

Case Report

Achondroplasia with multiple supplemental supernumerary teeth and multiple talon cusps: A rare case report

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ABSTRACT

Achondroplasia is the most common cause of dwarfism, which is inherited as an autosomal dominant disorder, caused by genetic mutation in fibroblast growth factor 3, leading to defective maturation of chondrocytes. It is known to be associated with various oral and dental manifestations such as delayed dental development, midfacial hypoplasia and constricted maxilla with a relatively large mandible, resulting in skeletal/dental Class III malocclusion, posterior crossbite, anterior reverse jet and anterior overbite. However, the association of achondroplasia with talon cusp and supernumerary teeth has never been reported in the literature. We hereby reported a case of achondroplasia associated with such unusual findings. Moreover, all the three variants of talon cusp, i.e., “true talon,” “semitalon” and “trace talon” are observed in the present case, which makes it a unique one. Further double talon cusps were noticed in the palatal aspect of maxillary central incisors.

Key Words: Achondroplasia, dental, dwarfism, supernumerary teeth

Received: October 2015

Accepted: January 2017

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INTRODUCTION

Achondroplasia is an autosomal dominant disorder and is the most common cause of dwarfism. It is caused by a genetic mutation in fibroblast growth factor 3 (FGFR3), causing defective maturation of chondrocytes and consequently the abnormal development of bone. FGFR3 gene is involved in converting cartilage into bone. Hence, any mutations in this gene can affect normal bone development. More than 90% of cases are of sporadic origin and are usually associated with increased paternal age at the time of conception. Homozygous achondroplasia has also been reported, which is usually lethal in the neonatal period and affects 25% of offspring of matings between heterozygous achondroplasia parents.^[1] Various orofacial findings in achondroplasia

have been reported in the literature, including delayed dental development, midfacial hypoplasia, constricted maxilla with relatively large mandible, resulting in skeletal/dental Class III malocclusion, posterior crossbite, anterior reverse jet and anterior overbite.^[2] Other features reported are macroglossia,^[2,3] oligodontia,^[3] delayed eruption of teeth,^[3] benign migratory glossitis^[4] and posterior open bite.^[4]

We hereby reported a case of achondroplasia associated with unusual dental findings like multiple supplemental supernumerary teeth and multiple talon cusps, which according to our knowledge, have never been reported in the English literature. Moreover, all the three variants of talon cusp, i.e., “true talon,” “semitalon,” and “trace talon” were observed in

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How to cite this article: Raviraj J, Suman V, Suresh D, Kartik K. Achondroplasia with multiple supplemental supernumerary teeth and multiple talon cusps: A rare case report. Dent Res J 2017;14:219-22.

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the present case, which has also never been reported before. Further double talon cusps were noticed in the palatal aspects of maxillary central incisors.

CASE REPORT

A 26-year-old dwarf, born to non-consanguineous parents, reported with a chief complaint of discolored teeth, with a desire to have them cleaned. The patient had undergone extraction of maxillary left and right first molars as they were grossly decayed. His medical history was noncontributory, and both the parents were of normal stature. On physical examination, the patient had short stature (110 cm), brachycephaly, frontal bossing, depressed nasal bridge, ocular hypertelorism, restricted elbow movements, prominent mandible with a concave facial profile, short stubby trident hands, bowed legs and partial hearing loss [Figure 1a-d].

Intraoral examination revealed macroglossia, clinically missing maxillary left and right first molars, multiple supplemental supernumerary premolars in maxillary and mandibular arches, multiple talon cusps involving maxillary and mandibular anteriors, a deep palate and a Class III malocclusion [Figure 2a-e]. The patient had moderate stains and calculi.

Posterior-anterior skull revealed a broad skull indicative of brachycephaly. Wrist radiograph revealed trident configuration and panoramic radiograph revealed multiple talon cusps involving maxillary and mandibular anteriors, multiple supernumerary teeth and a distomolar in the maxillary right quadrant and narrow sigmoid notch on either ramus of the mandible [Figure 3a-c]. Chest radiograph revealed normally developed clavicles.

Based on clinicoradiologic evaluation, a diagnosis of achondroplasia was considered.

