Anodontia of Permanent Teeth — A Case Report

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ABSTRACT

Anodontia is a rare genetic disorder which represents the congenital absence of all teeth in primary, permanent or both dentitions. Anodontia is usually a part of a syndrome and seldom occurs as an isolated entity. It is commonly associated with complex pathology known as Ectodermal dysplasia which originates from the abnormalities during the early stages of embryonic development, and usually transmitted as an X-linked recessive disorder. In this report a case of 10 year old boy is presented who had complete set of primary dentition except maxillary lateral incisors, but surprisingly complete absence of permanent dentition in radiological observation. In this case, anodontia is not associated with any syndrome which is a rare finding.

Key words: Anodontia, Ectodermal dysplasia

INTRODUCTION

Anodontia, which represents the congenital absence of all teeth in the primary and/or the permanent dentition is a rare condition. Total or complete Anodontia means complete absence of the deciduous and the permanent dentition or either of them. Partial anodontia or hypodontia, also termed as oligodontia, is more common form of anodontia and involves congenital absence of one or more teeth. Pseudoanodontia or false anodontia occurs, when teeth are absent clinically because of impaction, delayed eruption, exfoliation or extraction. There is no single etiology for Anodontia, although tooth agenesis is occasionally caused by environment factors, but in majority of cases a definite familial trend has been reported. The currently implicated genes include the PAX9, MSX1, AXIN2 and He-Zhao deficiency, which is associated with an unknown gene that maps to chromosome 10q11.2. There are several syndromes that are characterized by partial or complete anodontia, some of them are isolated cleft lip/palate, Pierre Robin sequence, Van der Woude syndrome, MSX1 mutation, hypohidrotic ectodermal dysplasia (EDA or HED), Ectrodactyly-ectodermal dysplasia-clefting syndrome (EEC), Cleft lip palate ectodermal dysplasia syndrome (CLPEDI), incontinentia pigmenti (IP, Bloch-Sulzberger Syndrome), Hypohidrotic ectodermal dysplasia and immune deficiency (HEDID), Oral facial digital syndrome type I (OFDI), Witkop tooth-nail syndrome, Fried syndrome, Böök syndrome (PHC), Hair-nail-skin-teeth dysplasias, Rieger syndrome, Holoprosen cephal, Down’s syndrome (trisomi 21), Wolf-Hirschhorn syndrome (deletion 4p), Kabuki syndrome, Diastrophic dysplasia (DTD) Hemifacial microsomia and Recessive incisor hypodontia (RIH).

CASE REPORT

A 10 year old boy reported to the Department of Pedodontics and Preventive Dentistry, College of Dental Sciences, Davangere with a complaint of inability to chew food and also complained of retained milk teeth and unerupted permanent teeth. Clinical examination revealed that intraorally there was the presence of conical shaped complete set of primary mandibular and maxillary teeth but with missing right and left lateral incisors. There was also the presence of anterior crossbite with mesial step molar relation on both sides. Dentition was characterized with generalized spacing. Extra oral examination revealed concave profile with hypoplastic maxilla and a relative prognathic
mandible. No other abnormalities as such were observed which are associated with typical ectodermal dysplasia.

Radiographic examination revealed erupted complete set of primary teeth except the maxillary lateral incisors and absence of permanent dentition.

DISCUSSION

Only a few similar cases have been reported in the literature. Anodontia is an extremely rare condition, especially in females. It has been reported as a manifestation of one of the most severe forms of ectodermal dysplasia of which more than 120 different variations are known. Among them X-linked hypohidrotic Ectodermal dysplasia is most common. In the complete Anodontia cases reported, many other abnormalities were found to be present, such as developmental disturbances of the hair all over the body, the absence of hair, lack of sweat glands and the absence of the sense of smell or taste. In nearly every case so reported, heredity seemed to play a prominent part. In present report there was no abnormalities related to hair, sweat glands and taste and also heredity did not have a significant role for the occurrence of the condition of Anodontia of permanent teeth.

Cautely reported the case of a young man aged 20 years, who had only deciduous teeth present. Both jaws were abnormal, the mandible especially, being small, underdeveloped and out of proportion to the rest of the face. His hair and sweat glands were quite normal. He had a brother and sister who were similarly affected. The mother and father and former generations as far as could be ascertained were affected.

As stated in this case, which was similar to the one reported by Cautley but maxilla was underdeveloped than mandible with the appearance of relative prognathism. No other siblings were affected with this condition and no other relatives as far as could be traced were affected.

Anodontia can be diagnosed by taking a panoramic radiograph at about 4 years of age, because teeth, if any can be expected to be visible on radiograph at this age. In the present case diagnosis was made by the observation of complete absence of permanent teeth with in the jaws in a panoramic radiograph.

Beierle and Jorgenson found 14 cases of complete Anodontia of primary and permanent teeth. They also described 3 cases in which primary teeth were present but the permanent teeth were missing. Clare in 1922 reported eight cases of Anodontia of the permanent dentition. Life time preservation of primary teeth and alveolar ridges is a problem rarely encountered by the clinician. Based on the reported cases of complete Anodontia is more frequent than Anodontia of permanent teeth only. The objective of treatment should be to maintain primary teeth by protecting the thin enamel cap and dentin from occlusal abrasion and trauma.

In this case the main mode of management is of course motivation of the patient as the chief complaint of the patient was difficulty in chewing food resulting in reduced intake. Parents were informed that his permanent teeth were not present and they were supposed to visit dental clinic regularly for check up and also
informed about the oral hygiene practice for the maintenance of primary dentition.

The future treatment planned for this patient after the exfoliation of primary dentition was the construction of complete dentures to carryout regular functions of the oral cavity and musculature of mouth.

CONCLUSION

As it is seen in the literature, there are many cases that have been reported of Anodontia. It is also seen that complete Anodontia is seen frequently rather than only of permanent dentition. Reports have shown that not always this condition is associated with heredity as seen in the present case. Treatment objective in this case was to maintain primary teeth by protecting the thin enamel cap and dentin from occlusal abrasion and trauma. So ultimately in this case treatment that was planned was to maintain primary teeth as long as possible followed by prosthetic replacement by complete dentures for rehabilitation of the patient.

REFERENCES

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