# Conservative Treatment of Multiple Keratocystic Odontogenic Tumors in a Young Patient with Nevoid Basal Cell Carcinoma Syndrome by Decompression: A 7-year Follow-up Study

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*Multiple keratocystic odontogenic tumors (KCOT) occurred in a young child is challenging problem in the field of pediatric dentistry, and might have been related to nevoid basal cell carcinoma syndrome (NBCCS). Because of high recurrence rate of KCOTs, complete surgical resection is generally accepted as definitive treatment. However, complete surgical resection could induce negative effect on the development of permanent teeth and growth of jaw. Herein, we reported successful treatment case of young KCOT patient with NBCCS. Although multiple KCOTs occurred continually, the majority of the lesions healed well by decompression and important anatomical structures and permanent teeth were successfully preserved. The purpose of this paper is to report more conservative treatment of multiple keratocystic odontogenic tumors (KCOTs) by repeated decompressions with later peripheral ostectomy during a 7-year follow-up.*

***Key words: Keratocystic odontogenic tumor, Nevoid basal cell carcinoma syndrome, Decompression.***

## INTRODUCTION

Philipsen used the term of odontogenic keratocyst (OKC) for the first time in 1956[1]. In 2005, however, the World Health Organization (WHO) changed the term of OKC to keratocystic odontogenic tumor (KCOT) due to better reflection of its neoplastic nature [2]. The presence of multiple KCOTs in children can be the first sign of nevoid basal cell carcinoma syndrome (NBCCS), also known as Gorlin–Goltz syndrome [3]. Gorlin and Goltz first reported this syndrome in 1960 [4]. It is defined as the occurrence of nevoid basal cell carcinomas, multiple KCOTs in the jaws and bifid ribs. Evans *et al* suggested that NBCCS should be diagnosed using the clinical criteria. These criteria include more than 3 palmar or plantar pits, congenital skeletal anomaly, calcification of the falx cerebri and cardiac or ovarian fibroma [5]. NBCCS is inherited in a dominant autosomal genetic manner[6]. The *PTCH* gene is known to be involved in the development of NBCCS[6]. KCOT is characterized by its high recurrence rate of 2.5–62.5%, which is increased further in the presence of NBCCS[7-9].

In order to decrease the high recurrence, the epithelium of the lesion should be removed completely[10]. However, radical treatment might result in damage to adjacent anatomical structures, extraction of involved teeth, destruction of the adjacent bone, and numbness caused by damage to adjacent nerve tissue. Therefore, radical resection may not be the best treatment option for young patients in terms of preservation of the tooth involved in the cystic area.

However, few reports of conservative treatment in young patients with NBCCS exist. The aim of this study was to evaluate and report conservative treatment of KCOT in a young patient with NBCCS, namely initial decompression and peripheral ostectomy.

## Case report

A 6-year-old female with a cystic lesion of the mandibular left primary molar was referred to the Department of Pediatric Dentistry, Sun Dental Hospital. She had no significant medical and dental history. There was no familial history except her father who received enucleation of two mandibular cysts 20 years ago.

Intraoral examination revealed mild buccal swelling in the mandibular left first primary molar region. In panoramic radiograph, a cyst-like radiolucency is observed below the mandibular left first primary molar area. However, the lesion was actually confirmed to be associated with the first premolar (Fig. 1A).

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Considering the patient's age, and to preserve the adjacent tooth, decompression treatment was planned. Under local anesthesia, a tube was inserted for decompression of the cyst. A biopsy specimen was taken from the lesion wall. Histopathological examination revealed a lining of parakeratinized squamous epithelium with a palisaded basal cell layer (Fig. 1B). The diagnosis was confirmed histologically as KCOT.