Hypochondroplasia and thanatophoric dysplasia are the two disorders that can be considered as a differential diagnosis for the present case. Hypochondroplasia is a syndrome which is very much similar to achondroplasia but all the features are milder and are usually not manifested until late childhood. Thanatophoric dysplasia is another cause for short-limb dwarfism which results due to a mutation in FGFR3 gene. There are two types: Type I is characterized by micromelia with bowed femur and rarely with clover leaf deformity, and Type II is characterized by micromelia with straight femurs and



Figure 1: (a) Photograph showing a rhizomelic dwarf. (b) Facial photograph showing ocular hypertelorism. (c) Lateral facial profile showing large head, frontal bossing, depressed nasal bridge and concave facial profile. (d) Bowed legs.

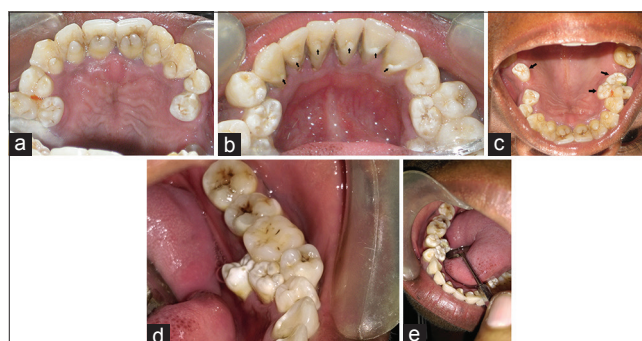


Figure 2: (a) Multiple talon cusps involving the palatal aspect of maxillary anteriors. Note the double talon cusps involving maxillary centrals. (b) Trace talon cusps involving the lingual surface of mandibular anteriors (black arrows). (c) Supplemental supernumeraries of maxillary arch (black arrows). (d and e) Supplemental supernumeraries of mandibular arch.

is commonly associated with clover leaf deformity of skull. Other features of this disorder include macrocephaly, short ribs, brachydactyly, narrow thorax, and distinctive facial features. However, the majority of the clinical features in the present case were in favor of achondroplasia, and all the features were evident in the early childhood.

DISCUSSION

Achondroplasia is an autosomal dominant disorder. However, sporadic cases have also been reported. Both parents of the patient were unaffected and were of normal stature, thereby ruling out the possibility of autosomal dominant pattern of inheritance. Hence, the present case can be considered sporadic in origin.

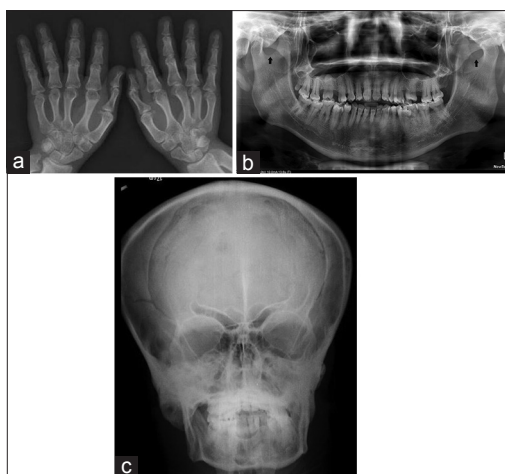


Figure 3: (a) Wrist radiograph showing “trident hand” configuration. (b) Panoramic radiograph showing multiple talon cusps involving the maxillary and mandibular arches and a distomolar in the first quadrant. Note the narrow sigmoid notch on either side (black arrows). (c) Posterior anterior skull revealing a broad skull, suggestive of brachycephaly.

Clinical features of achondroplasia include rhizomelic dwarfism with a disproportionate short stature, frontal bossing, saddle nose deformity, midfacial hypoplasia, trident hand configuration, bowed legs, lumbar lordosis, genu varum, hyperextensibility of joints except for the elbow, and normal intelligence, productivity and lifespan.^[2,5] The majority of clinical features of the present case were consistent with the literature except for the absence of genu varum and lumbar lordosis.

Although various dental findings were associated with achondroplasia, supernumerary teeth and talon cusp were never reported earlier. According to our knowledge, this is the first case report to describe such association.

