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Oral Rehabilitation of Patients with Ectodermal Dysplasia: Cases Series

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Abstract

Ectodermal dysplasia (ED) is a rare hereditary disease. The ectoderm-derived tissues and organs such as dental, alveolar bone, hair, nails, eyebrows and skin consist of various anomalies and developmental disorders. Patients with ED are developing functional and aesthetic problems due to structural and functional problems, broken jaws and teeth. In these patients, it is recommended to apply by modifying the prosthetic treatment. In our case report, were tried raising living standards providing rehabilitation ED with six cases. For the treatment of ED patients, in terms of oral and teeth health requires a multidisciplinary and professionals approach.

Keywords: Ectodermal dysplasia, hypodontia, partial dentures.

I. Introduction

Ectodermal dysplasia (ED) syndrome defines a wide group of heterogeneous diseases which is rare (7 in 10000) and hereditary. This syndrome where two or more ectoderm-derived tissues are primarily affected, which is characterised by common structural and functional disorders or deficiencies in tissues, is congenital, diffused and non-progressive. Especially skin, hair, nails, eccrine glands and teeth are affected. It is more frequently seen in males than females. Death can also be witnessed, albeit rarely, due to the high level of body temperature. (1)

Until today some 200 sub-groups are known and some 30 gene mutation have been detected which cause the disease. In most cases ED is responsible for the mutation of only 4 genes (EDA1, EDAR, EDARADD and WNT10A) (2,3).Frequently ED has been defined in three major groups, namely anhidrotic (Christ–Siemens–Touraine syndrome), hypohydrotic and hidrotic (Clouston syndrome). Anhidrotic ED is transmitted autosomal recessively, characterised by the lack of sweat glands and sebaceous glands and is more rare compared to the other types; hypohydrotic type ED shows these findings only slightly. Sweat glands and sebaceous glands are formed normally in hidrotic type which is transmitted autosomal dominantly (4-6).

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The most frequent type is hypohydrotic ectodermal dysplasia. It shows the features of X-dependent recessive, autosomal recessive or autosomal dominant transmission. X-dependent hypohydrotic ectodermal dysplasia (HED) causes mutation in EDA1 gene which codes ecto-dysplasia. In humans EDA1 is found in several tissues, and in bones in neuro-ectoderm, thymus, embryologic and fetal development periods including several epithelial tissues. Clinical features of HED can be listed as thin hair and eyebrows, creased and dry skin, missing and malformed teeth, and hypoplasia of sweat, sebaceous, Meibomian, lacrimal and mammary glands (7,8). Clinically hyperhidrosis, hypertrichosis and skull anomalies can be witnessed. Patients frequently have a smaller facial structure than normal. In oral area clinical features such as anodontia, hypodontia and conic teeth can be observed. Anodontia is also characterised with the lack of crest development (4,9). Insufficient growth of alveolar crests causes reduction of facial height, formation of protruding lip structure and a sunken appearance in the 1/3 of the face (10). In the teeth enamel dysplasia, tubercula disorders, mikrodontia, crossed closing can be seen (11). Tooth crowns are small and abnormal-shaped. Maxillary incisors and canines are always conic and sharp. Enamel layer is thin and narrows towards cervical of teeth. Taurodontism is common in milk second molars (12).

In this case presentation, the treatment of 6 child patients with ectodermal dysplasia who applied to our clinique due to lack of teeth and aesthetic complaints is examined.

CASE 1

A female child patient of 14 age applied to Department of Paediatric Dentistry, Faculty of Dentistry, Dicle University with aesthetic and functional complaints due to missing teeth. The patient was previously diagnosed as HED; it was seen that the results of intraoral, extra oral and radiological examination did not coincide with this disease. From the medical history obtained from family of the patient it was found out that her father and mother were not relatives, other male child was healthy, but the mother suffered from congenital teeth deficiencies.

