Multiple Odontogenic Keratocyst – A Case Report

Sushruth Nayak,*
Rajat Agarwal,**
Rakesh Kumar Manne, ***
Prachi Nayak, M.D.S.****

ABSTRACT

Although odontogenic keratocysts are common in clinical practice, the simultaneous occurrence of multiple cysts in a patient is rare. We report a case of an otherwise healthy individual who developed multiple odontogenic keratocyst.

Key Words: Odontogenic keratocyst, Syndrome

INTRODUCTION

Usually multiple odontogenic keratocysts (OKC's) occur as a component of nevoid basal cell carcinoma syndrome (NBCCS) with concommitant 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] We discuss the possibility that the current case is a partial expression of NBCCS.

CASE REPORT

A 13yr old female patient reported with a complaint of pain in relation to lower right posterior jaw region. Pain was present since fifteen days, which was continuous, severe in type and used to aggregate by lying down. The patient was otherwise healthy with an unremarkable medical history. No relevant findings were present on extra oral examination other than a swelling of the left eye which was due an insect bite. (Fig-1) Intra oral examination revealed mild swelling in right retro molar region and anterior mandible which was ovoid in shape with firm consistency.

Orthopantomogram (OPG) was taken, which revealed a well-corticated bilocular radiolucent lesion at the left side mandible, posterior to the canine region. The surrounding teeth, 32 and 33 were impacted. The OPG also shows two well corticated unilocular radioluncies, one at the periapical region of 31, 41 and 42 and other one at the right molar-ramus region associated with unerupted 48. (Fig-2) The radiographic diagnosis of multiple Dentigerous cyst was made. Complete enucleation of the cyst was done with the extraction of the associated teeth. The wound healed uneventfully. No recurrence is reported since 8 months.

The specimen was subjected to histopathological examination. Hematoxylin and eosin stained section showed the presence of cystic lining and connective tissue wall. The cystic lining showed the presence of parakeratinised stratified squamous epithelium with corrugated surface lining. Basal cells were columnar with the palisading arrangement of the nuclei. (Fig-3) Presence of satellite cysts were noted within the connective tissue. (Fig-4) The features were suggestive of Odontogenic keratocyst.

Author's affiliations: *MDS, Asst. Professor, Department of Oral and Maxillofacial Pathology & Microbiology, **MDS, Reader, Department of Oral and Maxillofacial Surgery, ***MDS, Asst. Professor,Department of Oral Medicine and Radiology, ****MDS, Asst. Professor, Department of Oral and Maxillofacial Pathology & Microbiology

Reprints Requests: Dr. Sushruth Nayak., M.D.S, Assistant professor Department of Oral and Maxillofacial Pathology & Microbiology, Vyas Dental College and Hospital, Jodhpur, Rajasthan, Phone: 09784579396, E-mail: drsushruthnayak@yahoo.co.in

DISCUSSION

Multiple OKC's usually occur as a component of NBCCS[1], orofacial digital syndrome[2], Noonan syndrome[3], Ehlers – Danlos syndrome[4], Simpson – Golabi – Behmel syndrome[5] or other syndromes. Our patient was apparently healthy and had no features suggestive of these syndromes, such as basal cell carcinoma, skeletal or orofacial defects, stunted growth, bleeding diathesis, hyper extensible skin, hyper mobile joints or other congenital abnormalities associated with overgrowth.

Based on histopathologic studies, parakeratinization and satellite cysts are more frequent among OKC's associated with NBCCS than in solitary keratocysts[6, 7]. In the present case, lining of OKC revealed the presence of parakeratinised epithelium and satellite cysts indicating NBCCS association.

The biological behavior of OKC's associated with NBCCS is more aggressive and these cysts have higher recurrence rates (82%) compared with solitary keratocysts (61%). The higher recurrence rates are attributed to epithelial remnants of the cystic lining or the satellite cysts left behind following surgery[8]. A recurrent OKC can be a new cyst that originates from epithelial residue or a microcyst left behind in the overlying mucosa. This is reinforced by the fact that OKC's can recur in bone grafts if overlying mucosa is not completely exised[8, 9].

