Case Report

Prosthetic Management of Patient with Hereditary Ectodermal Dysplasia

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Abstract

Ectodermal dysplasia is a hereditary condition characterized by absence or defect in the structure of ectodermal origin. A 16 year old female presented to our department with some of her teeth missing and desiring of replacement with artificial substitute. Since the patient was young, minimally invasive mode of treatment in the form of flexible denture was opted so that the growth is not interfered and patient can be reviewed and adjustment could be done in follow up till further definitive form of treatment can be rendered.

Introduction

Ectodermal dysplasia (ED) represents a large group of inherited disorders characterized by a primary defect in the structures of ectodermal origin [1]. It was first reported in a paper by Thurman in 1848. The term “Ectodermal Dysplasia” was coined by Weech in 1929 [2,3].

ED represents a large and complex group of diseases comprising more than 170 different clinical conditions. Its Prevalence is estimated to be 7 cases in 10000 births [4]. It is a genetic anomaly, which can be inherited through either parents or manifested via gene mutations [5].

Two different entities of hereditary ectodermal dysplasias have been reported; the Hidrotic (Clouston’s syndrome) and Hypohidrotic/ Anhydrotic (Christ-Siemens-Touraine syndrome) forms. The difference between the two is in terms of sweat gland manifestations. Hidrotic ectodermal dysplasia has an autosomal dominant inheritance where sweat glands are normal which are absent in Anhidrotic ectodermal dysplasia [6,7].

An hydrotic ectodermal dysplasia is a rare X-linked condition characterized by the classical triad of hypodontia, hypohidrosis and hypotrichosis. These patients generally have prominent supraorbital ridges, frontal bossing, and a saddle nose. The maxillae may be underdeveloped and the lips thick and prominent. The nose may appear pinched, and the alae nasi hypoplastic. The patient may resemble an edentulous old person. Some patients do not produce tears. The nails are usually normal. The skin of an infant may appear hypo pigmented. Maculopapular eruptions may occur during infancy. The characteristic dental defect in this syndrome is peg-shaped or conical front teeth, which cannot be distinguished from incisors. Both the deciduous and permanent teeth are affected. Anodontia may occur, but hypodontia with misshapen teeth is common, and these teeth may be hypoplastic [8].

In these patients, the appearance is extremely important since it ultimately affects their self esteem. Therefore prosthetic treatment is of great value for these patients for functional, psychologic as well as psychosocial point of view. The most frequent prosthetic treatment for the dental management of ED is removable prosthodontics.

‘This clinical report describes about the non invasive approach for the prosthodontic management of a young patient with an hydrotic ectodermal dysplasia.

Case Report

A 16 year old female presented to department of prosthodontics with the complaint of unaesthetic appearance due to missing teeth. There was no history of extraction of teeth in the past. The patient also gave history of lack of sweating and dryness of skin.

Extra oral Examination revealed sparse hair, frontal bossing, depressed nasal bridge, protuberant lips and loss of facial height.

Extraoral examination revealed decreased salivary flow and dry intraoral mucosa. Cone shaped teeth were present in 11 21 13 23 region, 12 14 22, 24, 27, 31, 32, 41, 42, 43, 45, 46 were missing, retained 85 and space of approx. 9mmpresent between 11 and 21 (Fig. 1)

Panoramic radiograph revealed no evidence of impacted tooth.

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Since the patient was young and not willing for orthodontic treatment, minimal invasive approach was selected for prosthetic rehabilitation. Maxillary left and right central incisors were morphologically restored with composite resin laminates as shown in (fig 2) Primary impressions were made with irreversible hydrocolloid impression material and casts were prepared with type 3 dental stone. Custom tray was prepared for lower arch and border molding was done. Final impression was made with light body polyvinyl siloxane and master cast was made. Jaw relation was done and cast was mounted on semi adjustable articulator. Teeth arrangement was done followed by tryin. After careful evaluation, the maxillary and mandibular prosthesis were fabricated in valplast resin by injection molding technique. The dentures were then retrieved, finished and polished.

Prior to insertion, the RPD were immersed in warm water for a minute to allow smooth initial insertion and adapt to underlying tissues and denture insertion was done as shown in (fig 3)

Follow up was done after 24 hr, 1 week, 1 month and 6 month.

Discussion

In cases of anhydrotic ectodermal dysplasia, the most common oral characteristic is hypodontia or anodontia [9]. Our patient presented with hypodontia with characteristically thin and underdeveloped alveolar ridge covered with thin mucosa and topped by a cord of movable connective tissue. These features along with decreased quantity of saliva are main problem encountered during treatment of such cases [10]. Prosthetic treatment modes using RPD, complete denture, and dental implants are the primary treatment options in patient with ED [11].

There is no definitive time to begin the treatment. Till and Marques suggested that initial prosthesis could be delivered when a child starts the school. Pigno et al suggested that initial prosthesis should be delivered before the school age of the patient [12]. The intraoral prosthesis can be modified during growth spurts or rapid growth periods [13]. For young patients like in our case, the use of flexible removable partial denture (RPD) is a reversible treatment that can significantly improve functions and esthetics without jeopardizing compromised dentitions.

Partials denture made from the Valplast resin have several advantages over cast partial denture and rigid acrylic dentures [14]. These dentures are esthetically superior because they are practically invisible. Translucency of the material picks up underlying tissue tones, making it almost impossible to detect in the mouth [7].

The presence of metal clasps on anterior teeth may cause esthetic problems with the use of cast partial denture which can be avoided with flexible dentures as no clasping is visible on tooth surfaces improving aesthetics.

The material is exceptionally strong and flexible. Even if there is a little bit of bending, it comes back to the original shape and position. It has good biocompatibility because the material is free of monomer and metal. Flexible dentures will not cause sore spots due to negative reaction to acrylic resins and will absorb small amounts of water to make the denture more soft tissue compatible [7].
Flexible dentures designed correctly do not engage the abutment teeth alone for support and retention. The gentle motion of the partial over the gum tissue produces a massaging effect that can prolong the healthier condition of the gums. There is no need of modification of the remaining teeth to receive occlusal rests or metal clasps as for cast partial dentures. Rebasing (changing the entire plastic/tissue area except the acrylic teeth) is possible [16].

However, it has certain drawbacks as well. Being a plastic material, it cannot be made into thin sections like metal. Therefore it is likely to break if cut into thin sections. It does not conduct heat and cold like metal. Therefore, the patient may not enjoy certain foods such as hot soup or ice cream. Since flexible dentures utilize the gaps (because of some missing teeth) for the "Retento-Grip Tissue-bearing Technique" for retention, the remaining teeth have to be in fairly good periodontal health [17]. In the patients with periodontally compromised dentition, it exerts unfavorable forces on the present teeth that further aggravate the periodontal problem. Also it requires special instruments (knives and polishing kit) to make the adjustment. A flexible denture is very difficult to repair if fractured [16].

**Summary**

Treatment of young patients with flexible removable partial is an acceptable modality, which improves function, speech, esthetics and psychosocial condition. However, its long-term success depends on regular recall appointments and meticulous maintenance of oral and prosthetic hygiene.

**Reference**