In extra oral examination the hair and eyebrows of the patient were thin. The patient told that her hair could be easily broken. Dryness in hand palms and weakness in nails was observed. Her skin was soft and dry. There was hypodontia. The patient especially complained about the intervals between her front teeth.

As a result of intraoral examination, it was observed that the teeth numbered 17,16,53,11,21,23,26,27,47,46,45,43,71,81,43,44,85,46 and 47 existed. In radiological examination it was observed that the teeth numbered 18, 28 and 48 existed and their crown formation was completed. No other embedded teeth were observed in the patient. Decay lesions in the 17,16,26,27,36,46 and 85 numbered teeth of the patient were restored with composite fillings (Filtek™ Ultimate 3M ESPE). There was diastema between maxillary central incisors. As a result of the evaluations, it was decided that the diastema of the patient should be closed first. Measure was taken from maxilla and moving walksplint was planned for the maxilla. Diastema was closed with finger spring which is a moving walksplint. After the diastema was closed measure was taken from maxilla. Then removable partial dentures was produced for maxilla and adapted to the mouth. For color harmony of the prosthesis of the patient polishing was applied to the existing teeth. The patient has been given appointment for 6 month intervals (Figure 1-5).

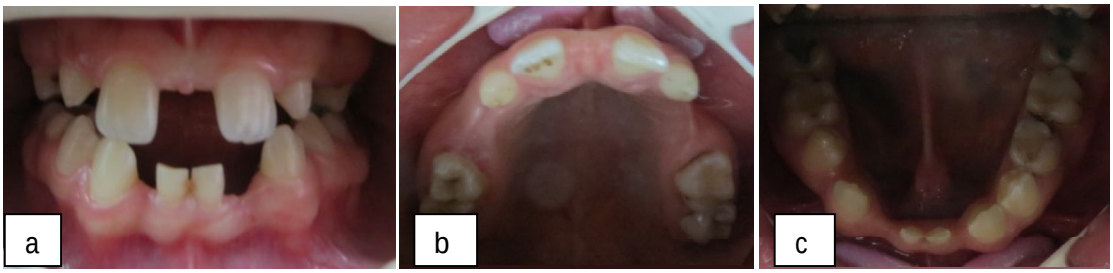


Figure: 1 a, b, c: Intra-oral view of the patient before treatment

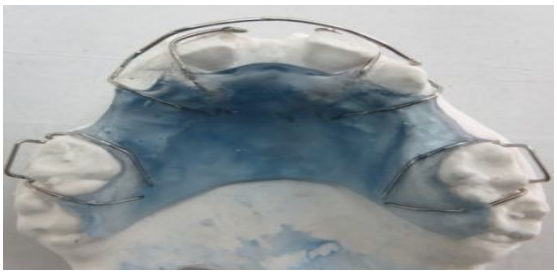
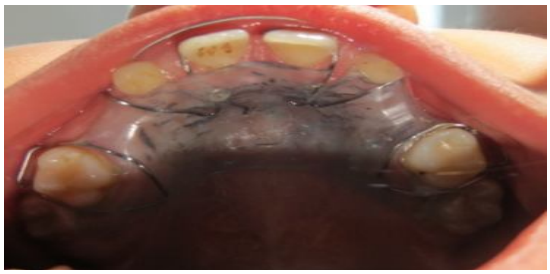



Figure 2: Model view of orthodontic walksplint. **Figure 3:** View of the patient with orthodontic walksplint.

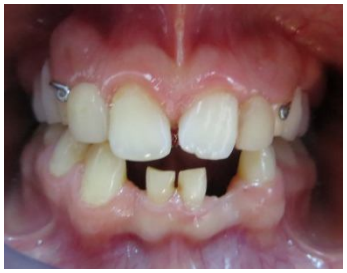


Figure 4: Intra-oral view of the patient after treatment.

