doi: 10.7213/archivesoforalresearch.09.001.RC05 ISSN 2236-8035
Archives of Oral Research, v. 9, n. 1, p. 111-115, Jan/Apr. 2013

Non-syndromic oligodontia in permanent dentition: a rare case report

Oligodontia não sindrômica na dentição permanente: relato de caso raro

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Abstract

Objective: Tooth agenesis is one of the most common congenital anomalies seen in humans. Although absence of one or more teeth is common, absence of multiple teeth is rare. Oligodontia is a rare developmental anomaly, involving agenesis of six or more permanent teeth, excluding the third molars. The reported prevalence of oligodontia in permanent dentition is 0.14%. Oligodontia can be presented as an isolated condition or as a part of a syndrome. **Discussion**: The present case report highlights a unique case of non-syndromic oligodontia, with agenesis of four permanent incisors, left permanent canine and right second premolar in the mandibular arch and its management with a novel fixed functional prosthetic appliance. **Conclusion**: Prosthetic rehabilitation is an urgent need for these kind of patients so that they do not suffer from masticatory and esthetic problems which can eventually lower the self esteem of individuals.

Keywords: Oligodontia. Missing mandibular incisors. Agenesis permanent canine.

Resumo

***Objetivo**: A agenesia dentária é uma das anomalias congênitas mais comuns em humanos. Embora a ausência de um ou mais dentes seja comum, a ausência de vários dentes é rara. A oligodontia é uma anomalia de desenvolvimento rara, envolvendo agenesia de seis ou mais dentes permanentes, excluindo os terceiros molares. A prevalência de oligodontia na dentição permanente é de 0,14%. A oligodontia pode apresentar-se como uma condição isolada ou como parte de uma síndrome. **Discussão**: O presente relato destaca um caso único de oligodontia não sindrômica, com agenesia de quatro incisivos permanentes, canino permanente esquerdo e segundo pré-molar direito no arco inferior, e seu manejo com um novo aparelho de prótese fixa funcional. **Conclusão**: A reabilitação protética é uma necessidade urgente para este tipo de paciente para que ele não sofra de problemas de mastigação e estética, os quais podem eventualmente reduzir a autoestima do indivíduo.*

***Palavras-chave**: Oligodontia. Incisivos inferiores ausentes. Agenesia do canino permanente.*

Introduction

Tooth agenesis is the most prevalent craniofacial congenital malformation in humans (1). Various terms used to explain absence of teeth are hypodontia, oligodontia and anodontia. Hypodontia is the developmental absence of one or more teeth. Oligodontia refers to agenesis of six or more teeth excluding third molars, and anodontia is the complete absence of teeth (2). The incidence of oligodontia is reported to be from 0.08% to 0.16%. The most commonly missing permanent teeth are the third molars (9-37%), mandibular second premolars (< 3%), maxillary lateral incisors (< 2%), maxillary second premolars and mandibular incisors (< 1%) (3).

The exact etiology of agenesis of teeth is not clear but genetic factors are thought to play a definite role. Grahnen stated that in majority of cases, oligodontia is mainly determined by a dominant autosomal gene pattern with incomplete penetrance of the trait and variable expressivity (4). Anomalies in the development of the mandibular symphysis, shortened arches as an evolutionary trend and endocrinal disturbances may affect dental tissues responsible for forming tooth buds, thus resulting in congenital absence of lower incisors (5). The reported environmental factors associated with agenesis of teeth are localized infection of the jaw (which might destroy the tooth buds), any disturbance of the endocrinal system, systemic infection (rubella), trauma in apical region of dentoalveolar process (due to fractures or while doing extraction of deciduous teeth), chemical substances or drugs (use of thalidomide or chemotherapy), radiation therapy or disturbances in jaw innervations (6). Clinicians often claim that incidence of dental agenesis has increased during recent decades (7). However there is no evidence to say whether this is an evolutionary trend or an observation as a result of more advanced screening and diagnosis of oral anomalies.

Oligodontia may occur as a feature of a specific disease such as (i) anhidrotic ectodermal dysplasia, (ii) incontinentia pigmenti (an X-linked genetic disorder that affects the ectodermal structures and is associated with congenital tooth absence and abnormal form), (iii) Down's syndrome, (iv) Pierre Robin syndrome, (v) Ehlers-Danlos syndrome, (vi) Limb mammary syndrome, and (vii) Kabuki syndrome (8, 9). It can also present as an isolated condition (non-syndromic) and has been linked to mutations of MSX1, PAX9 and EDA genes (10). However the pattern of tooth absence is not just influenced by the gene affected but the type of mutation in gene as well (11, 12).

There is a wide arena of treatment options available ranging from removable/fixed prosthesis to dental implants at a later age. This paper presents a unique case of non-syndromic oligodontia, with agenesis of four permanent incisors, left permanent canine and right second premolar in the mandibular arch and its management.

