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Non-Syndromic Oligodontia: A Rare Case Report

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ABSTRACT
Dental agenesis is a term referred to the absence of one or more teeth. However, oligodontia is a severe type of tooth agenesis involving six or more congenitally missing teeth, excluding the third molars. Oligodontia has a low prevalence and is a very rare condition. The aim was to show this case report of a 13-year-old female patient who presented oligodontia with absence of six permanent teeth and condylar atrophy on left side. The patient had no history of any syndrome or systemic disease according to the anamnesis. Is very important to know oligodontia features to perform a carefully treatment plan.
Key words: *Agenesis, Anodontia, Developmental Anomalies, Hypodontia, Oligodontia, Oral Abnormalities*

INTRODUCTION

Dental agenesis is the most common developmental anomaly in humans, often presenting a significant clinical problem. It is classified according to the number of missing permanent teeth excluding the third molars[1,2]. Hypodontia is used to describe the absence of one or few teeth, Oligodontia is used for agenesis of numerous teeth (more than six teeth) excluding the third molars and anodontia is the extreme of oligodontia where there is total absence of any dental structure[1,3,4,5].

Oligodontia is also known as partial anodontia, severe or advance anodontia. Some of them also refer this as selective tooth agenesis. According to different authors, the frequency of hypodontia is

1- 10% and oligodontia 0.1-0.9%. Anodontia occurs very rarely (17 cases were described over the last 50 years). Oligodontia may occur as a part of a genetic syndrome, as a non syndromic isolated familial trait, as an infrequent finding or as an isolated condition that has been linked to mutations of the MSX1andm PAX9.[5,6,7]

The third molar (M3) represents the tooth most affected with agenesis having a prevalence rate of 20.7%2. In contrast, permanent second molar (M2) agenesis is a rare occurrence, found in only 2 of 6,000 consecutive orthodontic patients (0.03%) [1,4,8]. Excluding the third molars, the prevalence rate of tooth agenesis is reported as 4.3 to 7.8%4, [5,9]. The mandibular second premolar (MnP2) is the tooth most often absent, with a relative frequency of 2.2 to 4.1% [4, 5]. In fact, the MnP2 is highly variable developmentally. Besides the high prevalence of agenesis, the MnP2 often shows significantly retarded development, especially when there is agenesis of other permanent teeth [6]. Despite the fact that the mean initial calcification age for MnP2 is 3 years (varying from 2y3m to3y7m) [7], its development can be suppressed until 6 years8, and some published reports show radiographic appearance of the MnP2 after the age of 9 and even at 13 years old [9, 10]. In addition, the MnP2 accounts for approximately 24% of all impacted teeth, excluding the third molars.[11]

The most frequent malposition reported for the unerupted MnP2 is distoangular development, with a prevalence rate of 0.2% in dental clinic patients[12]. This malposition was found to be associated with agenesis of the contralateral MnP2 Molecular studies have revealed that the instructive and permissive tissue interactions during mouse tooth development described above are mainly mediated by growth factor signalling. Development from initiation to eruption is governed by a sequential and reciprocal signalling process rather than simple one-way messages. The signalling involves all major signalling pathways, including transforming growth factor b (TGFb), fibroblast growth factor (FGF), sonic heghehog (Shh), anhidrotic ectodermal dysplasia (Eda), and epidermal growth factor (EGF) signalling, and studies with mouse mutants have shown that they are needed simultaneously during critical stages of development.[9,10,11,12]

Msx1 and Pax9 are transcription factors intimately involved in the genetic networks regulating tooth development. Msx1 contains a homeobox which binds to specific target sequences in the DNA but is also capable to proteins interaction. Msx1 has often been considered rather as a repressor than activator of gene expression. Pax9 belongs to the paired-box containing transcription factor family, and is one of the earliest mesenchymal markers of the future tooth forming positions in mouse. Pax9 is regulated by epithelial signals, especially FGF8, and it apparently regulates reciprocal signalling from the mesenchyme. In mice with hypomorphic Pax9 mutations, a partial failure of tooth development was observed, affecting in a dose-dependent manner the third molars and incisors and to a smaller extent the other molars. The ameloblast differentiation and dentinogenesis were also affected.[10,11,12]

It has been suggested that the key role of Msx1 and Pax9 is to facilitate the bud to cap stage transition. There is signals emanating from the

epithelium and mesenchymal during tooth development and molecular regulation Mesenchymal Msx1 expression is initially activated by the epithelial bone morphogenetic protein 4 (BMP4) signal, and needed for a reciprocal BMP4 signal from the mesenchyme. BMP4 and Msx1 thus form an autoregulatory loop. BMP4 signal to the epithelium is crucial for the formation of the epithelial signalling centre, the enamel knot, and the arrest of the development in Msx1 null mutant teeth can be rescued by external BMP4 or transgenically activated BMP4 expression. The expression of Pax9 is apparently needed to maintain and, by the synergism with Msx1, to enhance this loop and also needed later in tooth development.[9,10,11,12]

This article aims at presenting a case report of a non-syndromic young girl with several dental anomalies, contributing evidence to an understanding of genetically controlled dental anomaly patterns.

