Clinical management of ectodermal dysplasia with long term follow up: two case reports

Deniz Çetiner* / Umay Engel** / Gülay Tüter*** / Mehmet Yalim****

The present study describes the characteristics and clinical management of two patients with ectodermal dysplasia with long term follow-up. Dental treatments depend on the severity of disorder, therefore, treatment varies according to the age, growth and development of the stomatognathic system of the patient. It is important that the patient and dentist understand continued monitoring for dental problems is necessary. This provides improved aesthetics, function and emotional development.
J Clin Pediatr Dent 25(3): 187-190, 2001

INTRODUCTION

Ectodermal dysplasia (ED) is the heterogeneous group of disorders involving one or more ectodermal structures.[1,2] It is characterised by the total or partial absence of the sweat glands. The most common type is hydrotic ectodermal dysplasia. Major clinical findings of the disease are sparse hair, dryness of the skin, irregularities in nails, hypodontia or anodontia, hypohydrosis and hypotrichosis. The tooth crowns are often conical shape, the roots are short and the pulpal chambers are large. [3-5] Saddle-shaped nose, protrusive lips, malformed and protruding ears, wrinkles around eyes, pigmentation disturbances, disorders in sexual and mental developments are the other features of this disease. Additionally, dysphonia and hoarseness of the voice may be determined due to the atrophic pharyngeal and laryngeal mucosa.[1]

ED was redefined by Freire Maia[1,2] as a pathogenic developmental defect, which effects the ectodermal structures at the embryological level. The disorder is determined by the x-linked recessive gene, so the frequency and severity of the disease are more observed in males than females.[3] The aim of this report is to describe the dental abnormalities and oral rehabilitation with overdenture prothesis of the two cases in a long term follow-up.

* Deniz Çetiner, DDS, PhD, Resident, Gazi University, School of Dentistry, Department of Periodontology, Ankara, Turkey.
** Umay Engel, DDS, PhD, Resident, Gazi University, School of Dentistry, Department of Periodontology, Ankara, Turkey.
*** Gülay Tüter, DDS, PhD, Resident, Gazi University, School of Dentistry, Department of Periodontology, Ankara, Turkey.
**** Mehmet Yalim, DDS, PhD, Professor, Gazi University, School of Dentistry, Department of Periodontology, Ankara, Turkey.

Send all correspondence to Dr. Deniz Çetiner, 7. cadde, 60/13, 06490 Bahçelievler, Ankara, Turkey.

Telephone +90 312 2137715
Fax: +90 312 2121646
e-mail: denizcet@hotmail.com

Case I

The male patient was born in 1983 and was first referred to the Gazi University, Faculty of Dentistry for examination and treatment of his disorder in 1989. He exhibited hypodontia, phonetics and nutritional problems. In addition sparse hair, thin eye-brows was noted clinically (Figure 1). The tongue was in normal shape and size, and the saliva was in normal quality and quantity.

In radiological examination, maxillary left and right primary cuspids and second molars were detected. In mandible, only primary right and left cuspids were observed and no dental structure except the germs of mandibular left end right cuspids was presented. In maxilla permanent tooth germs were detected except right and left lateral teeth, first and second bicuspids.

It was determined that the patient could not perspire. He did not want to leave cool places and he had a fever. Familiar history revealed that the parents were healthy. They were not relatives and no symptoms of ED were seen in any of the others. Pregnancy and the birth were also normal.

A conventional acrylic removable overdenture was prepared for the patient to restore the function and aesthetics. Because of the changes in oral structures, it was decided to change the maxillary and mandibular overdentures in 6 months.

At 15 years of age (1998) orthodontic treatment managed to close the maxillary midline diastema (Figure 2). Inadequate oral hygiene was determined due to orthodontic appliances and caused difficulty in brushing. Gingival hyperplasia was diagnosed, and the diastema increased the hyperplasia (Figure 3). Then, a gingivectomy was done. There were no permanent teeth in mandible and a new overdenture was prepared for the

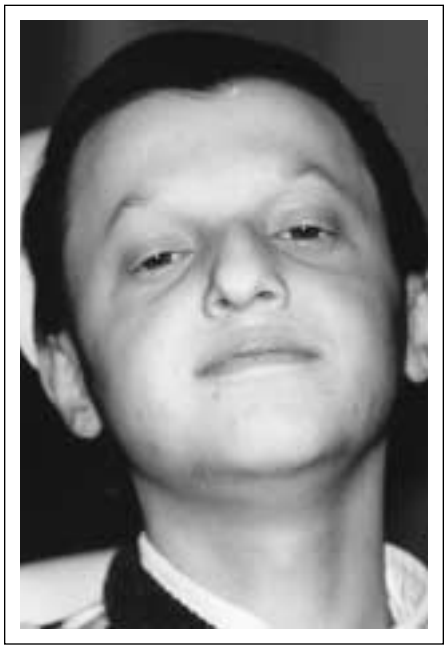

Figure 1. Physical appearance of the patient (case 1).