Teeth present in addition to normal set of teeth are considered to be supernumerary and are of 3 types: Supplemental, tuberculate, and conoid forms. Supplemental supernumerary refers to the presence of extra tooth/teeth apart from the normal series and resembling the anatomy of any of the tooth/teeth belonging to the normal set of teeth, which were reported in the maxillary and mandibular arches in the present case. Common syndromes associated with supernumerary teeth are cleidocranial dysostosis, Gardner syndrome, Anderson-Fabry disease, Hallermann-Streiff syndrome, Oro-facial-digital syndrome type 1, Sturge-Weber syndrome, Down syndrome, Ehlers-Danlos syndrome, Apert syndrome, Crouzon syndrome and Ito syndrome.^[6,7] However,

an association of supernumerary teeth with achondroplasia has never been reported.

Talon cusp is a developmental anomaly wherein an accessory cusp-like structure is seen projecting from cingulum or cemento-enamel junction of the anterior teeth. Syndromes associated with talon cusp are Rubinstein-Taybi syndrome, incontinentia pigmenta chromians, Mohr syndrome, Sturge-Weber syndrome, Ellis-van Creveld syndrome, and hypomelanosis of Ito and Alagille syndrome. Double talon cusps were observed in relation to maxillary central incisors in the present case. Double talon cusps in the present case were different from previous cases in the literature. The double talon cusps reported in the literature were associated with fused teeth, and in other cases, a talon cusp was present on the labial and lingual aspect each.^[8] However, two talon cusps were observed on the palatal aspect of maxillary central incisors, and such type of double talon cusp has not been reported earlier. Hattab *et al.*^[9] classified talon cusps into three types based on degree of cusp formation and extension: Type 1 is “true talon,” type 2 is “semitalon,” and type 3 is “trace talon.” In the present case, all the maxillary incisors had true talons; maxillary canines had semitalons and mandibular incisors had trace talons. This case report is again unique since all the three variants of talon cusps were observed in a single patient. Other oral manifestations of the present case included macroglossia, deep arched palate and dental Class III malocclusion, consistent with the literature.^[2]

Systemic management of achondroplasia includes limb lengthening procedures to improve stature and growth hormone therapy to increase the growth rate.^[10] Dental management includes orthodontic correction of Class III malocclusion, and open bite and crossbite. The present case was subjected to oral prophylaxis for oral hygiene improvement and esthetic purpose. No further medical and dental treatments were considered for the present case as the patient did not show interest in any kind of treatment.

In conclusion, we hereby reported a unique case of achondroplasia with unusual dental findings, namely multiple supplemental supernumerary teeth and multiple talon cusps with all the three variants which have never been reported in the literature.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for

his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

The authors of this manuscript declare no conflicts of interest, real or perceived, financial or nonfinancial in this article.

REFERENCES

1. Rousseau F, Bonaventure J, Legeai-Mallet L, Pelet A, Rozet JM, Maroteaux P, *et al.* Mutations in the gene encoding fibroblast growth factor receptor-3 in achondroplasia. *Nature* 1994;371:252-4.
2. Al-Saleem A, Al-Jobair A. Achondroplasia: Craniofacial manifestations and considerations in dental management. *Saudi Dent J* 2010;22:195-9.
3. Kale L, Khambete N, Sodhi S, Kumar R. Achondroplasia with oligodontia: Report of a rare case. *J Oral MaxillofacPathol* 2013;17:451-4.
4. Celenk P, Arici S, Celenk C. Oral findings in a typical case of achondroplasia. *J Int Med Res* 2003;31:236-8.
5. Cohen MM Jr. Some chondrodysplasias with short limbs: Molecular perspectives. *Am J Med Genet* 2002;112:304-13.
6. Ramakrishna A, Rajashekarappa KB. Multiple supplemental supernumerary premolars: Unusual presentation in a nonsyndrome patient. *Case Rep Dent* 2013;2013:614807.
7. Hattab FN. Double talon cusps on supernumerary tooth fused to maxillary central incisor: Review of literature and report of case. *J ClinExp Dent* 2014;6:e400-7.
8. Cho SY. Talon cusps in mandibular incisors: Report of eight rare cases. *J Indian SocPedodPrev Dent* 2014;32:185-9.
9. Hattab FN, Yassin OM, al-Nimri KS. Talon cusp in permanent dentition associated with other dental anomalies: Review of literature and reports of seven cases. *ASDC J Dent Child* 1996;63:368-76.
10. Seino Y, Yamanaka Y, Shinohara M, Ikegami S, Koike M, Miyazawa M, *et al.* Growth hormone therapy in achondroplasia. *Horm Res* 2000;53Suppl 3:53-6.