Case Report **Olgu Sunumu**
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EARLY PROSTHETIC MANAGEMENT OF CHILDREN WITH ECTODERMAL DYSPLASIA: A REPORT OF THREE CASES

Ektodermal Displazili Çocuklarda Erken Protetik Tedavi: Üç Olgu Sunumu

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ABSTRACT

Ectodermal dysplasia (ED) is a group of congenital anomalies characterized by defects of two or more ectoderm-derived structures, such as hair, teeth and nails. The aim of this paper is to describe the characteristics and prosthodontic treatment of three cases with ED. Case 1: A 6-year-old boy was brought to our clinic with complaint of tooth ageneses. In clinical and radiographical examination, patient presented anodontia of permanent teeth. Medical history revealed that the child has cutaneous dyshidrosis and hypohidrosis. Based on the findings, the patient was diagnosed as Anhydrotic ED. To provide function, fonation and aesthetics, a complete denture for lower jaw and partial denture for upper jaw were made. Case 2-3: Two brothers (aged 8, 9) were broght to our clinic with complaints of dental caries, dental pain and missing teeth. They had a medical history including hypohidrosis and problems with thermoregulation, and both of the patients presented with oligodontia. Based on the findings, brothers were diagnosed as Familial ED. Firstly, extractions and restorations were completed after which partial dentures were made to gain desired function. It is important to note that early prosthetic intervention gives oppurtunity to the child to develop physically, emotionally and socially like other healthy individuals.

***Keywords**: Child, dental care, ectodermal dysplasia*

ÖZ

Ektordermal displazi (ED) saç, diş, tırnak ve tükürük bezleri gibi ektoderm kökenli dokuların iki veya daha fazlasında görülen bozukluk ile karakterize konjenital bir anomalidir. Bu makalenin amacı; ED görülen üç hastanın klinik bulgularını ve protetik tedavilerini anlatmaktır. Olgu 1: 6 yaşındaki erkek hasta diş eksikliği şikayeti ile kliniğimize başvurmuştur. Yapılan klinik ve radyografik muayenede, hastanın daimi dişlerinde anadonti olduğu tespit edilmiştir. Hastanın medikal hikayesinde, kutanöz dishidrozis ve hipohidroz olduğu öğrenilmiştir. Bu bulgular sonucunda, hastaya anhidrotik ED tanısı konulmuştur. Fonksiyon, fonasyon ve estetiğin sağlanması amacıyla alt çeneye total protez, üst çeneye ise parsiyel protez yapılmıştır. Olgu 2-3: İki erkek kardeş hasta (yaş 8 ve 9) diş çürüğü, diş ağrısı ve diş eksikliği şikayetleri ile kliniğimize başvurmuştur. Medikal hikayelerinden, hipohidrozis ve termoregülasyon gibi problemlere sahip olduğu öğrenilmiştir ve her iki hastada da oligodonti görülmüştür. Bu bulgular ışığında, kardeşlere Ailesel ED tanısı konulmuştur. Öncelikle hastaların çekim ve restorasyonları tamamlanmıştır. Ardından istenen fonksiyonun geri kazandırılması amacıyla parsiyel protezler yapılmıştır. Erken protetik tedavilerin bu hastalarda, diğer sağlıklı bireyler gibi fiziksel, duygusal ve sosyal açıdan gelişmeleri için fırsat oluşturduğu unutulmamalıdır.

***Anahtar Kelimeler**: Çocuk, diş tedavisi, ektodermal displazi*

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INTRODUCTION

Ectodermal dysplasia (ED) describes a wide range and complex group of disorders defined by abnormal development of the skin and appendages (hair, nails, teeth, and sweat glands). Affected multiple organs are developed from the primordial external germ layer (1). The relationship between disease genes and embryo development genes, shows embryo development genes shows embryo development network (2). ED is a feature of impaired development of ectodermal appendages. The epithelium and the mesenchyme are two adjacent tissue layers, and by a series of inductive interactions between these layers growth of ectodermal appendages growth is regulated (3).The disease is classified as hypohidrotic (anhidrotic) and hidrotic ectodermal dysplasia (HED) according to the number and functions of sweat glands. The most common form is HED and X-linked recessive pattern is most commonly inherited (1).

Child mortality rate of HED is about 2-20%, and timely diagnosis and treatment plays important role (4). Hypohidrosis, oligodontia and hypotrichosis are the clinical characteristics of the HED patients. Patients might also have the following symptoms: salivary and sweat glands alterations, skin scaling, fever episodes, asthma, difficulty in breathing, loss of hearing, wrinkles around the mouth and eyes, periorbital hyperpigmentation, prominent lips, hypoplastic maxilla, heat intolerance, strabismus, conjunctivitis, cataracts as well as polydactyly and syndactyly (4-6). Oral symptoms include oral xerostomia, peg shaped teeth, microdontia both in primary and permanent teeth, amelogenesis imperfecta, hypodontia, oligodontia or anodontia, taurodontism, horizontal bone resorption (7).

