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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 9 year old girl is presented who had few set of primary dentition, but surprisingly complete absence of permanent dentition except tooth buds of mandibular canine which were radiographically observed. In this case, anodontia is not associated with any syndrome which is a rare finding.

Key words: ***Anodontia, oligodontia***

INTRODUCTION

Anodontia is a genetic disorder commonly defined as the absence of all teeth, and is rarely encountered in a pure form without any associated abnormalities. Rare but more common than complete anodontia are hypodontia and oligodontia. Hypodontia is genetic in origin and usually involves the absence of from 1 to 6 teeth. Oligodontia is genetic as well and is the term most commonly used to describe conditions in which more than six teeth are missing. These conditions may involve either the primary or permanent sets of teeth, but most cases involve the permanent teeth. These phenomena are associated with a group of non-progressive skin and nerve syndromes called the ectodermal dysplasias. Anodontia, especially, is usually part of a syndrome and seldom occurs as an isolated entity Hypodontia affecting the primary dentition is rare (prevalence rate of <0. 5%) and has been observed to afflict both sexes equally.[1]

In the permanent dentition, hypodontia has a prevalence of 1.6% to 9.6%, excluding agenesis of the third molars. Oligodontia has a population prevalence of 0.3% in the permanent dentition. It occurs more frequently in girls at a ratio of 3:2. Agenesis of only the third molars has prevalence between 9% and 37%. In the deciduous dentition, hypodontia occurs less often (0.1%-0.9%) and has no significant sex distribution.[2] The mandibular second premolar is the most frequently absent tooth after the third molar, followed by the maxillary lateral incisor and the maxillary second premolar. Agenesis

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of maxillary central incisors, canines, or first permanent molars seems to be rather exceptional.

Definition/diagnosis criteria: A tooth is defined as congenitally missing if it has not erupted in the oral cavity and is not visible in the radiograph. All primary teeth have erupted by the age of 3 and all permanent teeth except third molars between the ages of 12 and 14. Therefore 3-4 year old children are suitable for diagnosis of congenitally missing primary teeth by clinical examination and 12-14 year old children, for diagnosis of permanent teeth excluding the third molars.[3] The use of panaromic radiography is recommended, together with clinical examination for the detection or confirmation of dental development and performing the diagnosis of hypodontia. [4]

CASE REPORT:

A 9 year old girl reported to our department with a chief complaint of missing teeth. Her past dental history revealed that she had undergone incision and drainage of swelling extra orally below lower border of mandible. Her past medical history was non contributory and family history revealed that she was born to non **consanguineous** marriage with normal delivery and mother did not suffer from any disease during pregnancy. On general physical examination her weight was 18 kg, height 3 feet 11 inches and she was well oriented and reactive. On extra oral examination she had a concave profile with retrognathic mandible (Fig 1) there was a scar present on the right side of the lower orbital margin which was pinchable and was present since birth. On intra oral examination she had spacing present between both upper and lower deciduous central incisors (Fig 2). There was high upper frenal attachment, reverse overjet and knife edge pattern of the ridge. Teeth present were 51, 53, 54, 61, 63, 64, 71, 72, 73, 74, 81, 82, 83 & 84.

No other abnormalities as such were observed which are associated with typical ectodermal dysplasia. Radiographic examination (OPG) revealed erupted set of primary teeth except the maxillary lateral incisors and absence of permanent dentition except developing 33 and 43 (Fig 3). A provisional diagnosis of partial anodontia was given with differential diagnosis of ectodermal dysplasia; Rieger syndrome and Witkop syndrome were considered.

Pediatric consultation was taken regarding general health status of the child. Complete set of investigations were done which included radiographic examination of chest (Fig 4) as well as phalanges (Fig 5), routine examination of blood including serum calcium, alkaline phosphate, TSH, T3, T4. The findings of these investigations were normal. Final diagnosis of Non syndromic partial anodontia was given. Full moth rehabilitation was planned after the exfoliation of primary dentition.

DISCUSSION:

Oligodontia is the term used most commonly in describing the phenomenon of congenitally missing teeth. Oligodontia has been classified as isolated or nonsyndromic and syndromic hypodontia.[5] Although oligodontia can occur over with 60 different syndromes, these anomalies can occur without any syndrome or systemic disease. However, oligodontia is seen more common in non-syndromic or familial form than syndromic form.[6]

The biologic basis for the congenital absence of permanent teeth is partially explained by the failure of the lingual or distal proliferation of the tooth bud cells from the dental lamina. The causes of hypodontia are attributed to environmental factors such as irradiation, tumours, trauma, hormonal influences, rubella, and thalidomide or to hereditary genetic dominant factors, or to both. Familial tooth

agenesis is transmitted as an autosomal dominant, recessive, or X-linked condition. Affected members within a family often exhibit significant variability with regard to the location, symmetry and number of teeth involved. Residual teeth can vary in their size, shape or rate of development and the permanent dentition is more affected than the primary dentition.[7]

