Oral Rehabilitation of Hypohidrotic Ectodermal Dysplasia – A Case Report and Review

Chetan Patidar[1,*], Anand Shigli[2], Divya S Sharma[3], Vinaya K Kulkarni[4], Sanjay Sharma[5]

[1]Senior Lecturer, [2]Dean, Professor & HOD, [3,4]Professor & HOD, [5]Reader, Modern Dental College & Research Centre, Indore, Madhya Pradesh

***Corresponding Author:**
Chetan Patidar
Senior Lecturer, Modern Dental College & Research Centre, Indore, Madhya Pradesh
Email: drchetanpatidar@hotmail.com

Abstract

Ectodermal dysplasia is a hereditary disorder that occurs due to consequence of disturbances in the ectoderm of the developing embryo. It is usually accompanied by lack of sweat glands and a partial or complete absence of primary and/or permanent dentition. A case report illustrating the prosthetic rehabilitation by flexible and removable partial denture for a girl child of 9 years with Ectodermal dysplasia associated with congenitally missing primary and permanent teeth, mid facial defect and dental caries is presented. However oral rehabilitation of such case particularly in a child is often difficult, treatment should be accomplished by a multidisciplinary team.

Keywords: Anodontia, Ectodermal dysplasia, Hereditary disorder, Prosthetic rehabilitation, Partial denture

Introduction

The term Ectodermal dysplasia was coined by Weech[1] (1929). Ectodermal dysplasia syndrome is a heterogeneous group of inherited disorders, comprising more than 170 different clinical conditions with incidence of 1:100,000.[2,3] The syndrome was first described by Thurnam[3] (1848)and later by Darwin in 19th century. Both autosomal dominant and autosomal recessive type of syndrome show similar clinical inheritance.[3,4] The manifestations of which can be present in more than one derivative tissues primarily the skin, hair, eccrine glands, and teeth.[5,6] The etiology of ectodermal dysplasia is the genetic mutations in the ectodysplasin-A and ectodysplasin-A receptor genes are responsible for X-linked and autosomal hypohidrotic ectodermal dysplasia.[7]

Freire-Maia and Pinheiro[8] (1982) were the first to classify the ectodermal dysplasias, which was again updated in 1994.[9] Their original classification system divided the condition into various subgroups depending on the presence or absence of clinical findings such as (1) hair anomalies or trichodysplasias, (2) dental abnormalities, (3) nail abnormalities or onychodysplasias, and (4) eccrine gland dysfunction or dyshidrosis. Overall, the condition were classified into either group A disorders, which were manifested by defects in at least 2 of the 4 classic ectodermal structures as defined above, with or without other defects, and group B disorders, which were manifested by a defect in one classic ectodermal structure (1-4 from above) in combination with a defect in another ectodermal structure. Priolo and Lagana[10] (2001) reclassified and divided ectodermal dysplasias into 2 main functional groups: (1) defects in developmental regulation/ epithelialmesenchymal interaction and (2) defects in cytoskeleton maintenance and cell stability. Lamartine[11] (2003) classified the condition into 4-functional groups depending on underlying pathophysiologic defect: (1) cell-to-cell communication and signaling, (2) adhesion, (3) development, and (4) other. Clinically, the condition was broadly classified as two major types depending on the functionality of the sweat glands: (a) X-linked anhidrotic or hypohidrotic, where sweat glands are either absent or significantly reduced in number (Christ-Siemens- Touraine syndrome), and (b) hidrotic, where sweat glands are normal and the condition is inherited as autosomal dominant (Clouston's syndrome). The dentition and hair are affected similarly in both types.[7] A differential diagnosis can be Alopecia areata, Werner syndrome, incontinentia pigmenti, focal dermal hypoplasia, familial simple anhidrosis, and dyskeratosis congenita are some of the conditions.[12] Classical triad of ectodermal dysplasia consists of reduced hair follicles (varying from sparse scalp hair to complete alopecia), brittle nails with palmo-plantar hyperkeratosis (usually accompanied by reduced number of sweat glands), oligodontia/ anodontia of primary and/or permanent dentition with or without cleft lip and palate. Lack of alveolar ridge as a result of oligodontia is characterized by reduced vertical dimension of the lower face. Affected child's face appear older to his/her age.[13] Edentulous dental arches not only leads to masticatory problems in affected child but also affect the craniofacial growth as well.[13,14] A cephalomatric study by Vierucci et al[15] have shown significant differences in craniofacial features of affected and unaffected children with Ectodermal dysplasia.

