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Oral Rehabilitation of a 4-Year-Old Child with Metaphyseal Chondrodysplasia: A Case Report

Mansi Gandhy Baviskar[1]*, Ajit Baviskar[2]

1. Pediatric and Preventive dental department, School of Dentistry, DY Patildeemed to be University, Navi Mumbai, India School of Dentistry, India
2. Emergency Medicine department, School of Medicine, DY Patil deemed to be University, Navi Mumbai, India

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**** Corresponding author:***
Pediatric and Preventive dental department, School of Dentistry, DY Patildeemed to be University, Navi Mumbai, India School of Dentistry, India

Email: mansi.baviskar@dypatil.edu

A B S T R A C T

Metaphyseal chondrodysplasia is a rare genetic disorder with an incidence of 3-6 c per one million births. It is characterized by short long bones, cupping of metaph and phalanges, and many other variable components. There are over 100 type identified metaphyseal dysplasia targeting various genes, and many more that unidentified. This case report describes oral rehabilitation of a 4-year-old child wit unknown type of metaphyseal dysplasia with dental involvement.

Keywords: Metaphyseal Chondrodysplasia; Genetic; Dental; Oral Rehabilitat Anaesthesia; General

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INTRODUCTION

Metaphyseal chondrodysplasia is a rare genetic, heterogenous group of intrinsic skeletal dysplasia that may show an autosomal dominant or recessive transmission. There are over 400 types of skeletal dysplasia with over 100 distinct types of chondrodysplasia. The Schmid type, Pyle type, Spahr type, and Jansen type of achondroplasia have been more commonly documented [1-3]. They vary from mild distortion of the cartilaginous structures to severe malformations.

Metaphyseal chondrodysplasia is characterized by metaphyseal irregularities; hence, it can be mistaken for rickets [4]. The type of chondrodysplasia is very difficult to pin as it requires gene sequencing and testing that are not easily available and affordable in developing countries [5]. Literature is scarce regarding the various types of chondrodysplasias, and any correlation with dental anomaly. Literature has shown odontochondrodysplasia to be associated with dentinogenesis imperfecta [6].

Herein, we report a case of metaphyseal chondrodysplasia with dental abnormality. To the best of the authors' knowledge, this is the first known case of documented oral rehabilitation of a child with this abnormality.

CASE PRESENTATION

A 4-year-old child was brought by his father

with the chief complaint of carious broken teeth. The father complained that his child was unable to chew food and that food had to be mashed to paste.

Medical history:

The child was suspected with skeletal dysplasia on prenatal ultrasonography that revealed short bones and developmental delay in the 7th month of pregnancy. Delivery was normal, and the birth weight was 3.3kg. The child was breastfed until 2 years and had no bottle feeding thereafter.

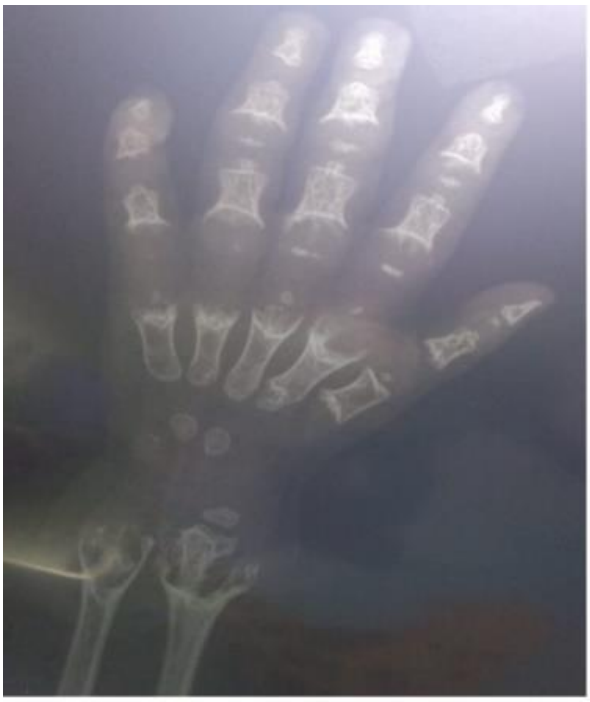

Fig 1. Hand wrist X-ray

Radiograph of the pelvis and both hips showed dysplastic metaphyses with cupping and cavitary changes [6]. Ultrasonography of the abdomen revealed dilation of renal pelvis and mild to moderate hydronephrosis of the left kidney. Renal profile revealed low serum creatinine level (0.3mg/dl) and low serum uric acid level (3.3mg/dl). Type of metaphyseal chondrodysplasia was not determined even after targeted gene sequencing (Medgenome) that was done when the child was 7 months old.

