

Case Report

Congenital absence of all permanent second molars: a rare case report and literature review

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Abstract: Congenital absence of permanent second molar is an extremely rare condition, which may appear as part of a syndrome or as a non-syndromic form. Non-syndromic second molar agenesis combined with other types of teeth agenesis has occasionally been described in the literature but isolated cases are rarely observed. It might be caused by a number of complex interactions between genetic, epigenetic and environmental factors during the process of dental development. This report presents an isolated case of hypodontia with absence of bilateral maxillary and mandibular permanent second molars in a healthy 25-year-old male patient and a literature review on the prevalence, etiology, and restoration of the missing teeth.

Keywords: Hypodontia, congenitally missing teeth, multifactorial etiology, evolutionary

Introduction

Hypodontia is a common dental anomaly in population. Patients with hypodontia, may suffer from unfavorable appearance, malocclusion, periodontal damage and other problems. Accurate diagnosis of hypodontia requires radiographic, clinical, and dental cast examinations to distinguish whether the tooth is extracted, impacted, or congenitally absence. The prevalence of congenitally missing permanent teeth has been investigated in numerous studies. Reports of the prevalence of hypodontia in the literature vary widely, ranging from 0.1% to 2.4% for the primary dentition and 0.15% to 16.2% for the permanent dentition excluding the third molars [1].

According to most studies, the most commonly missing teeth were found to be the mandibular second premolars followed by the maxillary lateral incisors and the maxillary second premolars [2-4], while some other reports stated the maxillary lateral incisor might be the most commonly missing tooth [5-7]. Sheikhi *et al.* [8] re-

vealed that the least commonly missing teeth were first and second molars of both jaws, followed by mandibular canine, which agrees with several other studies conducted by Fekonja *et al.* [9] and Endo *et al.* [6]. Our report highlights a rare occurrence of hypodontia in an unusual site. This report presents a unique case of hypodontia with absence of bilateral maxillary and mandibular permanent second molars in a healthy 25-year-old male patient and a literature review on the prevalence, etiology, and restoration of the missing teeth.

Case report

A 25-year-old male student from Zhejiang University presented to Stomatology Hospital affiliated to Zhejiang University School of Medicine in January 2015, with a complain of bleeding when brushing his teeth. This study was conducted in accordance with the declaration of Helsinki. This study was conducted with approval from the Ethics Committee of Zhejiang University. Written informed consent was obtained from all participants. This was his first

Congenital absence of all permanent second molars



Figure 1. Intraoral view of maxillary arch showing the absences of 17 and 27.



Figure 2. Intraoral view of mandibular arch showing the absences of 37 and 47.



Figure 3. Panoramic radiograph showing the absences of 17, 27, 37 and 47.

dental visit. He was in good health with no history of trauma, extraction, systemic disease or any syndrome. No history of consanguineous marriage is reported between his parents. His mother gave no history of exposure to radiation or any medications during pregnancy. Physical examination revealed a face with normal facial profile and normal skeletal dental base relations. No abnormality in his skin and hair were found.

Intraoral examination (**Figures 1 and 2**) revealed no bilateral maxillary and mandibular permanent second molars. In order to detect permanent second molars, palpation of the buccal

sulcus and the lingual side of the alveolar process failed to indicate the presence of unerupted teeth. During clinical examination, a class I molar relationship with a centered dental midline and deep anterior overbite was presented. All other teeth were shaped in normality, and no decay was found in any tooth. Lingual side of mandibular anterior teeth deposited small amount of plaque and calculus, and gingival margin appeared slightly swollen. Oral mucosa appeared normal.

A panoramic radiograph (**Figure 3**) was taken to detect the presence and the location of the permanent second molar and to check for any other anomalies. The radiograph confirmed absence of all permanent second molars, and all third molar germs were also missing.

Finally based on above findings, non-syndromic hypodontia associate with chronic gingivitis as final diagnosis was justified. The treatment for the patient at his first visit was given supragingival scaling and oral health education. The patient didn't have problem of masticatory function, so no restoration was taken for the missing permanent second teeth eventually.