Removable prosthesis is the treatment for the dental management of ectodermal dysplasia especially

in growing child. However it is difficult to fabricate denture for a child with ectodermal dysplasia due to lack of alveolar bone; therefore, restoring function and appearance is more challenging than usual.[14] Follow-up by a multidisciplinary team involving pediatric dentistry, orthodontics, prosthodontics, and oral-maxillofacial surgery specialists is advocated to be the most appropriate approach in such cases.[16]

This case report aimed to describe the prosthetic rehabilitation of 9 year girl child with hypohidrotic ectodermal dysplasia associated with severe hypodontia.

Case Report

A 9-year-old girl reported with her parents to the Department of Pedodontics and Preventive dentistry, Modern Dental College and Research Centre, Indore with the complaint of multiple missing teeth since childhood. On detailed history gathering from parents revealed that the patient was the only child and there were no other such cases with similar findings among close relatives. Patients were more concern about her inability to chew due to missing teeth. The parents also gave a history of delayed eruption of teeth and retention of primary front teeth. Further history gathering revealed less sweating and more sensitiveness towards heat. There was no history of consanguineous marriage between the parents and grandparents.

On extra oral examination, the patient had dry skin, fine textured hair, outwardly placed ears and flattened nasal bridge. Both upper and lower eyelids showed sparse eyelashes. Retrognathic mandible with convex profile and lower lip trap is seen (Fig. 1). Intraoral examination revealed multiple missing primary teeth no. 71,72,74,75,81,82,84 and 85. Dental caries in teeth no. 55,54,53,63,65; Coronal discoloration of 51 secondary to dental trauma 3 years back, increased overjet, thin alveolar ridges, reduced bone height and sulcus depth in the edentulous regions of lower jaw (Fig. 2, 3 & 4). Panoramic radiography revealed congenitally bilaterally missing dental follicles of teeth no. 15,25,17,27,35,45,32,31,41 and 42 (Fig. 5).

In order to improve appearance, mastication, and speech, a removable and flexible partial dentures for mandible arch were suggested to be the treatment of choice. At first treatment appointment, restoration with decayed teeth and pulpectomy for 51 was started. Following that an alginate (Algitex, DPI) impression was taken. Acrylic base plate was used to take jaw relation and try-in was done, any interference in occlusion and jaw movement are adjusted in the try -in phase (Fig. 6). Heat cured flexible denture material (D-FLEX^{TM}, Myerson Duraflex) was used for fabrication of partial denture from (Fig. 7). After the final insertion (Fig. 8), routine hygiene instructions for the dentures were given to both the child and her parents. The patient was advised for soft diet initially for few days. Parents were also instructed to remove the dentures at night to promote healing of the oral tissue. Completion of restorative treatment was done in subsequent visits. However after initial lack of compliance, the child tolerated the dentures well. To allow dynamic growth and development, the patient was advised for follow up visits every month initially and then three monthly. Good retention of prosthesis was observed and the parents reported an appreciable improvement in mastication at recall visit after three months. Parents were given anticipatory guidance regarding need of continued follow-ups for modification or replacement of the prosthesis to maintain harmony with developing jaws. Parents were further referred to speech therapist for speech development.