The occurrence of multiple OKC's may be the first and only manifestations of NBCCS[9]. Multiple OKC's can occur a decade before other symptoms associated with NBCCS[9,10] and clinical manifestations of NBCCS may remain hidden in the earlier years of life[11]. Thus, a dentist may well be the first to detect this syndrome. The possibility of our young patient developing other features of NBCCS in the future cannot be excluded. The relatively early occurrence of multiple OKC's may be due to a genetic defect or mutation in the human patched gene[12].

In the treatment of recurrent OKC's associated with NBCCS, overlying surface epithelium should be excised along with the cystic lining to prevent recurrences from residual epithelial islands and microcysts. In addition, use of carnoy's solution following cyst enucleation and cryosurgery is advocated to kill epithelial remnants and dental lamina within the osseous margin and, thus prevent recurrences[13, 14].

In conclusion, in any patient with multiple OKC's the possibility of NBCCS must be considered. A complete clinical and histopathological examination must be performed to detect any features associated with this syndrome. As OKC's 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 genetic counseling[15].

REFERENCES

1. McGrath CJ, Myall RW. Conservative management of recurrent keratocysts in Basal-cell naevus syndrome. Aust Dent J 1997; 42(6): 399-403.
2. Lindeboom JA, Kroon FH, de Vires J, van den Akker HP. Multiple recurrent and de novo odontogenic keratocysts associated with oral- facial digital syndrome. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2003; 95(4): 458-462.
3. Connor JM, Evans DA, Goose DH. Multiple odontogenic keratocysts in a case of the Noonan syndrome. Br J Oral Surg 1982; 20(3): 213-216.
4. Carr RJ, Green DM. Multiple odontogenic keratocysts in a patient with type II (mitis) Ehler-Danlos syndrome. Br J Oral Maxillofac Surg 1988; 26(3): 205-214.
5. Krimmel M, Reinert S. Multiple odontogenic keratocysts in mental retardation- overgrowth (Simpson – Golabi – Behmel) syndrome. Br J Oral Maxillofac Surg 2000; 38(3): 221-223.
6. Dominguez FV, Keszler A. Comparative study of keratocysts, associated and non associated with nevoid basal cell carcinoma syndrome. J Oral Pathol 1988; 17(1): 39-42.
7. Todd R. Molecular approaches to the diagnosis of sporadic and nevoid basal cell carcinoma syndrome associated odontogenic keratocysts. Oral Maxillofac Surg Clin N Am 2003; 15: 447-461.
8. Stoelinga PJ. Excision of the overlying, attached mucosa, in conjunction with cyst enucleation and treatment of the bony defect with carnoy solution. Oral Maxillofac Surg Clin N Am 2003; 15: 407-414.

9. Yeo JF, Loh FC. Multiple odontogenic keratocysts of the jaws. Case report. Aust Dent J 1989; 34(6): 503-506.
10. El Murtadi A, Grehan D, Toner M, McCartan BE. Proliferating cell nuclear antigen staining in syndrome and non syndrome odontogenic keratocysts. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1996; 81(2): 217-220.
11. Chiang ML, Huang WH. Odontogenic keratocyst clinically mimicking an eruption cyst: report of a case. J Oral Pathol Med 2004; 33(6): 373-375.
12. Lench NJ, Telford EA, High AS, MarkhamAF, Wicking C, Wainwright BJ. Characterisation of human patched germ line mutations in nevoid basal cell carcinoma syndrome. Hum Genet 1997; 100(5-6): 497-502.
13. Schmidt BL. The use of liquid nitrogen cryotherapy in the management of the odontogenic keratocyst. Oral Maxillofac Surg Clin N Am 2003; 15: 393-405.
14. Schmidt BL, Pogrel MA. The use of enucleation and liquid nitrogen cryotherapy in the management of the odontogenic keratocysts. J Oral Maxillofac Surg 2001; 59(7): 720-725.
15. Ahn SG, Lim YS, Kim DK, Kim SG, Lee SH, Yoon JH. Nevoid basal cell carcinoma syndrome: a retrospective analysis of 33 affected Korean individuals. Int J Oral Maxillofac Surg 2004;33(5):458-462.

