International Journal of Biomedical and Advance Research
ISSN: 2229-3809 (Online); 2455-0558 (Print)
Journal DOI: https://doi.org/10.7439/ijbar
CODEN: IJBABN

Case Report

Ectodermal Dysplasia Syndrome with comprehensive Oral rehabilitation - A Case Report

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Abstract

Ectodermal dysplasia (ED) is a hereditary disorder characterized by abnormal development of ectodermal derivatives resulting in abnormal morphological as well as functional presentation of sweat glands, scalp hair, nails, skin pigmentation, and craniofacial structures. Orofacial manifestation of the ectodermal dysplasia may varies according to their chromosomal inheritance pattern with most frequently reported ED syndrome is X-linked hypohidrotic dysplasia and women being the more affected. The oral feature of the syndrome include anodontia or hypodontia, hypoplastic conical teeth, underdevelopment of the alveolar ridges, and extra oral feature are frontal bossing, depressed nasal bridge, protuberant lips, and hypotrichosis which results in compromised aesthetics as well as masticatory function. Dental appearance in ED patients is extremely important because it can affect their self-esteem and is also a feel good factor for family members. Prosthodontics treatment of ED is extremely important to restore these functions which includes fixed, removable, or implant supported prostheses.

Keywords: Ectodermal dysplasia (ED), Dental Aesthetics, Prosthetics Rehabilitation.

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***Article History:**
Received: 09/04/2020
Revised: 29/05/2020
Accepted: 07/06/2020
DOI: https://doi.org/10.7439/ijbar.v11i6.5397

QR Code

How to cite: Mahakur M, Prasanna Sahoo P. K, Dash J. K, Ray P, Sahu M. and Janarthan T. Ectodermal Dysplasia Syndrome with comprehensive Oral rehabilitation - A Case Report. *International Journal of Biomedical and Advance Research* 2020; 11(06): e5397. Doi: 10.7439/ijbar.v11i6.5397 Available from: https://ssjournals.com/index.php/ijbar/article/view/5397

1. Introduction

Ectodermal dysplasia (ED) is a hereditary disorder characterized by abnormal development of certain tissues and structures of ectodermal origin like the sweat glands, scalp hair, nails, skin pigmentation, and craniofacial structures [1]. According to Freire-Maiaz if any person who exhibits at least two of the following features: (1) trichodysplasia (abnormal hair), (2) abnormal dentition, (3) onychodysplasia (abnormal nails), and (4) dyshidrosis (abnormal or missing sweat glands) is considered to have ectodermal dysplasia syndrome) [2].

The oral feature of the syndrome include anodontia or hypodontia, hypoplastic conical teeth, underdevelopment of the alveolar ridges, and extra oral feature are frontal bossing, depressed nasal bridge, protuberant lips, and hypotrichosis[4]. Ectoder-mal dysplasia might be inherited as autosomal-dominant, autosomal-recessive, and X-linked modes. There are more than 100 different ED syndromes reported [1,3]. The most frequently reported ED syndrome is X-linked hypohidrotic dysplasia and women being the more affected [5]. Dental appearance in ED patients is extremely important because it can affect their self-esteem and is also a feel good factor for family members. Prosthodontics treatment of ED can include fixed, removable, or implant supported prostheses [6]. The treatment planning depends upon the patient needs and growth stage.

2. Case study

9-year-old boy was reported to the Department of Paediatric Dentistry, SCB Dental College & hospital

Cuttack, with chief complaint of missing teeth, difficulty in speech, eating and ugly appearance. It was a case of proband as there was no previous family history. The past medical history shows less sweating and recurrent fever in the childhood with no relevant systemic illness.