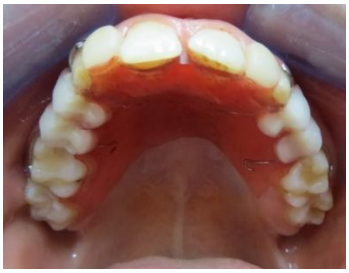


Figure 5: Intra-oral view of the patient after prosthetic treatment

CASE 2

Clinical examination of the 12 years old boy with Ectodermal dysplasia showed that he suffered from missing teeth, polydiestemia and conical teeth. It was observed that especially there were missing teeth in the maxilla and mandible. Extra-oral examination of the patient revealed dry skin, saddle nose, abnormal hair, protruding lips (Figure 9). It was learned that it was not a kin marriage and no problems existed in other members of the family. After prosthetic evaluation, it was decided to apply removable partial denture to the maxilla in order to meet functional and aesthetic expectations as the patient had not completed his growth and development. Detailed information was given to and approval was obtained from the parents of the patient (Figure 6-10).

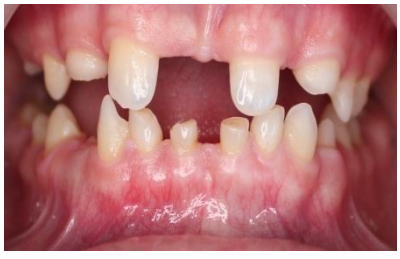


Figure 6: Intra-oral view of the patient before treatment

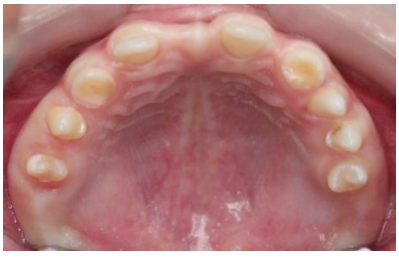
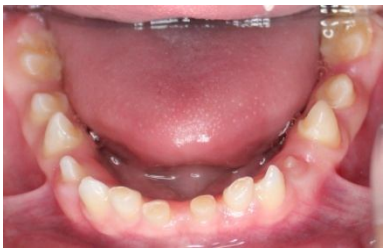


Figure 7: View of the maxilla before treatment



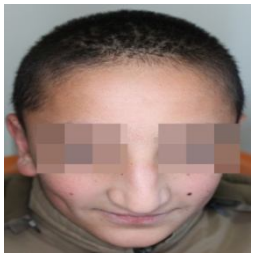


Figure 8: View of the mandible before treatment. **Figure 9:** Profile view of the patent before treatment

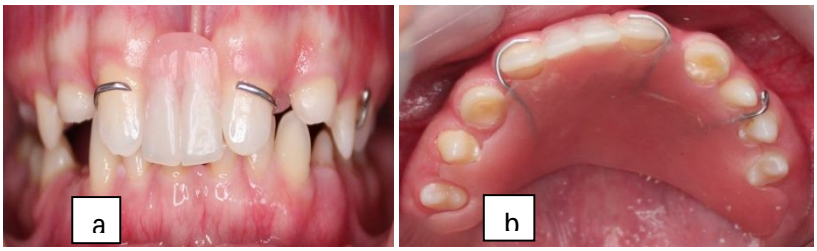


Figure 10a,b: Intra-oral view after treatment

After oral preparations were completed, preliminary impressions were made with irreversible hydrocolloid (Hydrocolor 5, Zhermack, Zhermack SpA., Badia Polesine, Italy) and personal impression spoon was obtained for the maxilla from acrylic resin on the models. Using this personal impression spoon second impression was made with hydrocolloid impression material (Hydrocolor 5, Zhermack, Zhermack SpA., Badia Polesine, Italy). Laboratory procedures of the removable partial denture were performed with known methods. Vertical size and occlusion control was performed and delivered to the patient (Figure 10a,b). Attention was paid to produce the prosthesis so as to ensure the normal vertical size of the patient. As a result of the prosthetic treatment the lost function, aesthetic and phonation was restored and no problems were experienced in the acceptance of the prosthesis by the patient. Taking into consideration the physical growth of the patient, his appointments are going to check on for prosthesis control and follow up oral hygiene in six month 6 a monthly basis.