The patient was instructed to do in-home irrigation using normal saline daily. The radiodensity and the size of the lesion were evaluated by radiologic image monthly. As the radiolucency of the lesion decreased, the drainage tube was removed 3 months after the operation (Fig. 1C). However, the lesion recurred 6 months after removal of the tube (Fig. 1D). The adjacent primary tooth was extracted, and marsupialization was performed. To keep the window open, iodoform gauze was inserted. The gauze had been changed once a week until an obturator delivery. Six months following the marsupialization procedure, the lesion in the area of the mandibular first premolar had disappeared, and the tooth bud of the first premolar was repositioned (Fig. 2A).

A radiographic examination performed at this time revealed the presence of another cystic lesion in the mandibular canine region (Fig. 2A). The mandibular left primary canine was extracted, and decompression was performed. The findings of the histopathological examination were similar to those of the first lesion.

Two years after the first operation, the lesion on the mandibular left canine had healed (Fig. 2B). However, further multiple cystic lesions were found on the mandibular right canine, the left second molar, and the maxillary right second molar (Fig. 2B). Histopathological results also indicated KCOT, and decompression was performed. The lesion on the right canine disappeared after 1 year of previous decompression. However, radiographic examination revealed that the dimensions of the lesion on the maxillary right second molar had not diminished (Fig. 2C). Because of the recurrence and limited surgical access, the impacted maxillary right second molar was enucleated in pieces through the maxillary buccal mucosa, and soft tissue curettage was performed under general anesthesia.

Four years following the first operation, another radiolucent lesion was detected on the mandibular right second molar (Fig. 2D). The treatment was postponed at that time because of difficulty accessing the lesion due to the severe lingual inclination of the tooth. Decompression was repeated (Fig. 3A), resulting in evident reduction of the lesion (Fig. 3B).

The patient had a mild hypertelorism and an intracranial calcification was also evident (Fig. 3C). Dermatologic examination revealed three small palmar pits (Fig. 3D). Therefore, this patient was diagnosed with NBCCS according to the criteria of Evans *et al*.

## DISCUSSION

The aim of this study was to evaluate and report the treatment of KCOT with decompression and later peripheral ostectomy in a young patient with NBCCS.

The appropriate treatment of KCOT in a young patient with NBCCS is controversial [1,7-12], however, many recent reports have indicated a tendency towards application of conservative treatment [1,7,8,12].

Because the patient was young, decompression and later peripheral ostectomy were performed as a conservative treatment approach. Teeth, anatomical structures, bone and soft tissue associated with the lesion could be successfully preserved with this approach.

**Fig. 1. Progressive panoramic radiographs and histology (A) Panoramic view shows a round-shaped cystic lesion near the mandibular left first primary molar, but it is associated with the first premolar. (B) Histopathological examination of the cyst revealed a lining of parakeratinized squamous epithelium with a palisaded basal cell layer (H&E; magnification, ´400). (C) The size of the lesion was decreased at 3 months after the operation. (D) The lesion recurred 9 months following the operation**

