

Non Syndromic True Generalized Microdontia with Multiple Talons Cusp - Unusual Case Report.

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Abstract: Variation in size of the teeth can be either in the form of microdontia or macrodontia. Microdontia literally means small teeth. This condition is present in three forms involving single tooth, multiple teeth in large jaws and True Generalized Microdontia (TGM), in which all the teeth are smaller than normal. Very rarely, microdontia may affect all teeth, but seen in cases of pituitary dwarfism. The talons cusp is a projection of tooth structure usually of the cingulum of the maxillary incisors and rarely seen in rest of the teeth. This article emphasis a rare case report of true generalized Microdontia with multiple talons cusp in a non syndromic patient.

Keywords: microdontia, talons cusp, pituitary dwarfism, syndromes, eagles talon.

I. Introduction

Microdontia is the developmental alteration of the tooth involving the tooth sizes that are smaller than normal, outside the usual limits of variation. Boyle states that in general microdontia, the teeth are small with short crowns and normal contact areas between the teeth are frequently missing [1]. There are three types of microdontia, 1. Microdontia involving single tooth, 2. Relative generalized Microdontia due to relatively smaller teeth in large jaws, 3. True Generalized Microdontia, in which all the teeth are smaller than normal [2]. The third type is rare and been reported in pituitary dwarfism, radiation or chemotherapeutic treatment during the developmental stage of the teeth [3] and in Fanconis anemia [4]. Syndromes with Microdontia are seen examples being orofacial digital syndrome, oculo-mandibulo-facial syndrome, gorlin-chaudhry moss syndrome, Williams' syndrome, rothmund-thomson syndrome, hallermann streiff syndrome and chromosome 13 syndromes [5].

The talons cusp is the developmental tooth anomaly seen as a projection of tooth structure from cervical third to the lingual aspect due to alteration of the inner enamel epithelium [6]. Sometimes it is associated with syndromes like Rubinstein Taybi syndrome, Mohr syndrome, Sturge Weber syndrome, Ellis van Creveld syndrome, and incontinentia pig-menti [7, 8]. Certain racial predilections especially in Asians, Inuit, Native Americans and Arabs have also been reported [9]. This article emphasis a rare case report of true generalized Microdontia with multiple talons cusp in a non syndromic patient.

II. Case History

A 15 year old girl reported with the complaint of small teeth .Her parents noted small teeth since the eruption of the permanent teeth. Her mother had the same complaint and was treated earlier. The patient was short stature of 4.5 feet height similar to her mother, normal in appearance. (FIG-1). On external examination, her limbs, hands, hair, nail and skin appears normal. Intellectual and scholastic performance was normal. Her medical history revealed a growth hormone deficiency value of 0.30ng/mL at 3years and 2.10ng/mL at 11 years.

The intra oral examination shows normal soft tissues but the teeth are smaller in size (FIG-2). Diagnostic cast were made (FIG-3). Othopantomogram shows small permanent teeth in normal bone (FIG-4). Occlusal view and periapical radiographs show the talons cusps in the anterior teeth. (FIG-5, 6). The patient had permanent dentition with class I occlusion with spacing between teeth. Comparison of the patient's teeth measurement was done with anatomic average dental measurements. Over all the dentition was smaller than that of the average adult (Table-1). All maxillary anterior teeth and mandibular lateral incisors, canine and premolar showed lingual projections suggestive of talons cusp.

Except for the dental abnormality in the form of true generalized microdontia and multiple talon cusp, no other abnormal clinical feature were observed. A diagnosis of non syndromic occurrence of true generalized Microdontia with multiple talons cusp was made. The spacing was corrected with fixed prosthetic treatment.

III. Discussion

The initiating factors responsible for microdontia remain obscure. Mutation in developmental regularity of genes is the known cause for variety of dental defects [10]. Both genetic and environmental factors are involved in the complex etiology of microdontia. The variation in size of a tooth arises during the period when the tooth form is determined by the enamel organ and Hertwig root sheath at the bell stage. Studies showed that different regions of the oral epithelium rather than the underlying ectomesenchyme are responsible for the shape [11]. Studies by Alborg proved that generalized Microdontia can result from intrauterine growth retardation [12]. This patient has growth hormone deficiency which would have results in overall reduction in her growth with a decrease in tooth size. The various syndromes associated with TGM are Gorlin-Chaudhry-Moss syndrome, Williams syndrome, trisomy 13 syndrome, oculo mandibulo facial syndrome.