Extra oral finding shows dry skin, sparse hair on the head, eyebrow and eye lashes with hypoplastic mid face, prominent lips, frontal bossing, and prognathic mandible. (Figure 1)

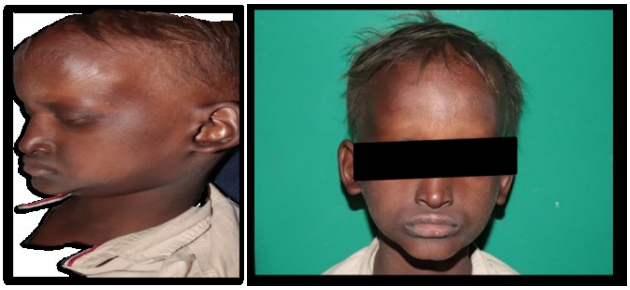

Figure 1: Extraoral findings (lateral view & front view)

The intraoral examination revealed upper jaw having four peg shaped anterior teeth with spacing between them and one posterior first molar on both site and other teeth was absent but the lower jaw was totally edentulous with thin alveolar ridge and reduced vertical height. (Figure 2)

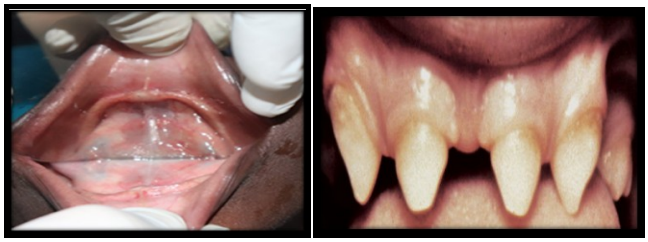

Figure 2: Intraoral findings - (Lower jaw & upper Jaw)

Panoramic radiograph showed presence of right upper central and lateral incisor, left upper central and lateral incisor with impacted upper right and left canine and two impacted left canine & premolar on completely edentulous mandibular arch (Figure 3).

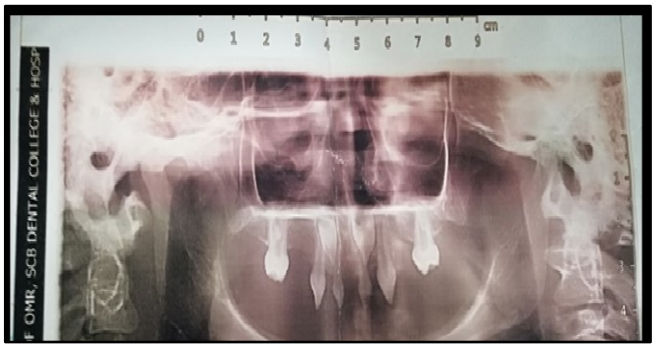

Figure 3: Panoromic radiograph findings

After considering hispast medical history and clinical as well as radiological features, it is diagnosed as a case of ectodermal dysplasia an-hidrotic type.

Treatment was planned with consideration of improvements in aesthetics, speech, masticatory function, jaw growth, nutrition, social wellbeing, feel good factor for family. In upper arch segmental fixed prosthesis and lower arch complete denture prosthesis were planned.

3. Procedure

Two set of study cast were made with alginate impression and dental stone. The peg shaped teeth of upper jaw were so nicely shaped and spaced hence without any crown preparation making it as abutment we made a temporary fixed partial denture of white acrylic for measurement of jaw relation and patients motivation. These teeth being immature and having wide pulp chamber are contraindicated for crown cutting. The fixed denture made was segmental without crossing the midline as it can hinder the growth of jaw. (Figure 4)