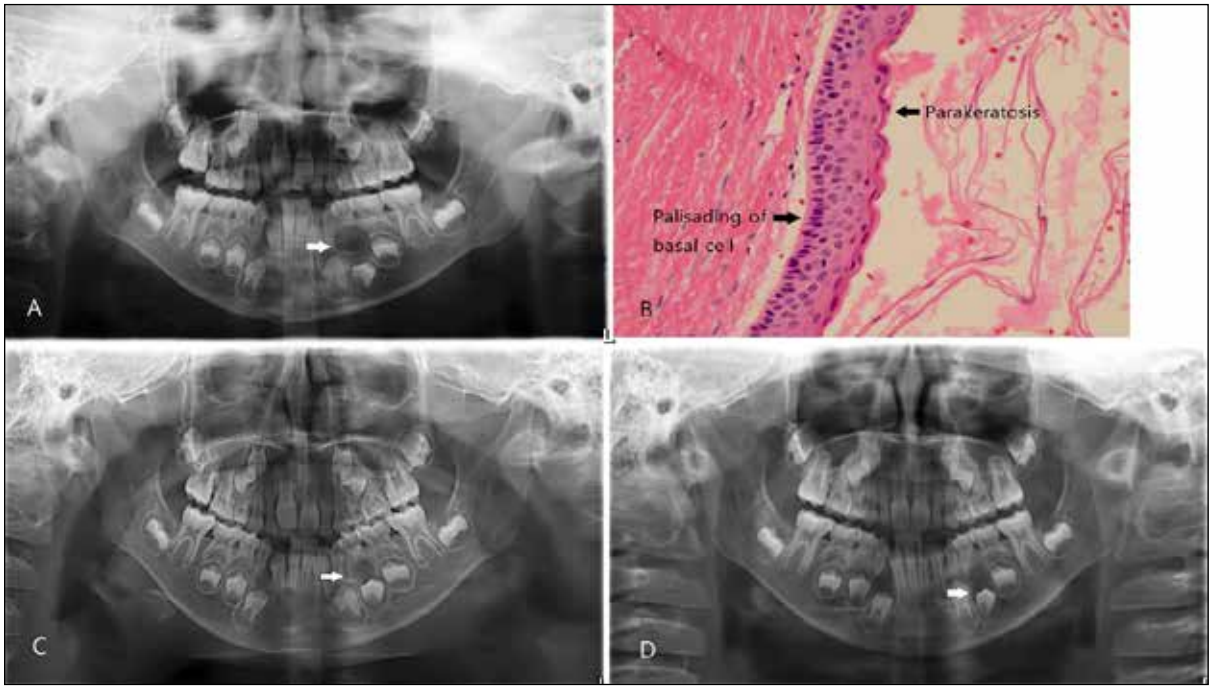


**Fig. 2. Follow up panoramic radiographs (A) The lesion of the mandibular left primary molar had disappeared after 6 month of previous decompression, however, another lesion was observed in the mandibular right and left canine area. (B) The radiolucency of the mandibular left canine area had disappeared after of previous decompression (black arrow), however, additional three radiolucent lesions were developed at the maxillary right second molar, mandibular left second molar and mandibular right canine (white arrows). (C) The radiolucency of the mandibular right canine area had disappeared after 1 year of previous decompression (black arrow), however, the lesion on the right second molar did not diminish in size (white arrow). (D) Another radiolucent lesion on the mandibular right second molar was detected (4 years after the first visit).**