Several studies have shown that *MSX1*and *PAX9 genes* play a role in early tooth development. *PAX9* is a paired domain transcription factor that plays a critical role in odontogenesis. All mutations of *PAX9* identified to date have been associated with nonsyndromic form of tooth agenesis.[8, 9, 10]

The condition should be considered as it may result in abnormal occlusion, altered facial appearance which may cause psychological distress, difficulty in mastication and speech. Dental clinicians should keep in mind that there are good possibilities with conventional prosthodontic techniques to help patients with dental abnormalities. Treatment depends on extent of hypodontia and should consist of interdisciplinary approach. Therefore early diagnosis is important in this condition. Case of tooth agenesis should be recorded with complete clinical history including medical and radiological investigations to rule out any syndrome.[11]

CONCLUSION

Oligodontia is frequent finding in many syndromes, but in this case it was not associated with any syndrome which is a rare finding. So as an oral physician if we come across any case having multiple congenital missing teeth it may not be always associated with multiple other abnormalities as seen in syndromes.

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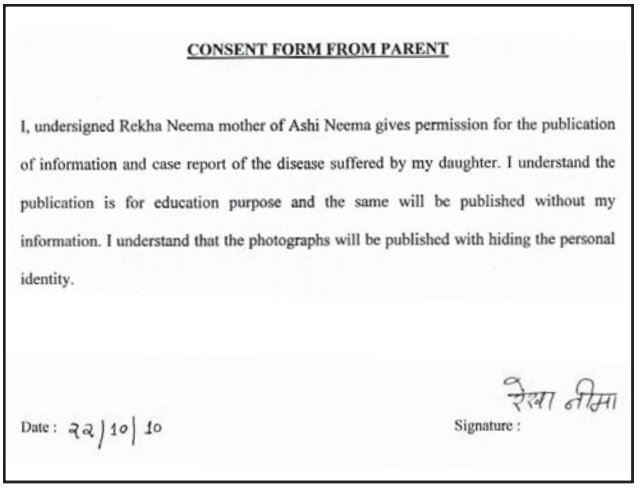

CONSENT FORM FROM PARENT

I, undersigned Rekha Neema mother of Ashi Neema gives permission for the publication of information and case report of the disease suffered by my daughter. I understand the publication is for education purpose and the same will be published without my information. I understand that the photographs will be published with hiding the personal identity.

Date : २२/10/10

Signature : रेखा नीमा

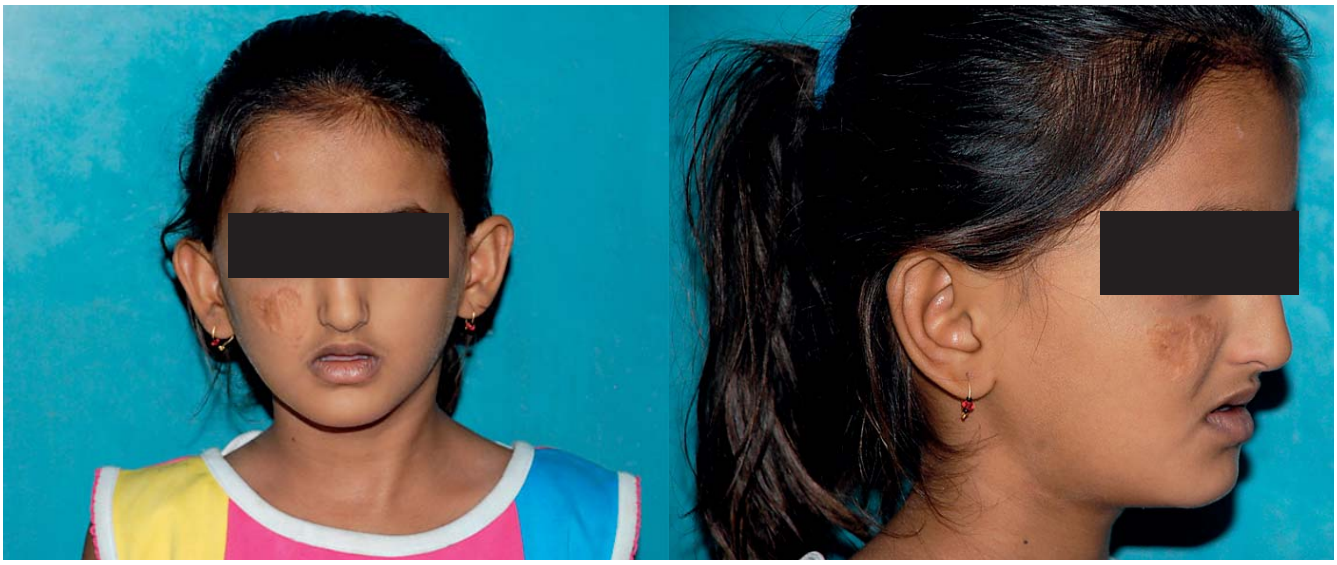

Figure 1: Concave profile with retrognathic mandible.

