IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 17, Issue 9 Ver. 9 (September. 2018), PP 84-87
www.iosrjournals.org

Ectodermal Displasia; A Case Report

Abhinav Agarwal[1], Kaushik Kumar Pandey[2], Pratibha Katiyar[3], Fauzia Tarannum[4] , Mujtaba Ashraf [5], Anant Agarwal

PG student, Department of Prosthodontics, Career Post Graduate Institute of Dental Sciences, Lucknow[1] , Assistant Professor, Department of Prosthodontics, Career Post Graduate Institute of Dental Sciences, Lucknow [2], Associate Professor, Department of Prosthodontics, Career Post Graduate Institute of Dental Sciences,Lucknow[3] , Assistant Professor, Department of4 Prosthodontics, Career Post Graduate Institute of Dental Sciences,Lucknow[4] , PG student, Department of Prosthodontics, Career Post Graduate Institute of Dental Sciences,Lucknow[5] , PG student, Department of Prosthodontics, Career Post Graduate Institute of Dental Sciences

Corresponding Author: Kaushik Kumar Pandey

Abstract: *It is right of every individual to look good and have proper nutrition. In certain hereditiry condition like Ectodermal displasia, patient has difficulty in eating and esthetics is also poor. Ectodermaal dysplasia is congenital disorder which affects body part of ectodermal origin like hair, nails, teeth and sweatglands. In this case report prosthetic rehabilitation of female patient suffering from hypohydroitic ectodermal dysplazia is explained.*

Keywords: *ectodeermal displasia, hypohydriotic ectodermal displasia, congenital disorder, missing teeth*

Date of Submission: 10-09-2018 Date of acceptance: 27-09-2018

I. Introduction

Genetically transmitted diseases are often common to some patients. Hereditary Ectodermal Displasia is rare group of disorder affecting mainly childeren. It is a x linked recessive trait, in which female is carrier and male predilection. Ectodermal Displasia(ED) is first described by Thurman and The term Ectodermal dysplasia was coined by Weech (1929). Synonyms of ED are Anhydrotic Ectodermal Dysplasia, Ectodermal Dysplasia Anhidrotic And Christ-Siemens-Touraine Syndrome. In this sweat glands are either absent or reduced in number.The condition is thought to occur in approximately 1 of 1 of 100000 live births. Freire maia and pinherio described 117 varieties of ED with multiple combination of abnormal ectodermally derived structures. Hypohydrotic ED resultant defective formation of ectodysplasin protein. In ED, the clinical features shows sparse hair, scanty eyebrow and eye lashes, depressed nasal bridge, frontal bossing, reduced vertical facial height and depth and marked resorption of maxillary and mandibular alveolar ridges.[1-4] Intraoral findings are congenitally mising teeth with retained decidous teeth. In anterior region of maxilla and mandible teeth are conical in shape. Other features are mid face hypoplasia, pre-oricular skin shows fine wrinkling with hyperpigmentation, reduced salivary secretions and voice quality is also compromised. Freire-Maia and Pinheiro (1982) were the first to classify the ectodermal dysplasias, which was again updated in 1994.[2] They 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. There were various other classification of ED depending upon clinical features and combination of involved ectodermal structures. 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). In the presented case report, prosthodontic rehabilitation of ectodermal displasia patient is described.[3, 5, 6, 7]

CASE REPORT:-

A 20 year old female patient was reported to the department of Prosthodontics, of Career Post Graduate Institute of Dental Sciences & Hospital Lucknow, with chief complaint of difficulty in eating and bad appearance in relation to upper and lower front region. The patient was a student. Patient had given the dental history of congenitally missing some permanent teeth and retained deciduous teeth. Intraoral examination of

upper arch revealed missing all permanent teeth except 11, 21 and Retained maxillary deciduous teeth are B C D E. In lower arch missing teeth was 31 and 41. The patient's diagnosis was done as hypohydroitic ectodermal dysplasia. Main features of hypohydroitic ectodermal dysplasia was found in patient; frontal bossing, less hair (hypotrichosis), congenitally missing teeth (hypodontia), lack of breast development. Thus On the basis of patient's current condition; a simplified treatment approach was planned. On radiographic investigation it was found that deciduous teeth should not be extracted and can be used as abutment. Individual crown is planned on permanent teeth 11, 21 and deciduous teeth B, C, D, and E. in mandibular arch 4 unit fixed bridge is planned on 32 and 42 to restore 31 and 41.(Fig. 1, 2, 3)

