

CASE REPORT



Nonsyndromic oligodontia - A rare case report

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Abstract

Oligodontia is a rare congenital disorder which clinically presents as missing six or more teeth in deciduous, permanent, or both dentitions. Usually, oligodontia is seen as a part of few syndromes rarely occurs as an isolated entity. Msh homeobox 1 and paired box 9 genes are found to be responsible for nonsyndromic oligodontia. We present a case report a 17-year-old male patient, who had the absence of multiple teeth. General physical examination revealed there was no abnormality in either hair or nails, perspiration was normal and no congenital clefts of lip or palate were seen suggesting that he was not suffering from any syndromes. Hence, in our case, oligodontia was not associated with any syndrome which is a rare finding.

Keywords: Congenital, developmental, nonsyndromic, oligodontia**Introduction**

Various developmental anomalies of teeth can occur. These anomalies may be either primary or arise secondary to environmental influences (e.g., concrescence, hypercementosis, and dilaceration).^[1]

Several terms are used in the discussion of the variations in a number of teeth of teeth. Anodontia is a condition in which there is complete absence development of the tooth. Hypodontia is a condition in which there is lack of development of one or more teeth; oligodontia (a subdivision of hypodontia) is a condition in which there is a lack of development of six or more teeth. In hyperdontia, there is the development of more number of teeth than normal. These terminologies are applied to teeth that failed to erupt or develop and should not be used to describe teeth that developed but are impacted or have been removed.^[2]

Oligodontia is a rare finding which is rarely reported in India. The prevalence rate of oligodontia in India is about 0.3% in the permanent dentition. Prevalence is more in females than males with a ratio of 3:2. Hypodontia in deciduous dentition is rare (0.1-0.9%) and shows no significant gender predilection. Most frequently missing is the mandibular second premolar after the third molar, followed by maxillary lateral incisor and the maxillary second premolar. The absence of maxillary central incisors, canines, or the first permanent molars is relatively uncommon.^[3]

Hypodontia and oligodontia can be further classified as isolated or nonsyndromic hypodontia/oligodontia and syndromic hypodontia/oligodontia or hypodontia/oligodontia which is associated with syndromes.

Typical dental findings are decreased number of teeth, reduced tooth size, anomalies of tooth morphology, and delayed eruption. Lack of teeth in young patients can cause cosmetic, functional, and psychological problems particularly if the teeth of the anterior region are involved.^[4] We present a case of 17-year-old patient with multiple congenitally missing teeth.

Case Report

A 17-year-old male patient came with a complaint of missing multiple teeth [Figure 1]. History of presenting illness revealed the teeth did not erupt after the shedding of deciduous teeth 5 years back. Family history revealed that his parents were a consanguineous marriage. No other member in the family had similar problem of missing multiple teeth. There was no history of extractions or trauma. No abnormalities were detected on extraoral examination. Intraoral examinations revealed that teeth number 12, 13, 14, 15, 22, 23, 31, 34, 35, 41, 44, and 45 were missing [Figures 2 and 3].

The patient was advised for an orthopantomograph which showed missing multiple teeth [Figure 4]. Since there were no other clinical features of any syndrome, we have a provisional

diagnosis as nonsyndromic oligodontia. Differential diagnosis of ectodermal dysplasia, Rieger syndrome and Van der Woude syndrome was given.

To rule out other etiologies, complete blood investigations, serum Ca, alkaline phosphate, and thyroid function tests were advised. The findings of these investigations were normal. Based on the reports, nonsyndromic oligodontia was given as final diagnosis.

The patient was referred to the department of oral and maxillofacial surgery for the further management [Figure 5].

Discussion

Etiology of congenital oligodontia is thought to be due to the failure of the lingual or distal proliferation of the cells of tooth bud from the dental lamina. Predisposing factors of hypodontia/oligodontia are environmental factors such as radiation, tumors, trauma, hormonal factors, and viral infections like rubella drugs like thalidomide or to hereditary.^[5] Mutation in genes Msh homeobox 1 (MSX1) and paired box 9 (PAX9) plays an important key factor in early development of the tooth. PAX9 is a paired domain transcription factor which plays a major role in odontogenesis.^[5]

Etiopathogenesis

Exact cause of oligodontia is unknown. Multiple factors have been reported in previous literature.^[6] Heredity is one of the main etiological factors. Other factors include environmental factors such as viral infections and irradiation or chemotherapy can affect the formation permanent teeth. However, most of the cases are caused by genetic factors. Most of the earlier literature shows the heritable nature of congenitally missing teeth. Genetic factors may be autosomal dominant or recessive, and it is obvious that in many cases multiple genetic and environmental factors are acting together. The appearance of multiple cases among relatives and higher incidence in identical than in nonidentical twins has shown the importance of genetic factors. It has been reported that several genes which, when mutated may cause congenital absence of multiple teeth.^[6]

The MSX1 gene is located in the chromosome number 4 which is involved in several epithelium–mesenchymal processes of embryogenesis. This gene is commonly expressed in the mesenchyme of the tooth buds, during its early stages.

The PAX9 gene is located on the chromosome 14 and belongs to a family of genes which are involved in transcription factors which act during the initiation of the embryo's development. Expression of PAX9 gene is required for dental mesenchyme to condense around the epithelium of developing tooth bud.

Protein products of the PAX9 and MSX1 genes play a major role in the maintenance and expressions of mesenchymal bone morphogenetic protein 4, which initiates the morphogenesis of the dental organ. Defects in the PAX9 and mutations in the MSX1 gene have been identified in humans, thus, once there is an expression defect of these genes, there might be arrested of



Figure 1: Profile picture



Figure 2: Intraoral view showing missing multiple maxillary teeth



Figure 3: Intraoral view showing missing multiple mandibular teeth

tooth bud development, leading to lack of development of tooth bud.^[6]

Oligodontia may cause various disturbances such as abnormal occlusion, altered facial appearance which can lead to psychological problems, difficulty in mastication and speech. Dental management may vary depending on the severity of the disease and requires a multidisciplinary approach. Early diagnosis



Figure 4: Orthopantomograph showing missing multiple teeth



Figure 5: Orthopantomograph showing implants placed in mandibular arch

of the condition is essential in such cases with a complete clinical history, medical, and radiological investigations to rule out any syndrome.^[7]

Conclusion

Oligodontia is mostly considered to be associated with several syndromes, but nonsyndromic aspect of oligodontia should also be taken into consideration. Thus, dental team should aim at managing patients with oligodontia, as early as possible and achieve both prosthetic and esthetic functionality of teeth.

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