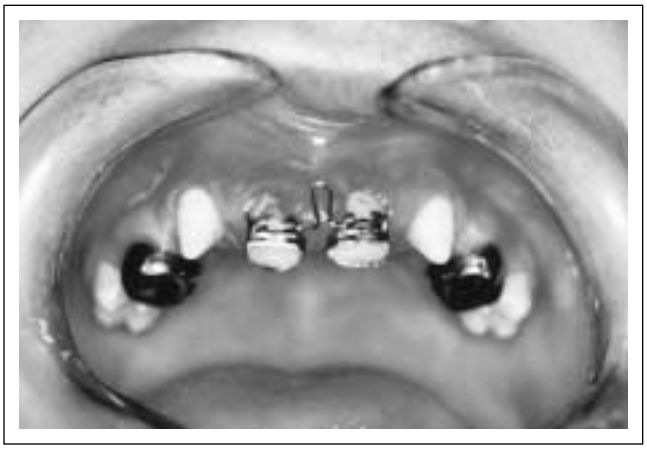

Figure 2. Orthodontic treatment was applied to close maxillary midline diastema.

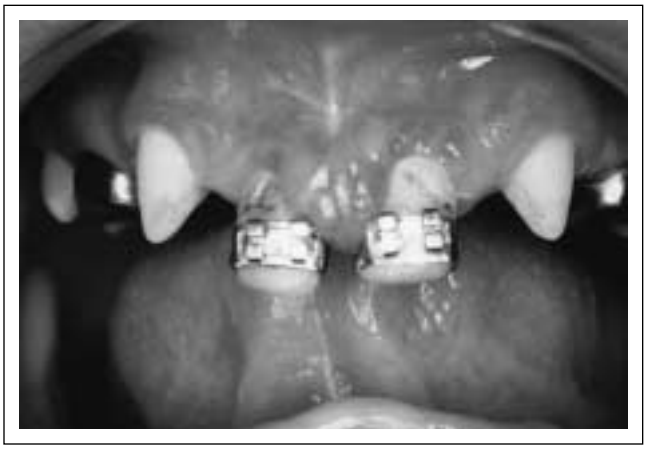

Figure 3. Gingival hyperplasia was occurred due to improper oral hygiene.

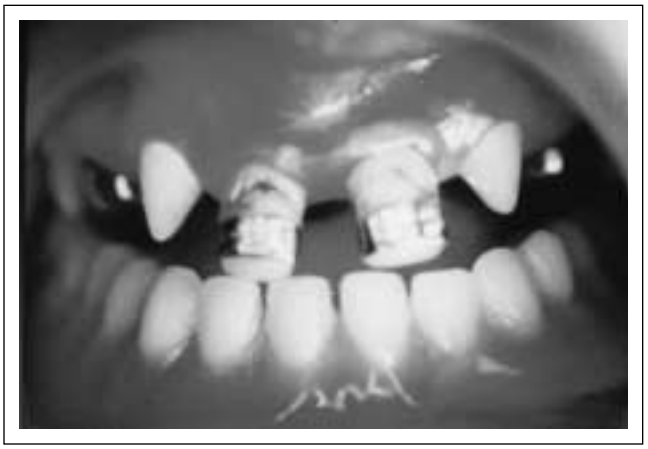

Figure 4. View of mandibular overdenture and maxillary arch after gingivectomy.

patient. Regular recalls were scheduled for 6 months, to monitor the patient (Fig 4).

Case II

A 21 year old girl was first referred to the Gazi University Faculty of Dentistry, Department of Periodontology in 1994 complaining of hypodontia, gingival bleeding, disfonation and inability to chew properly. Clinical examination revealed that the midface was depressed, the cheekbones were high and the lips were protuberant. The patient exhibited the other classic characteristics of ED; hypohidrosis and hypothrichosis. She has very light with fine hair and inability to sweat (Figure 5). There was hyperkeratosis of the palms of the hands (Figure 6). Maxillary right first and second incisors and left first incisor, canine and first molar teeth were observed. In the mandible, left and right second incisors, canines and left first molar teeth were present. All teeth were conical shaped. There were diastemas among them (Figure 7).

Her dental history determined that there was missing teeth in her primary dentition. She revealed hyperthermia that pointed to hypohidrosis. Her parents were in a good health, relatives. No similar cases among relatives have been identified in her family.

