

# Achondropasia and Dentigerous Cyst- A Coincidental Finding or any Relationship?

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Sir,

Achondroplasia, an autosomal dominant disorder, is the most common form of chondrodysplasia associated with mutations of the transmembrane receptor Fibroblast Growth Factor Receptor 3 (FGFR3) [1]. The presence of characteristic craniofacial features like midface hypoplasia and dental malocclusions along with the need for special precautions during treatment poses a major challenge for the maxillofacial surgeon. Bilateral dentigerous cysts and its management in a patient of achondroplasia have not been reported in literature till date. We report a case of bilateral dentigerous cyst in an eight year old male child with achondroplasia.

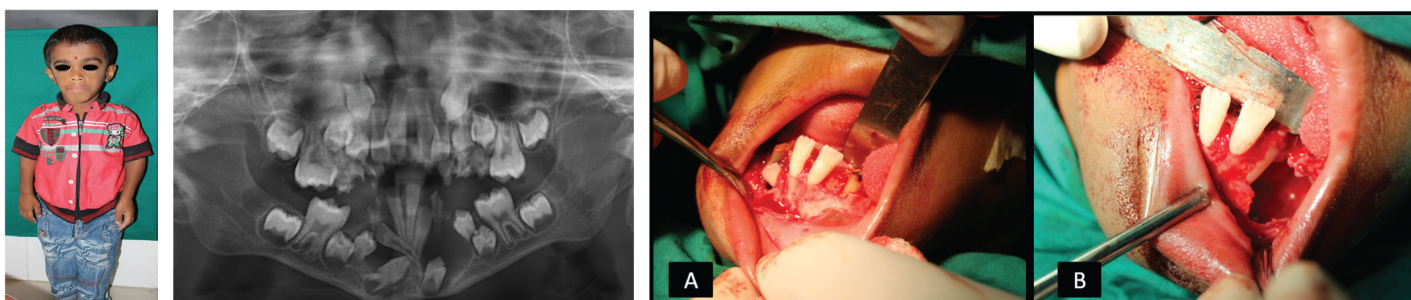
An eight year old male patient, a diagnosed case of achondroplasia [Table/Fig-1], born to non-consanguineous normal parents reported to the Department of Oral and Maxillofacial Surgery with the complaint of pain and swelling on both the sides of the lower jaw since one month. He presented with all the characteristic features of achondroplasia like disproportionate short stature (108 cm), rhizomelic shortening of the arms and legs, a trident hand configuration and craniofacial manifestations like midfacial hypoplasia, frontal bossing, flat nasal bridge and a concave facial profile. Intraoral examination revealed bilateral hard swellings in the cuspid- bicuspid regions involving 73, 74 and 83, 84. All the teeth were grossly decayed. The panoramic radiograph showed bilateral unilocular well defined radiolucency associated with bilateral unerupted mandibular canines [Table/Fig-2]. Histopathologic examination of the Fine needle aspiration biopsy was suggestive of a dentigerous cyst.

Written informed consent of the guardian was obtained. Enucleation of the dentigerous cysts with extraction of the overlying teeth (83, 84, 42, 33, 73, 74) was performed under general anaesthesia

[Table/Fig-3a&b,4]. In our case, enucleation of the cyst was planned. Marsupialization of the cyst generally requires a prolonged treatment period and follow up to which the guardians did not agree. Maintenance of oral hygiene in presence of an open cystic cavity was also considered difficult in a young child. The surgery was done under general anaesthesia due to the presence of bilateral cysts in a young uncooperative child. Preoperative assessment to rule out the risk of difficult intubation like limited mouth opening, macroglossia, tonsillar hyperplasia, limited neck extension, instability of the cervical spine and other neurological abnormalities was performed. The patient was administered routine pre and post operative medications. The procedure was uneventful and the patient has been asymptomatic till date [Table/Fig-5,6]. Histopathological examination of the post operative specimen confirmed the diagnosis of dentigerous cyst [Table/Fig-7].

The clinical features of achondroplasia include disproportionate short stature with rhizomelic shortening of limbs, normal trunk length, short stubby trident hands, spinal stenosis and lumbar lordosis [2]. Craniofacial features include macrocephaly, frontal bossing, depressed nasal bridge, maxillary hypoplasia upper-airway obstruction, otitis media and sinusitis and foramen magnum stenosis resulting in hydrocephalus [2].

Dentofacial manifestations of achondroplasia reported in literature include dental malocclusion, midface hypoplasia, protrusive maxillary incisors, anterior open bite, maxillary hypoplasia and relative mandibular prognathism [2]. Delayed dental development has been attributed to altered bone growth. Data related to delayed eruption of primary and permanent tooth is controversial and rare instances of developmentally absent teeth have been reported [3]. To our



[Table/Fig-1]: Child with achondroplasia [Table/Fig-2]: Pre operative panoramic radiograph showing bilateral dentigerous cyst  
[Table/Fig-3a,b]: Bilateral dentigerous cyst enucleation



[Table/Fig-4]: Excised cyst lining and extracted teeth [Table/Fig-5]: Postoperative photograph at six months follow up [Table/Fig-6]: Postoperative panoramic radiograph at six months follow up [Table/Fig-7]: Photomicrograph of dentigerous cyst (20X) showing 2-4 layers of cystic lining made up of stratified squamous epithelium with underlying connective tissue

knowledge, bilateral dentigerous cysts in a patient of achondroplasia have not been reported till date.

Dentigerous cysts develop around the crown of an unerupted or impacted tooth [4]. They are the most common developmental cysts of jaw and usually presents in the second and third decades of life with very few cases reported in children younger than 10 y of age [4].

It is mostly found in association with impacted mandibular third molar teeth though may involve maxillary canine, maxillary third molars, and rarely central incisors [5]. Bilateral or multiple dentigerous cysts are rare and are typically associated with a developmental syndrome like cleidocranial dysplasia, Maroteaux Lamy and basal cell naevus syndrome [6].

Management of dentigerous cyst depends on its size and location [7]. Enucleation of the cyst with primary closure and marsupialization are the usual treatment options [7].

Craniofacial features of patients with achondroplasia predispose them to several complications during operative procedures [8]. Important problems to be anticipated when the achondroplastic patient is taken under general anaesthesia includes difficulty in mask ventilation, difficulty in intubation and risk of cervico-medullary compression and spinal cord ischemia during neck extension due to foramen magnum stenosis [8,9]. Narrow nasal cavities, midface hypoplasia, adenotonsillar hypertrophy and hypotonia contribute to upper airway obstruction and obstructive sleep apnoea [8].

This case is of clinical interest since the occurrence of bilateral dentigerous cysts in relation to both the unerupted mandibular canines have not been previously reported in a case of achondroplasia. Management of dentigerous cyst under general anaesthesia in a paediatric patient of achondroplasia requires special consideration towards preoperative assessment for airway management and adequate postoperative monitoring.

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