

Dentistry

CLINICAL PRESENTATION AND MANAGEMENT OF ECTODERMAL DYSPLASIA

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ABSTRACT

This article reviewed the clinical and radiologic aspects of ectodermal dysplasia as well as the dental treatment, and describe the oral rehabilitation. Patients with this disease often need complex dental treatment. The options for a definitive treatment plan may include fixed, removable, or implant-supported prostheses, singly or in combination. This clinical report describes the diagnosis and treatment of ectodermal dysplasia in the six patients (three males and three females, aged 9 to 22 years) with ectodermal dysplasia. Ectodermal dysplasia patients had a clinical examination and they had undergone radiographic and Steiner's analysis, and respiratory capability test before being assessment and treated. The prosthodontic treatment included interim removable partial dentures fabricated to establish an acceptable therapeutic occlusal vertical dimension, followed by definitive overlay removable partial dentures. As a result comprehensive and multidisciplinary team approach necessary to these patients for improve their dental, masticatory, growing, and orthognathic conditions.

Introduction

The ectodermal dysplasias (ED) are a large and complex nosological group of diseases, first described by Thurnam in 1848 (11). ED is a congenital syndrome characterized chiefly by abnormalities of tissues of ectodermal origin, namely skin, nails, hair, and teeth (4, 14, 15).

Findings give the clinicians the chance to redefine ED not simply as a result of a general 'ectodermal' abnormal development, but, more precisely, as systemic pathologic conditions (11).

The clinical manifestations of ED cause affected individuals considerable problems socially. Dental treatments of the clinical traits of ED can have a profound impact on these patients. The ability to look and feel like their peers is imperative for their psychological development. The literature has demonstrated the benefits that corrective dentistry has for the self-esteem and social

well-being of these patients (1, 2, 3).

This clinical research describes the condition and management of ectodermal dysplasia.

Materials and Methods

The all of cases (three males and three females, aged 9 to 22 years) were referred to our clinic for treatment management of the oral rehabilitations of their ED. Each patient had the benefit of a rigorous clinical examination for diagnosis and therapy. All major symptoms of hypohydrotic type ED were present such as hypodontia, curve and thin hair (trichodysplasia), and smooth skin (hypohidrosis). The ED may be inherited by autosomal-dominant, autosomal-recessive, or x-linked genetic transmission (**Fig. 1**) (6, 8, 12). The pedigrees of the case one and two were showed at the (**Fig. 2**).

All of patients at the lateral cephalogram showed as orthodontically, the vertical di-

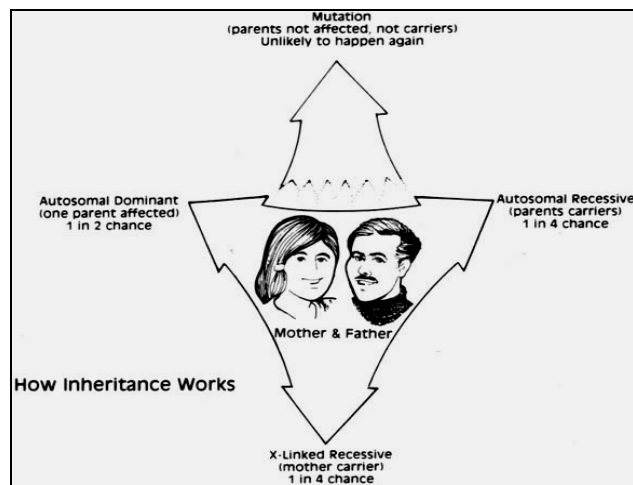


Fig. 1. Inheritance pattern associated with ectodermal dysplasias (6).

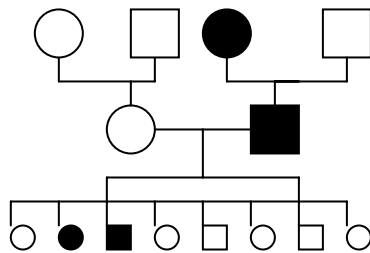


Fig. 2. The pedigrees of the patients.

mension was low, as expected, because of the posterior material lack, according to Steiner analysis Sella Nasion-Gonion, sagittal relationship were normal. The panoramic radiograph, confirmed that there is an absence of permanent teeth and, hypodontic appearance (Fig. 3), (Fig. 4), (Table 1).

The patients were informed the treatment with dental implants would need to be preceded by a bone-grafting procedure. At that time they refused to undergo this treatment for economic reasons. The treatment plan consisting of application of maxillary and mandible fixed and removable overlay prosthesis were discussed with the patients

according to Table 2 (3).

Finally, regular recalls were scheduled for 12 months to make necessary adjustments. They were very happy with esthetic and functional result.

Results and Discussion

ED is a rare, inherited, multi-system disorder. In our patients, after a discussion of the family and medical history, it was found that the parents of some of the patients were related to each other and they had a similar aspect of ED in their parents, which confirmed the hereditary nature of ED.