Case report

An 8-year-old girl reported to the Department of Pediatric Dentistry with the chief complaint of missing lower front teeth. The parents had observed this condition since early childhood but did not seek any dental consultation, and there was no history of any previous trauma or extraction. Past medical and

family histories were noncontributory. On extraoral examination a well balanced face, with an apparent eye defect and a convex facial profile was observed. Intraoral examination revealed (i) mixed dentition stage, with (ii) mesial step molar relationship, (iii) root stumps of 52, 61, 62, 73, 84, (iv) missing 41, 45, 31, 33 and (v) multiple carious teeth (Figure 1, 2). The alveolar ridge in the mandibular anterior region was typically knife edged (Figure 2), suggesting absence of mandibular anterior teeth. To ascertain the provisional diagnosis, an orthopantomograph was advised and revealed no evidence of development of 31, 32, 33, 41, 42, 45 (Figure 3). Parents were informed about the agenesis of six mandibular permanent teeth.

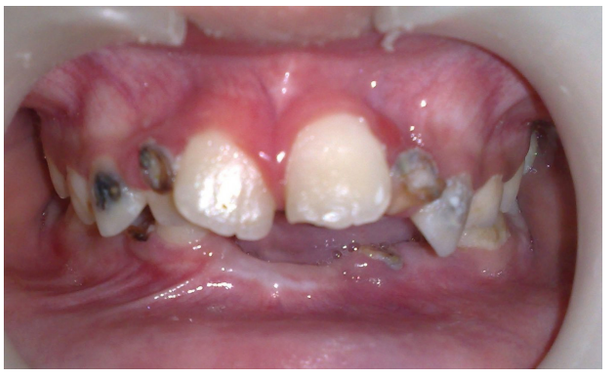

Figure 1 - Preoperative photograph of teeth in occlusion

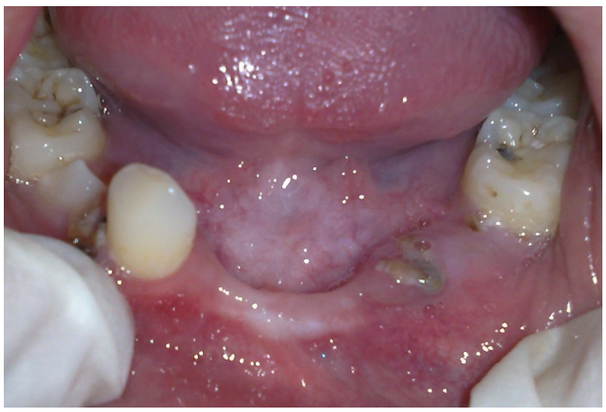

Figure 2 - Preoperative photograph of mandibular arch

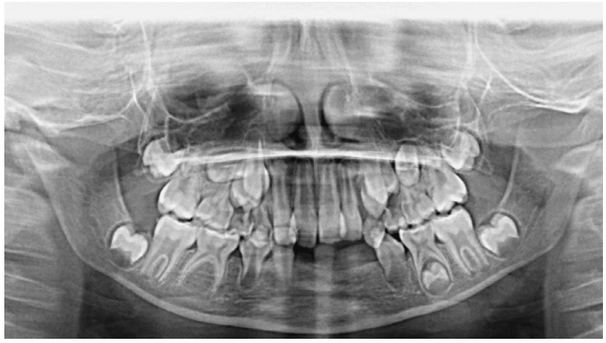

Figure 3 - Orthopantomograph of the patient

To rule out any syndromic involvement, the patient was referred to Department of Pediatrics, and the medical evaluation of the child revealed absence of any syndrome. Further, Karyotyping (Figure 4) and Cyto SNP assay were done to detect any chromosomal aberrations responsible for agenesis of the teeth; however no significant findings could be ascertained.

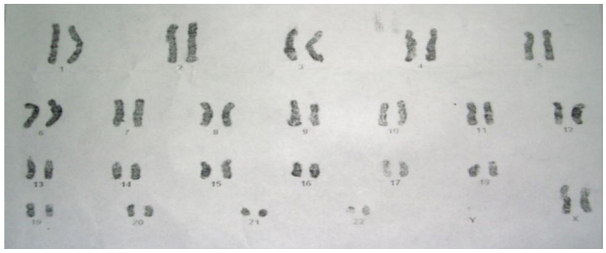

Figure 4 - Karyogram of the patient without any abnormalities

The treatment plan comprising of thorough oral prophylaxis, placement of pit and fissure sealants, restoration of carious teeth, extraction of root stumps of deciduous teeth, followed by the fabrication of a fixed prosthesis for the missing teeth was discussed with the parents. After obtaining informed consent from the parents, the necessary restorative and preventive treatment was carried out. Following this an intermediate fixed prosthetic replacement of the congenitally missing mandibular teeth was done using acrylic teeth attached to a lingual arch (Figure 5, 6). The patient was instructed to maintain good oral hygiene and parents were asked to adhere to a regular follow-up dental visit schedule.