CASE 3

A female child patient at the age of 4 applied to Department of Paediatric Dentistry, Faculty of Dentistry, Dicle University with aesthetic and functional problems caused by lost teeth. The patient was previously diagnosed as HED; it was seen that the results of clinical and radiological examination showed that the patient suffered from missing teeth and conical teeth anomaly which coincided with this disease. From the medical history obtained from family of the patient, it was found out that her father and mother were not relatives, and her sister also suffered from similar problems. Extra oral examination showed that the patient did not have any problems with hair and eyebrows, but she explained that her hair could be easily broken. In addition, dryness in skin and weakness in nails was witnessed.

Inside the mouth of the patient 55,53,51,61,63,65,71,73,81 and 84 numbered teeth was observed; in the panoramic film the germ of teeth numbered 16, 13, 11, 21, 23, 26, 31, 32, 34, 36, 41, 44 and 46 was observed. The parents were informed about the treatment process and planning of the patient. The treatment stages in case 2 were followed and removable partial denture was produced to both mandible and maxilla. Attention was paid to produce the prosthesis so as to ensure the normal vertical size of the patient. As a result of the prosthetic treatment the lost function, aesthetic and phonation was restored and no problems were experienced in the acceptance of the prosthesis by the patient. Taking into consideration the physical growth of the patient, her appointments are going to check on for prosthesis control and follow up oral hygiene in six month 6 a monthly basis. (Figure 11-13)

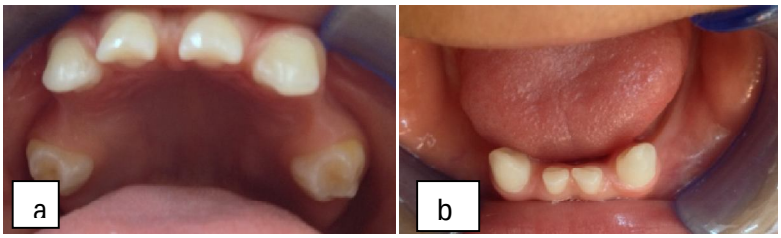


Figure 11a,b: Intra-oral view of the patient before treatment

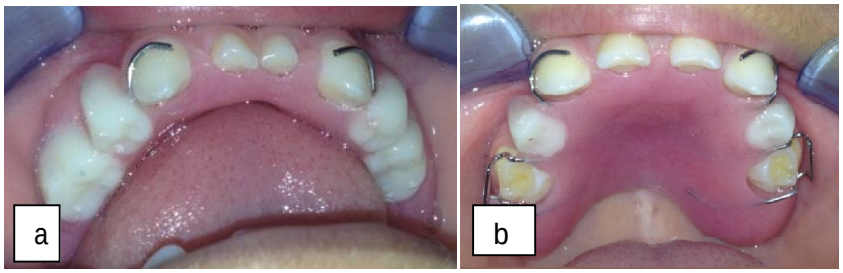


Figure 12a,b: Intra-oral view of the patient after treatment

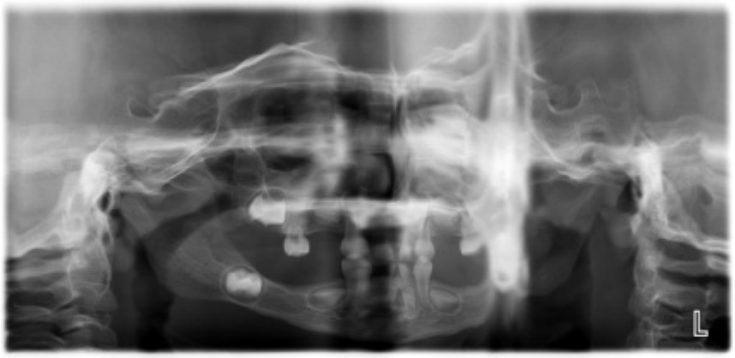
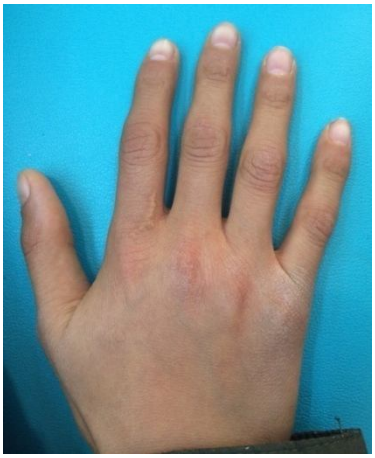


