



Ectodermal Dysplasia with Oligodontia: A Rare Case—Rehabilitation by Prosthetic Management

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Abstract

Ectodermal dysplasia (ED) is an uncommon genetic disorder in which there are congenital birth defects (abnormalities) of 2 or more ectodermal structures beside occasional dysplasia of mesodermal tissues. The occurrence of ED is said to be about 1 in 10,000 to 100,000 births. We report a case of hypohidrotic ectodermal dysplasia in an 8 year old girl who exhibited severe hypodontia and was successfully rehabilitated with conventional partial dentures in both maxillary and mandibular arches. Prosthodontic treatment majorly affects aesthetics and functions, facilitates psychological development and improves emotional condition and social life of the patient.

Keywords: Rehabilitation; Prosthetic; Ectodermal Dysplasia

Introduction

The National Foundation for Ectodermal Dysplasia (NFED) describe ectodermal dysplasia (ED) as an uncommon genetic disorder in which there are congenital birth defects (abnormalities) of 2 or more ectodermal structures beside occasional dysplasia of mesodermal tissues [1,2]. The occurrence of ED is said to be about 1 in 10,000 to 100,000 births [2]. There are 2 categories of ED, the hidrotic and hypohidrotic forms. In both forms, dentition and hair are similarly affected, but the hereditary patterns and manifestations of the nail and sweat glands tend to differ [3,4]. Clinical features of ED consist of numerous facial abnormalities like prominent forehead, saddle nose, thick lips, linear wise and hyper pigmentations around the eyes, hypoxemia and anodontia [2]. The primary and permanent dentition hypodontia is the second most common oral symptom related to hypoplasia of the alveolar bone structures and poorly formed alveolar ridges. The teeth that do develop are often conical in shape, making it difficult to use them as removable partial denture abutments, but sometimes offering the possibility of using them as abutments for complete overdenture. The most common option for restoring oral structures in childhood is prosthetic treatment with removable total dentures with acrylic denture base, partial dentures and tooth - retained overdentures [5].

The purpose of this article was to develop the characteristics and management of ED in a young patient by means of prosthetic rehabilitation and assess its effect on the social life of the patient.

Case Report

An 8 year-old girl was referred to the Department of Pediatric Dentistry, Kerman University of Medical Sciences, accompanied by her mother with the complaint of unaesthetic appearance due to the conical anterior teeth. The general medical history was non-contributory. The patient was seen by a clinical geneticist who made a provisional diagnosis of ectodermal dysplasia. There were no other cases of ectodermal dysplasia in the family history. The extra oral examination (Figure 1) revealed soft, thin and dry skin with a hyperpigmentation around the right ear, prominent chin, depressed midface, depressed nasal bridge, frontal bossing, thin upper lip, an aged profile with increased nasolabial fold and pseudo class III jaw relationship, thin and sparse hair, with sparse eyelashes eyebrows. The patient exhibited no abnormality of nails or sweat glands.

The intraoral examination (Figure 2) revealed caries lesion in teeth #M, #N, #O, #P, #Q, #R, #K, #L, #S, #T, #B, #A, #I, #J, #30, #19 and #3. Teeth #K and #T were infraocclusion. The central maxillary incisors were conical (figure 1) Her oral hygiene was poor. The color of alveolar mucosa and gingiva was normal.



Figure 1: Frontal view of the patient showing conical maxillary central teeth.



Figure 2: Intraoral photograph before and after treatment.

Radiographic examination (Figure 3) revealed oligodontia with missing of eight permanent maxillary teeth (#4, #5, #6, #7, #10, #11, #12, #13) and ten permanent mandibular teeth (#18, #20, #21, #23, #24, #25, #26, #28, #29, #31) and impaction of permanent mandibular canines and maxillary second molars. It seems that tooth #27 has an ectopic eruption. Radiographic examination confirmed the clinical findings and revealed ankyloses of teeth #K and #T.

Following the application of the preventive program, tooth #3 was restored using composite resin (Filtek Z250 -3M ESPE, USA). Tooth #14 was sealed with fissure sealant (Clinpro™, 3M ESPE, USA). Celluloid strip crowns (3M, USA) were used to build up two anterior conical-shaped teeth using shade A2 composite resin (G-aenial anterior; GC Corp). In teeth #M and #R, pulpectomy was performed. Then, tooth #R restored with composite resin and tooth

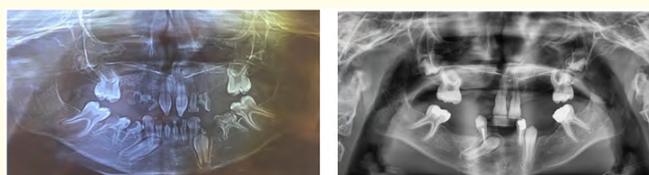


Figure 3: Panoramic radiograph revealed absence of eighteen teeth except third molars.