CASE REPORT

A 13-year-old female patient patient reported to my private Pediatric Dental Clinics reporting absence of some teeth. Through a digital panoramic radiograph the existence of multiple agenesis of permanent dentition was revealed. In the radiograph agenesis of tooth 1.5, 2.5,3.4, 3.5, 4.4,4.5 (Fig. 1,2) was identified, with a small dimension of maxilla. Also, a slight condylar asymmetry with a small size and a slight stylohyoid ligament ossification was noted in the left side. After this, a foot radiograph was taken to determine if the condition had a relation with an osteopetrosis; however, normal findings were noted. During anamnesis the patient reported she had no trauma history, previous tooth extraction, orthodontic treatment or complications during pregnancy or birth. The patient´s mother informed that there was no history of syndromic or systemic disease. (Fig. 3,4,5,6,7)

At general examination no alterations or systemic diseases were identified, with facial symmetry, no palpable lymph nodes and both jaws were normal. Clinically, in the intraoral examination no caries and the absence of the same teeth were observed with tooth rotation of 1.3, 2.3 and 4.3. No presence of periodontal disease was noted.

The patient was examined to rule out syndromes associated with oligodontia. She was normal in his facial appearance and did not show any physical or skeletal abnormality. Radiological examinations of the clavicles, vertebral skeleton, skull and chest were found to be normal. Ophthalmological and neurological examination of the patient revealed no pathological symptoms and showed no signs of mental retardation. Hematological and biochemical findings were within the normal limits.