Figure 13: Panoramic view of the patient.

CASE 4

A male patient at the age of 14 applied to Department of Paediatric Dentistry, Faculty of Dentistry, Dicle University with aesthetic and functional problems caused by lost teeth. The patient was previously diagnosed as HED; it was seen that the results of clinical and radiological examination showed that the patient suffered from missing teeth and conical teeth anomaly which coincided with this disease. From the medical history obtained from family of the patient it was found out that her father and mother were not relatives, and other children did not have any health problems. The parents of the patient told that he suffered from sweating and hearing problems. In addition, dryness in skin, weakness in nails was detected. It was also reported that he had frequent upper respiratory tract infections.

As a result of the intraoral examination, it was observed that 17,16,15,14,53,13,11,21, 23,63,24,65,26,27,71,32,73, 34,75,36,37,81,42,83,44,85,46 and 47 numbered teeth existed. Radiological examination revealed 38 and 48 numbered tooth germs. The 14, 71, 81 and 75 numbered teeth of the patient were restored with composite fillings (Filtek™ Ultimate 3M ESPE). Periodontal treatment of the patient was conducted and polishing was applied to the teeth. The patient was called for dentist control with 6 month intervals (Figure 14-17).



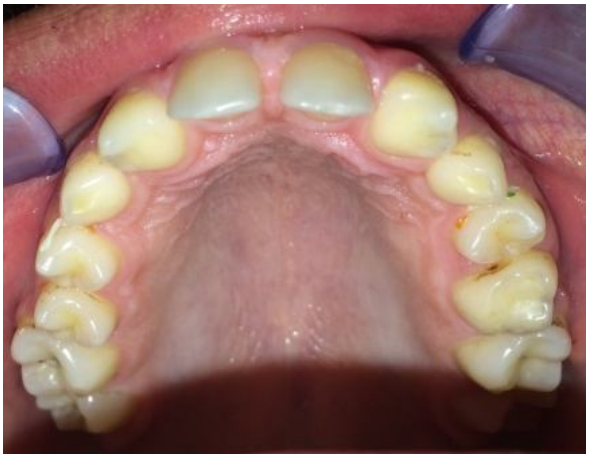


Figure 14: Skin and nail view of the patient. **Figure 15a:** Intra-oral view of the patient before treatment.

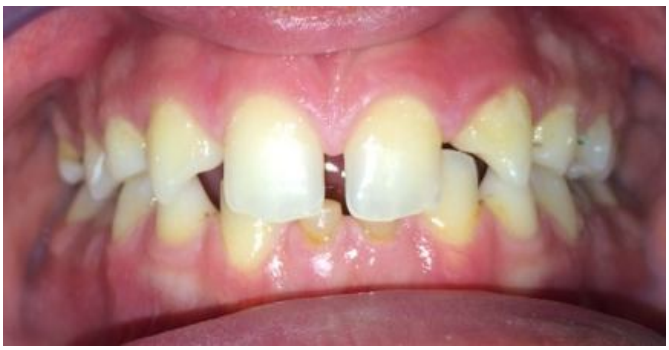
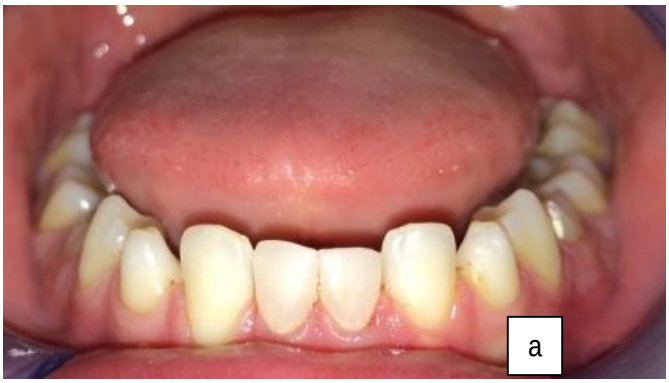


Figure 15b: Intra-oral view of the patient before treatment.



