Case Report

Nonsyndromic Oligodontia in Siblings

V. Vinothini, A. Sanguida, K. R. Prem Lal[1], G. S. Prathima

Departments of Paedodontics and Preventive Dentistry and [1]Oral Pathology and Microbiology, Indira Gandhi Institute of Dental Sciences, Sri Balaji Vidyapeeth, Puducherry, India

Abstract

Tooth agenesis is a condition where the teeth are missing due to developmental failure. Congenital tooth agenesis can be either hypodontia or oligodontia. Oligodontia can occur either as an isolated condition or it can be associated with other genetic syndromes. The exact etiology of oligodontia is unknown. A multidisciplinary staged approach of the management is required that includes endodontic, restorative, surgical, and orthodontic procedures to improve the esthetics and function. The present article reports a rare case of oligodontia in siblings identified and treated in the mixed dentition with 1-year clinical follow-up.

Keywords: *Agenesis, ankyloglossia, familial, hypodontia*

Introduction

Congenital agenesis of one or more permanent teeth is one of the most common oral anomalies. Congenital absence of six or more teeth excluding third molars is termed as oligodontia, and it may or may not be associated with syndromes or severe systemic abnormalities. Oligodontia is commonly associated with syndromes such as anhidrotic ectodermal dysplasia, van der Woude syndrome, Down syndrome, Pierre Robin syndrome, and Ehlers–Danlos syndrome.[1,2] Dental features in oligodontia include microdontia, ectopic eruption, taurodontism, enamel hypoplasia, and delayed eruption. Problems associated are those with esthetics, mastication, speech, functional problems such as malocclusion, periodontal damage, inhibition of alveolar growth, and consequent psychological impact.[3] Hence, early diagnosis is important and treatment decisions are made based on the age, esthetic need for rehabilitation, and condition of the remaining teeth.

Case Report

A 9-year-old female and her 7-year-old male sibling reported to the department of pedodontics. The girl had come for a routine dental checkup, and the boy with a chief complaint of broken upper front teeth while playing. Their medical histories were noncontributory, and family history revealed that they were born of nonconsanguineous marriage. Their father had multiple congenitally missing teeth in both primary and permanent dentitions; his remaining teeth were affected with caries; and he had been wearing complete dentures. General and extraoral examinations of the children did not reveal any other abnormalities. The girl's past dental records revealed root canal treatment in tooth number 26 and extraction of teeth numbers 54 and 64. The boy's past dental records revealed restoration in maxillary incisors and extraction of grossly decayed tooth number 64. Intraoral examination of the girl revealed the presence of the following primary teeth: 52, 53, 55, 62, 63, 65, 71, 72, 73, 74, 75, 81, 82, 83, 84, and 85 and the following permanent teeth: 11, 16, 21, 26, 36, and 46. All teeth except 11, 21, 71, 72, 73, 81, 82, and 83 were grossly carious. She also had ankyloglossia (Kotlow's class III).[4] The child also reported difficulty in pronouncing certain words. The panoramic radiograph revealed the absence of 17 permanent teeth excluding third molars: two maxillary lateral incisors, four mandibular incisors, one canine (23), all eight premolars, and two maxillary second molars [Figure 1].

Intraoral examination of the boy revealed the presence of all primary teeth except tooth number 64 and all permanent first molars. Teeth numbers 51 and 54 were root stumps, and 53, 55, 65, 74, 75, 84, and 85

Address for correspondence: Dr. V. Vinothini, Department of Paedodontics and Preventive Dentistry, Gandhi Institute of Dental Sciences, Sri Balaji Vidyapeth, Pillayarkuppam, Puducherry, India. E-mail: vinothinibds@gmail.com

How to cite this article: Vinothini V, Sanguida A, Prem Lal KR, Prathima GS. Nonsyndromic oligodontia in siblings. J Indian Acad Dent Spec Res 2018;5:52-4.

Access this article online

Quick Response Code:

Website: www.jiadsr.org

DOI: 10.4103/jiadsr.jiadsr_10_18

were grossly decayed. Panoramic radiograph revealed the absence of 14 teeth excluding third molars: two maxillary lateral incisors, four mandibular incisors, and all eight premolars [Figure 2].

Following discussion of the treatment plan with the parents, complete rehabilitation of the dentition was done which included extraction of root stumps, pit and fissure sealants, restorations, pulpectomy, stainless steel crowns, and a functional band and loop space maintainer in 74 for the boy [Figure 3] and restorations, stainless steel crowns, and laser-assisted lingual frenectomy for the girl [Figure 4]. The parents were advised to come for regular follow-up to monitor the eruption of the permanent teeth. Figure 5 shows the status of the dentition of the children 1 year after a complete rehabilitation.

