ORIGINAL ARTICLE

NON SYNDROMATIC OLIGODONTIA IN PERMANENT DENTITION: A RARE CASE REPORT

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ABSTRACT

Congenital absence of teeth is the most commonly seen developmental dental anomaly in pediatric dental clinic, although agenesis of first permanent molar has the least frequency. Its prevalence in the maxilla ranges from(0.2 to 1.5)% and in the mandible no data had been reported. The etiology of this anomaly is not exactly specified; in general it may occur in association with oligodontia or other dental disturbances. Despite the rare occurrence of first permanent molars' agenesis, it has a significant effect on the treatment plan outcome, as it plays a major role in food mastication, vertical dimension support of the face, and anchorage of teeth against orthodontic forces. In this study we have described the clinical and radiographic findings of a rare case report among Jordanian pediatric population of congenitally missing bilateral maxillary and mandibular first permanent molars.

Key words: *Agenesis, First permanent molars, Pediatric, Anomaly, Mandibular.*

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INTRODUCTION

Tooth agenesis is the most common developmental dental anomaly in humans.[1] Different terms are used to explain the absence of teeth, including hypodontia, oligodontia and anodontia.[1]

A congenitally missing tooth is diagnosed when it can't be identified on a radiograph and there was no history of extraction.[2]

In the general population the incidence of permanent tooth agenesis ranges from 1.6-9.6%, excluding third molars, while in primary teeth it ranges from 0.5-0.9% with a female to male ratio of 3:2 respectively.[3]

Several hypotheses have been reported to explain tooth agenesis, and general opinion is that the permanent first molars are always present and never missing. Agenesis of all first permanent molars has been reported in this case report, that falsifies the previous hypothesis.[3]

The most commonly missing tooth in frequency is third molar, mandibular second premolar, maxillary lateral incisor, maxillary second premolar and mandibular incisors.[4]

The etiology of tooth agenesis is not clear, but it may occur due to genetic and environmental factors such as infection of the jaw, trauma, rubella infection during pregnancy, chemical drugs and radiotherapy.[5]

Many studies have found a relation between tooth agenesis and mutations to genes:MSX1,PAX9,AXIN2,EDA,SPRY2,TGFA,SPRY4,WNT10A and others.

Oligodontia is also associated with genetic syndromes like: Ectodermal dysplasia, Down Syndrome, Pierre Robin syndrome, Ehler Danlos syndrome, Kabuki syndrome and syndromatic clefts.[1-5] Individuals with oligodontia tend to have deep bite, poor gingival contour and overeruption of the opposing tooth. Moreover they have difficulty in chewing. Treatment plan needs experienced dental specialists to maintain the existing teeth, improve esthetics and speech, and promote emotional well-being of the child.[1-5]

Despite the low prevalence rate of permanent first molar agenesis, it has significant problems that affect the treatment plan and outcome, because it plays an important role in mastication, supporting the vertical dimension of the face and anchorage of teeth against orthodontic forces.[6]

CASE REPORT

A 10 years old male patient was referred to the pediatric dentistry department at King Hussein Medical Center in the Royal Jordanian Medical Services, with presentation of multiple missing teeth. History was taken from the mother. She reported absence of any systemic disease or syndromes, but both the father and the sister had similar presentation, with different

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set of missing teeth. Past medical and family history was unremarkable. The mother had uncomplicated pregnancy, normal vaginal delivery, and no history of neonatal ICU.

Extraoral examination didn't show any abnormalities, however intraoral examination revealed that the patient was in the late mixed dentition stage, had fair oral hygiene, deep anterior overbite, retained lower primary central incisors, a stainless steel crown on tooth 84, large amalgam filling on 75, carious 74, a supplemental maxillary central incisor on the right side, and absence of 16,26,31,36,41,46. The patient had no history of trauma or extraction of the permanent teeth.(figures 1,2,3)

The following teeth were present intraorally:

15 54 12 11 11 21 22 23 64

85 84 43 42 81 71 32 73 74 75

A panoramic x-ray was taken and the diagnosis of missing teeth was confirmed ; all first permanent molars, mandibular central incisors, maxillary and mandibular left second premolars, maxillary right and left second molars were absent, and the buds of third molars were not visible.

The patient was referred to the genetic department of Queen Rania specialty hospital for children, The physician note stated that there was no specific syndromic association, but as both father and sister shared the same phenotype, we are dealing with an autosomal dominant disorder; wherein our patient and his sister got the abnormality from their father by vertical transmission. Case diagnosis was familial non-syndromic oligodontia .

Treatment of carious teeth, stabilization of oral hygiene, and regular follow up will be maintained in our pediatric dental clinic, orthodontic intervention will be postponed until the eruption of all permanent teeth, and further team work of prosthodontic and implantology specialists will be included.

DISCUSSION

Oligodontia is a rare developmental dental anomaly characterized by the absence of six or more teeth. The exact etiology for oligodontia is unknown, various factors have been described in the literature.[12]

Oligodontia has been classified as isolated or non-syndomic hypodontia, and syndromic hypodontia,[10] Hereditary or familial distribution has been suggested as the primary cause.[4]

Environmental factors such as trauma, radiotherapy, chemotherapy, hormonal and metabolic influences, and iatrogenic removal of a tooth germ during extraction of primary teeth may contribute.