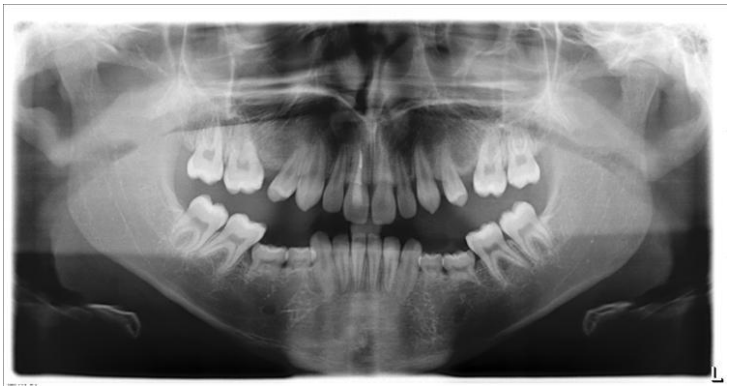

Fig: 1 2 missing upper premolars, and 4 missing lower premolar

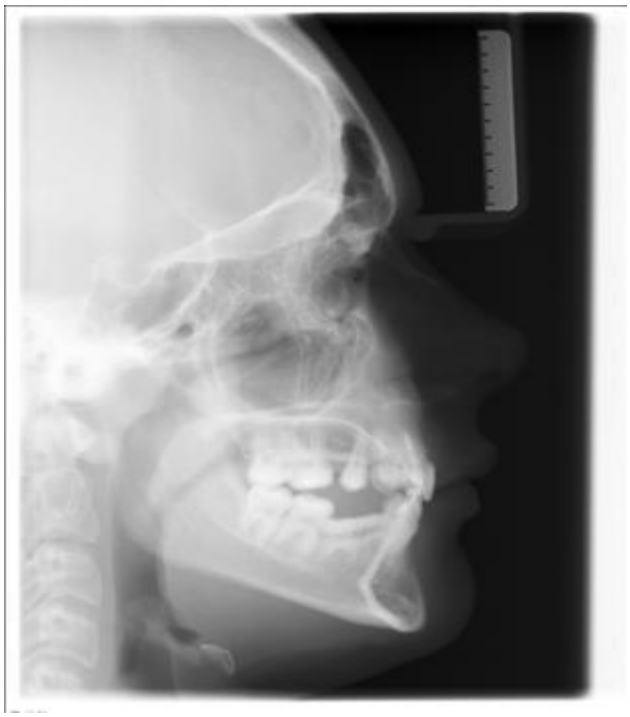

Fig: 2 Ceph view

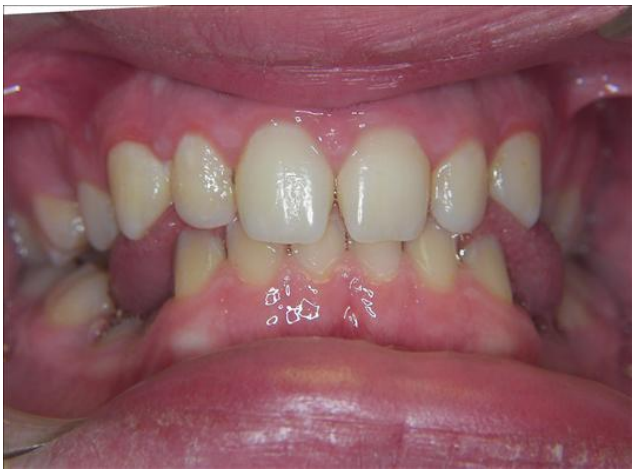

Fig:3 Front view

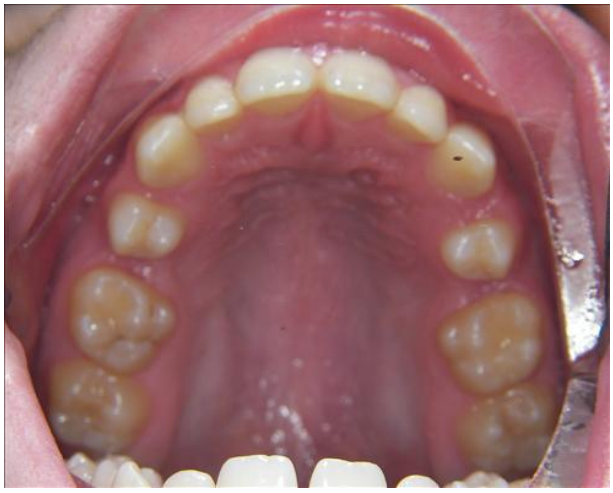

Fig: 4 Upper arch

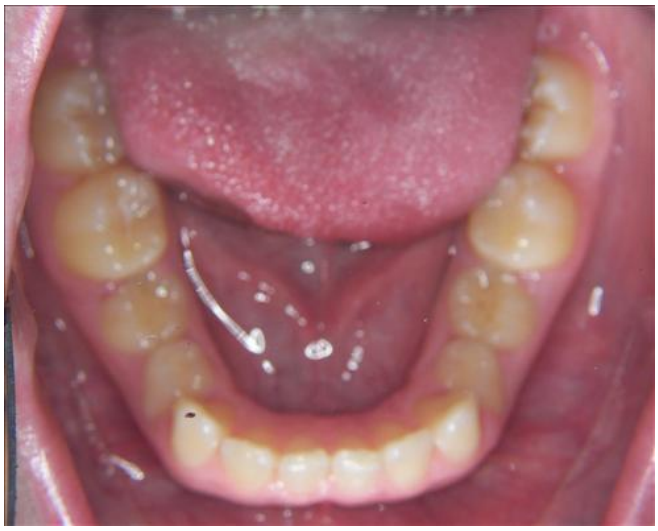

Fig 5 Lower arch

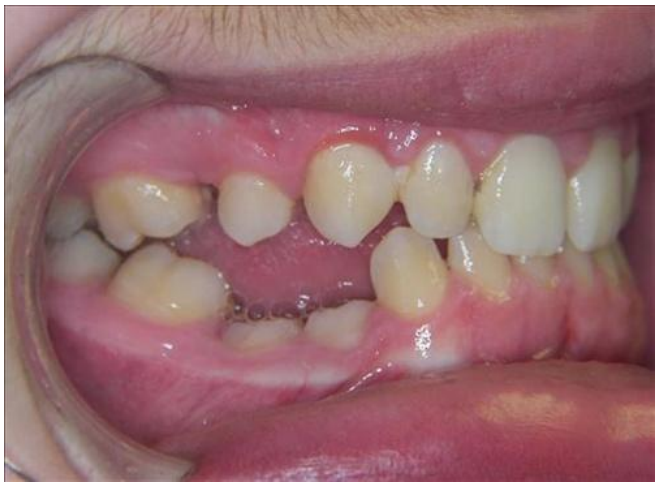

Fig: 6 Right retained primary teeth are ankylosed

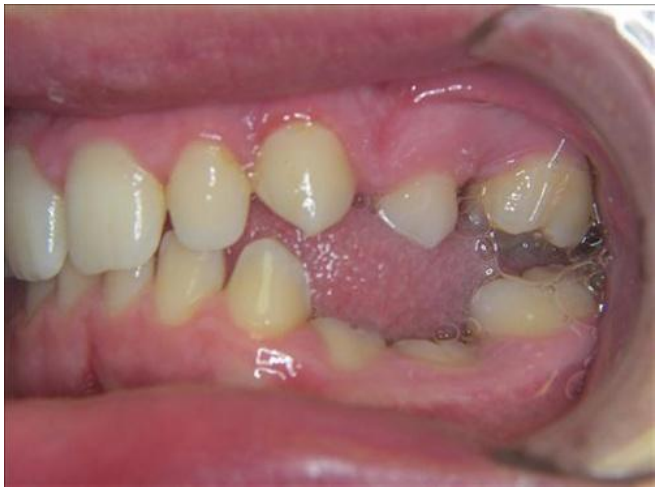

Fig: 7 Left retained primary teeth are ankylosed

DISCUSSION

A tooth may be considered to be developmentally missing when it cannot be discerned clinically or radiographically and no history exists of its extraction.[3] Hypodontia/oligodontia that may result insignificant psychological, dental, aesthetic and functional problems is classified as isolated or nonsyndromic, where as hypodontia/oligodontia and syndromic hypodontia/oligodontia or hypodontia/ oligodontia are associated with syndromes. Dhanrajani classified hypodontia according to the severe 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, indicates "severe hypodontia". Oligodontia is the absence of multiple teeth, usually associated with systemic disorders.[13]

Hypodontia and oligodontia are classified as isolated or non-syndromic hypodontia/oligodontia and syndromic hypodontia/oligodontia or hypodontia/oligodontia associated with syndromes.3 Most often oligodontia appears as part of some congenital syndromes that affect several organ systems.3 Oligodontia can occur in association with various genetic syndromes, such as ectodermal dysplasia, incontinentia pigmenti, Down syndrome, Rieger syndrome, Wolf-Hirschhorn syndrome, Van der Woude syndrome, Ectrodactyly- ectodermal dysplasia-clefting syndrome, Cleft lip palate ectodermal dysplasia syndrome, Oral facial digital syndrome type I, Witkop tooth-nail syndrome, Fried syndrome, Hair- nail- skin- teeth dysplasias.

In a survey conducted by Muller et al, found that girls had a higher rate of congenitally missing permanent teeth than boys. [14] Numerous studies have appeared on the prevalence of hypodontia in different countries, showing some variation in populations, on continents and among races. Family studies have shown the frequency of hypodontia and peg-shaped lateral incisor(s) in parents and sibs of the probands to be significantly higher than in the general population.