Discussion

Oligodontia can be isolated (oligodontia – I) or occur as a part of a syndrome (oligodontia – S).[5] The condition shows female predilection (3:2) with the incidence varying from 0.08% to 0.16%.[6] Isolated oligodontia may be familial or may manifest *de novo*. Environmental factors such as localized infection, radiotherapy, chemotherapy, trauma, or injudicious use of certain drugs could be causes for sporadic cases. Familial oligodontia may result from single dominant gene defect, X linked, or recessive.[7] About 50% of siblings would be affected if autosomal dominant inheritance had full penetrance.[8] In a systematic review and meta-analysis of the genetic background of nonsyndromic oligodontia, it was found that genes PAX9, EDA, MSX1, AXIN2, EDARADD, NEMO, and KRT17 in decreasing order of frequency had the potential to cause nonsyndromic oligodontia.[9] It has also been found that half of the mutations causing isolated nonsyndromic oligodontia are found in the gene WNT10A.[10] The prevalence of agenesis is more common in the permanent dentition (1%–10%), while in primary dentition, it is 0.5%–1%.[11] The present case report on siblings is an interesting one as more than ten permanent teeth, excluding third molars, were missing involving both the arches and occurring bilaterally. The father was also affected. It was familial and not associated with any other systemic abnormalities. It was not possible to construct a pedigree chart as parents could not give correct information regarding the occurrence of agenesis in other relatives. Another uniqueness of this report is the association of ankyloglossia with