He gave a history of fracture of left femur 6 months earlier due to a fall; the fracture was treated under general anesthesia and showed normal healing. The child was able to walk normally without any support. Bone density test was done thereafter but T and Z scores are not available for the Indian population at this age group although reports showed that absolute bone mineral density appeared low at 0.78g/cm^2.

Family history:

The child' parents had a non-consanguineous marriage [7]. The child had an older female sibling who had no abnormalities.

Oral examination:

Oral examination revealed multiple root pieces in the upper and lower jaw (Fig. 2).

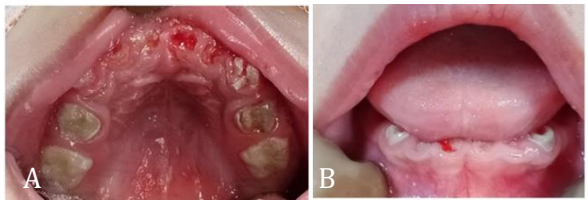

Fig 2. Upper (A) and lower (B) dental arch

The child was uncooperative so the panoramic radiograph was not clear but showed presence of only one-third of the root length. It was unclear whether it was because of early resorption or malformation. Extraction of 15 root pieces and restoration with stainless-steel crowns were planned for 5 teeth in a 2-stage manner under general anesthesia upon consultation with a pediatrician and pediatric orthopedic surgeon so as to prevent the risk of mandibular fracture since extraction of all 10 mandibular root pieces was required (Fig. 3).

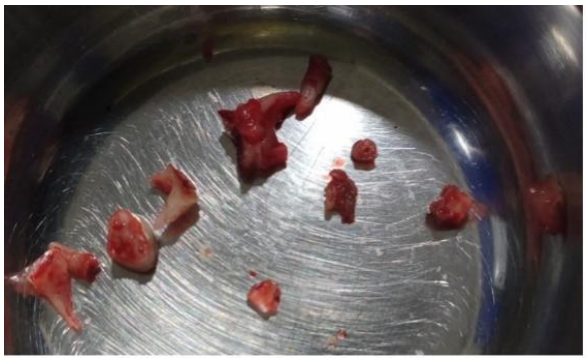

Fig 3. Extracted resorbed mandibular teeth

Stage 1: Formocresol pulpotomy and

restoration with stainless-steel crown for the upper left quadrant. Extraction of maxillary anterior incisors was also performed. Extraction of mandibular right quadrant teeth from the mandibular central incisor to primary second molar teeth was also performed. Bleeding was arrested by applying hemostatic Gelfoam and 30 vicryl resorbable sutures.

Stage 2: This stage was done 1 month after the first stage to ensure bone healing. Formocresol pulpotomy of upper right quadrant teeth with stainless-steel crown and extraction of the remaining mandibular root pieces were done. A maxillary putty impression was made for a functional space maintainer (Groper's appliance) (Fig. 4).

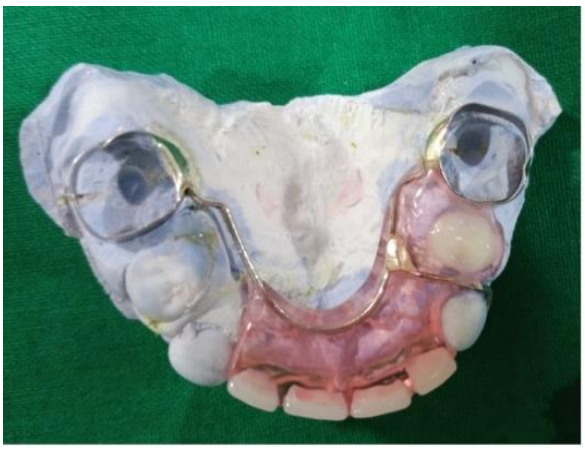

Fig 4. Groper's appliance

The child was recalled after 1 week, and the Groper's appliance was cemented. Healing of lower jaw was satisfactory. One month after treatment, primary impression with compound was made, custom tray was fabricated, and final impression was made after border molding with a putty-wash impression material. The child who was tense but cooperative was taught tongue movements in a fun manner to try to get an accurate impression.

This was followed by jaw-relation recording, teeth setting using custom modified teeth of A1 shade. The final complete denture was fitted (Fig. 5). The child was taught tongue moving exercises to prevent the denture from lifting, talking clearly with the appliance, and eating with the denture over the next 2 weeks so as to complete the treatment and rehabilitation of the child who had now become quite friendly (Fig. 6).