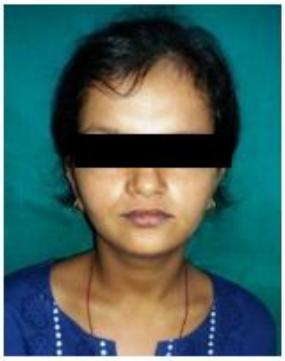

Fig. 1 pretreatment front view of patient

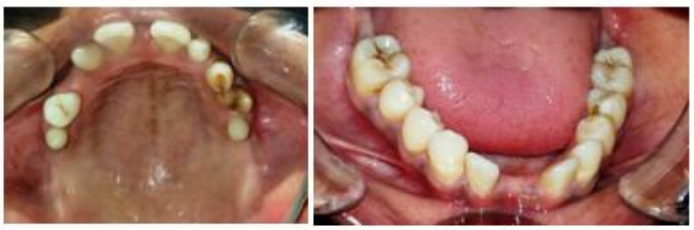

Fig.2 and Fig 3: showing missing all permanent teeth except 11 and retained deciduous BCDE in maxillary arch and missing 31, 41 in Mandibular arch

Treatment Steps :-

Diagnostic mounting:-

Diagnostic impression was taken with alginate impression material and cast was poured with type III gypsum product. Diagnostic face bow record was taken and with the help of inter- occlusal record cast was mounted. Vertical raised 2 mm and hard splint was given to patient for 45 days. Diagnostic wax- up was done and putty index made.

Teeth Preparation:-

Patient was referred to department of periodontics for scaling and root planning. Patient was further referred to department of endodontics for root canal treatment of permanent teeth 11, 21 and deciduous teeth B, C, D. Mandibular teeth 32 and 42 was prepared first and 4 unit temporary bridge was made. Maxillary permanent teeth 11, 21 and deciduous teeth B, C, D, E were prepared and temporization was done. Patient was recalled after one week and evaluated for pain and intraoral swelling, sinus. Final impression with light body and putty was taken by using putty wash technique. (Fig. 4, 5, 6)

Lab Procedure and fixed teeth trial:-

Models were retrieved and final face bow record was taken. After getting centric interocclusal record with alu wax; cast were mounted on Hanau wide view semi-adjustable articulator. In lab metal copings were fabricated for metal ceramic fixed prosthesis. Coping trial was done with minor adjustments. Unglazed trial and final cementation was done after verification of shade, esthetics and phonetics. (Fig. 7, 8, 9)

Follow up:-

Patient was recalled after 1 week, 6 week and I year interval. Patient was satisfied with the prosthesis. (Fig. 10, 11, 12)

Fig. 4 & Fig. 5: teeth preparation mandibular and maxillary

Fig. 6: Face bow record

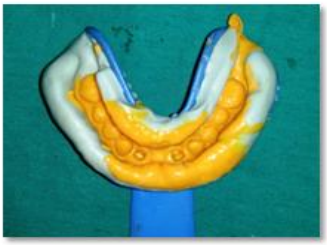

Fig. 7: FINAL IMPRESSION

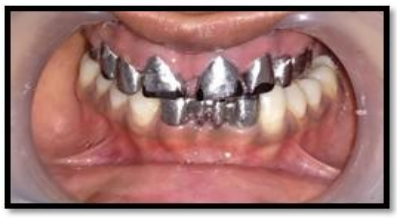

Fig. 8: Coping trial

Fig. 9:- Final prosthesis on semi-adjustable articulator

Fig. 10: Before

Fig. 11: After

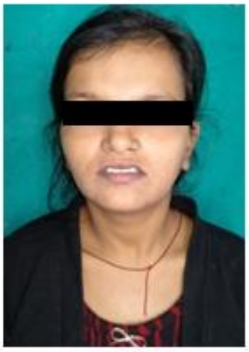

Fig. 12: Confident Patient with smile correction

II. Discussion

Prosthetic rehabilitation of ectodermal dysplasia patient is challenging for both clinician and patients. The main motive of clinician is to provide a good prosthesis, so that the esthetics, form, function, phonetics, sagittal and vertical jaw relationship during growing stage of patient should be proper.[7-10]

