

Bilateral Dentigerous Cysts in a Non-Syndromic Patient: Literature Review and Report of a Case

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Abstract

Introduction: Dentigerous cysts (DCs) are the most common developmental cysts of the jaws, mostly associated with impacted third molars and canines. Multiple or bilateral DCs are rare and typically occur in association with some syndromes including cleidocranial dysplasia and Gorlin-Goltz. The occurrence of multiple DCs is rare in the absence of these syndromes.

Case Presentation: A 28-year-old male was referred for pain and mobility of the left mandibular second molar. On radiological examination, follicular enlargement in relation to the unerupted third molar was found. Another unilocular radiolucent area was seen in relation to the right-side unerupted third molar. A provisional diagnosis of multiple odontogenic keratocysts was made; however, the histopathological studies revealed DC on both sides. The patient's medical history was non-significant, and no associated syndromes were present. Bilateral DCs are rare in the absence of a syndrome, and to date, only 53 cases have been reported in the literature. Here, we report the unusual bilateral occurrence of DCs associated with unerupted mandibular third molars in a non-syndromic patient with a review of the literature for this unusual finding. The one-year follow-up radiograph of the patient showed a favorable osseous formation with no evidence of recurrence of the cysts.

Conclusion: A comprehensive radiographic examination, especially for mandibular third molars, is essential for correct diagnosis and management of the lesions. Furthermore, systematic clinical examinations should be performed to rule out any associated syndrome.

Key Words: Dentigerous Cyst, Radiography, Syndrome

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Introduction

A dentigerous cyst (DC) is an odontogenic cyst that is attached to the neck of an unerupted tooth at the cemento-enamel junction (CEJ). When cysts are small, they are usually discovered in radiographic examinations that are performed to investigate other symptoms or failure in tooth eruption [1]. Since the cyst may increase in size, the best treatment is surgical removal of the lesion and the

involved teeth or decompression to salvage the teeth [2].

Most DCs are solitary. Bilateral and multiple cysts are usually found in association with a number of syndromes including cleidocranial dysplasia, Maroteaux-Lamy syndrome, Gorlin-Goltz syndrome, and mucopolysaccharidosis. In the absence of these syndromes, bilateral DCs are rare [3,4].

As noted in Table 1, there have been 53 cases of multiple non-syndromic cysts reported in the literature from 1941 to 2018, and we only found 20 reports of non-syndromic bilateral DCs associated

with mandibular third molars.

Here, we report the unusual occurrence of non-syndromic bilateral DCs associated with unerupted mandibular third molars.

Table 1. Results of the literature review on bilateral dentigerous cysts (DCs)

Author	Year	Gender	Age	Location	Treatment
Boyko	1941	M	*	Mandibular third molars	Enucleation
Myers	1943	F	19	Mandibular third molars	Enucleation
Tam	1955	M	7	Mandibular first molars	Enucleation
Henefer	1964	F	52	Maxillary third molars	Enucleation
Stanback	1970	M	9	Mandibular first molars	Enucleation
Callaghan	1973	M	38	Mandibular third molars	Enucleation
Burton	1980	F	57	Mandibular third molars	Enucleation
Swerdloff	1980	F	7	Mandibular first molars	Enucleation
Crinzi	1982	F	15	Mandibular third molars	Enucleation
McDonnell	1988	M	15	Mandibular second premolars and second molars	Enucleation
Eidinger	1989	M	15	Mandibular first molars	Enucleation
ONeil	1989	M	5	Mandibular first molars	Enucleation
Banderas	1996	M	38	Mandibular third molars	Enucleation
Carr	1996	M	7	Permanent mandibular first molars	Enucleation
Sands	1998	F	3	Mandibular central incisors and first molars	Enucleation
Ko	1999	M	42	Mandibular third molars	Enucleation
De Biase	2001	M	8	Mandibular first molars	Enucleation
Shah	2002	M	39	Mandibular third molars	Spontaneous regression
Ustuner	2003	M	6	Maxillary canines	Enucleation
Batra	2004	F	15	Mandibular third molars and second premolars	Enucleation
Frietas	2006	M	13	Maxillary third molars and mandibular second molars	Enucleation
Yamalik	2007	M	51	Mandibular third molars	Enucleation
Shahrabi	2007	M	37	Maxillary and mandibular canines	Surgical resection
Fregnani	2008	M	5	Permanent mandibular first molars	Marsupialization
Chew	2008	F	30	Mandibular third molars	Spontaneous regression
Maurette	2008	M	7	Permanent mandibular first molars	Decompression
Cury	2009	M	5	Permanent mandibular first molars	Enucleation
Saluja	2010	M	22	Maxillary premolars and mandibular premolars	Enucleation
Krishna	2010	F	12	Maxillary canines and unilateral mandibular lateral incisor	Enucleation
Tikeakar	2010	M	11	Second premolars	Enucleation after marsupialization
Prasad	2010	F	12	maxillary canines	Enucleation
Grewal	2011	M	11	Maxillary canines	Enucleation
Shirazian	2011	M	10	Mandibular second premolars	Marsupialization
Tamgadge	2011	M	10	Maxillary premolars	Enucleation
Reddy	2011	F	11	Mandibular first premolars	Enucleation
Ishihara	2012	M	13	Mandibular premolars	Enucleation