Discussion

Congenital missing teeth constitutes one of the most common developmental abnormalities seen in human populations. To describe dental anomalies, several different terms are being used. Hypodontia is defined as an absence of less than six permanent teeth, excluding third molars, whereas the absence of six or more teeth is referred to oligodontia. The most severe form of congenital missing teeth is anodontia denoting absence of the entire permanent or primary dentition. Hypodontia may occur either as part of a syndrome or as a non-syndromic form, which can be differentiated by conducting through physical examination of hairs, nails, sweat glands, eyes and to check for any congenital disorders [10]. If hypodontia is a part of a syndrome there are usually changes on the skin, hairs, nails, eyes, or skeleton. On the contrary, the patient presented here does not have peg-shaped teeth, dry-scaly skin, straw-like hair, frontal bossing, wide spaced eyes or any congenital disorders. He appears to be a perfectly normal youth, without history of missing teeth. The features in this patient are not consistent with any specific forms of ectodermal

Congenital absence of all permanent second molars

dysplasia or syndrome. Therefore the present case is diagnosed as non-syndrome hypodontia.

Little data were available in the literature because the incidence of permanent second molar agenesis is extremely low in the cases reported, ranging from 0% to 3.4% [3, 4, 6, 11, 12]. Aktan *et al.* [11] found 34 cases of second molar agenesis out of 10,057 Turkish population (0.6%), Whereas Cantekin *et al.* [12] could not detect any cases among 1,291 patients. In their meta-analysis, Khalaf *et al.* [2] demonstrated the overall prevalence of mandibular and maxillary second molar agenesis accounted for 1.8% and 1.5% relatively.

It was previously reported that single second molar agenesis is more predominant than multiple second molar agenesis and mostly occurs with other types of dental anomalies such as congenital absence of other teeth, microdontia, delayed tooth formation [5]. Second molar agenesis in previous studies seldom involves all permanent second molars. Few studies reported isolated absence of all permanent second molars. In our case there was neither congenital absence of other permanent teeth nor other dental anomalies associated with second molar agenesis.

The exact etiology of teeth agenesis is not clear, but in the majority of cases, genetics plays a crucial role in congenital dental agenesis. Many investigators have suggested that both *MSX1*, *PAX9* and *AXIN2* or genes are essential for early tooth development [13-15]. A number of subtle traits are apparent within human pedigrees possessing identifiable gene mutations in association with hypodontia. *MSX1* function is predominantly associated with premolar and occasionally molar agenesis, while *PAX9* mutations are almost always associated with missing molar and only occasionally premolar teeth. In contrast, hypodontia associated with *AXIN2* mutations involves a wider range of tooth types [16].

However, Brook *et al.* [17] advocated a single multifactorial theory that heredity was not absolutely the transmission way of hypodontia. It is caused by a number of complex interactions between genetic, epigenetic and environmental factors during the process of dental development. Several studies supported this

complex interaction of congenital missing teeth [18, 19]. The pattern of congenitally missing teeth can differ between monozygotic twins, possibly pointing to additional underlying mechanisms [19], such as epigenetic factors which might be implied by simultaneous occurrence of two anomalies [20]. This multifactorial etiology can include environmental factors as well, including multiagent chemotherapy, radiation therapy, fractures, surgical procedures on the jaws. However, this case showed the absence of all permanent second molars, and none of environmental factors mentioned above attributed to hypodontia in the present case.

Nevertheless, an evolutionary trend towards fewer teeth has been proposed as a contributing factor in hypodontia [1]. Those which are vulnerable to absence are actually valueless organs for modern people. Without any selective advantage, these teeth will be eliminated inevitably in the process of evolution. Just like multifactorial etiology resulting in impacted third molars or absence of third molars, one of factors for the present case might be evolutionary to adapt with the gradually shrinking size of the jaws.

The treatment of congenitally missing teeth is comprehensive. A number of factors should be taken into account for treatment planning: the condition of the deciduous teeth, the patient's occlusion, facial growth pattern, and preferences of the patient. It might represent an interdisciplinary challenge for specialists in oral and maxillofacial surgery, operative dentistry, pediatric dentistry, orthodontics and prosthodontics [21, 22]. The principal aim of treatment in such cases is to replace the missing teeth and improve the patient's appearance, speech, and masticatory efficiency. Absence of teeth may be managed with fixed/removable prostheses and dental implants. With development of dental implantology, implant-supported prostheses may be chosen increasingly for treatment of congenitally missing teeth. However, implantation during childhood should be restricted to those patients who may not be conventionally provided with an aesthetically and functionally satisfying prosthodontic restoration because of the extent of missing teeth present.

However, the present case was found occasionally, and it involved neither aesthetic nor func-

Congenital absence of all permanent second molars

tional problem. The patient has been accustomed to the masticatory efficiency for a dozen year, which differs from that of tooth loss resulting from various factors acquired. So it is unnecessary to give prosthodontic restoration for the missing teeth in the present case, and the patient doesn't want to restore them at all.

Disclosure of conflict of interest

None.

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