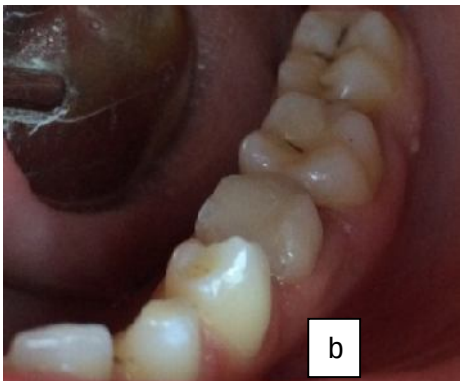


Figure 16a,b: View of the patient after restorative treatment of 71, 81(a) and 75(b) numbered teeth.

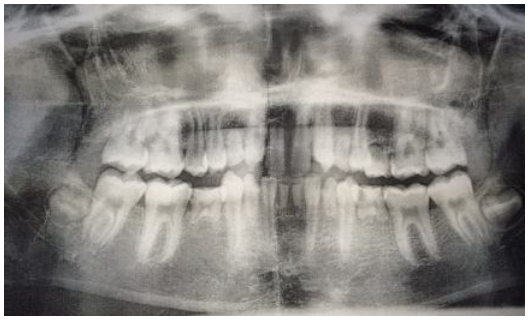


Figure 17: Panoramic view of the patient.

CASE 5

A male patient at the age of 10 applied to Department of Paediatric Dentistry, Faculty of Dentistry, Dicle University with aesthetic and functional problems caused by lost teeth. The patient was previously diagnosed as HED; it was seen that the results of clinical and radiological examination showed that the patient suffered from missing teeth and conical teeth anomaly which coincided with this disease. From the medical history obtained from family of the patient it was found out that her father and mother were not relatives, and other children did not have any health problems. Dryness in skin, thin hair and weakness in nails were detected in the patient. It was also reported that he had frequent upper respiratory tract infections.

As a result of the intraoral examination, it was observed that 16,53,63,36,85 and 46 numbered teeth existed. Stainless steel crown was produced for the 85 numbered tooth of the patient. After oral preparations were completed, 2, 3 stages of the case were performed. Vertical size and occlusion control was made and delivered to the patient. The patient was called for dentist control with 6 month intervals (Figure 18-20).

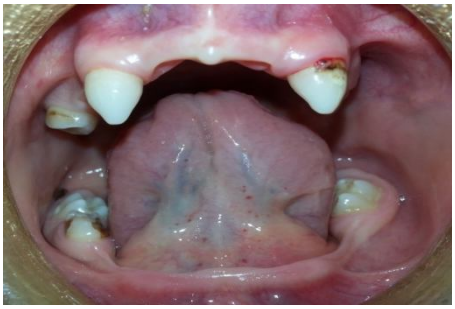


Figure 18: Intra-oral view of the patient.

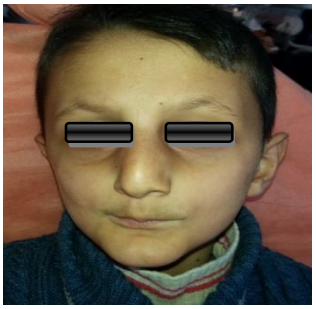


Figure 19: Profile view of the patient before treatment.