Initially, a periodontal treatment was performed. Mandibular right and left lateral incisors exhibited mobility of about 1 mm in bucco-lingual direction due to periodontitis. Gingiva was bluish red and bright. Scaling was done after the consent of the patient. Orthodontic consultation and treatment were required, but she did not cooperate sufficiently to receive treatment. A conventional overdenture was the choice of treatment for this patient to preserve remaining dentition and to restore function and aesthetics. But she did not accept the treatment. Only prophylaxis could be performed.

One year later, when she came for periodic monitoring to our clinic, it was observed that she was still uncooperative and had her mandibular first molar, left and

Figure 5. Facial view of the patient (case 2).

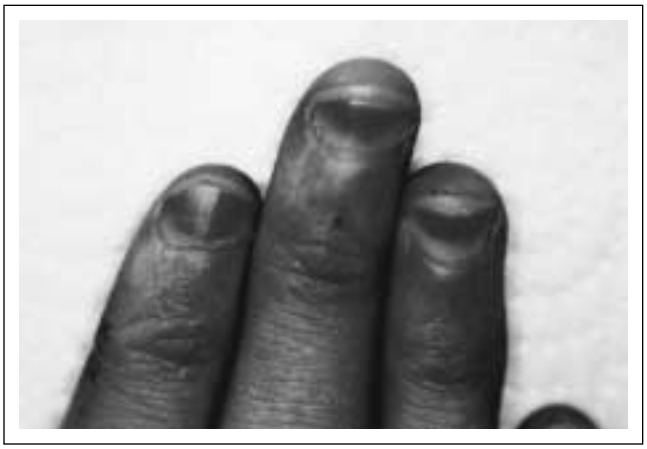

Figure 6. Dystrophic nails of the patient.

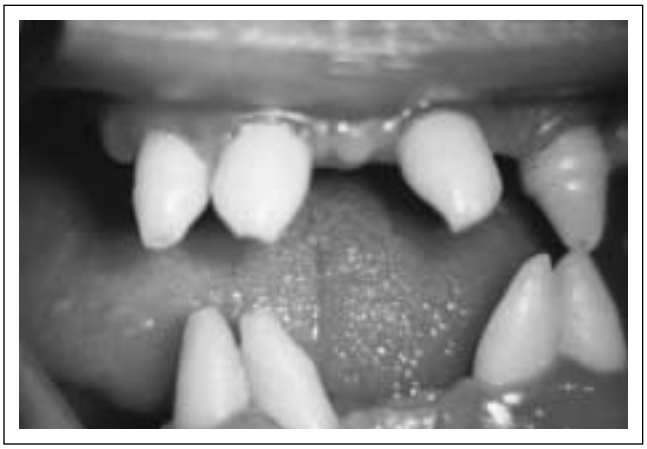

Figure 7. Anterior view of maxillary and mandibular conical incisors.

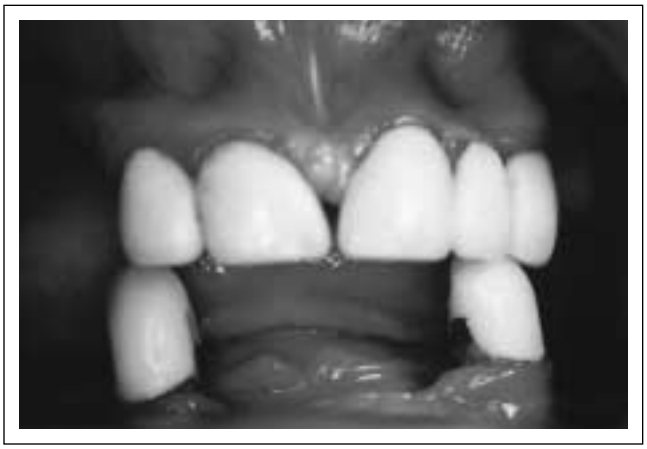

Figure 8. The dentition is shown after cementation of the veneers.

right lateral incisors extracted. No treatment was performed. Also consequent arrangements were made to review her every 6 months to monitor general growth and dentition.

In 1996, she arranged for an overdenture (Figure 8). However, she began to feel discomfort due to changes in soft tissue structures with improper oral hygiene, and could not tolerate this type of treatment. She rarely used her dentures. Oral hygiene instructions were given and the objectives of the treatment were explained to her family. Follow-up of the patient were periodically continued for 3 months.

Six months later, she began to use her overdenture and improvements in her social development as a consequence of the dental treatment was clearly seen (Figure 9). She was able to enjoy a better quality diet because of her new ability to chew. Her aesthetic appearance was improved and this helped her to improve psychologically.

