

MOLAR-LIKE INCISORS: A RARE CASE OF MULTIPLE ANOMALIES OF THE DEVELOPING DENTITION

¹SAEEDA ABDULLAH, BDS, MCPS, FCPS

²ANSER MAXOOD, BDS, MSC, FRACDS

ABSTRACT

A rare case of a 9-year-old child is presented. The child was brought to the dental department of Children Hospital of Pakistan Institute of Medical Sciences, Islamabad in 2005, with severe toothache. His painful tooth was a mobile left lower central incisor which was cavitated, abnormal in shape and bigger in size. His other erupted teeth were also of abnormal shape (lower anterior teeth resembled molars and upper anteriors resembled premolars), hypocalcified and cavitated. The mobile tooth was extracted under local anaesthesia. The extracted tooth had a hollow crown without any root. The panoramic radiograph of the patient showed generalized macrodontia, abnormal shaped teeth with almost no roots in both primary and permanent dentition. There were missing primary and permanent anterior teeth in both jaws possibly resulting in impaction and ectopic position of the other teeth. Due to uncooperative behaviour of the patient's father investigations of any systemic involvement could not be performed. It is believed that no similar case has been previously reported in literature.

Key Words: Macrodontia, Severe Hypodontia, Oligodontia, Ectopic Eruption, Molar-like incisors, Generalized Macrodontia.

INTRODUCTION

Dental anomalies have been known to occur in humans due to a variety of genetic and environmental factors. Combinations of dental anomalies are mostly known to be associated with specific syndromes, though the combinations of dental anomalies have also been reported in patients with no generalized abnormality or disease. Some cases of multiple dental anomalies occur in families, while others have been reported in individuals with no other family member showing any such anomaly.¹

Macrodontia (enlarged tooth/teeth) is a rare anomaly of size and shape that results from disturbance at morpho-differentiation stage of the tooth development. Macrodontia is usually associated with systemic condition such as insulin-resistant diabetes² or syndromes such as otodental syndrome,³ facial hemi-hyperplasia, KBG syndrome, Ekman-Westborg-Julin syndrome, XYY syndrome.²⁻⁹ Macrodontia can be classified into "true generalized," "relative generalized," and "isolated" type. In true generalized macrodontia, all teeth are larger

than normal, and the condition may be associated with pituitary gigantism.¹⁰ Relative-generalized macrodontia refers to the presence of normal or slightly larger teeth in smaller jaws. Isolated macrodontia refers to macrodontia of a single tooth, and is an extremely rare condition that may be seen as a simple enlargement of all tooth-related structures or may be related with morphological anomalies. This type of macrodontia is more frequently found in incisors and canines.^{11,12} Depending on their size and morphology, macrodonts can create a variety of functional and esthetic problems that may require endodontic, prosthetic, surgical, and/or orthodontic treatment.¹³

Hypodontia or congenital tooth absence is a deficiency in tooth number. It results from disturbance at initiation (bud stage) of the tooth development. A tooth is considered to be congenitally missing when it cannot be identified clinically or radiographically, with no history of its extraction. Hypodontia/oligodontia may result in significant psychological, dental aesthetic and functional problems. It is classified as isolated, non-syndrome and syndromic.¹³ Hypodontia is also classified according to the severity of the condition. The term "mild-to-moderate hypodontia" is used to denote agenesis of two to five teeth, while the absence of six or more teeth, excluding the third molars, is termed as "severe hypodontia". Oligodontia is the absence of multiple teeth, usually associated with systemic disor-

¹ Assistant Professor, Dental Department, PIMS Islamabad, Pakistan
Email: dr_saeeda@hotmail.com

² Professor & Head of the Dental Department, PIMS, Islamabad, Pakistan

Received for Publication: April 3, 2014

Revision Received: May 14, 2014

Accepted: May 16, 2014

ders.¹⁴ Anodontia is the term used for absence of all of the teeth, usually seen with ectodermal dysplasia).¹⁵ The prevalence of permanent tooth agenesis ranges between 1.6% and 9.6%, and that of primary teeth between 0.5% and 0.9%.¹⁶

