Non-Syndromic Multiple Dentigerous Cysts

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Abstract

Dentigerous cysts are common cysts of the jaws. They are associated with the crowns of permanent teeth, most frequently associated with impacted mandibular third molars. Bilateral dentigerous cysts are rare and generally occur in association with a developmental syndrome or systemic disease, such as mucopolysaccharidosis and cleidocranial dysplasia. Bilateral dentigerous cysts in the absence of a syndrome are rare and to date only 21 cases have been described. The following is a report of a case of unusual multiple non-syndromic dentigerous cysts associated with mandibular third molar and maxillary third molar and a review of the literature.

Keywords: Dentigerous cyst, unerupted teeth, enucleation

Introduction

Dentigerous cysts are common odontogenic cysts of the jaws which enclose the crown and are attached to the neck of an unerupted tooth. In other words, a dentigerous cyst is an epithelium- lined developmental cavity at the cementoenamel junction of an unerupted/ impacted tooth. They account for approximately 24% of all true cysts of the jaws, thus being the second most common jaw cysts after radicular cysts. They generally appear during tooth development in young patients.

The cyst arises from the separation of the follicle from the crown of an unerupted tooth.[2] Mandibular 3rd molar and maxillary canine are the most frequently involved. A typical dentigerous cyst presents clinically as an asymptomatic swelling unless secondarily infected.

It may often be discovered when radiographs are taken to investigate a failure of tooth eruption, a missing tooth or a malalignment. The cyst appears as a unilocular radiolucency with well defined sclerotic margins enclosing the crown of an unerupted tooth. A dentigerous cyst can be suspected when the follicular space is more than 5 mm. It is not uncommon to notice root resorption of the teeth in the region of the cyst. Hence, patients may have complaint of teeth mobility associated with jaw swelling.

Most dentigerous cysts are solitary. Bilateral & multiple cysts are usually found in association with a number of syndromes including basal cell nevus syndrome, cleidocranial dysplasia & Maroteaux- Lamy syndrome. In the absence of these syndromes, bilateral dentigerous cysts associated with mandibular molars is rare. There have been only 21 cases of multiple non-syndromic dentigerous cysts till 2011 to the best of our knowledge. Non syndromic bilateral dentigerous cysts in a single jaw are rare and non syndromic multiple dentigerous cysts in both jaws are rarer still.

This article presents an unusual case of multiple dentigerous cysts involving both maxillary and mandibular impacted third molars in a non-syndromic patient.

Case Report

A 40- year- old male patient reported to us with a complaint of swelling on the right side of the face. The swelling was noticed a year back but there had been no change in its size since then. Extraoral examination revealed a diffuse swelling on the posterior right mandibular body region measuring about 2×3 cm in size. Intraoral examination revealed a well- circumscribed swelling extending anteriorly from the mandibular right second molar region to the right mandibular angle region posteriorly. The swelling was hard in consistency, non- tender and non- fluctuant. Both buccal and lingual cortex expansion was evident. Mucosa over the swelling appeared normal and there was no evidence of any discharge from the swelling. Right mandibular 2nd and 3rd molar were Grade-II mobile and non- tender on percussion. Patient did not have any signs of paresthesia.

Evaluation of the orthopantamogram revealed a horizontally impaced right mandibular third molar. A large unilocular radiolucency with thin radio opaque border was noted extending from the right retromolar region to the mandibular 1st molar region. The radiolucency was attached to the cementoenamel junction of the impacted mandibular 3rd molar. The radiolucency extended inferiorly upto the inferior border of the mandible causing thinning of the lower border. However, the lower border appeared intact with no pathological fracture. The mandibular canal appeared to be pushed inferiorly by the lesion. Root resorption was present with respect to the mandibular 1st and 2nd molar.

On visualizing other areas of the orthopantamogram, bilateral impacted maxillary 3rd molars were noted. Another small unilocular radiolucency was noted in the right maxillary posterior region arising from the impacted right maxillary 3rd molar extending up to the distal aspect of the maxillary right 2nd molar. Based on the above radiological findings, the patient was re-evaluated clinically to confirm the findings. No observable swelling was evident in the right maxillary 3rd molar region but the mucosa was mildly compressible. The right maxillary 2nd molar was having Grade I mobility. A wide bore aspiration was done for both the lesions which yielded a yellowish straw coloured fluid with shiny crystals and protein estimation was done for the same. The values for protein estimation for the maxillary and the mandibular lesions were 8.45 gm/dl and 8.25 gm/dl respectively.

Based on the clinical, radiological examination and protein estimation, a provisional diagnosis of multiple dentigerous cysts was made. Incisional biopsy was done for the lesion in the mandible as the lesion in the right maxilla was inaccessible. The lower 2nd molar was extracted and the underlying lining of the lesion was submitted for histopathological examination which was reported to be an infected dentigerous cyst.

Since, the patient did not present with any other signs and symptoms such as basal cell tumours, rib anomalies, ophthalmic, neurologic or sexual abnormalities (characteristic of basal cell nevus syndrome) or corneal opacification, mental deterioration, dysostosis multiplex, vertebral anomalies (characteristic of Maroteaux-Lamy syndrome) or clavicle anomalies, delayed eruption process and multiple supernumerary teeth (characteristic of cleidocranial dysplasia), the patient was diagnosed to be non- syndromic.