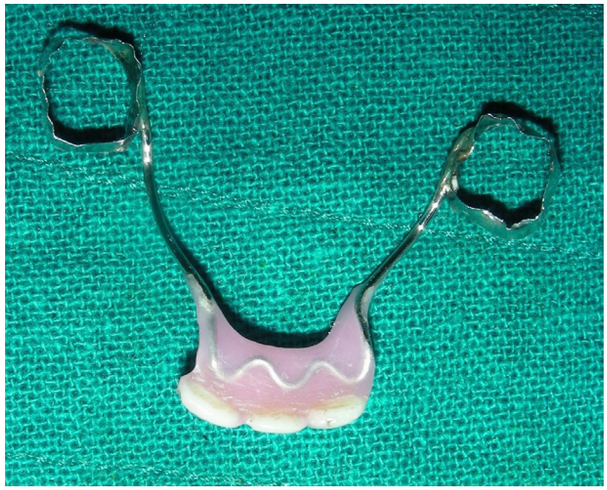

Figure 5 - Photograph of appliance before insertion

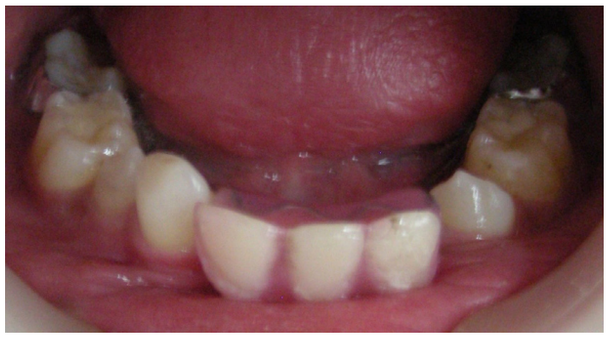

Figure 6 - Photograph depicting appliance after cementation

Discussion

A tooth may be considered to be congenitally missing if it cannot be discerned clinically or radiographically, and no history exists of its extraction (13). Radiographically the diagnosis of tooth agenesis can be made after the age of six years (Pirinen and Thesleff) as all permanent tooth crowns – except the third molars – appear to have begun their mineralization by this age (14). The absence of six missing permanent teeth is very rare (0.14%) (15). The child in our case report was eight years old, and no evidence of calcification of the six permanent teeth could be seen, thereby ascertaining the agenesis of the aforementioned teeth.

Different theories have been put forward to explain the etiology of agenesis of teeth. Kjaer and coworkers (16) proposed a theory stating that tooth agenesis may result from an abnormality in peripheral nerve supply, in the overlying epithelium (as seen in ectodermal dysplasia) or in supporting bone. They demonstrated that teeth located near peripheral nerve endings are the most affected by agenesis and hypodontia could be associated with missing mandibular canal. However, in this case report, the overlying mucosa was found to be normal and radiographic examination of mandible showed the presence of both mental foramen and mandibular canal, suggesting a normal neural innervation. Tooth patterning has also been explained by two hypothetical models, the field theory and the clone theory. The absence of mandibular incisors along with one mandibular canine and contralateral premolar in our case report was unique and could not be explained by any of these models as it would require bilateral absence of teeth.

To date three genes, namely, the homeobox gene MSX-1, paired domain transcription factor PAX 9, and EDA, have been associated with non-syndromic oligodontia (17). The known mutations in these genes are possible to screen but the analysis is available at select centres for research, so the test to rule out gene mutations could not be done for the present case of hypodontia discussed in this article.

In this case report, oligodontia could not be associated with any syndrome as the thorough assessment by the pediatrician, Karyotyping and Cyto SNP assay did not reveal any significant findings. None of the family members suffered from a similar condition, suggesting absence of hereditary basis to the present defect. The prenatal and postnatal histories were noncontributory to suggest any environmental cause. Agenesis of mandibular incisors has been reported (5, 18) but unilateral absence of a second premolar and contralateral canine is rare.

The treatment options available for dental prosthesis vary from removable/fixed prosthesis to dental implants at a later age as dental implants can be given once the period of active growth is over in order to prevent infraocclusion of the implant. So the choice of treatment in the present case was that of a removable or a fixed prosthesis but as the child was school-going and did not want a removable prosthesis, hence the decision for fabrication of a fixed prosthesis was made.

The design of the appliance comprised of a wire mesh over the ridge area (clearance of 1.5 mm) of missing teeth, soldered to the bands on permanent molars. Keeping in mind the space required for erupting premolar on left side, three incisors were

placed in heat cure acrylic over the mesh. A periodic recall was advised every three months when the appliance would be removed and fluoride application would be done on banded molars to prevent their decalcification. The parents were informed that prosthesis would need to be replaced as per growth changes from time to time and at a later age dental implants would be a better treatment option for complete rehabilitation.

Conclusion

A patient with missing teeth suffers not only from masticatory and esthetic problems but psychological stress as well, as it can lower the self-esteem of an otherwise healthy individual. A Pediatric dentist might be the first one to witness a case of hypodontia and therefore dental rehabilitation requires orthodontic and prosthetic intervention at the appropriate time.

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Recebido 12/07/2013
Received: *07/12/2013*

Aceito: 22/08/2013
Accepted: *08/22/2013*