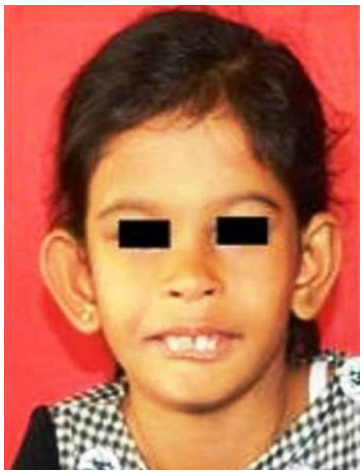

Fig. 1: Facial view of the patient

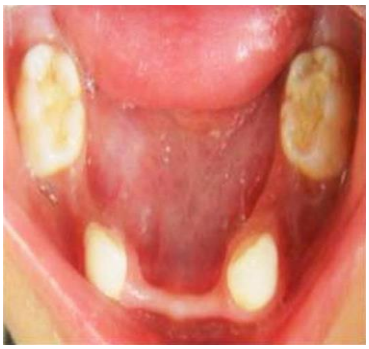

Fig. 2: Intraoral view of the lower jaw

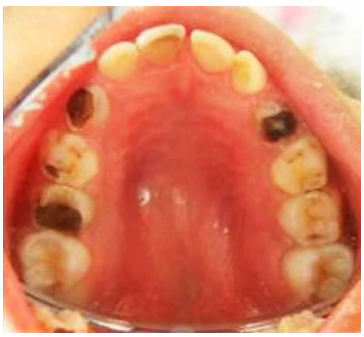

Fig. 3: Intraoral view of the upper jaw

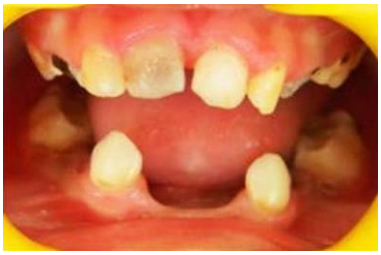

Fig. 4: Intraoral view before treatment

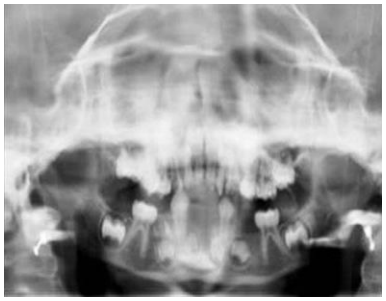

Fig. 5: Panoramic radiograph confirming multiple congenitally missing teeth

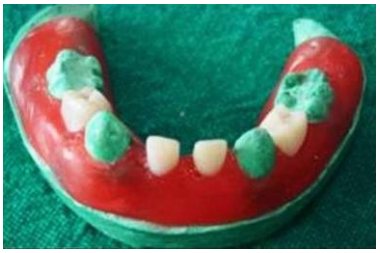

Fig. 6: Acrylic bases plate with acrylic teeth in modeling wax

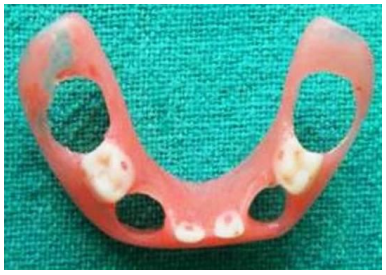

Fig. 7: Finished prosthesis

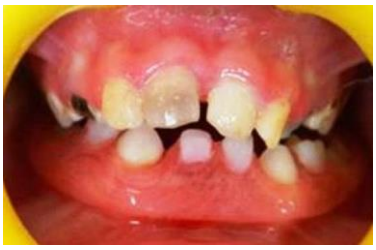

Fig. 8: Intraoral view after treatment

Discussion

It is now very well documented that dental findings in ectodermal dysplasia may range from hypodontia to anodontia of the primary or permanent teeth. Comprehensive rehabilitation of patient with ectodermal dysplasia is required to improve both the vertical and sagittal craniofacial relationship during growth and development in order to provide improved esthetics, speech, and masticatory efficiency. Removable prostheses are the most common treatment method.[17]

Although dental literature describes many conventional prosthetic approaches to the clinical management of these patients, osseointegration is also documented as a safe and predictable method for replacement of missing teeth. Dental implants are considered to be a treatment option especially in combination with implant supported dentures for adolescents over 12 years of age are recommended as a treatment choice in literature.[18] In situations where implant therapy is indicated, the main problem is insufficient bone; if bone atrophy progresses in these already alveolar- deficient patients, implant placement may not be possible without bone grafting.[13] Further such reconstruction surgery required during implant placement is subjected to a greater risk of failure like ankylosis of teeth when compared to removable prosthetic treatment, also the surgical procedures as psychological non acceptance in pediatric patients[16,19]. In the present case, implant placement to replace missing teeth was not the treatment choice considering about the amount of remaining growth and development and lack of sufficient alveolar bone. Consequences of the delay in start of management of patient with ectodermal dysplasia in regards to speech, mastication, and psychological development were determined to be significant. Till and Marquez[20] recommend that an initial prosthesis can be planned when the child get admission in school, so that pateint may enjoy a better appearance and will have time to adapt to the prosthesis. Dental prostheses significantly help improve the tone of the facial expression and muscles of mastication that may compensate for the reduced vertical growth of craniofacial complex.[21]

As seen in present case, the efficacy of mastication and dietary patterns improved significantly with removable prosthesis.