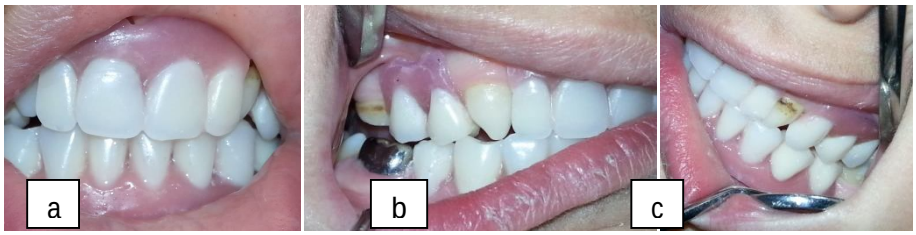


Figure 20a,b,c: Intra-oral view of the patient after treatment.

CASE 6

Male patient at the age of 17 who was diagnosed with ectodermal dysplasia applied to our clinique with complaint of missing teeth. In clinical and radiological examination, it was detected that maxilla had lost teeth, polydiestemia and conical teeth, and there were no teeth in the mandible, as the closing was Class III. In addition, extra-oral examination of the patient revealed dry skin, saddle nose, abnormal hair, and protruding lips. After prosthetic evaluation it was decided to apply fixed and removable partial denture for maxilla, and total denture for mandible so as to meet aesthetic, functional and phonetic expectations. Parents were informed in detail about the planning of the treatment of the patient and their approval was obtained. The prosthesis stages of the patient were conducted by following the stages identical to those in case 2. (Figure 21-23).

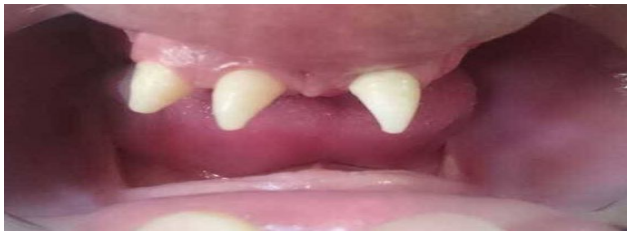


Figure 21: Intraoral view of the patient

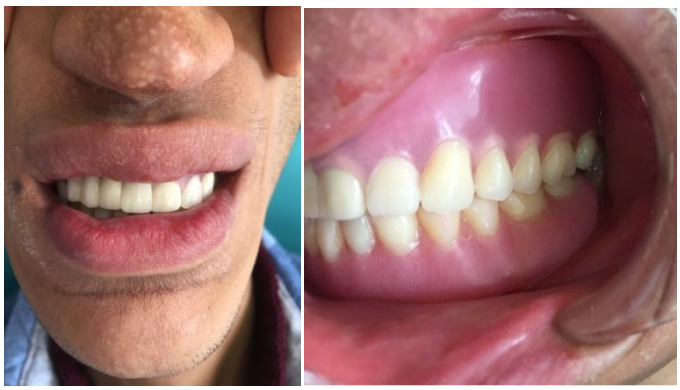


Figure 22a,b: Intraoral view of the patient after treatment

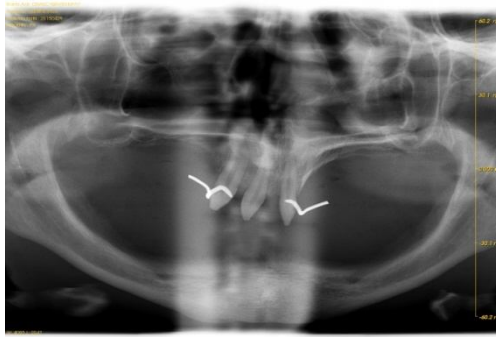


Figure 23: Panoramic view of the patient.

Discussion

Ectodermal dysplasia is a rare and genetically transmitted disease affecting ectodermal derived tissues such as skin, hair, nail and saliva glands, which affect two or more tissues (13). Ectodermal dysplasia, which is a genetic disease, is transmitted genetically depending on X chromosome; it is more frequently reported in males. It is heterozygote in female cases who carry normal X chromosome and an unhealthy chromosome, and they generally do not show all symptoms of the disease (14, 15). Genetic transmission model shows autosomal dominant, autosomal recessive and X-chromosome-dependent recessive characters (5, 16).