Inheritance of HED is mostly as autosomal dominant disorder. In those patients, sweat glands are normal. However dystrophic nails, sparse hair and hyperkeratotic palms and soles are observed. Treatment includes protection from high ambient temperature (7).

Herein, we aimed to present treatment of a patient with hypohidrotic and two patients with hidrotic ectodermal dysplasia.

CASE 1

A 6-year-old boy was brought to Kırıkkale University, Department of Pediatric Dentistry, with a complaint of tooth ageneses. The child exhibited the typical features of ED including sparse hair and dry skin. In clinical and radiographic examination, anodontia of permanent teeth was observed. There were only primary maxillary canines (peg shaped) in the oral cavity (Figure 1). The parents stated that the child had a difficulty of speaking and eating properly because of missing teeth. Also medical history revealed that the child has cutaneous dyshidrosis and hypohidrosis and subsequent problems with thermoregulation. Based on clinical and radiographic findings, the patient was diagnosed as anhydrotic ED. To provide function, fonation and aesthetics, it was decided to make complete denture for lower jaw and partial denture for upper jaw (Figure 2). Due to the presence of anxiety in children, maxillary canines could not be converted to lateral teeth. Only incisal edge of the teeth was removed.

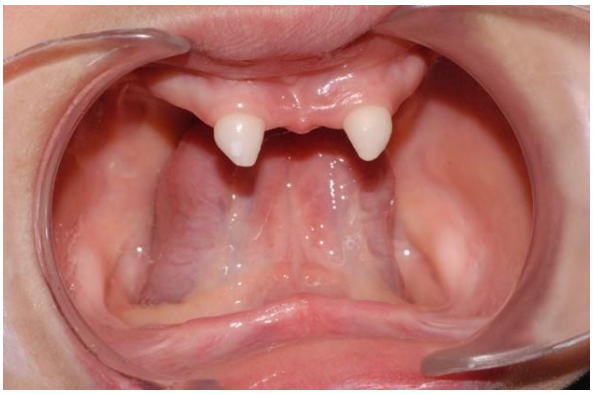

Figure 1. Intraoral image of Case 1 before treatment

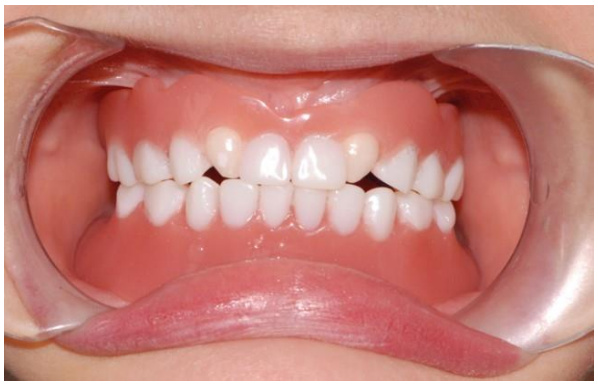

Figure 2. Prosthetic management of Case 1

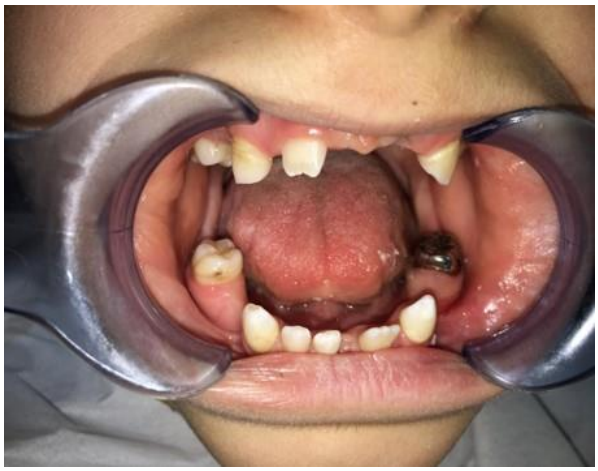

Figure 3. Intraoral image of Case 2 before treatment

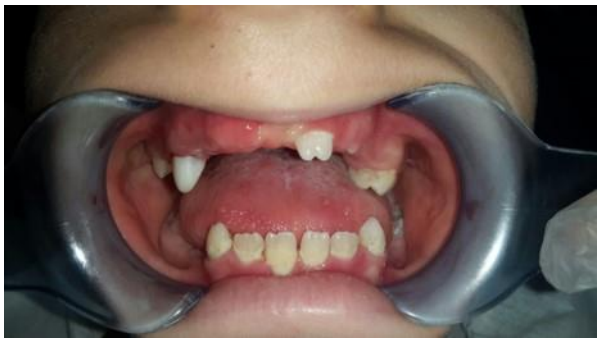

Figure 4. Intraoral image of Case 3 before treatment

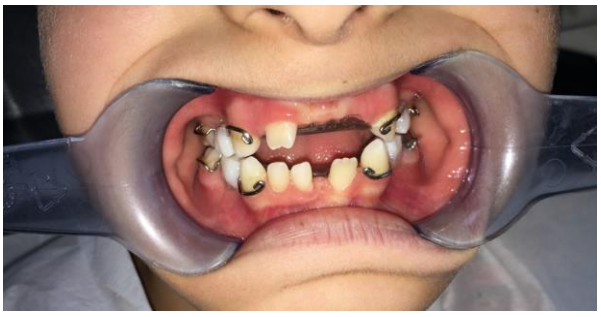

Figure 5. Prosthetic management of Case 2

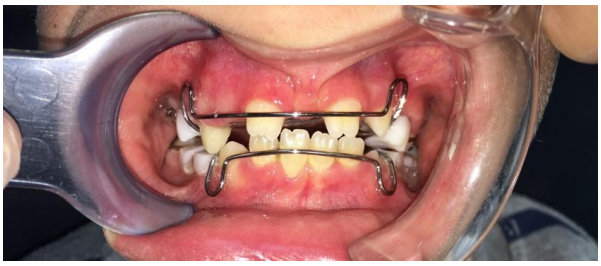

Figure 6. Prosthetic management of Case 3

CASE 2 and 3

Two brothers, aged 8 and 9, were brought to Kırıkkale Universtiy, Department of Pediatric Dentistry, with complaints of dental caries, dental pain and missing teeth. They had a medical history including hypohidrosis and subsequent problems with thermoregulation. In clinical and radiographic examination, it was observed that both of the patients had oligodontia. Orthopantomograph revealed the absence of permanent left central incisor, lateral incisors, first and second premolars and molars in the maxillar jaw; second premolars and first and second molars in the mandibular jaw in younger brother (Figure 3). In the elder brother, permanent lateral incisors, second premolars in the maxillar jaw; second premolars and molars in the mandibular jaw were absent (Figure 4). Based on the clinical and radiographic findings, brothers were diagnosed as Familial ED. In both patients, all caries lesions were removed and the teeth were restored with composite resin (3M ESPE, FiltekTM Ultimate). Pulpectomy treatment was applied for the left primary molar tooth of younger brother with BioAggregate (DiaRoot) material. After dental treatment was completed, partial dentures were made to gain desired function considering growth and development of the patients (Figure 5,6).