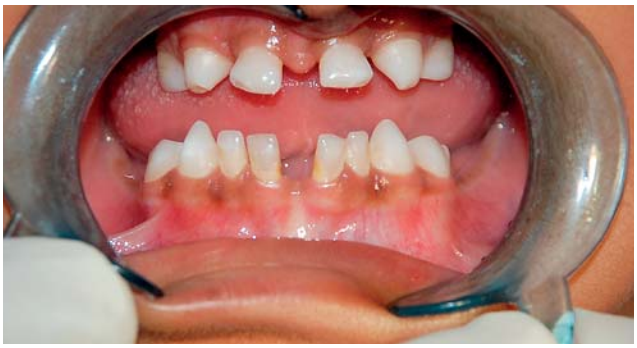

Figure 2: Spacing present between both upper and lower deciduous central incisors.

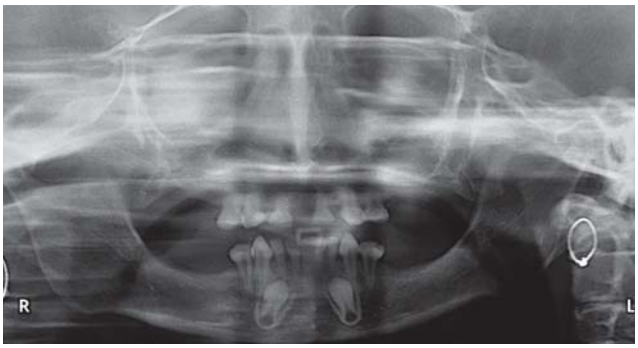

Figure 3: O. P. G

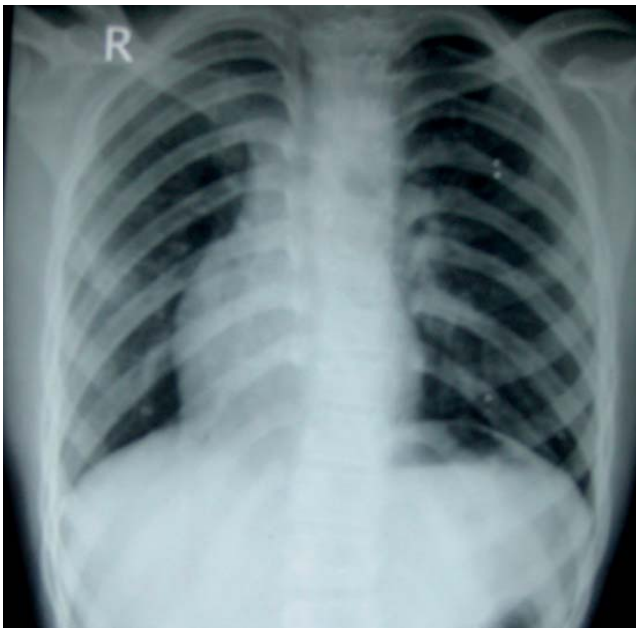

Figure 4: Chest X-ray.

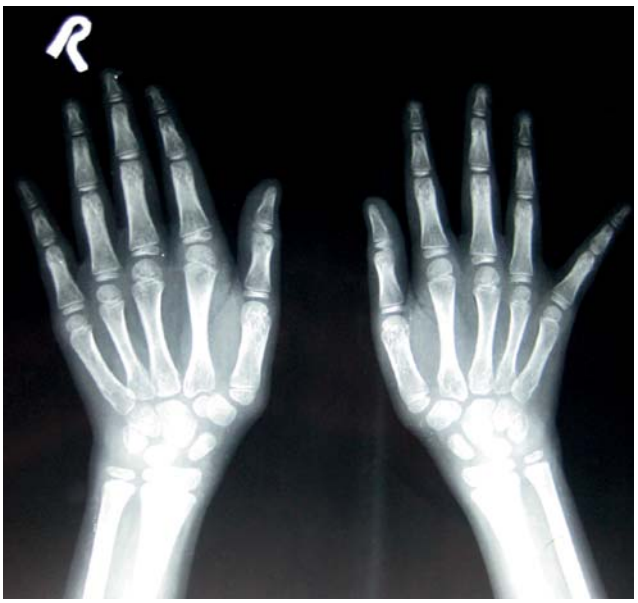

Figure 5: Hand wrist radiograph.