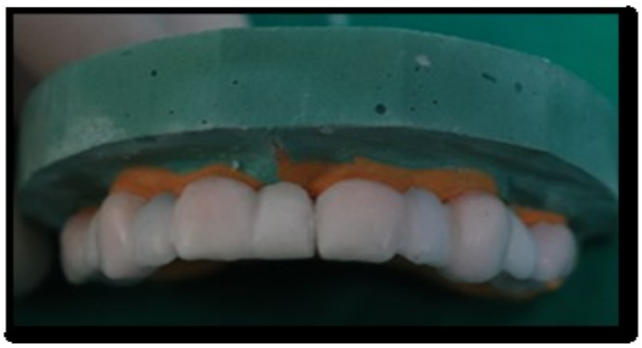

Figure 4: Cast mock up type 2

After upper arch lower jaw treatment was started with fabrication of secondary tray, then boarder molding was done, secondary impression was taken with Zinc oxide paste, secondary cast poured with die stone, over the secondary cast occlusal rim of modelling wax and then assessment of vertical jaw relation, horizontal jaw relation and orientation jaw relation were performed, then the cast was sent for tooth setting to lab. Try-in was done after checking all the parameters and patients satisfaction we finalized the lower denture and delivered the same after 3 days with all instruction of maintenance and follow up every 6 month advised for adjustment or replacement of old denture. (Figure 5)

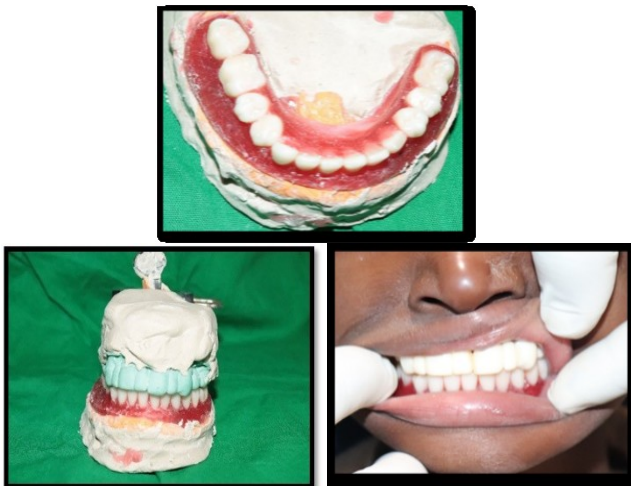

Figure 5: Lower jaw complete denture try in

4. Discussion

Oral rehabilitation of growing patients with ectodermal dysplasia is necessary to improve esthetics, speech, and masticatory efficiency as well as sagittal and vertical skeletal relationships during craniofacial growth and development [7]. Prosthodontics treatment of ED can include fixed, removable, or implant-supported prosthesis. The removable prosthesis is most common but Implant-supported denture can also be planned for adolescents over 15 years (1989 Conference on Implantology) but the main disadvantage is insufficient bone which can be corrected with bone grafting [8]. Early prosthetic treatment is generally recommended from the age of 3 to 4 years if the child is cooperative [9]. This early intervention will have positive effects on self-confidence, facial aesthetics, speech and masticatory function.

However, due to growth of individuals, regular follow up with adjustment and replacement of removable partial or complete dentures is required. With consideration to decreased vertical dimension of occlusion or any abnormal mandibular posture [10]. Dryness of the oralmucosa, under-developed maxillary tuberosities and alveolar ridges are problematic factors for resistance and stability of the dentures in ectodermal dysplasia patients [11]. The atypical conical anterior teeth may not be suitable for removable partial denture stability, so they can be used as abutments for over dentures that we have done in this case [12]. But in this type of case special care must be taken to keep the harmony in the occlusion. Prosthetic treatment should be performed as soon as possible to prevent the atrophy of the alveolar ridges due to oligodontia or anodontia which successively may leads to, reduced vertical dimension, prominent chin, and class III in-term axillary relationship.

5. Conclusion

Oral rehabilitation of ectodermal dysplasia patients is a privileged to pedodontics but a definite treatment plan with multidisciplinary approach among the paediatrician, prosthodontics, orthodontics and oral maxillofacial surgeon is highly needed. Improvement in function, speech, aesthetics and psychosocial condition is the main aim to treat a young patient. However long-term success of treatment depends on regular recall visit of every six month for relining and replacement of denture with meticulous maintenance of oral and prosthetic hygiene.

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