Enucleation and curettage was planned for both the maxillary and mandibular lesions. The impacted mandibular and maxillary 3rd molars, maxillary 2nd and mandibular 1st molars were extracted followed by surgical enucleation and curettage. Since the patient had a propensity to develop dentigerous cysts, the impacted 3rd molar in the left maxillary posterior region was extracted prophylactically.

Histopathological Report

Examination of stained H & E sections showed presence of epithelial lining and connective tissue capsule. Epithelial lining in both the maxillary and the mandibular lesions were 2-4 layer thick resembling reduced enamel epithelium. The connective tissue capsule showed wavy dense collagen fibres, fibroblasts, few chronic inflammatory cells, cholesterol clefts with giant cells, blood vessels and extravasated RBCs. It suggested that both the maxillary and mandibular specimens were infected dentigerous cysts.

Discussion

Dentigerous cysts are very common developmental cysts and are generally solitary.[1] The substantial majority of the dentigerous cysts involve the mandibular 3rd molar and maxillary permanent canine, followed by mandibular premolars, maxillary 3rd molars and rarely maxillary premolars. Studies have shown that incidence rate of dentigerous cysts involving maxillary premolar was 2.7% as compared to 45.7%

involving the mandibular 3rd molar. Dentigerous cyst occurs most commonly during the 2nd and 3rd decades of life.[4] A significant percentage of cases also occurred in the 4th, 5th and 6th decades (17%, 17% and 12% respectively). Only a very small percentage of dentigerous cysts occurred in the 1st or the 7th decade of life. Males have a higher incidence of dentigerous cysts than females (1.6: 1). The reason for this gender predilection is not clear. Daley et al suggested that it might be related to a smaller jaw size and a greater tendency for prophylactic extraction of 3rd molars in females. Caucasians are more likely to develop dentigerous cysts than Asians.[3]

Although dentigerous cysts are common developmental cysts, bilateral dentigerous cysts are extremely rare and hardly reported. Bilateral or multiple dentigerous cysts are usually associated with the Maroteaux- Lamy syndrome, cleidocranial dysplasia, basal cell nevus syndrome. They are sometimes suggested to be induced by certain drugs. The combined effect of cyclosporine and calcium channel blocker has been reported to cause bilateral dentigerous cyst. Pleomorphism in chromosome 1qh+ has also been reported with this condition. In our case, there were no clinically evident features of any of these syndromes.[4] Also, there was no history of drug intake such as cyclosporine and calcium channel blocker.

The exact histogenesis of dentigerous cyst is not known. It is stated that the dentigerous cyst develops around the crown of an unerupted tooth by accumulation of fluid either between the reduced enamel epithelium and enamel or in between the layers of enamel organ. This is due to the pressure exerted by the erupting tooth on an impacted follicle, which obstructs the venous outflow and thereby induces rapid transudation of serum from across the capillary wall. Toller stated that the likely origin of the dentigerous cyst is due to the breakdown of the proliferating cells of the follicle after impeded eruption. These breakdown products results in increased osmotic tension and hence cyst formation. Bloch suggested that the origin of the dentigerous cyst is from the overlying necrotic deciduous tooth. The resultant periapical inflammation will spread to the involved follicle of the unerupted predecessor; an inflammatory exudate ensues and results in dentigerous cyst formation. Most of the authors have reported the presence of carious or discoloured deciduous teeth in relation to the cyst development. This suggests that periapical inflammatory exudates might be one of the risk factors for cyst development.[4]

There have been no reported cases of non-syndromic, bilateral dentigerous cysts occurring in all the four quadrants. Since cysts can attain a considerable size with minimal or no symptoms, early detection and removal is important to reduce morbidity. It is therefore important to perform radiological examination of all unerupted teeth. While bite- wing and periapical radiography is typically performed, this series of radiographs may fail to delineate the full extent of the lesion. A panoramic radiograph supplemented with skull series or more advanced imaging such as computed tomography may permit a better delineation of the extent of the lesion and its relationship to adjacent anatomical structures.[2]

Removal of the associated tooth and surgical enucleation and curettage of the soft tissue component is definitive therapy in most instances.[4]

Conclusion

Dentigerous cysts are the most common odontogenic cysts associated with an unerupted tooth. Multiple dentigerous cysts are of rare occurrence and are usually associated with various syndromes. But non-syndromic multiple dentigerous cysts may also occur. It is mandatory to perform a thorough clinical and systemic examination to rule out any associated syndrome especially in patients who present with multiple dentigerous cysts. It is certain by this case report that visualizing all areas of a radiograph is important to avoid missing out an additional lesion that may be clinically inevident.

Enucleation and curettage along with the removal of impacted teeth usually suffices in most cases. Since these cysts are usually asymptomatic, patients tend to present in advanced stage. Dentigerous cysts are also known to give rise to cystic ameloblastoma, epidermoid carcinomas and mucoepidermoid carcinomas which would require aggressive surgical resection.[6] Early diagnosis and timely treatment can avoid aggressive surgeries thereby minimizing morbidity and improving the patient's quality of life.

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Legends

Fig. 1: Pre-operative OPG
Fig. 2 (a&b) Surgical enucleation of the maxillary & mandibular cysts
Fig. 3: Histopathological Sections of the specimens
Fig. 4: Post- operative OPG