DISCUSSION

ED is estimated to be seen in 1-7 of 100,000 births, and up to 3 years of age in male patients mortality rate could be seen about 28% (8). In ED patients two or more ectodermal structures have congenital birth defects (9) and several genetic patterns including autosomal-dominant, autosomal-recessive and X-linked modes might be inherited (10).

More than 170 different clinical conditions are represented by ED as a large and complex group of diseases (11). There are 2 major types of ED; (a) X-linked anhidrotic or hypohidrotic ED, where sweat glands are absent or significantly reduced (Christ-Siemens- Touraine syndrome), (b) autosomal dominant inherited hidrotic ED, where sweat glands are normal (Clouston's syndrome) (12). Skin disorder is the most frequent abnormality in ED (93%), followed by hair and nail disorders (86%) (13). In hypohidrotic patients, hair disorders (91%) and dental anomalies (89%) are observed most commonly after hypohidrosis (100%) (14).

Hypodontia and anodontia are the most common dental anomalies of ED. These anomalies affect the patient aesthetically and functionally, also cause psychosocial problems for the patient (15). Dental appearance in ED patients is extremely important because it may affect their self-esteem. Due to rapid growth in early ages, the use of removable partial or complete dentures are indicated for these patients. After growth is complete, dental implants could be used for stabilizing dental prostheses. In the long-term prognosis for oral rehabilitation, osseo-integrated implants are great choices that will provide major improvement (15).

Early prosthetic intervention in children, allows healthy development of chewing, speaking and swallowing for healthy facial support and allows the formation of advanced temporomandibular joint function (10). However, the most important point to be considered before the initiation of treatment is the cooperation of the child (10, 13). Cooperation of children in this report was moderate and removable dentures were made to avoid growth and development problems. Follow-up appointments were scheduled and the dentures were considered to be renewed every 6 months until growth is complete.

Pediatric dentists play an important role in oral health of ED patients. A multidisciplinary approach involving various clinical modalities is important to render a comprehensive dental care. Treatment decisions depend on patient's needs, intraoral situation and cooperation of the patient.

Conflict of interest: The authors declare that they have no conflict of interest.

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