Ectodermal dysplasia is very common in India. It should be diagnosed in early stage so that the line of treatment is proper and accurate. The diagnosis is based on familial history, changes in teeth shape, missing teeth, less hair, less sweat glands, changes in nail shape and color. First of all, the patient is suffering from ectodermal dysplasia has to be rehabilitated with removable prosthesis. Removable prosthesis can be complete denture, removable partial denture, or overdenture. In mixed dentition, crown root ratio is evaluated; for fixed dental prosthesis. All abutment teeth should be evaluated in terms of crown root ratio, periodontal health, and healthy occlusion. Primary retained teeth can be abutment in fixed dental prosthesis, if it has fulfilled all criteria. After 12 years age, implant supported fixed dental prosthesis can be the given to the patient, if sufficient bone is there for implant placement. In atrophic conditions, the only option for implant placement is bone grafting. Many patients are not ready for implant supported prosthesis due to surgical interventions, high treatment cost and duration of treatment. So prosthetic management of Ectodermal Dysplasia patients depends mainly on complete denture, tooth supported overdenture with and without attachments and fixed dental prosthesis.[1, 2,9,11]

In this case report, fixed dental prosthesis was planned for rehabilitation of mandibular anterior and maxillary full arch. Following corrections were done with fixed dental prosthesis; maxillary anterior midline space closer, correction of proportion of maxillary tooth shape and size, shade of the teeth, chewing efficiency enhanced, rehabilitation of mandibular missing anterior teeth. Patient's self confidence was drastically improved due to smile correction. Patient is satisfied with the prosthesis in terms of esthetics and function.

III. Conclusion

The patients suffering from Ectodermal Dysplasia have multiple missing teeth and retained deciduous teeth. These young patients look like old edentulous patients. These patients lack self confidence and have poor smile and masticatory efficiency. Prosthetic rehabilitation of these patients should be done with following; removable partial denture, complete denture, overdenture, tooth supported or implant supported fixed dental prosthesis.

References

[1]. Weech AA. Hereditary ectodermal dysplasia (congenital ectodermal defect). Am J Dis Child. 1929;37:766-90.
[2]. Rahul Prasad, Abdulaziz, Abdullah Al-Kheraif, Nidhi Kathuria, V.N.V. Madhav, S.V. Bhide. Ravikumar Ramakrishnaiah. Ectodermal Dysplasia: Dental Management and Complete Denture Therapy. World Applied Sciences Journal 20 (3): 423-428, 2012.
[3]. Tarjan, I., K. Gabris and N. Rozsa, 2005. Early prosthetic treatment of patients with ectodermal dysplasia: a clinical report. J. Prosthet Dent. 93: 419-424.
[4]. Hickey, A.J. and T.J. Vergo, Jr. 2001. Prosthetic treatments for patients with ectodermal dysplasia. J. Prosthet. Dent., 86: 364-368
[5]. Lo Muzio L, Bucci P, Carile F, Riccitiello F, Scotti C, Coccia E, et al. Prosthetic rehabilitation of child affected from anhydrotic ectodermal dysplasia: Acase report. J Contemp Dent Pract 2005; 6:120-6.
[6]. Tarjan I, Gabris K. Rozsa N. Early prosthetic treatment of patients with ectodermal dysplasia:a clinical report. J Prosthet Dent 2005; 93:419-24.
[7]. Ramesh K, VinolaD, John JB. Hypohidrotic ectodermal dysplasia: diagnostic aids and a report of 5 cases. J Indian SocpedodPrev Dent 2010; 28:47-54.
[8]. 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.
[9]. Prasad M, Prathyusha. Ectodermal dysplasia and partial anodontia: A case report. J Med Sci Res. 2015;3(1):40-42
[10]. Thurnam J. Two cases in which the skin, hair and teeth were very imperfectly developed. Proc RM Chir Soc.1848;31:71-82.
[11]. 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.

Dr. SanthiVadugu" 'Guillain-Barre Syndrome' with some distinct dispositions (DLE, TB meningitis, Hearing Loss) – A novel presentation." IOSR Journal of Dental and Medical Sciences (IOSR-JDMS), vol. 17, no. 9, 2018, pp 35-37.