Aher	2013	M	24	Maxillary and mandibular third molars	Enucleation
Byantal	2013	M	13	Maxillary mesiodens	Enucleation
Imada	2014	F	42	Mandibular third molars	Enucleation after marsupialization
Subash	2014	M	14	Maxillary and mandibular second molars	Enucleation
Morais	2014	M	15	Mandibular third molars	Enucleation
Naik	2014	M	25	Mandibular third molars	Enucleation
Vassiliou	2015	F	38	Mandibular third molars	Enucleation
Kaushik	2015	M	*	Maxillary and mandibular canines	Enucleation
Sanjay	2015	F	24	Mandibular canines	Enucleation
Devi	2015	M	17	Maxillary central and lateral incisors and mandibular canines and first premolars	Enucleation
Jeon	2016	M	15	Maxillary and mandibular third molars	Enucleation
Esmaelizadeh	2016	M	8	Mandibular second molars	Enucleation
Iyengar	2017	M	7	Mandibular first molars	Enucleation
Shruthi	2017	F	15	Mandibular third molars	Enucleation
Khandeparker	2018	M	10	Maxillary second premolars and canines	Enucleation
Gogula	2018	M	40	Mandibular third molars	Enucleation
Mehdizadeh	2018	M	28	Mandibular third molars	Enucleation after marsupialization

M=Male, F=Female, *: not defined in the literature

Case Presentation

A 28-year-old male was referred to an oral and maxillofacial surgeon for pain and mobility of the left mandibular second molar. In the intraoral examination, third molars were clinically absent and there were bilateral mild swellings in association with unerupted third molars. The patient's medical history was non-significant and no associated syndromes were present. On radiological examination, follicular enlargement was detected on the contralateral side of the mandible in relation to the unerupted third molar. A thin sclerotic border surrounded the well-defined unilocular radiolucent area that was present on the right side of the mandible in relation to the third molar, and a larger radiolucent area was evident on the left side of the mandible in relation to the third molar (Figure 1). A provisional diagnosis of multiple odontogenic keratocysts was made; however, ameloblastoma and ameloblastic fibroma were also considered in the differential diagnoses. For the left cyst, incisional biopsy and marsupialization were performed, and the left



Figure 1. Preoperative panoramic view

mandibular second molar was extracted because of severe root resorption and poor prognosis of the endodontic treatment. The surgical specimens were sent to the Department of Oral and Maxillofacial Pathology of Shahid Beheshti University of Medical Sciences for diagnosis. The microscopic sections showed a cystic lesion lined by a

nonkeratinized stratified squamous epithelium. The epithelium-connective tissue interface was flat. The wall of the cyst consisted of a loose fibrous connective tissue. A mild infiltration of chronic inflammatory cells was also seen in the connective tissue wall (Figure 2). These findings suggested the diagnosis of DC. Surgical enucleation of the right and left cysts was selected as the treatment of

were seen during the surgery on both sides. The same histological result was obtained regarding the second specimen; therefore, the diagnosis of DC was confirmed (Figure 4). The patient was followed for 2 months (Figure 5) and 8 months (Figure 6) after the second surgery; there was no evidence of recurrence of the cysts with a favorable osseous formation.