ED patients typically affect the hair, teeth, nails, and/or skin. ED is primarily characterized by partial or complete absence of certain sweat glands (eccrine glands), causing lack of or diminished sweating (anhidrosis or hypohidrosis), heat intolerance, and fever; abnormally sparse hair (hypotrichosis); and absence (hypodontia) and/or malformation of certain teeth. Many affected infants and children experience recurrent attacks of wheezing and breathlessness (asthma); respiratory infections; chronic inflammation of the nasal passages (atrophic rhinitis); scaling, itchy (pruritic) skin rashes (eczema); and/or other findings (14, 15) as our cases aspects.

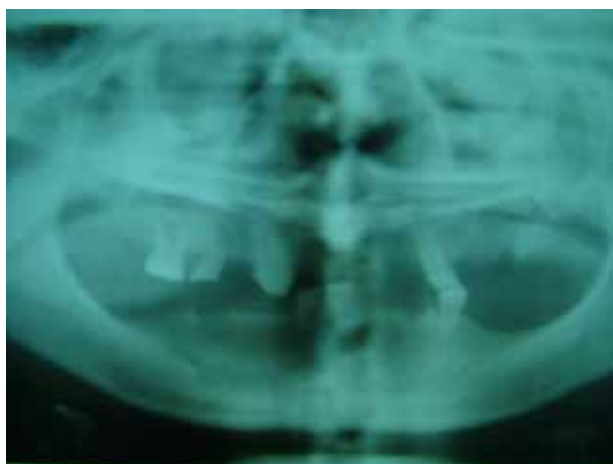


Fig. 3. Panoramic radiograph of Case 2 before prosthetic treatment.



Fig. 4. Cephalometric radiograph of Case 1 before prosthetic treatment.

TABLE 1

Results report of cephalometric analyses performed on the patients

Patients	Age	Clinical Face	SNA	SNB	ANB	SND	SNPog	SNGoGn	Upper lip	Lower Lip	Skeletal-Class
Case 1	20 (male)	Short retrused	77	80.5	-3.5	83	87	19.5	-6	-4	III
Case 2	22 (female)	Short retrused	76.5	78.5	-2	81.5	85	26	-9	-7	III
Case 3	9 (female)	Short retrused	78.5	77.5	+1	78	83	25.5	-2.5	-4	III
Case 4	12 (male)	Short retrused	72	87	-15	89	86	19.5	-3.5	-3.5	III
Case 5	18 (male)	Short Retrused	78	81	-3	82	88	19.0	-6	-5	III
Case 6	21 (female)	Short retrused	77	78	-1	83	86	25.5	-8	-6	III

TABLE 2

Tentative treatment plan for EDA and Oligodontia (3)

Age (years)	Treatment
6-12	Prophylaxis, supervised occlusal development
12-14	Orthodontic treatment, maintenance care
14-15	Semipermanent prosthetic treatment, ie, composite-retained onlay fixed partial dentures and/or removable partial dentures with cast-metal framework
15-20	Maintenance care; tomographic examination to analyze the quality and quantity of bone in the edentulous areas; offer to refer for genetic counselling; offer to refer to a dermatologist when needed
20-25	Conventional and/or implant-supported fixed prosthodontics at patient's demand; follow-up and maintenance care



Fig. 5. Full face of patient (Case 1).

In modern curricula for dental management, emphasis is on the 'whole body' concept. ED is a typical example in this regard, requiring a multidisciplinary approach for diagnosis and treatment. This study has enabled us to demonstrate a relationship between all major symptoms of hypohydrotic type ED were present such as hypodontia, thin hair (trichodysplasia) and, smooth skin (hypohidrosis) and EDs. Furthermore, the results support previous findings of an association between all major symptoms (10).

Cases of the study were oligodontia patients are usually referred to specialized

prosthodontist, orthodontist, pedodontist, and operative dentistry as multidisciplinary approach at an early age for treatment planning in this study.

Steiner analyses revealed a facial height reduction and concavity in 6 of the patients compared with normal values of Turkish persons (7, 9). Also found were maxillary reduction, labial retrusion, chin prominence and nasolabial and chin reinforcement (Fig. 5). In agreement with preceding a research, (5, 13). The researchers think that these measures may be unreliable because they vary according to tooth agenesis and the severity of ED. In agreement with prior research, sagittally underdeveloped maxillary retrusion and vertical dentoalveolar development are related to severe hypodontia.

Proper treatment sequencing is important to achieve the desired functional and esthetic results. For the patient described above, enhanced esthetics was possible because the clinical crown and root form were favorable for complete-coverage restorations.

Dental management allow these patients to preventive and supportive aesthetic activity as a result avoid social problems that are associated with partial or full dentures, particularly in young people. It also has been suggested that dental and maxillo-facial skeletal maturity. Excellent oral hygiene is crucial to the successful treatment of these patients. The patients should use



Fig. 6. Dentition after insertion of prostheses (Case 1).

topical fluoride daily for prophylaxis against caries.

The major goal of dental management is to provide patients comfort like other healthy individuals. Extensive prosthodontic treatment in growing individuals should preferably be performed with a multidisciplinary team approach. In treatment that is planned and performed over long periods, this approach has the advantages of continuity and shared responsibility for therapy decisions. Depending on their ages and their abnormalities, patients underwent either dental or prosthodontic (**Fig. 6**), or orthodontic treatment.

We couldn't certain about clinical associated with views of ED. However we believed every research will provide to improve for knowledge, treatment and care of ED.

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