#M restored with amalgam restoration (Valiant PhD, Dentsply Caulk, Milford, DE, USA). Vital pulp therapy with Cem Cement ((BioniqueDent, Tehran, Iran) was done in tooth #19 and after 24 h restored with an amalgam buildup. Class II amalgam restoration was performed on tooth #30. Hopeless teeth including primary molars in both arch and teeth #Q, #P, #O and #N were extracted (figure 2) The stages of fabrication of removable partial dentures included: Maxillary and mandibular impressions were taken using alginate (Blue Print X-creme, Dentsply International). When properly fitting custom tray was ready and border molding was done, final impression was made. Jaw relationship was recorded using temporary base and wax rim and vertical dimension was determined with the usual 2-4 mm freeway space. During the trial appointment, the arranged teeth were checked in the mouth. After laboratory processing, the patient was delivered partial dentures with wire clasps (Figure 4) Instructions were given at the delivery appointment to maintain proper oral hygiene. Continuous follow - ups for the adjustment or replacement of old dentures were planned every three months. The child was provided with removable partial upper and lower dentures to improve appearance, chewing and speech. Throughout the procedures, behavior management techniques such as tell-show-do have been used. The girl's socialization skills were improved by increased self - esteem (figure 4). Further recalls have taken place every three months.



Figure 4: Frontal view showing improved facial appearance after insertion of dentures (a) Intraoral photograph with insertion of the acrylic partial dentures (b).

Discussion and Conclusion

Molecular etiologies of HED consist of mutations in genes involved in ectodysplasin/NF - kappaB or Wnt/beta - catenin, both of which are involved in the development of ectodermal appendages [6]. The skin on most parts of the body can be abnormally thin, dry, and soft with an unusual absence of pigmentation (hypopigmentation). Although, the skin around the eyes (periorbital) can be darkly pigmented (hyperpigmentation) and thinly wrinkled, appearing prematurely aged. However, the literature has described a decrease in the salivary glands of the intraoral mucosa, in the present case the amount of saliva was sufficient for the adhesion of the dentures. Teeth if present, are often conical in shape, malformed, widely spaced, delayed in eruption and vary in morphology with large pulps [7].

Children and adults with HED are challenged by their heat. Most of them do well with simple measures like wet clothing, air conditioning, wet bands, etc. Dental restoration has a great psychological advantage and it is very helpful to implement dentures early. Early prosthetic therapy for children with ectodermal dysplasia offers a unique opportunity for cooperative effort between the pedodontist and the prosthodontist, as pedodontists are better trained in child psychological management. Treatment with prosthodontics may begin at an early age of 3 - 4 years as it improves the growth and development of orofacial structures [8].

Conflict of Interest

The authors of this manuscript certify that they have no financial or other competing interest concerning this article.

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Bibliography

- Hickey Aj and Vergo Jr TJ. "Prosthetic treatments for patients with ectodermal dysplasia". *The Journal of Prosthetic Dentistry* 86.4 (2001): 364-368.
- Ansari G., et al. "Dental management of ectodermal dysplasia syndrome at an early age: a case report". *Journal of Dental School* 35.3 (2017): 115-117.
- Tarjan I., et al. "Early prosthetic treatment of patients with ectodermal dysplasia: a clinical report". *The Journal of prosthetic dentistry* 93.5 (2005): 419-424.
- Rao L and Gounder R. "Prosthetic rehabilitation of a patient with Type-I ectodermal dysplasia: A case report". *Journal of Dr NTR University of Health Sciences* 4.2 (2015): 124.
- Dimova-Gabrovska M., et al. "Removable prosthetic treatment in children-literature review". *Journal of Imab* 24.3 (2018): 2172-2176.
- Schnabl D., et al. "Prosthetic rehabilitation of patients with hypohidrotic ectodermal dysplasia: A systematic review". *Journal of Oral Rehabilitation* 45.7 (2018): 555-570.
- Paschos E., et al. "Clinical management of hypohidrotic ectodermal dysplasia with anodontia: case report". *Journal of Clinical Pediatric Dentistry* 27.1 (2002): 5-8.
- Prasad PG., et al. "Hypohidrotic ectodermal dysplasia: a report of two cases in a family". *Journal of Evolution of Medical and Dental Sciences* 4.4 (2015): 716-721.

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