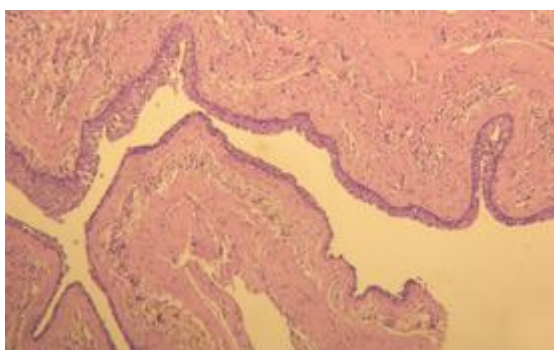


Figure 2. Photomicrograph of the left-side lesion shows features of a dentigerous cyst (DC); Hematoxylin and eosin (H & E) stain, $\times 40$ magnification

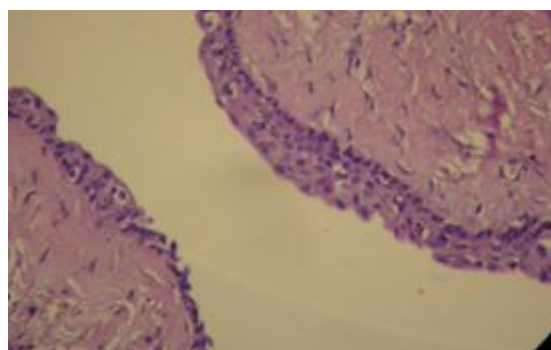


Figure 4. Photomicrograph of the right-side lesion shows features of a dentigerous cyst (DC); Hematoxylin and eosin (H & E) stain, $\times 400$ magnification

choice. At first, the left cyst was enucleated, and endodontic treatment was performed for the left mandibular first molar (Figure 3). Then, the right



Figure 3. Panoramic view after incisional biopsy and marsupialization of the left cyst. The left mandibular second molar was extracted, and endodontic treatment was performed for the left mandibular first molar

cyst was enucleated. The attachments of the cysts' wall to the CEJ of the mandibular third molars

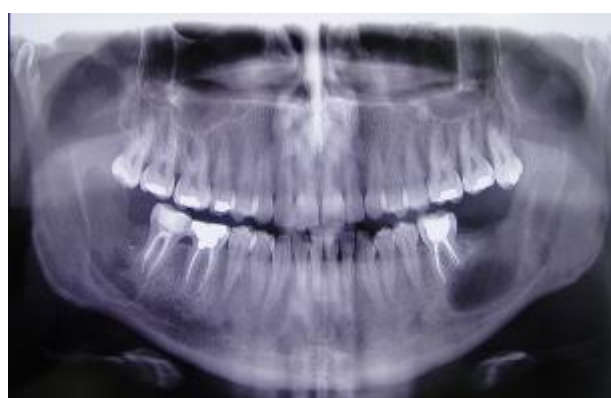


Figure 5. Panoramic view 2 months after enucleation of the right cyst and 7 months after enucleation of the left cyst. Endodontic treatment was performed for the right mandibular second molar

Discussion

A dentigerous cyst can be defined as one which encircles the crown of an unerupted tooth, expands the follicle and is attached to the CEJ. Although DC may be



Figure 6. Panoramic view one year after enucleation of the right cyst and 8 months after enucleation of the left cyst

