IN BRIEF

- Ectodermal dysplasia is a rare condition in which patients exhibit anodontia and hypodontia intra-orally.
- The treatment of a 10-year-old patient by surgical, orthodontic and prosthodontic approaches is described.
- Osseointegrated implants and permanent prosthodontic treatment are planned when the patient is fully grown.

Orthodontic and prosthodontic treatment of ectodermal dysplasia – a case report

M. Yenisey,¹ A. Guler² and U. Unal³

In addition to its other symptoms, ectodermal dysplasia causes anodontia and hypodontia intraorally. Partial or total anodontia results in some loss of function, such as chewing, and affects aesthetics. Prosthodontic rehabilitation can be accomplished with fixed, overdenture, complete, or implant-retained prostheses. For rehabilitation, it is crucial to know the age, number and condition of present teeth, and the state of growth of the patient. A 10-year-old male patient who visited our clinic was treated by a multi-disciplinary team of surgeons, orthodontists, and prosthodontists. An overdenture was planned, and an implant-supported prosthesis was considered for when the patient had finished growing. A clasp retained over the denture was planned for prosthetic rehabilitation after considering his growth and the number and condition of his present teeth.

Ectodermal dysplasia syndrome was first described in the medical literature by Thurman, who reported two typical patients in 1848.¹ Individuals affected by ectodermal dysplasia syndromes have abnormalities of the glands, tooth buds, hair follicles, and nail development. Some ectodermal dysplasia syndromes are mild, while others are devastating. Other symptoms include deficient tears and saliva, poorly functioning mucous membranes, frequent respiratory infections, hearing or vision deficits, missing fingers or toes, cleft lip or palate, problems with the immune system, sensitivity to light, lack of breast development, and other abnormalities of the ectoderm. Lifespan can be affected in some rare ectodermal dyspla-

sia syndromes. However, there are very few documented examples of a person affected by ectodermal dysplasia syndrome dying because of an inability to perspire.^{1,2,8}

Anhydrotic ectodermal dysplasia is a rare X-linked condition. Affected males lack teeth, have hypotrichosis, and no sweat glands. Patches with and without sweat glands are seen in heterozygous females.^{1,3,8} Anhydrotic ectodermal dysplasia is a triad of hypodontia or anodontia, hypotrichosis,⁴ and hypohydrosis, and is associated with other problems that result from the defective development of structures of ectodermal origin.⁵ This is one of 132 known clinical ectodermal dysplasia syndromes.⁶ Anhydrotic ectodermal dysplasia occurs in all races, with an incidence of 1 to 7 per 100,000 live births.⁷

Patients with anhydrotic ectodermal dysplasia generally have prominent supra-orbital 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 hypopigmented. Maculopapular eruptions may occur during infancy. Asthma and

eczema are occasionally reported in these patients.^{1,8}

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 usual, and these teeth may be hypoplastic.^{1,9}

Early and extensive dental treatment is needed throughout childhood because of the absence of most of the deciduous and permanent dentition. A multidisciplinary team approach to treatment is recommended in these cases.¹⁰ Osseointegrated implants are an alternative treatment in older people with anhydrotic ectodermal dysplasia.^{11,12,16}

This clinical report describes a combined surgical, orthodontic, and prosthodontic approach to the treatment of a patient with anhydrotic ectodermal dysplasia.

CLINICAL REPORT

A 10-year-old boy with partial anodontia visited our clinic because he had difficulty chewing. The patient had been diagnosed with anhydrotic ectodermal dysplasia when he was 3-years-old.

¹Assistant Professor, ²Research Assistant, Department of Prosthodontics, Ondokuz Mayıs University, Faculty of Dentistry, 55139 Kurupelit-Samsun, Turkey

³Research Assistant, Department of Orthodontics, Ondokuz Mayıs University, Faculty of Dentistry, 55139 Kurupelit-Samsun, Turkey

*Correspondence to: Dr Murat Yenisey, Department of Prosthodontics, Ondokuz Mayıs University, Faculty of Dentistry, 55139 Kurupelit-Samsun, Turkey
Email: muratyen@omu.edu.tr
muyenisey@hotmail.com

Refereed Paper

doi:10.1038/sj.bdj.4811344

Received 27.09.02; Accepted 09.07.03

© British Dental Journal 2004; 196: 677–679

Fig. 1. Appearance of the patient's face before treatment

The patient had the typical characteristics of anhydrotic ectodermal dysplasia, such as protuberant lips, a saddle nose, fine sparse hair, and scant eyelashes and eyebrows (Fig. 1). He had hyperkeratosis on his palms and soles. His parents lacked these characteristics, while his teenage sister had some of the characteristics, such as a saddle nose, sparse hair, prominent supraorbital ridges, and conical, hypoplastic front teeth.