Gorlin-Chaudhry-Moss syndrome is characterized by a mild delay in physical development (growth retardation), short stature, mild mental retardation, and several physical abnormalities. In Gorlin-Chaudhry-Moss syndrome, premature closure of the fibrous joints (coronal sutures) between bones in the front (frontal bone) and sides (parietal bones) of the skull (craniosynostosis) may cause the head to appear abnormally short (brachycephaly). Individuals with Gorlin-Chaudhry-Moss syndrome may also have several abnormalities of the teeth. Some teeth may be absent (hypodontia), unusually small (microdontia) and/or abnormally shaped [13,14].

Williams' syndrome is a developmental disorder characterized by mild to moderate intellectual disability or learning problems, unique personality characteristics, distinctive facial features and cardiovascular problems. Young children with Williams' syndrome have distinctive facial features including a broad forehead, a short nose with a broad tip, full cheeks, and a wide mouth with full lips. Many affected people have dental problems such as small teeth, widely spaced, crooked or missing [15].

Trisomy 13 Syndrome is a rare chromosomal disorder in which all or a portion of chromosome 13 appears three times (trisomy) rather than twice in cells of the body affected infants and children. Developmental delays, profound mental retardation, unusually small eyes (microphthalmia), an abnormal groove in the upper lip (cleft lip), small teeth (microdontia) incomplete closure of the roof of the mouth (cleft palate), undescended testes (cryptorchidism) in affected males, and extra (supernumerary) fingers and toes (polydactyl) are seen in affected individuals [16]. Oculo-mandibulo-facial syndrome characterized by 'bird-like' facies with a beaked nose, brachycephaly and micrognathia. Microstomia with a shortened ramus and forward displacement of the temporomandibular joints is characteristic. A few teeth may even be present at birth (natal teeth) [17]. Our patient appears short statured with microdontia without any of the above abnormal clinical findings seen in syndromes

The term talon cusp was given by Mellor and Ripa in 1971, due to the appearance similar to an eagle's talon analogous to dens evaginatus [18]. The cause for talons teeth can be of genetic and environmental factors [7]. Hattab et al. postulated that the anomaly probably originated during the morph differentiation stage. Trauma to the tooth germ or endocrine disturbance could sometimes result in outward folding of the inner enamel epithelium and transient hyperplasia of the dental papilla [19]. Here the cause can be of endocrine growth hormone deficiency. The talon cusp may represent a spectrum of structural anomalies of normal cingulum, from an enlarged cingulum, a small accessory cusp, and to a fully formed talon cusp. It is mostly seen in the lingual surface of maxillary anterior teeth, though examples of facial cusps and even double cusps (facial and lingual) have been reported. [20].

The anomaly can be unilateral or bilateral; can be present with syndromes such as Rubinstein Taybi syndrome, Mohr syndrome, Sturge Weber syndrome, Ellis van Creveld syndrome, and incontinentia pigmenti. Mohr-Claussen syndrome, also called Oral-facial-digital syndrome II, is an autosomal recessive disorder comprising midline cleft lip, lobulated tongue, micrognathia, high/cleft palate, digital defects, absent incisors and ear defects. It is an extremely rare condition [9]. Rubinstein Taybi syndrome (RSTS) is an autosomal dominant condition comprising of developmental defects like short stature, facial and digital abnormalities, mental disability, and heart/kidney/eye defects. Distinctive facial features include broad nasal bridge and beaked nose, mandibular micrognathia, and arched brows. Risk of developing brain tumors and leukemia necessitates genetic testing and counseling. Talon cusp is a distinctive feature in this syndrome [21]. Ellis van Creveld syndrome (Chondroectodermal dysplasia) is an autosomal recessive disorder comprising dwarfism, heart defects, polydactyly and syndactyly. Oral anomalies include abnormal fusion of labial and alveolar mucosa, missing teeth, shovel shaped or taloned incisors, supernumerary teeth, early eruption etc [22].

Dental characteristics like talon cusps have been observed to have racial differences. Worldwide incidence is between 1 and 6%. A North Indian study had found the incidence to be 7.7%. Chinese populations seem to have higher proportion of talon cusps [23].

IV. Conclusion

TGM is the condition seen commonly in pituitary dwarfism, but along with the multiple talons cusp is rare. The wide variations in clinical manifestations in cases of non syndromic occurrence of dental anomalies are challenging. The dental malformation having both the size and shape of the teeth are rare which needs advanced research in future.

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Figure Legends



FIG 1. Showing short stature of the patient.



FIG 2-Intra oral examination showed missing 11 with talons cusp.