encountered in patients in a wide age range, it is mostly detected in patients between 10 and 30 years of age with a slight male predilection. The prevalence is higher among whites than in blacks [5]. DC particularly involves mandibular third molars, permanent maxillary canines, mandibular premolars, and maxillary third molars, respectively. Mourshed [4] stated that 1.44% of impacted teeth may undergo DC transformation. Bilateral DCs are extraordinarily rare. Bilateral or multiple DCs are usually associated with Maroteaux-Lamy syndrome (mucopolysaccharidosis type VI), cleidocranial dysplasia, and basal cell nevus syndrome (Gorlin-Goltz). These are developmental conditions that cause alterations in tooth eruption or development and are usually detected in young patients with stigmata of the syndromes [3,4]. Cleidocranial dysplasia is an autosomal dominant disease that results in the absence of clavicles, short stature, parietal and frontal bossing, maxillary micrognathia, delayed exfoliation of deciduous teeth, delayed eruption of the permanent dentition, and unerupted supernumerary teeth [6]. The Maroteaux-Lamy syndrome is one of the mucopolysaccharidoses (MPS); it is resulting from a genetic defect in the degradation of specific mucopolysaccharides. With this syndrome, dermatan sulfate accumulates in tissues and is excreted in the urine, characterized by severe skeletal dysplasia, short stature, motor dysfunction, and kyphosis. Dental features include

unerupted dentition, DCs, condylar defects, malocclusion, and gingival hyperplasia [7]. Gorlin-Goltz is an autosomal dominant syndrome which is characterized by numerous basal cell carcinomas, multiple odontogenic keratocysts (seen in 75% of patients), ribs and vertebrae anomalies, intracranial calcifications, and musculoskeletal malformation [8]. Multiple DCs occur in association with these syndromes and can develop at any site in the upper or lower jaws [3,4,6,8]. In our case, there were no clinically evident syndromes.

Sometimes, Bilateral or multiple DCs are suggested to be induced by prescribed drugs. The combined effect of cyclosporin and a calcium blocker is reported to cause bilateral DCs [8]. Pleomorphism in chromosome 1qh+ has also been reported with this condition [9].

After extensive literature search on bilateral DCs, only 53 reported cases were identified from 1941 to 2018 (Table 1). This finding reflects the uncommonness of the condition. The age range for the reported cases varies widely from 3 years to 57 years of age. The mean age of these cases was about 19 years. In all reported cases, including the present case, radiographic examination showed a unilocular radiolucent lesion associated with the crown of an unerupted tooth with well-defined sclerotic margins. Thirty-six out of 53 cases have been associated with mandibular molars, and 20 cases have been associated with third molars, similar to our case.

The patients frequently present with unerupted teeth or asymptomatic slow-growing swellings. Because the lesion is asymptomatic, delayed diagnosis is expectable. Since the cysts can reach a considerable size with minimal or no symptoms, their early detection and removal are important to reduce morbidity. Therefore, it is important to perform radiological examinations when encountering unerupted teeth. While bite-wing and periapical radiographs are typically used in the routine examination of patients with a healthy dentition, these radiographs may occasionally fail to depict the full extent of a lesion. A panoramic radiograph may be used for the initial examination [10].

Radiographic assessments provide valuable information. However, pathological analysis of the

lesion is necessary for the final diagnosis. Other lesions may have the same radiological features as DCs, such as odontogenic keratocysts and unicystic ameloblastomas [11]. After the radiographs were observed, these lesions were included in the differential diagnoses. Although tooth involvement, cortical expansion, and root resorption are features mostly related to DCs, other lesions were not excluded until the results of the pathological examinations were revealed. Odontogenic keratocysts cause less expansion in the bone compared to DCs and are less likely to cause root resorption. The mean age of patients with an odontogenic keratocyst is less than that of patients with a DC; the mean size of odontogenic keratocysts is larger than that of DCs. Furthermore, DCs are more likely to have a smooth periphery but odontogenic keratocysts mostly have a scalloped periphery [12]. It is not possible to differentiate unicystic ameloblastomas from DCs with clinical and radiographic examinations [13]. Removal of the associated tooth and enucleation of the soft tissue component comprise the final treatment in most cases. Larger lesions may be surgically drained and marsupialized to relieve the pressure within the cysts and to avoid injury to the involved permanent teeth. The recurrence of DCs is very uncommon [14].

Conclusion

Bilateral DCs are uncommon in the absence of a syndrome or a systemic disease. After searching the literature, only 53 cases were identified from 1941 to 2018. A comprehensive radiographic examination, especially for mandibular third molars, is essential for correct diagnosis and management of the lesions. Furthermore, systematic clinical examinations should be performed to rule out any associated syndrome.

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