Enamel hypoplasia is defined as an incomplete or defective formation of the organic enamel matrix of the teeth in the embryonic stage of the tooth. There are 2 types of enamel hypoplasia i.e. hereditary and environmental. Hereditary type enamel hypoplasia is an ectodermal disturbance that occurred during the embryonic development of the enamel. The mesodermal components are normal. Both the deciduous and permanent teeth are involved and only the enamel is affected. Environmental type enamel hypoplasia is caused by the environmental factors like nutritional deficiency (vitamins A, C, and D), skin diseases (measles, chickenpox, and scarlet fever), congenital syphilis, Hypocalcemia, birth injury, Local infection and trauma. Both the enamel and the dentin of deciduous or permanent teeth may be affected.¹⁷

Supernumerary teeth are extra teeth in addition to the regular number of teeth. They can appear in any area of the dental arch. Supernumerary teeth can be classified by shape and by position. The shapes include supplemental, tuberculate, conical, compound odontoma, complex odontoma. When classified by position, a supernumerary tooth may be referred to as a mesiodens, a paramolar, or a distomolar.

Root resorption is a physiologic event for the primary teeth. Root resorption seems to be initiated and regulated by the stellate reticulum and the dental follicle of the underlying permanent tooth via the secretion of stimulatory molecules, i.e. cytokines and transcription factors. The primary root resorption process is regulated in a manner similar to bone remodelling, involving the same receptor system known as RANK/RANKL (receptor activator of nuclear factor-kappa B/RANK Ligand). Primary teeth without a permanent successor eventually exfoliate as well, but the literature is vague on how resorption of the dentine, pulp and periodontal ligament of the primary teeth occurs.¹⁸⁻¹⁹

This case report presents clinical and radiographic findings of an unusual occurrence of a combination of dental anomalies in a 9-year-old male patient. The dental anomalies in this patient included multiple enamel hypoplasia, generalized macrodontia, root resorption, hypodontia and supernumerary teeth.

CASE REPORT

A 9-year-old child was brought to Children's Hospital (Pakistan Institute of Medical Sciences, Islamabad, Pakistan) in year 2005, with a chief complaint of pain

in a lower anterior tooth. On intra-oral examination; abnormal shaped teeth were found. The tooth in pain was mobile and although located at lower central incisor region, it resembled a molar in shape. The patient's other teeth were also bigger in sizes and of abnormal shape. According to the father of the child, eruption of primary teeth started at the age of approximately 4 years. Patient never visited a dentist before. None of the primary teeth had yet exfoliated. Patient was mentally retarded and was not going to school due to this reason. There was no history of any such abnormality or other systemic problem in other family members. Parents were first cousins and other siblings were normal.

A panoramic view radiograph was taken that showed some teeth at bottom of mandible. The child's mobile tooth was extracted after taking photographs. The quality of the photographs was compromised as the child was not cooperating for the photography and refused mouth retractor. Patient was referred to a medical specialist for possibility of any syndrome. His brain CT scan showed some abnormality at mastoid region with no other significant pathology. After physical examination and analysis of CT scan report, the paediatrician suggested a provisional diagnosis of Ellis Van Creveld Syndrome. However, its definite diagnosis required ultrasound of abdomen and Echocardiogram. Appointment was arranged for both the investigations; however, the child was not brought back for any of the above investigations. The father of young patient took all record of CT scan and other documents for a second opinion from another dentist; nothing remained available to share except for few photographs and a radiograph. Duplication of reports was not carried out before handing over reports to child's parents due to several reasons including lack of interest from parents of the child in diagnosing the problem, overload on the hospital, and that there was no proper computerized backup or duplication facility of reports at that time.

After a year, child was brought again for another painful, mobile tooth for extraction. Once again, it was tried to convince the father of that child to allow complete systemic examination, diagnosis and treatment, but he declined the permission. This child since then has visited the hospital 4 to 5 times just for extractions in subsequent three years. His teeth were extracted one at a time, whenever they were loose and hurting the patient. All extracted teeth were almost hollow without any roots at the time of extraction.

