKCs are the most common form of cystic lesions affecting the maxillofacial region.¹⁸ They are clinically aggressive lesions which are thought to arise from the dental lamina or its remnants.² OKCs constitute about 3% - 21.5% of odontogenic cysts.^{18,19,20} Several studies indicate a male predilection.^{11,19,20} However some studies do not correlate with this.^{1,21,22}

Some investigators have reported a bimodal age distribution with an additional peak in the fifth and sixth decades.²³

The term “multiple cysts” does not necessarily mean that the patient must have more than one cyst at a given time; rather it refers to the occurrence of cysts over the life time of the patient.

NBCCS is characterized by multiple OKCs, nevoid basal cell carcinomas of the skin, bifid ribs, calcification of the falx cerebri, and other features. Multiple OKCs can also occur as a component of orofacial digital syndrome, Ehler-Danlos syndrome, Noonan syndrome, Simpson-Golabi-Behmel syndrome, or other syndrome^{24,25}

There is no specific laboratory test to diagnose NBCCS, although the diagnosis is made clinically using the criteria suggested by Evans et al.²⁴ (Table 1) and Kimons et al.²⁵ (Table 2). However, there may be variations in the major diagnostic criteria for NBCCS in some populations due to genetic and geographic differences.

Table 1. Diagnostic criteria for nevoid basal cell carcinoma syndrome according to Evans et al.²⁴ (2 major or 1 major and 2 minor criteria should be satisfied for positive diagnosis).

Major Criteria	Minor Criteria
More than 2 basal cell carcinomas (BCCs), 1 BCC before 30 years of age; or more than 10 basal cell nevi	Congenital skeletal anomaly (e.g., bifid rib, fused, splayed or missing rib, wedged or fused vertebrae)
Any odontogenic keratocyst (proven on histology) or polyostotic bone cyst	Occipital-frontal circumference higher than the 97th percentile, with frontal bossing
3 or more palmar or plantar pits	Cardiac or ovarian fibroma
Ectopic calcification; lamellar or early (< 20 years of age) falx calcification	Medulloblastoma and Lymphomesenteric cysts
Family history of nevoid basal cell carcinoma syndrome	Congenital malformations, such as cleft lip or palate, polydactylism or eye anomaly (cataract, coloboma, microphthalmos)

Table 2: Diagnostic criteria for nevoid basal cell carcinoma syndrome according to Kimons et al.²⁵

Major criteria	Minor criteria
More than 2 Basal Cell Carcinomas (BCCs) or 1 BCC in a patient <20 years of age	Macrocephaly
Odontogenic keratocysts of the jaws (proven by histopathologic analysis)	Congenital malformations (e.g., cleft lip or palate, frontal bossing, coarse facies and moderate or severe hypertelorism)
3 or more palmar or plantar pits	Other skeletal abnormalities (e.g., Sprengel deformity, marked pectus deformity and marked syndactyly of the digits)
Bilamellar calcification of the falx cerebri	Radiologic abnormalities (e.g., bridging of the sella turcica, vertebral anomalies, modelling defects of the hands and feet, flame-shaped lucencies of the hands and the feet)
Bifid, fused or markedly splayed ribs	Ovarian fibroma or medulloblastoma (not applicable if patient is male)
A first-degree relative with NBCCS	

Our patient was apparently healthy and did not meet any of these diagnostic criteria for NBCCS, such as basal cell carcinoma, skeletal or orofacial defects, stunted growth, bleeding diathesis, hyperextensible skin and hypermobile joints or other congenital anomalies associated with overgrowth.

Ahlfors and colleagues have proposed that OKCs should be regarded as a benign cystic neoplasm rather than a developmental cyst, and Bataineh and al Qudah advocate jaw resection as the favorable treatment for an OKC. OKCs require complete removal. Aspectrum of treatments have been recommended, ranging from enucleation to resection without a continuity defect.²³

Postoperative follow-up with regular radiographic examination is important with OKCs because of the potential for recurrence. OKCs usually recur within five years after surgery, but they can recur more than 15 years later.²³

IV. Conclusion

Multiple OKC is considered as one of the important feature of NBCCS. So, in patients with multiple OKC a complete clinical examination and histopathologic analysis must be performed to detect any features associated with this syndrome. As OKCs are important and may be the first and only manifestation of NBCCS, the dentist may be the first to detect it and refer the patient to a clinical geneticist for counseling.

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