Figure 1: Extra oral view of the patient

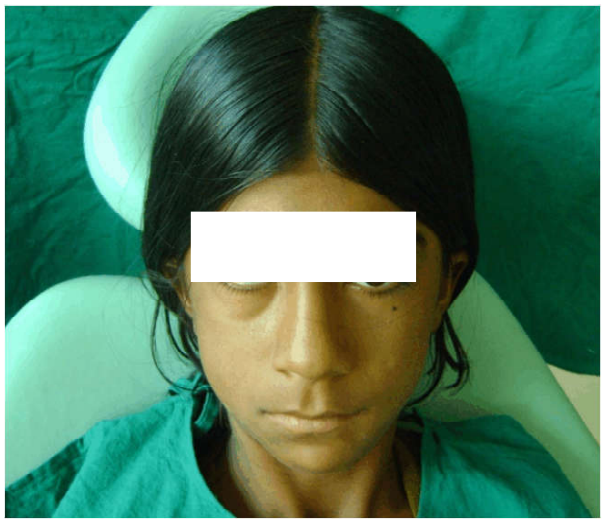

Figure 2: Orthopantomogram showing radiolucent lesions in anterior and posterior mandibular region

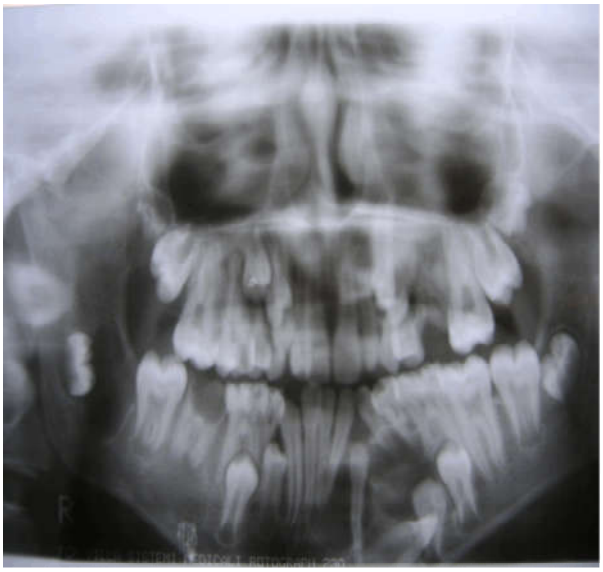

Figure 3: Section stained with hematoxylin and eosin (4×) showing cystic lining with parakeratinized stratified squamous epithelium of uniform 6–8-cell thickness

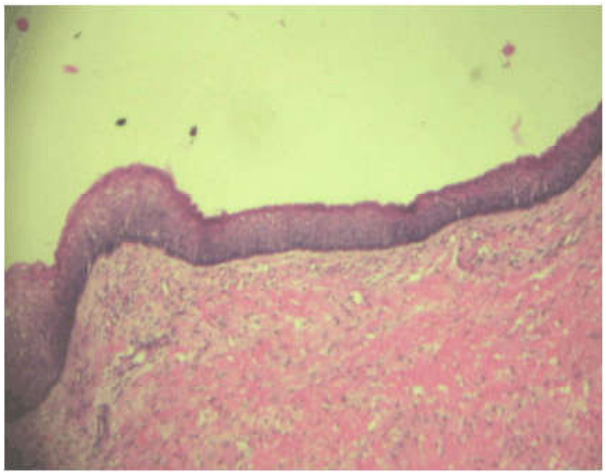

Figure 4: Section stained with hematoxylin and eosin (10×) showing a satellite cyst