No further visits of the patient have been recorded in the last 4-5 years. Patient could not be diagnosed for any syndrome or other systemic disease; however it was a rare case with multiple tooth abnormalities. Purpose of reporting of this case is to share an unusual case with dental community.



Fig 1: Frontal view of face

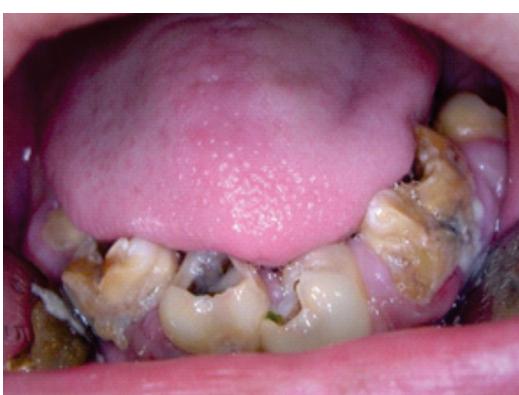


Fig 2: Intra oral view of mandibular teeth



Fig 3: Intra oral view of maxillary teeth

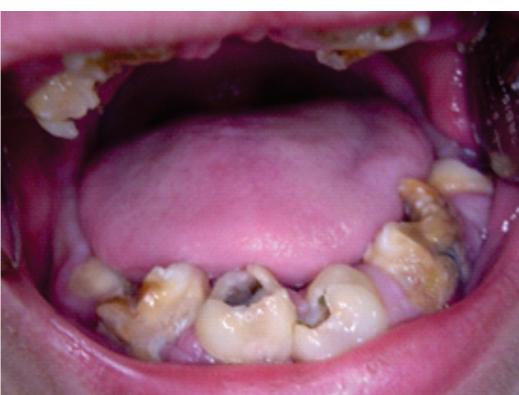


Fig 4: Any other Intra oral view mandibular teeth



Fig 5: Occlusion of the patient (Frontal view)

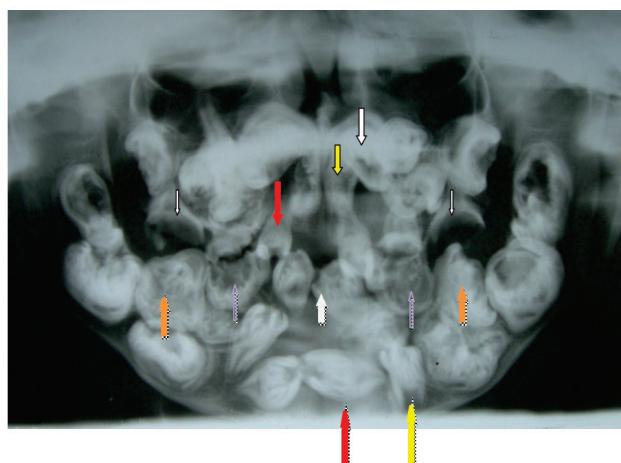


Fig 6: OPG of the patient

Maxilla: Red arrow = First premolar, Yellow arrow = Canine, White arrow = 2nd premolar, Narrow whitish arrows = second primary molars

Mandibular: Red arrow = First premolar, Yellow arrow = second premolar, White arrow = Primary first molar, Orange arrow = supernumery, Narrow whitish arrows = second primary molars.

DISCUSSION

Dental anomalies consist of a wide range of disorders including changes in the number, eruption, shape and size of teeth.²⁰ The developmental anomalies of teeth are caused during tooth development, whereas the acquired anomalies are caused after the tooth development is complete.²¹

The most common developmental and congenital dental anomaly is tooth agenesis. Congenitally missing teeth refers to teeth whose germ did not develop sufficiently to allow the differentiation to dental tissues.²²

The aetiology of macrodontia is not clear, but genetic and environmental factors have been proposed.¹ Generalized macrodontia has been associated with chromosomal anomalies like in XYY males (Klinefelter syndrome) and endocrine disorders such as pituitary gigantism and insulin resistant diabetes.¹³ Males demonstrate a higher frequency of macrodontia and hyperdontia, while females have a greater prevalence

of microdontia and hypodontia.¹ Localized macrodontia has been associated with syndromes like the oto-dental syndrome, especially in the posterior segment and patients with unilateral facial hyperplasia who exhibit macrodontia on the affected side.⁴

In the present case generalized macrodontia and abnormal shaped teeth in abnormal positions were found in normal jaw size. Radiograph confirmed the presence of macrodontia and abnormal shapes of both erupted and un-erupted teeth.