Conclusion

Although ectodermal dysplasia is a rare genetic disorder, the clinical manifestations cause considerable social problems. A careful and a thorough examination is required to analyze different aspects of growth and development of craniofacial complex. Main concern is to restore normal function in such patients. A multidisciplinary approach for treatment of ectodermal dysplasia cases not only improve esthetics and function of stomatognathic system but also favors normal growth.

References

1. Weech AA. Hereditary ectodermal dysplasia (congenital ectodermal defect). Am J Dis Child. 1929;37:766-90.
2. Lamartine J, Munhoz Essenfelder G, Kibar Z, Lanneluc I, Callouet E. Mutations in GJB6 cause hidrotic ectodermal dysplasia. Nat Genet. 2000;26(2):142–144.
3. Prasad M, Prathyusha. Ectodermal dysplasia and partial anodontia: A case report. J Med Sci Res. 2015;3(1):40-42.
4. Thurnam J. Two cases in which the skin, hair and teeth were very imperfectly developed. Proc RM Chir Soc. 1848;31:71-82.
5. Monreal AW, Ferguson BM, Headon DJ, Street SL, Overbeek PA. Mutations in the human homologue of mouse dl cause autosomal recessive and dominant hypohidrotic ectodermal dysplasia. Nat Genet. 1999;22(4):366–369.
6. Monreal AW, Zonana J, Ferguson B. Identification of a new splice form of the EDA1 gene permits detection of nearly all X-linked hypohidrotic ectodermal dysplasia mutations. Am J Hum Genet. 1998;63(2):380–389.
7. Tarjan I, Gabris K, Rozsa N. Early prosthetic treatment of patients with ectodermal dysplasia: a clinical report. J Prosthet Dent 2005;93:419-424.
8. Pinheiro M, Freire-Maia N. The ectodermal dysplasias. Arch Dermatol. Apr 1982;118(4):215-6.
9. Pinheiro M, Freire-Maia N. Ectodermal dysplasias: a clinical classification and a causal review. Am J Med Genet. Nov 1 1994;53(2):153-62.
10. Priolo M, Lagana C. Ectodermal dysplasias: a new clinical-genetic classification. J Med Genet. Sep 2001;38(9):579-85.
11. Lamartine J. Towards a new classification of ectodermal dysplasias. Clin Exp Dermatol. Jul 2003;28(4):351-5.
12. Lowry RB, Robinson GC, Miller JR. Hereditary ectodermal dysplasia. Symptoms, inheritance patterns, differential diagnosis, management. Clin Paediatric 1966;5:395-402.
13. Imirzalioglu P, Uckan S, Haydar SG. Surgical and prosthodontic treatment alternatives for children and adolescents with ectodermal dysplasia: a clinical report. J Prosthet Dent 2002;88:569-572.
14. Vieira KA, Teixeira MS, Guirado CG, Gaviao MB. Prosthodontic treatment of hypohidrotic ectodermal dysplasia with complete anodontia: case report. Quintessence Int 2007;38:75-80.
15. Vierucci S., Baccetti T., Tollaw I. Dental and craniofacial findings in hypohidritic ectodermal dysplasia during primary dentition phase. J Clinical Ped Dent 1994;18:291-297.
16. Lo Muzio L, Bucci P, Carile F, Riccitiello F, Scotti C, Coccia E, et al. Prosthetic rehabilitation of a child affected from anhydrotic ectodermal dysplasia: a case report. J Contemp Dent Pract 2005;6:120-126.
17. Mehmet Bani, Ali Melih Tezkirecioglu, Nese Akal, Tamer Tuzuner. Ectodermal Dysplasia with Anodontia: A Report of Two Cases, European Journal of Dentistry 2010;4:215-222.
18. Albert D. Guckes et al Use of endosseous implants in a 3-year-old child with ectodermal dysplasia: case report and 5-year follow-up, Pediatric Dentistry 1997;19:4:282-285.
19. Rad AS, Siadat H, Monzavi A, Mangoli AA. Full mouth rehabilitation of a hypohidrotic ectodermal dysplasia patient with dental implants: a clinical report. J Prosthodont 2007;16:209-213.
20. Till MJ, Marques AP. Ectodermal dysplasia: treatment considerations and case reports. Northwest Dent 1992;71:25-28.
21. Itthagarun A, King NM. Ectodermal dysplasia: a review and case report. Quintessence Int 1997;28:595-602.