Case Report

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Hypohidrotic Ectodermal Dysplasia with Partial Anodontia: A Case Report

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ABSTRACT

Background: Ectodermal dysplasia is a group of rare inherited disorders characterized by dysplasia of tissues of ectodermal origin. Hypodontia, in these patients, cause reduced alveolar bone growth with lack of development of the alveolar ridges and malformed teeth apart from other extraoral deformities, thus necessitating intervention and rehabilitation for better mastication , phonetics , esthetics and psychological well being. This case report presents a patient affected by hypohidrotic ectodermal dysplasia with partial anodontia and its management.

Keywords: Ectodermal Dysplasia , Hypohidrotic , Hypodontia , Over Denture.

INTRODUCTION

Ectodermal dysplasia is a group of rare inherited disorders characterized by dysplasia of tissues of ectodermal origin-primarily nail, teeth, skin and occasionally additional dysplasia of mesodermally derived tissues like part of eyes, ears and other organs[1]. Clinically ectodermal dysplasia may be divided into two broad categories: the Hypohidrotic form (x-linked recessive) and the Hydrotic form or Cloustone syndrome (Autosomal dominant).[2]

The Hypohydrotic Ectodermal dysplasia (Christ-Siemens Tourine syndrome) is the commonest form characterized by a triad of signs comprising sparse hair (hypotrichosis), abnormal or missing teeth (hypodontia or anodontia) and absence or decreased sweat because of the lack of sweat glands (anhidrosis or hypohidrosis).[3] Hypodontia, in these patients, cause reduced alveolar bone growth with lack of development of the alveolar ridges resulting in extremely narrow and concave ridges lingually[4]. Teeth, if present, are mostly conical in shape, malformed and widely spaced. Apart from these saddle nose, prominent lips, linear wrinkles, hyperpigmentation around the eyes, atopy, mild facial dysmorphic features and increased susceptibility to respiratory infections are characteristics of this condition.[4] These presentations are seen characteristically only in affected males.[5]

Prosthetic dental treatment for Ectodermal Dysplasia is necessary to improve mastication, facial aesthetics, speech, nutrition and the patient's emotional well being.[6] Various treatment modalities are adopted in each case and it varies from simple restorations to prostheses and implants.[7,8]

CASE REPORT

A 18 year old boy reported to the department of Conservative Dentistry and Endodontics , with

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the complaint of carious teeth , missing teeth , inability to eat and difficulty in speech. The family history of the patient did not show similar manifestation of condition in any other members of the family . The patient presented with classical features of ectodermal dysplasia : hypodontia, hypohidrosis , hypotrichosis , large forehead , saddle nose , reduced lower facial height , sparse scalp hair , missing eyelashes and eyebrows with protuberant lips.(Fig. 1)

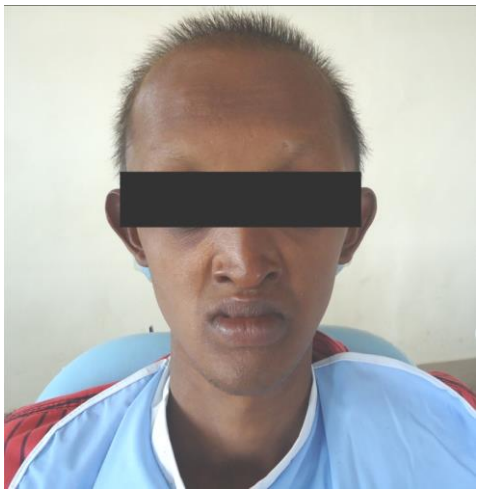

Fig 1: Hypohidrotic Ectodermal Dysplasia, extraoral features.

The intra-oral examination revealed presence of only 12, 16 and 23 in the maxillary arch as a result of which the edentulous alveolar ridges were deficient in both height and width(Fig. 2) . The mandibular ridge was of knife edge pattern and the maxillary ridge was depressed posteriorly with a shallow palate . The oral mucosa was dry and the appearance of the tongue was evidently large.