None of the patient's primary teeth were exfoliated; therefore it can be assumed that this patient was a case of hypodontia both with missing primary and permanent anterior teeth. It is possible that due to absence of his all primary mandibular anterior teeth, the primary molars lost the guidance to their path of eruption and because of their natural tendency to drift mesially, migrated in anterior mandibular region (erupted in place of lower incisors as shown in Fig 2 and 4). The reason for delayed eruption of his primary teeth may be that, the primary molars travelled a long distance from their original position to anterior mandibular segment. Teeth were cavitated, possibly due to hypocalcified enamel, poor oral hygiene and poor diet while abnormal short roots were probably resorbed due to underlying abnormally positioned teeth. These erupted mandibular teeth seem to be rotated as visible in the Fig 4. It can also be easily seen that mesiobuccal developmental groove of left lower anterior crown is on the distal side rather than on buccal side, which confirms its horizontal rotation at 90 degrees. Right lower anterior tooth was grossly carious and mesiobuccal developmental groove was not clearly visible due to large carious lesion. Radiograph also showed impacted teeth in mandibular anterior region. It can be assumed that these impacted teeth were permanent premolars (with red arrow in fig 6) and due to absence of permanent incisors they lost their guidance to correct path of eruption and resulted in impaction. In radiograph, mandibular tooth with yellow arrow would be second premolar. There are some supernumerary molar like teeth in mandible with orange arrow in radiograph (Fig 6). Similarly, it can be assumed that maxillary anterior permanent teeth were congenitally absent. Canine were impacted in anterior region (with yellow arrow in Fig 6). Maxillary teeth visible intra orally were first premolars (with red arrow in Fig 6), while white arrow in radiograph would be maxillary second premolar. Narrow arrows in both maxilla and mandible would be second primary molars, while an orange arrow in mandible indicates supernumerary teeth.

Cases of hypodontia/ oligodontia have been reported in literature previously.²³⁻²⁵ Akif (2007)²³ reported four cases of severe hypodontia/ oligodontia in with age

ranging from 11-19 years (two males and two females). He reported 11, 13, 14 and 19 congenitally missing permanent teeth (except for third molar) in these four cases. In his cases all the teeth present were normal, while in present case 12 teeth were missing and those present were hypocalcified and of larger than normal size. Pirinen (2001)²⁴ also reported 10, 16 and 20 absent permanent teeth in 3 siblings but again teeth present were of normal size and shapes. Abhinay (2013)²⁵ reported hypodontia and malformed roots with all molars having a single root.

There are so many other cases of hypodontia/ oligodontia and literature has shown studies which suggested a number of factors and genes responsible for hypodontia/ oligodontia, but mutations in very few human genes have been identified that result in hypodontia of specific teeth and associated tooth morphology. It has been suggested that epithelial signals such as bone morphogenetic proteins (BMP), fibroblast growth factor (FGF), sonic hedgehog gene (Shh) and wingless integration (Wnt) molecules determine the generation of distinct tooth shapes/classes. Reports suggest BMP4 directs the shape of incisors, whereas FGF8 directs the shape of molars.²⁶

Reardon (2010)²⁷ reported a case of macrodontia affecting all permanent teeth and exhibiting shovel-shaped maxillary/mandibular incisors, multi-tuberculate molars and premolars. Solanki (1996)²⁸ presented a case of Seip-Berardinelli syndrome in two sisters aged 12 and 14 years. The sisters presented with endocrinological disturbances along with dental manifestations including talon cusps, macrodontia, aberrant tooth morphology, and severe generalized crowding. Present case could not be diagnosed further due to lack of cooperation from child's parents.