The prevalence of permanent tooth agenesis ranges between 1.6% and 9.6%, and the prevalence of deciduous tooth agenesis is lower, ranging between 0.5 % and 0.9 %.[15]

Dental anomalies can results from many factors, including genetic and environmental ones. Although defects in certain genes have the highest incidence, etiological events in prenatal and postnatal periods have also been blamed for anomalies in tooth number, dimension, morphology, position, and structure [16,17] this case report, the patient presented oligodontia because it had absence of six teeth excluding third molars. The predominance in females was according with the stated with Mattheeuws et al.[18] Also, this case concurs with Aktan et al[19]. who reported that similar number of missed teeth was in both sides and maxilla was more affected. Celikoglu et al. [20]found that the most frequently missing teeth were the maxillary lateral incisors, followed by the mandibular second premolars and the mandibular central incisors; however, in this case only second premolars agrees with the previously raised. The other teeth compromised were not reported in the literature. Some authors had found a relation between oligodontia and osteopetrosis [21] however, this hypothesis was

discarded when foot radiograph density was observed.

In the present case, the alveolar process was affected which concurs with Tavajohi-Kermani et al[22]. The absence of Six permanent teeth causes some several clinical problems with the dimension of maxilla and mandible. The above, was supported by Mattheeuws et al [18]. The small size in maxilla concurs with Bu et al. [23]) who found a smaller dimension in patients with oligodontia. All this leads to a very complexdental treatment, agreeing with Renault [24]. Patient´s history and anamnesis concurs with a non-syndromic patient. Finally, is very important to know oligodontia features to perform a carefully treatment plan.

CONCLUSION

Oligodontia cases should be evaluated carefully for the presence of any syndromes and managed appropriately. Patients suffering from oligodontia may have severe functional, esthetic and psychological problems. Hence, the management of such patients generally requires a multidisciplinary approach.

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