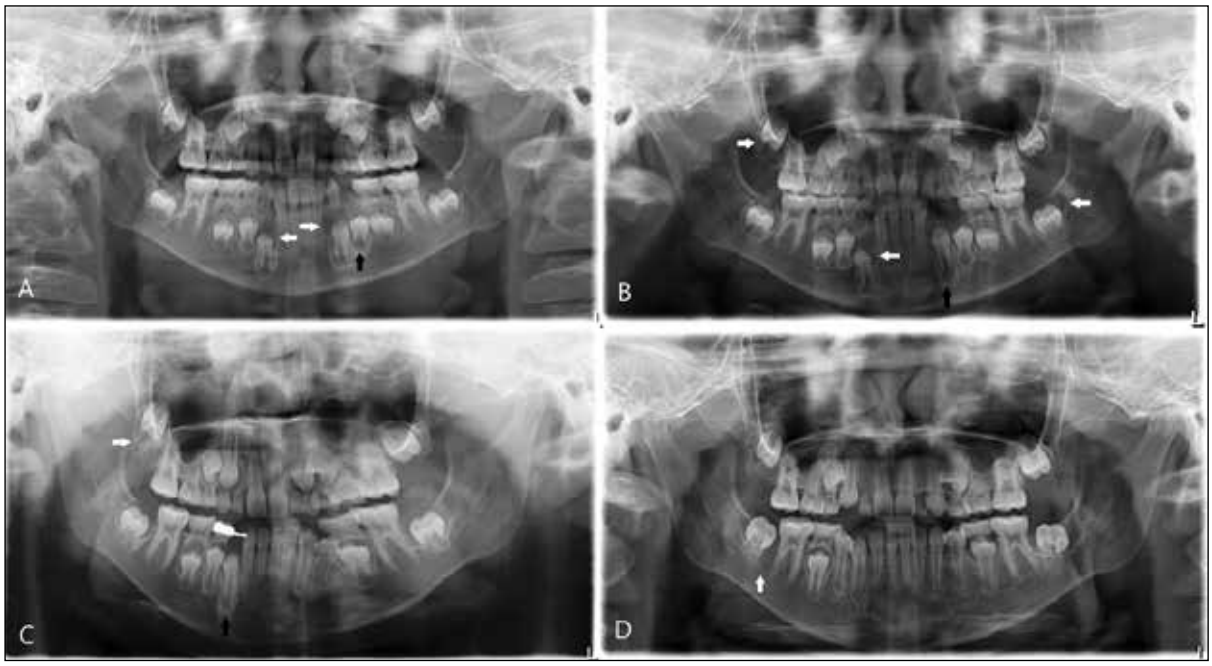
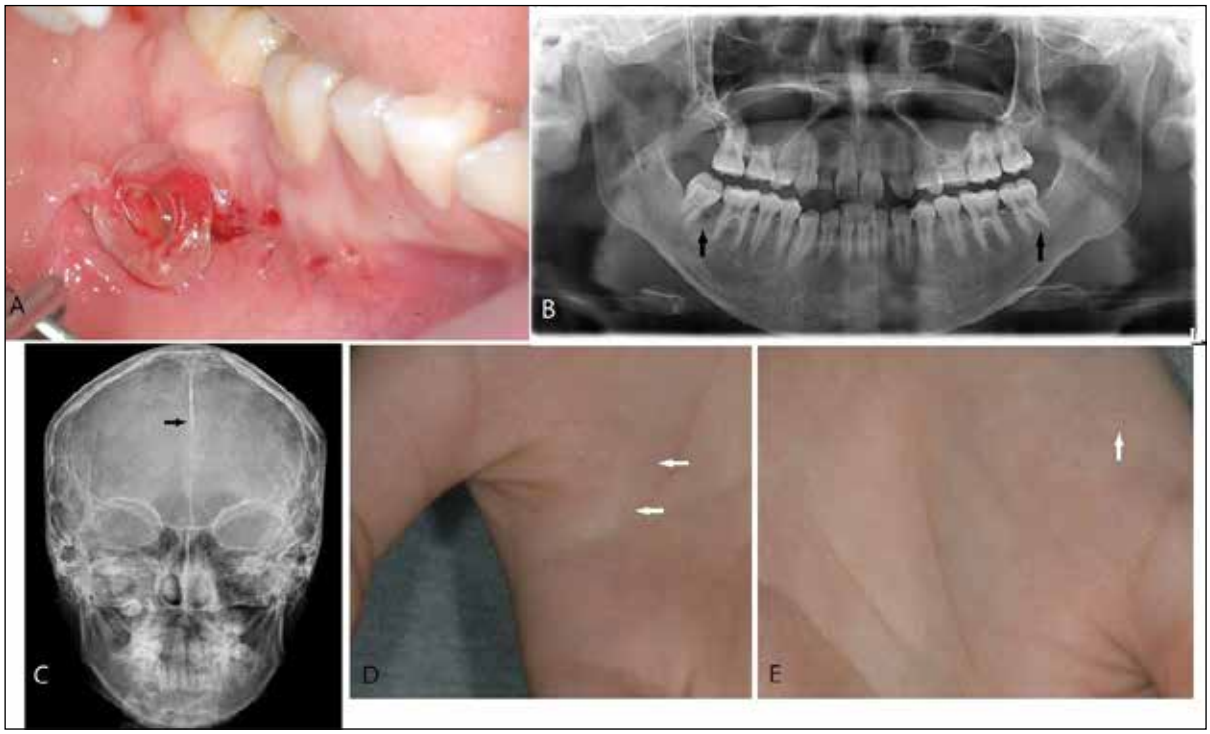