Shahoon (2010)²⁹ reported 9 missing teeth in an 18-year-old, otherwise healthy male patient. Missing teeth were maxillary both lateral incisors, mandibular first premolars of both sides, right mandibular second molar and all third molars. Due to missing mandibular first premolars, guidance for the eruption of mandibular second premolars was lost, resulted in impaction and distally migrated second premolars. In the present case, as hypodontia was present both in primary and permanent dentition, almost all primary molars and permanent premolars migrated from their original positions.²⁹

Komatsu (2012)³⁰ presented a case with multiple macrodontia and proposed a diagnosis of Ekman-Westborg-Julin trait. In Ekman-Westborg-Julin trait there are some other dental anomalies like multi-tuberculosis, dental evagination, tooth impaction along with macrodontia, while present case had only macrodontia

and hypo calcification of enamel. Anomalies of the developing dentition result in aesthetics and functional disturbances, malocclusion, crowding, disfigurement and psychological problems. Present case was unique as patient had hypoplasia, oligodontia, thin small roots and macrodontia. No similar case could be found in literature. The condition of both erupted and unerupted teeth was poor due to hypoplasia and short or absent roots. It is expected that dentition of this child will be of short duration if impacted teeth ever erupted. There is also a possibility of cyst development around these unerupted teeth. The patient has not visited this hospital again since last 4 years; so his current oral condition cannot be reported. However, the patient is likely to need dental prosthesis in future to restore function and esthetics.

SUMMARY

The present case was unique due to presence of generalized dental anomalies in normal sized jaw. Any treatment to save his erupted or unerupted teeth for was not possible because of short or absent roots. Due to possibility of pathological changes in impacted teeth, such patients requires prolonged follow-up.