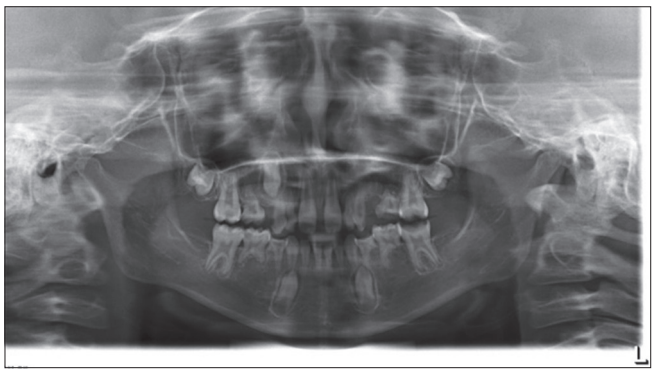

Figure 1: Panoramic radiographs of the female child

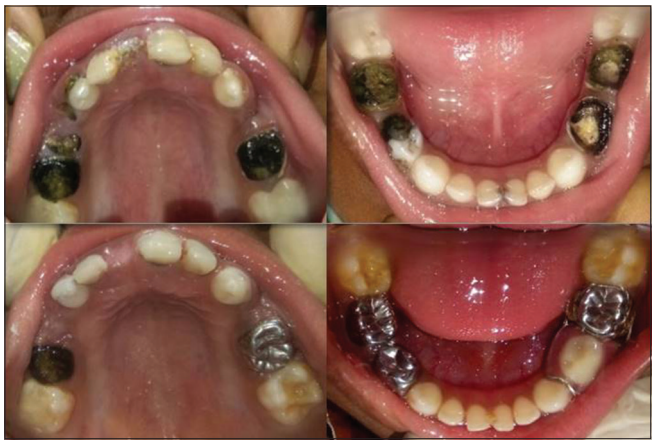

Figure 3: Preoperative and postoperative occlusal views of the male child

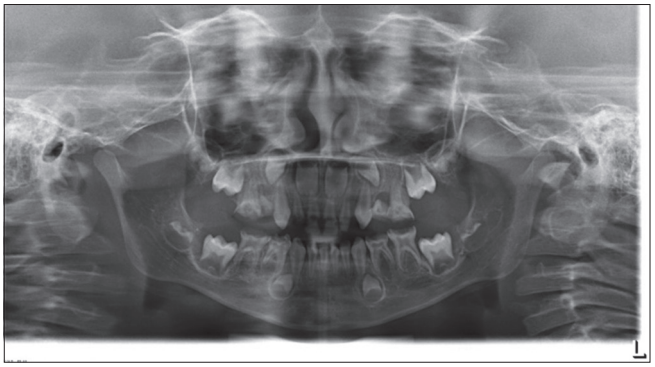

Figure 2: Panoramic radiographs of the male child

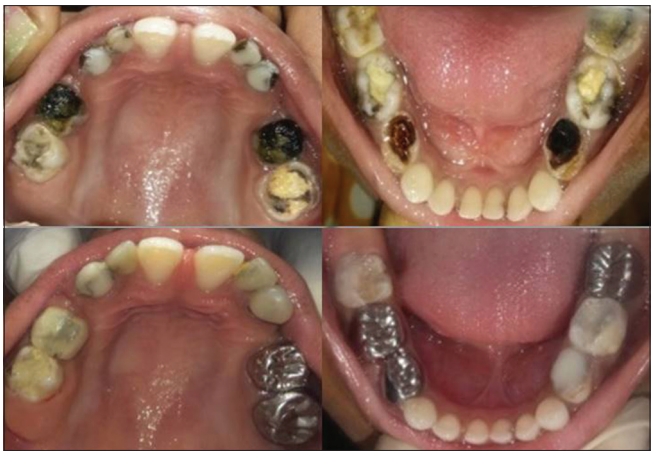

Figure 4: Preoperative and postoperative occlusal views of the female child

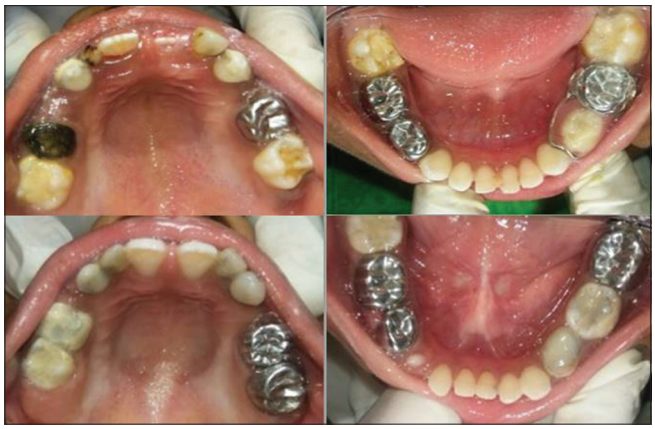

Figure 5: Occlusal views of the male child (Top) and occlusal views of the female child (Bottom) 1 year after rehabilitation

oligodontia in the female child. Ankyloglossia generally shows male preponderance.[12] The treatment objectives in the children in the present report were to maintain the primary teeth in good health for mastication, esthetics, preservation of arch length, and psychological well-being. Oligodontia is rare and when diagnosed parents should be made aware of this condition and explained the need for multidisciplinary approach in management. Future definitive treatment in these children would involve orthodontics, tooth transplantations, implants, fixed, and removable prostheses and would cover a 10–20-year period.[13] Genetic counseling is also necessary to discuss issues related to inheritance. Early diagnosis, thorough investigations, and patient cooperation are crucial to the successful management of oligodontia in growing children.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Rathore R, Deepshika R, Piyush A, Don V, Sumita K. A non-syndromic hereditary oligodontia-a rare case report. J Oral Med Oral Surg Oral Pathol Oral Radiol 2016;2:180-3.
2. Rakhshan V. Congenitally missing teeth (hypodontia): A review of the literature concerning the etiology, prevalence, risk factors, patterns and treatment. Dent Res J (Isfahan) 2015;12:1-3.
3. Ephraim R, Rajamani T, Feroz TM, Abraham S. Agenesis of multiple primary and permanent teeth unilaterally and its possible management. J Int Oral Health 2015;7:68-70.
4. Kotlow LA. Ankyloglossia (tongue-tie): A diagnostic and treatment quandary. Quintessence Int 1999;30:259-62.
5. Tangade P, Batra M. Non syndromic oligodontia: Case report. Ethiop J Health Sci 2012;22:219-21.
6. Hosur MB, Puranik RS, Vanaki SS. Oligodontia: A case report and review of literature. World J Dent 2011;2:259-62.
7. Closs LQ, Weissbluth MF, Nakamura E, Hermann FP. Esthetic and functional rehabilitation for oligodontia in the mixed dentition: Case report. J Dent Child 2012;79:193-6.
8. Alshame A, Ramalingam K, Abdalla K. Nonsyndromic familial oligodontia: A case series from Libya. Univ Res J Dent 2016;6:234.
9. Ruf S, Klimas D, Hönemann M, Jabir S. Genetic background of nonsyndromic oligodontia: A systematic review and meta-analysis. J Orofac Orthop 2013;74:295-308.
10. Dahllof G, Jacobsen PE, Martens L. Children with chronic health conditions: Implications for oral health. In: Koch G, Poulsen S, Espelid I, Haubek D, editors. Pediatric Dentistry: A clinical Approach. 3rd ed. Chichester, West Sussex (UK), Ames, Iowa: John Wiley and Sons Inc.; 2017. p. 358.
11. Guttal KS, Naikmasur VG, Bhargava P, Bathi RJ. Frequency of developmental dental anomalies in the Indian population. Eur J Dent 2010;4:263-9.
12. Naik SV, Ghousia S, Shashikran ND, Naik S, Shashihushan KK. Occurrence of partial ankyloglossia with isolated oligodontia in permanent dentition: A case report. Pediatr Dent J 2014;24:124-8.
13. Koch G, Thesleff I, Kreiborg S. Tooth development and disturbancesin number and shape of teeth. In: Koch G, Poulsen S, Espelid I, Haubek D, editors. Pediatric Dentistry: A Clinical Approach. 3rd ed. Chichester, West Sussex (UK), Ames, Iowa: John Wiley and Sons Inc.; 2017. p. 33.