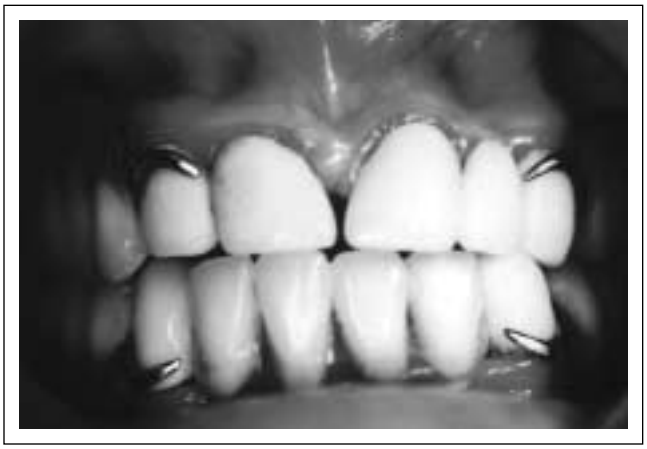

Figure 9. Intraoral view of partial overdenture.

DISCUSSION

Three main problems can be revealed in untreated patients; psychological problems, masticatory problems due to dental abnormalities, phonetically disorders because of abnormalities of orofacial structures.[1,6,7,8]

The features that were recorded in the present report agree with the results of previous studies.[3,5] In the first case, a mandibular primary incisor, maxillary primary lateral incisors and primary molars were

absent. There was a diastema between maxillary permanent first incisors. This was also detected among present teeth in second case. And in this case, only treatment opinion was the fixed and removable overdenture for both arches, because of her severe management problems and the age of the patient, made various treatment appliances difficult. In the first case, the beginning of treatment was in early ages of patient. So advanced treatment models could be applied easily.

In many cases, lack of permanent teeth were observed even the primary teeth were detected. However the presence of permanent teeth was reported in areas where no primary teeth had erupted. Two years follow-up of this patient has been previously documented.[9]

The successful treatment of these patients can be expected to help them both physically and psychologically.[1] Timing of treatment is also important so that young patients should receive dental treatment at an early age.[7] Dental treatments depend on the severity of disorder, therefore varies according to the age, growth and development of the stomatognathic system of the patient.[5] Orthodontic, prosthodontic and periodontal treatment appliances play an important role in the improvement of the attitude and self-confidence of the patient.[5,8,10] Although patients may have a limited cooperation, it is essential to prepare a long-term treatment plan including all types of treatment process to ensure that the patient with ED maintains an adequate level of oral health care.

ACKNOWLEDGEMENT

We would like to thank to Prof. Dr. Alev Alaçam for administrative assistance in preparing this manuscript.

REFERENCES

1. Itthagarun A, King NM. Ectodermal dysplasia: A review and case report. Quintessence Int 28: 595-602, 1997.
2. Hassed SJ, Kincannon JM, Arnold GL. Clouston syndrome: An ectodermal dysplasia without significant dental findings. Am J Med Genet 61: 274-276, 1996.
3. Vierucci S, Baccetti T, Tollaro I. Dental and craniofacial findings in hypohidrotic ectodermal dysplasia during the primary dentition phase. J Clin Pediatr Dent 18: 291-297, 1994.
4. Chitty LS, Dennis N, Baraitser M. Hidrotic ectodermal dysplasia of hair, teeth, and nails: case reports and review. J Med Genet 33: 707-710, 1996.
5. Bonilla ED, Guerra L, Luna O. Overdenture prosthesis for oral rehabilitation of hypohidrotic ectodermal dysplasia: A case report. Quintessence Int 28: 657-665,1997.
6. Bakri H, Rapp R, Hadeed G. Clinical management of ectodermal dysplasia. J Clin Pediatr Dent 19: 167-172, 1995.
7. Pigno MA, Blackman RB, Cronin RJ Jr, Cavazos E. Prosthodontic management of ectodermal dysplasia: a review of the literature. J Prosthet Dent 76: 541-545, 1996.
8. Shankly PE, Mackie IC, McCord FJ. The use of tricalcium phosphate to preserve alveolar bone in a patient with ectodermal dysplasia: a case report. Spec Care Dentist 19: 35-39, 1999.
9. Ulusu T, Alacam A, Iscan HN, Ucuncu N. The relation of ectodermal dysplasia and hypodontia. J Clin Pediatr Dent 15: 46-50, 1990.
10. NaBadalung DP. Prosthodontic rehabilitation of an anhidrotic ectodermal dysplasia patient: a clinical report. J Prosthet Dent 81: 499-502, 1999.