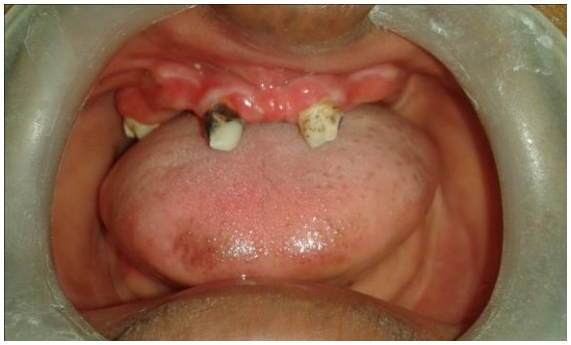

Fig 2: Intraoral findings.

Radiographic examination showed complete absence of permanent tooth buds and underdeveloped maxillary and mandibular alveolar ridges . 12 and 23 showed caries approximating pulp and 16 presented with moderately deep caries.(Fig. 3)

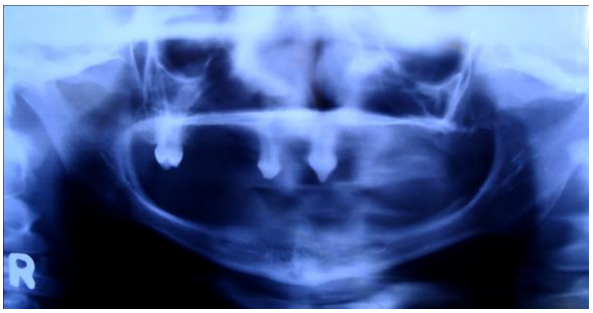

Fig 3: Orthopantomogram (OPG).

Caries excavation was done for the three teeth and it was found that 12 and 23 were pulpally involved. Root canal treatment was completed for the affected teeth and 16 was restored with Type 9 glass ionomer cement.(Fig.4)

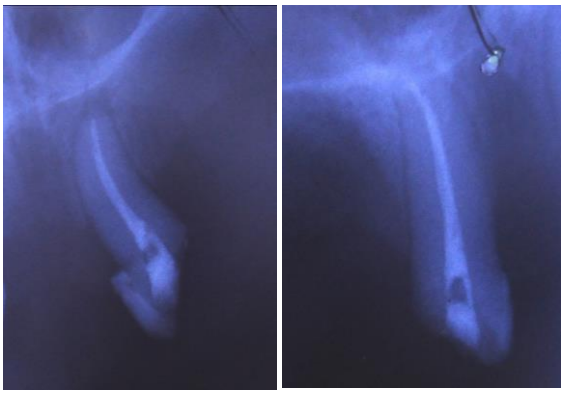

Fig 4: Endodontically treated 12 and 23

Metal copings were fabricated for all the three teeth after crown preparation keeping the finish line sub gingival.(Fig. 5)

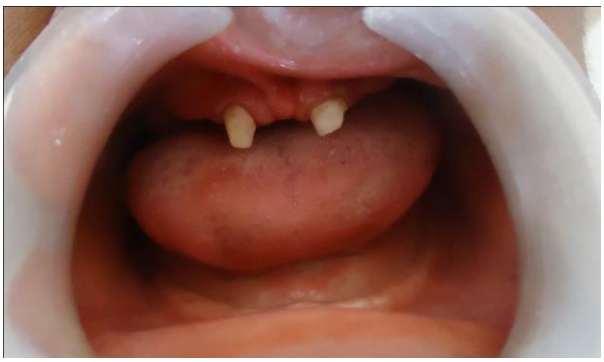

Fig 5: Crown preparation with subgingival finish lines.