REFERENCES

- 1 Suprabha BS, Sumanth KN, Boaz K, George T. An unusual case of non-syndromic occurrence of multiple dental anomalies. Indian J Dent Res 2009; 20: 385-87.
- 2 Holmes J, Tanner MS. Premature eruption and macrodontia associated with insulin resistant diabetes and pineal hyperplasia, Report of two cases. Br Dent J 1976; 141: 280-84.
- 3 Van Doorne L, Wackens G, De Maeseneer M, Deron P. Otodental syndrome. A case report. Int J Oral Maxillofac Surg 1998; 27: 121-24.
- 4 Shafer WG, Hine MK, Levy BM. A textbook of oral pathology. Philadelphia: WB Saunders, 1974.
- 5 MacMillan AR, Oliver AJ, Reade PC, Marshall DR. Regional macrodontia and regional bony enlargement associated with congenital infiltrating lipomatosis of the face presenting as unilateral facial hyperplasia. Brief review and case report. Int J Oral Maxillofac Surg 1990; 19: 283-86.
- 6 Rowe NH. Hemifacial hypertrophy. Review of the literature and addition of four cases. Oral Surg Oral Med Oral Pathol 1962; 15: 572-87.
- 7 Herrmann J, Pallister PD, Tiddy W, Opitz JM. The KBG syndrome: a syndrome of short stature, characteristic faces mental retardation, macrodontia and skeletal anomalies. Birth Defects Orig Artic Ser 1975; 11: 7-18.
- 8 Ekman-Westborg B, Julin P. Multiple anomalies in dental morphology: macrodontia, multituberculism, central cusps, and pulp invaginations: Report of a case. Oral Surg Oral Med Oral Pathol 1974; 38: 217-22.
- 9 Ebru CH, Canoglu, Alper A, Zafer C and Cehreli. Isolated bilateral macrodontia of mandibular second premolars: A case report. European Journal of Dentistry July 2012; (6): 330-34.
- 10 Groper JN. Macrodontia of a single tooth: review of literature and report of case. J Am Dent Assoc 1987; 114: 69.
- 11 Solanki M, Patil SS, Baweja DK, Noorani H, PkS. Talon cusps, macrodontia, and aberrant tooth morphology in Berardinelli-Seip syndrome. Oral Surg Oral Med Oral Pathol Oral Radiol Endod Jan 2008; 105(1): e41-7.
- 12 Shafer WG, Hine MK, Levy BM. A textbook of oral pathology. 4th edition, pp 40 Philadelphia: WB Saunders, 1983.
- 13 Mainali A, Denny C, Sumanth KN, Ongole R. Non syndromic generalised macrodontia. J Nepal Dent. Assoc. 2010; 11(2): 150-3.
- 14 Schalk-Van Der Weide Y, Bosman F. Tooth size in relatives of individuals with oligodontia. Arch Oral Biol 1996; 41: 469-72.
- 15 Prevalence of congenitally missing permanent teeth in Iran-Prevalence of congenitally missing permanent teeth in Iran Sheikhi M, Sadeghi MA, and Ghorbanizadeh S. Prevalence of congenitally missing permanent teeth in Iran. Dent Res J Dec 2012; 9(1): 105-11.
- 16 Zarrinnia K, Bassiouny MA. Combined aplasia of maxillary first molars and lateral incisors: a case report and management. J ClinPediatr Dent 2003; 27: 127-32.
- 17 Quinonez R, Hoover R, Wright JT. Transitional anterior esthetic restorations for patients with enamel defects. Pediatr Dent 2000; 22(1): 65-7.
- 18 Harokopakis HE. Physiologic root resorption in primary teeth: molecular and histological events. J Oral Sci March 2007; 49(1): 1-12.
- 19 Bille ML1, Nolting D, Kvety MJ, Kjaer I. Unexpected early apical resorption of primary molars and canines. Eur Arch Paediatr Dent Sept 2007; 8(3): 144-9.
- 20 Neville BW, Damm DD, Allen CM, Bouquot J. Oral and maxillofacial pathology. 3rd ed. St. Louis: Saunders; 2009.
- 21 White SC, Pharaoh MJ. Oral radiology: principles and interpretation. 6th ed. St. Louis: Mosby Elsevier; 2009.
- 22 Kapdan A, Kustarci A, Buldur B, Arslan D, Kapdan A. Dental anomalies in the primary dentition of Turkish children. Eur J Dent 2012; 6: 178-83.
- 23 KifSümbüllü M. Severe hypodontia: four case reports and review of the literature. Atatürk üniv. Di_hek. Fak. Derg. Cilt:17, say": 1, y"l: 2007: 51-4.
- 24 Pirinen S, Kentala A, Nieminen P, Varilo T, Thesleff I and Arte S. Recessively inherited lower incisor hypodontia. J Med Genet 2001; 38: 551-56.
- 25 Abhinay A, Gundappa Mohan, Miglani S and Nagar R. Asyndromichypodontia associated with tooth morphology alteration: A rare case report. J Conserv Dent. May 2013; 16(3): 269-71.
- 26 Arte S, Nieminen P, Pirinen S, Thesleff I, Peltonen L. Gene defect in hypodontia: Exclusion of EGF, EGFR, and FGF-3 as candidate genes. J Dent Res 1996; 75: 1346-352.
- 27 Reardon GT, Slayton LR, Norby C and Geneser T. Macrodontia, shovel-shaped incisors, and multituberculism: probable Ekman-Westborg-Julin trait Sep 2012; 79(3): 197-201.
- 28 Solanki M, Patil SS, Baweja DK, Noorani H, PkS. Talon cusps, macrodontia, and aberrant tooth morphology in Berardinelli-Seip syndrome. Oral Surg Oral Med Oral Pathol Oral Radiol Endod Jan 2008; 105(1): e41-7.
- 29 Shahoon H, Esmaeili M. Bilateral Intraosseous Migration of Mandibular Second Premolars in a Patient with Nine Missing Teeth. Journal of Dentistry, Tehran University of Medical Sciences, Tehran, Iran 2010; 7(1): 50-3.
- 30 Komatsu T, Tae KuriharaYumi Ito, Lee MC, Miyagi A and Ikeda M. Oral characteristics of a patient with Ekman-Westborg-Julin trait: a case history. Spec Care Dentist 2012; 32(2) 70-4.