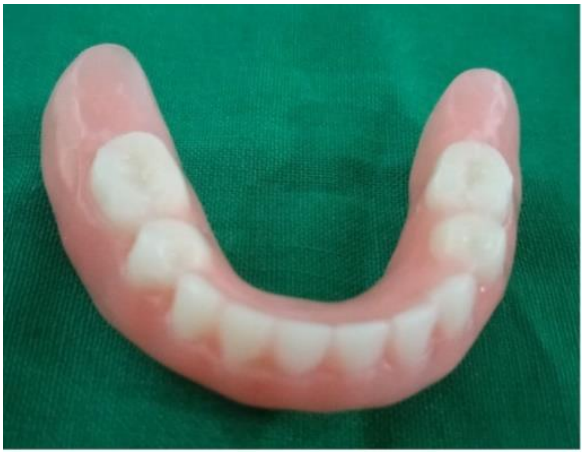

Fig 5. Lower complete denture

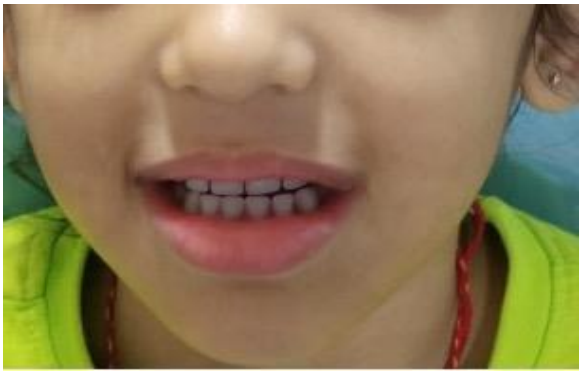

Fig 6. Postoperative complete rehabilitation profile

DISCUSSION

Metaphyseal chondrodysplasia is a rare genetic disorder with an incidence of 1-3 cases per one million and over 100 distinct types [2]. Its diagnosis and classification are difficult due to a number of unknown gene mutations. The child in this report was subjected to targeted gene sequencing and testing at the age of 7 months but the reports were inconclusive. The diagnosis was made by clinical and radiological examination. No further tests were conducted to classify the metaphyseal chondrodysplasia as the child had normal height, gait and intellect. Counseling was done in view of the hands' deformity, and parents were informed about the dangers of low bone density. The parents were advised to do symptomatic treatment and sent to a pediatric

dentist for restoration of broken teeth in their 4-year-old child.

On clinical examination, early childhood caries was suspected as the parents gave a history of breast feeding for 2 years [8]. As the child was uncooperative, it was initially perceived that the panoramic radiograph taken was of poor quality and unclear. An intraoral periapical radiograph of the anterior region revealed absence of roots (only one-third of the roots was present, even for the lower canine). This, along with the previous medical reports, made us suspect that the dental condition is related to the medical condition. A differential diagnosis of associated dentinogenesis imperfecta and dentin dysplasia was made. After going through literature, it was found that dentinogenesis imperfecta is associated with odontochondrodysplasia with similar general clinical and radiological features. The cause for this is a mutation on TRIP 11 mRNA but this was ruled out during targeted gene sequencing. Hence, we concluded that this was either a variant of odontochondrodysplasia targeting another gene or an entirely new form of metaphyseal dysplasia. After a session of counseling, the treatment plan was devised in 3 stages:

The procedure was performed in two stages under general anesthesia to try to reduce the risk of mandibular fracture as a result of low bone density and the fact that all the mandibular primary root pieces had to be extracted. The teeth that could be saved were pulpotomized and restored. Pulpectomy was not an option due to absence of roots and root canals. Also, formocresol pulpotomy was preferred for its mummifying effect on the remaining pulp tissue with the hope to stop any further root resorption. Restorations were finished with stainless-steel crown. This was followed by a rehabilitation phase using a fixed functional Groper's appliance and lower complete denture keeping in mind that the child was entering preschool [9,10]. The parents were concerned with clarity of speech and esthetics to prevent any psychological trauma to the child at school. The parents were also counselled about jaw development and that the lower denture my need to be replaced at regular intervals.

CONCLUSION

This case highlights the importance of thorough medical history and investigations in planning dental rehabilitation. It also emphasizes a holistic approach, with the support and advice of a team including a pediatrician and an orthopedic doctor, to provide the best treatment for a child with an unknown type of metaphyseal chondrodysplasia.

CONFLICT OF INTEREST STATEMENT

None declared.

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