In this case, based on the clinical and radiographic examination ten permanent teeth were missing. A supplemental maxillary right central incisor is pres-

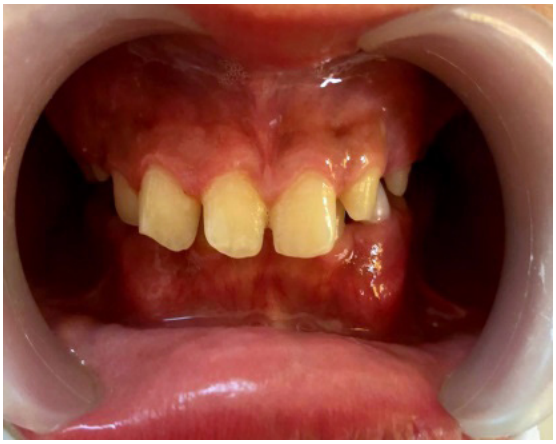

Fig 1: Frontal view of the patient's teeth

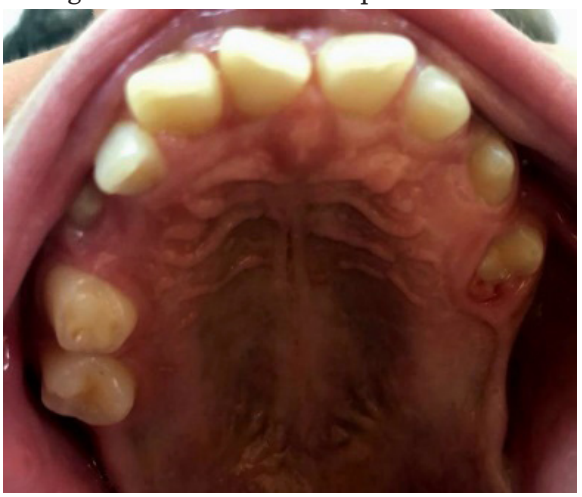

Fig 2: Intraoral picture of the maxillary arch.

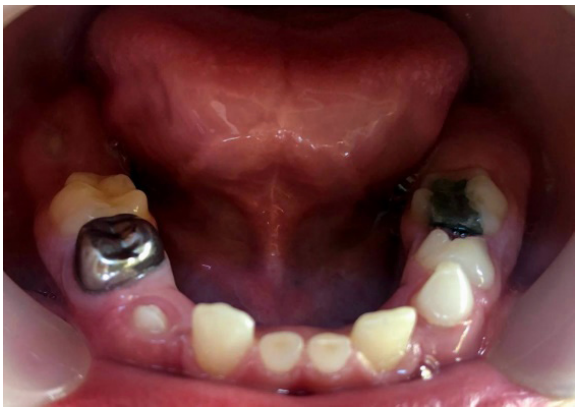

Fig 3: Intraoral picture of the mandibular arch.

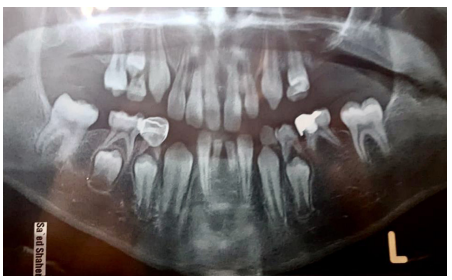

Fig 5: The panoramic radiograph showing agenesis of teeth:17,16,25,26,27,31,35,36,41,46

ent, no other dental anomalies like ectopic eruption or malformation of teeth was observed.

No systemic or any associated syndromes were noted, therefore this is a typical case of familial non syndromatic oligodontia.

Interestingly in this case we have 3 rare sets missing of teeth:

1. Bilateral permanent maxillary second molars (0.7- 0. 8)%.[15]
2. Bilateral mandibular central incisors (2.2)% .[15]
3. The four permanent first molars (0 - 0.02) %.[2]

The eruption time of permanent first molars in the oral cavity is considered to be the least variable among all permanent teeth.[2]

Usually at the age of 8-10 years, the first signs of third molars appear on radiographs and evaluated after 10 years of age.[14]

In familial oligodontia, the type of inheritance in majority of the families seems to be autosomal dominant, with incomplete and variable expressivity.[14] And this applies to the current reported case.

An autosomal recessive mode of inheritance is also possible. Ultimately, hypodontia carries an esthetic, functional, psychosocial and financial burden for affected individual.[5]

Treatment plan is usually complex and an interdisciplinary approach,including a pediatric dentist, an orthodontist and a prosthodontist is required.

The age of the patient, number and condition of retained teeth, number of missing teeth, condition of supporting tissues, occlusion and inter-occlusal space are all factors that should be taken into consideration while treatment planning.

Treatment options include orthodontic therapy, implants, adhesive techniques, removable and fixed prosthesis, and overdentures, and they are indicated depending on the condition and the age of the patient.[12]

CONCLUSION

Oligodontia presents an overwhelming challenge to both the patient and his family along with the dental professionals as it needs a well compliance and long term follow up in multiple dental disciplines.

This is a rare case of a familial non syndromic oligodontia with ten missing permanent teeth wherein improvement of the psychological aspect and self-confidence through restoration of esthetics, speech and masticatory function are our primary goal.

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CONTRIBUTIONS BY AUTHORS
All authors have contributed substantially.