The copings were luted with type 1 glass ionomer cement and an over denture was fabricated.(Fig. 6)

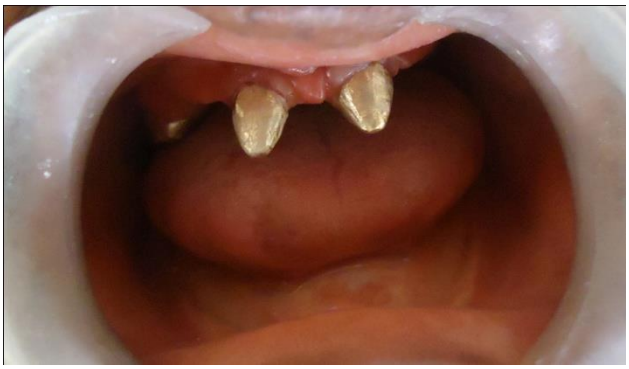

Fig 6: Metal copings luted.

The over denture was fabricated using small sized acrylic teeth to accommodate in the small sized jaws.(Fig. 7,8)

Fig 7: Overdenture.

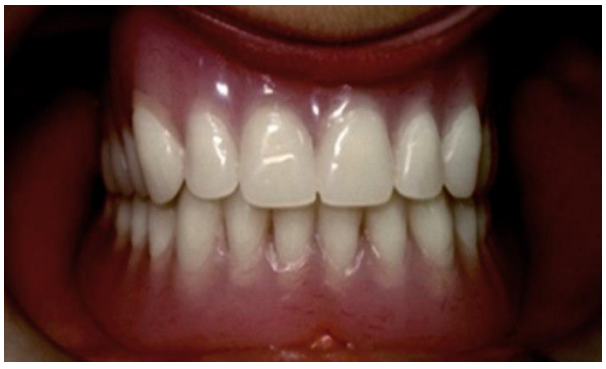

Fig 8: Post insertion.

Retention of the maxillary denture was good and the patient gradually adapted to the lower denture. The patient's mastication , phonetics , self confidence improved along with his socialization skills. Further recalls have been scheduled every three months.

DISCUSSION

Patients with ectodermal dysplasia need rehabilitation of the teeth to improve sagittal and vertical skeletal relationships during growth as well as improvement in speech, esthetics and masticatory efficiency[3]. Implant-supported denture is also suggested as the ideal reconstruction modality for adolescents over 12 years[9] but in this case the bone atrophy over the years resulted in inadequate bone support and rendered possibility of implants impossible.

Prosthetic replacements using removable dentures are the viable alternatives but they require proper follow up and the prosthesis should be adjusted and replaced when a decreased vertical dimension or an abnormal mandibular posture is detected due to growth.[10] In these patients , dry oral mucosa and under-developed alveolar ridges with no prominent tuberosities are problematic factors for resistance and stability of the dentures.[11] When fabricating dentures for these patients , special considerations should be made to obtain a wider distribution of occlusal loads by extending the denture base as much as possible.[12] Although the atypical conical anterior teeth are not always favourable for removable partial denture stability , they are effectively used as abutments for overdentures as in this case.

In this case , the additional metal copings on the retained teeth were prepared to

a) prevent further exposure of the teeth to the oral cavity and in turn prevent any recurrence of carious activity.

b) to prevent fracture of the crown structures during mastication due to lack of adequate tooth structure.

c) to preserve the residual bone height around the teeth thus maintaining the vertical height of the jaw

d) for enhancing the retention of the over denture.

Special impression making technique should be used for the fabrication of the over dentures. Oligodontia or anodontia almost in all cases lead to atrophy of the alveolar ridges as discussed before, reduced vertical dimension and class III intermaxillary relationship are the possible challenges in fabricating the over denture.[13,14]

CONCLUSION

Management of oral manifestations of ectodermal dysplasia presents a unique challenge for dental specialties and often require a multidisciplinary approach. Treatment of such edentulous patients with removable partial or complete denture is a time tested reasonable approach which is cost effective and improves function , speech , esthetics with psychosocial well being of the patient. However , its long- term success depends on regular reevaluation and refabrication with good maintenance of vertical dimension a and occlusal stability.

CONFLICTS OF INTEREST

The authors declare they have no potential conflict of interests regarding this article.

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