**Fig. 3. (A) Figure of maintenance tube inserted in the right second molar area after final decompression and peripheral ostectomy. (B) 7-year follow up panoramic view showed reduced radiolucency in mandibular right second molar and complete recovery in mandibular left second molar (C) Posterioanterior skull radiograph reveals calcification of the falx cerebri. (D) Two palmar pits on the right palm. (E) One palmar pit on the left palm.**



Decompression reduced the pressure of the cystic fluid. This procedure results in shrinkage of the cavity of the lesion and bone apposition around the lesion walls[1)]. Furthermore, the residual epithelium was thickened and more cohesive after decompression. In addition, decompression induced a change in the epithelial layer from parakeratinized to nonkeratinized. This facilitated loosening of the epithelium from the bone cavity and its removal at the time of the peripheral ostectomy[13].

In this case, six lesions in different areas were detected within 5 years and were treated by decompression and later peripheral ostectomy. The decompression period was 9 months on average.

Four of the six lesions showed a good prognosis. Decompression of the mandibular right second molar area was again performed recently, and this has resulted in evident reduction of the lesion.

Transformation of the epithelium is a time-dependent process. The drainage tube is also an important factor in the modulation of the epithelium because the drain causes an inflammation adjustment within the connective tissue, which can induce transformation of the epithelial layer [1].

August *et al.*[13)] suggested that decompression period of at least 9 months may be required to induce obvious epithelial modulation using an anti-cytokeratin-10 antibody.

The drainage tube was removed when the size of the first lesion had been reduced (as determined by x-ray examination) and the adjacent permanent tooth bud was repositioned 3 months after the tube insertion. However, this was too early for any change in the epithelial layer. The procedure was subsequently changed to marsupialization using an obturator, and the adjacent primary tooth was extracted. This led to a reduction in the size of the lesion; no recurrence has been observed.

The lesion of the maxillary right second molar area did not show a good prognosis. Accurate positioning of the tube end is difficult for lesions located near the maxillary sinus. This technique can be problematic because the surgeon can confuse the lumen of KCOTs for the inner side of the maxillary sinus.

However, the other lesions could be resolved by decompression with preservation of the adjacent permanent tooth. Consequently, decompression may still be the first treatment option in young patients due to its the advantages of preserving teeth, bone, and important anatomical structures related with the lesion. Decompression indeed reduces the size of the lesion especially in young patients in whom more aggressive treatment would result in a long-term surgical defect. Cooperation of patients is key factor to successful decompression treatment. This patient and her parents were cooperative and maintained the cleanliness of the area surrounding the tube. Therefore, decompression treatment was successful in this case.

The results of decompression treatment vary according to the positions of the lesion and adjacent anatomical structures that affect the surgical approach.

The patient was diagnosed as having NBCCS. She has three palmer pits on her hands at the time of writing; this number could increase as she ages [14].

Although nevoid basal cell carcinoma (NBCC) does not occur on the skin, dermatologic examination and regular follow-ups are needed because these skin abnormalities, similar to basal cell carcinoma, usually appear after puberty[15]. Therefore, the patient has been referred to a dermatologist and is undergoing continuous radiographic and clinical follow-ups to monitor the progress of the syndrome.

## CONCLUSION

We suggested that initial decompression and later peripheral ostectomy can be a good conservative treatment option for young patients with NBCCS. The success of treatment was influenced by the lesion location, the method of decompression, and the cooperation of patients. Long-term follow-up is mandatory because of the high recurrence rate. This patient has undergone regular follow-up for 7 years, and this will continue for the foreseeable future.

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