Intra-oral examination revealed a dry oral mucosa, protuberant lips, a thin vermillion line, and undeveloped alveolar ridges. There were two conical, hypoplastic anterior teeth in the right maxilla. Radiographic examination showed a conical impacted tooth in the middle of the left anterior maxilla (Fig. 2). The mandible was edentulous.

ORTHODONTIC TREATMENT

Denture planning was undertaken before orthodontic treatment. As bringing one tooth into the arch can affect the appearance, retention, and stabilisation of a denture, the unerupted incisor in the left anterior maxilla was erupted orthodontically to support and stabilise the planned overdenture. This was thought better than leaving the tooth unerupted in the bone.

To provide an anchor during the orthodontic treatment, an immediate denture was prepared with a gap to allow growth of the impacted anterior tooth.

A button was placed on the upper prosthesis. The mucosa over the impacted tooth was opened surgically and another button was placed on the tooth. Elastics were attached between the buttons to apply a 60-gram force (Fig. 3). During the orthodontic treatment, the patient was seen once a month. The orthodontic treatment was completed in 7 months (Fig. 4).

PROSTHODONTIC TREATMENT

After the orthodontic treatment, a diagnostic cast was prepared and the denture was

planned. We decided to apply a complete denture for the mandible and an overdenture for the maxilla. In the evaluation, the coronal structures of the existing teeth, axial wall inclination, and relative positions were considered to determine the path of insertion, retention, and stabilisation of the overdenture, to minimize plaque accumulation on the hypoplastic teeth, and to optimise the crown/root ratio of the fabricated metal copings. While preparing the teeth for the metal copings, the slopes of the axial walls were made nearly parallel. An impression was taken with an individual tray using medium-viscosity silicone impression material (Xantopren® M, Bayer Dental, Germany). The copings were cast with a Cr-Ni-based metal alloy (Wirrolloy®, BEGO, Germany).

To promote good retention, horizontal grooves were made on the mesial and distal

surfaces of the coping of the right maxillary central incisor, the mesial surface of the coping of the left maxillary central incisor, and on the distal surface of the coping of the left maxillary lateral incisor. The copings were cemented to the teeth using polycarboxylate cement (Harvard CC carboxylate cement, Harvard Dental-GmbH, Berlin, Germany) (Fig. 5). As the patient had a dry mucosa, the impression was taken using rapid-setting silicone impression material with high elasticity.^{14,15} Bilateral balanced occlusion was developed using anatomic (33 degree cusp angle) acrylic teeth. The maxillary overdenture and complete mandibular denture were prepared by conventional methods using heat-polymerizing acrylic resin.

Retention was achieved by clasps that engaged in the horizontal grooves. The clasps that fitted in the coping grooves to

Fig. 2 The two conical anterior teeth in the right maxilla

Fig. 3 Elastics were attached between the buttons to apply a 60-gram force

Fig. 4 Appearance of the other conical anterior teeth in the left maxilla after orthodontic treatment

Fig. 5 Cemented copings showing the retention grooves on the mesial aspects of the incisors

Fig. 6 Appearance of the clasps in the upper overdenture

Fig. 7 Finished prostheses in the patient's mouth

Fig. 8 Appearance of the patient after treatment

us to make adjustments. Osseointegrated implants and permanent prosthodontic treatment are planned for when the patient is fully grown.

Transitional overdentures are advantageous from the perspective of psychological development, and with patients feeling more secure with the aesthetic appearance of an overdenture.

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help retain the overdenture were bent over the cast and placed in the overdenture during the acrylic resin application phase (Fig. 6). The clasps were activated once the dentures had been adjusted to fit the patient (Fig. 7).

The patient was seen 48 hours later for adjustment, then at one and two weeks (Fig. 8), and 1, 3, and 6 months. The patient was followed up annually. No major complications occurred.

DISCUSSION

In many cases of anhydrotic ectodermal dysplasia, the most common oral characteristic is hypodontia or anodontia.¹⁶ In these cases, the advantages of existing teeth with regard to retention, stability,

function, and the phonetics of the denture should be considered. In addition, existing teeth help protect the proprioception mechanism and prevent the formation of residual alveolar ridges. Prosthetic rehabilitation of patients with ectodermal dysplasia is a routine process.

Endosseous implants can be considered as an alternative treatment.^{11,12,13} Considering the age and potential growth of our patient, it was deemed better to postpone osseointegrated implants.

In the present case, clasps were preferred over precision attachments because of the ease of care and fabrication. As the physical development of our 10-year-old male patient was incomplete, a transitional overdenture was planned, which allowed