In a study, 68% kin marriage was detected in family histories of the ED patients (17). In our study, despite the fact that there are no kin marriages, it was detected that in one case the sibling also suffered from similar problems. In addition, in another case the mother is found to have suffered from congenital missing tooth. Basic findings of this syndrome are deformities related to skin, teeth, nails and hair (18). In recent studies it was reported that 49% of the cases had recurrent otitis media and 43% had voice problems. It was reported that these cases were prone to acute pharyngo-laryngitis and pulmonary infections (19). In one case with HED findings Goyal et al. detected temporomandibular joint ankyloses and cleft palate anomalies (20). In addition, it was reported that 75% of these patients suffered from ophthalmological problems. These problems were hyperpigmentation (88.9%), eyebrow anomalies (88.9%) and xerophthalmia (61.1%). In all of our findings skin, nail, tooth and hair anomalies can be found. In addition, in our findings parents reported frequent formation of upper respiratory tract infections. In 4 cases hearing problems are also reported.

Hypodontia or anadontia can be frequently seen in patients with Ectodermal dysplasia. It is reported that with early rehabilitation of these patients function (chewing, swallowing), phonation and aesthetic development can continue. Complete or partial prosthesis made for this purpose can be effective (1). In all of our findings, missing teeth were witnessed which had negative impact on the development of chin and lips. Alveolar crest looks thinner and lips appear more protruding. For these reasons the aesthetic look of the face was also compromised. In order to cure the function, phonation and aesthetic of our cases, oral rehabilitation was made with partial prosthesis and restorative fillings.

Oral rehabilitation of patients with ED can be provided with fixed or moving prosthesis, overdenture prosthesis, crown coatings, implant-supported prosthesis or composite restorations. Convenient treatment options must be determined for the patient. In this manner the prosthesis will support the phonation of the patient and their chewing food functions (22).

Oral rehabilitation is more difficult in children with ED which requires a multidisciplinary approach (17). Prosthetic rehabilitation of patients with ED can include over denture, complete or partial moving prosthesis and fixed prosthesis supported by teeth or implants. In growth and development stage, using moving prosthesis is recommended over implant or fixed prosthesis. In patients with ED below18 years of age there are some ideas which support application of implants. In addition, at the age of 6, transverse growth of front section of mandible is accepted as stable. Transverse growth in maxilla can continue up to 20 years of age; thus, it is recommended that implant application should be postponed until growth and development period is completed. Implant can be opted for in planning of treatment of ED patients. However, taking skeletal and dental development into consideration, implant can be applied by choosing a proper site (10). In our cases, as skeletal and dental development continued, fixed prosthetic and implant treatment was not applied.

Hearing disorders can be witnessed in patients with ED. In such cases prosthetic and surgical steps might be needed and planned for the estimated aesthetic and function requirements. Due to maxillary hypoplasia and hypodontia, supraeruption can be witnessed in mandibular dento-gingival complex which causes restrictions in restoration area. Alveolectomy is applied for implant and prosthetic treatment which will be performed in the sizes between restricted dental archs (23).

Early rehabilitation of patients with ED will cure the lost aesthetic, function and phonation and decrease social anxiety. Particularly in schooling period ED can cause psychological problems in the children and their parents (24). The patients with ED are also affected both creates tissue malformations and the quality of life of patients (25).

Conclusion

Dental anomalies are among main findings in ectodermal dysplasia. In ectodermal dysplasia cases missing teeth is frequently observed. For this reason, protective treatment in the existing teeth of the patient is an important application so as to prevent possible tooth losses. Due to missing teeth the patents can suffer from functional, phonetical and aesthetic problems. Conservative and prosthetic treatments in dentistry can be useful in eliminating the problems that the patient can suffer in their social and psychological lives.

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