FIG 3. Maxillary and mandibular cast shows multiple talons cusp.

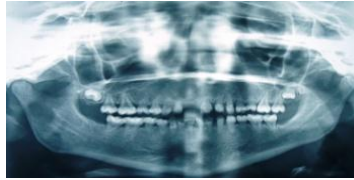


FIG 4. OPG shows smaller permanent teeth in normal jaw.

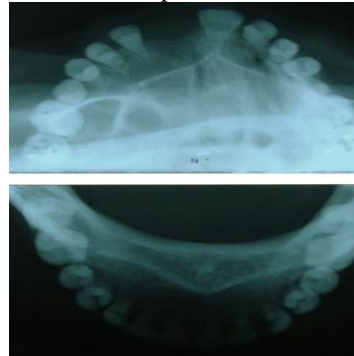


FIG 5- Occlusal view shows multiple talons cusp

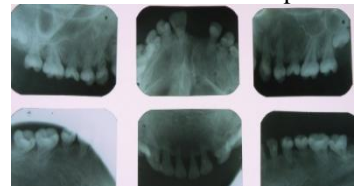


FIG 6- Periapical radiographs shows multiple talons cusp

Table 1

Table 1. Comparison of mesiodistal(MD), bucco or labiolingual(LL) and cervicoincisal or cervico occlusal(C) crown dimensions with an average of the maxillary and mandibular teeth. Tooth numbered by FDI system.

Tooth no.	average	Patient	Tooth no.	average	Patient
11	MD/LL/C 8.5/7.0/10.5	Missing	21	MD/LL/C 8.5/7.0/10.5	MD/LL/C 7.0/2.0/5.0
12	MD/LL/C 6.5/6.0/9	MD/LL/C 5.0/1.0/4.9	22	MD/LL/C 6.6/6.0/9.0	MD/LL/C 5.0/1.5/4.0
13	MD/LL/C 7.5/8.0/10	MD/LL/C 5.0/2.0/3.5	23	MD/LL/C 7.5/8.0/10.0	MD/LL/C 5.0/2.5/5.0
14	MD/LL/C 7.0/9.0/8.5	MD/LL/C 5.5/4.5/3.5	24	MD/LL/C 7.0/9.0/8.5	MD/LL/C 5.5/5.0/4.0
15	MD/LL/C 7.0/8.5/9.0	MD/LL/C 5.0/5.0/2.0	25	MD/LL/C 7.0/9.0/8.5	MD/LL/C 5.0/5.5/2.0
16	MD/LL/C 10.0/11.0/7.5	MD/LL/C 8.5/6.5/3.0	26	MD/LL/C 10.0/7.5/11.0	MD/LL/C 8.0/6.0/4.0
17	MD/LL/C 9.0/11.0/7.0	MD/LL/C 6.0/6.0/2.5	27	MD/LL/C 9.0/11.0/7.0	MD/LL/C 8.0/7.0/4.0
31	MD/LL/C 5.0/6.0/9.0	MD/LL/C 4.5/1.0/3.5	41	MD/LL/C 5.0/6.0/9.0	MD/LL/C 5.0/1.0/3.5
32	MD/LL/C 5.5/6.5/9.5	MD/LL/C 4.5/1.0/4.0	42	MD/LL/C 5.5/6.5/9.5	MD/LL/C 5.5/1.0/4.5
33	MD/LL/C 7.0/7.5/11.0	MD/LL/C 3.5/1.0/5.0	43	MD/LL/C 7.0/7.5/11.0	MD/LL/C 4.5/1.5/4.0
34	MD/LL/C 7.0/7.5/8.5	MD/LL/C 5.0/4.0/4.0	44	MD/LL/C 7.0/7.5/8.5	MD/LL/C 5.0/4.5/4.0
35	MD/LL/C 7.0/8.0/8.0	MD/LL/C 5.5/4.0/3.0	45	MD/LL/C 7.0/8.0/8.0	MD/LL/C 5.5/3.5/3.0
36	MD/LL/C 11.0/10.5/7.5	MD/LL/C 7.5/6.0/4.0	46	MD/LL/C 11.0/10.5/7.5	MD/LL/C 8.5/6.0/3.5
37	MD/LL/C 10.5/10/7.0	MD/LL/C 7.5/5.0/3.5	47	MD/LL/C 10.5/10/7	MD/LL/C 7.5/5/3.5

Anatomical average taken from wheeler textbook of dental anatomy and physiology edition 7. Patient – measurement in millimeters were taken at widest part of clinical crown on diagnostic cast