Ectodermal Dysplasia: A Case Report of Aesthetic and Functional Rehabilitation

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Abstract

During the development of embryo ectodermal dysplasia occurs as a consequence of disturbances of the endoderm. Clinical features include hypotrichosis and palmoplantar hyperkeratosis or alopecia,a triad of nail dystrophy,a partial or complete absence of primary and/or permanent dentition,and lack of sweat glands. In this study, the syndrome ectodermal dysplasia shows a severe case of anodontia. Since the oral rehabilitation of these cases is often difficult; particularly in pediatric patients, treatment should be administered by a multidisciplinary team involving pediatric dentistry, orthodontics, prosthodontics, and oral-maxillofacial surgery.

Keywords: *Ectodermal Dysplasia; Aesthetic & Functional Rehabilitation; Anodontia.*

Introduction

Ectodermal dysplasia is a heterogeneous group of inherited disorders, resulting from the abnormal development of two or more tissues at a time, derived from embryonic ectoderm.[1] Analyzing several ectodermal dysplasia cases reveals that the syndrome is an X-linked recessive trait. The first clinical cases of Ectodermal dysplasia were reported in early 1792 by Danz, who described the congenital absence of teeth and hair in two Jewish boys. The prevalence of the syndrome is more in the case of females than males. In some cases, the mother of the patient exhibited hypodontia/conical teeth. Syndrome ectodermal dysplasia is divided into two categories and among two categories anhidrotic type is more severe manifestation. Clinical features include palmoplantar hyperkeratosis, alopecia, and dystrophy of nails some may found with partial or complete absence of dentition.

Finn reviewed 82 cases of anhidrotic ectodermal dysplasia and found that 5.4% had more mandibular teeth than maxillary and 63.5% had more maxillary teeth than mandibular teeth. And many of the teeth are conical in shape.[3] There is always a lack of development of alveolar ridge in anodontia patients. The vertical dimension of the face is also reduced along with dryness of oral mucosa and malformation of existing teeth.

Histopathology: The epidermis of Ectodermal dysplasia is thin with effacement of rete ridges around it. Sebaceous glands, Apocrine glands are variably reduced and mucous glands of the upper respiratory tract may be sparse. Light and scanning electron microscope findings of hair shaft abnormalities are longitudinal clefts or grooves and transverse fissuring, the bulb of the hair shaft is dystrophic. Also, mandibular X-ray shows dental aplasia or hypoplasia. Other histopathology features including epidermal and follicular orthokeratotic hyperkeratosis, apocrine ducts enter follicles at abnormal locations, and comedo formation.

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Differential Diagnosis: Differential diagnosis including Alopecia Areata, Aplasia Cutis, Incontinentia Pigmenti, Congenital focal dermal hypoplasia syndrome, Pachyonychia Congenital, Naegeli-Franceschetti-Jadassohn Syndrome.

Case Report: An 8-year-old female patient visited the Department of Pedodontics and Preventive Dentistry, Institute of Dental Sciences, Bhubaneswar, India, with the chief complaint of the absence of teeth in her oral cavity since childhood and 7 primary teeth erupted in the oral cavity with dryness of the mouth. There was no history of birth complications during his delivery, and no other live family member presented a similar condition.

And other medical history was taken including vocal function, bleeding disorders, other syndromes associated with this. All the medical history examined were normal.

On clinical examination, the skin appears to be dry (Figure 1A)and there is a mild elevation of temperature. Hair examination showed thin and less hairs. On examination of eye dryness and reduced lubrication was seen. Brittle nails were observed on finger examination. Ears were large and gives an older look (Figure 1B) gives an older look as compared with those of his age with a normal intelligence and partial Anodontia (Figure 1C). Intraoral examination revealed dry mucous membranes and reduced vertical heights of both the arches were of considerable significance (Figure 1D & E). On radiographic examination, there is the presence of tooth buds of permanent molars. In this case, the treatment modalities include temporary prosthesis(Figure 2 A to D). After there is a proper growth of arch and face, implants and permanent restoration will be provided.

According to the history and investigation treatment modalities had planned and appointment for the patient was scheduled.

Appointment I- proper case history with intraoral and extraoral photographs of the patient and OPG had taken. Non-pharmacological behavior management had performed by demonstration of instruments and friendly communication with the patient. Preliminary impression had taken using alginate and perforated impression tray. The patient was recalled after 7 days.

Appointment II- Border molding procedure and secondary impression had taken using ZOE impression material. Other lab procedures including denture base, occlusal rim, and articulation had done.

Appointment III- Most important procedure for the removable prosthesis 'Jaw relation' including anteroposterior, horizontal, and vertical relation had performed. Lab work, teeth setting on occlusal rim had completed.

Appointment IV- In this appointment patient had recalled for teeth trying, masticatory movement of jaws, and speech analysis. After that, all the lab work as well as dearticulating, packing, dewaxing, curing, finishing, and polishing of the denture had done.

Appointment V- Adjustment of denture had done and the removable denture had delivered to the patient with the postoperative instructions.

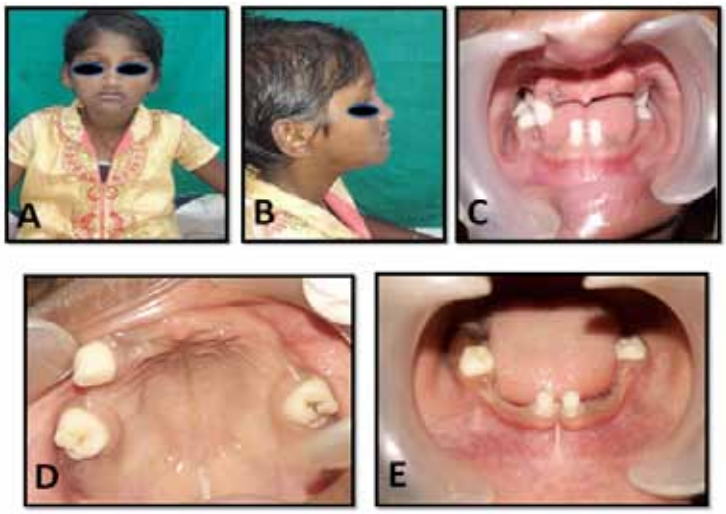

Figure 1A to E : (A) Dry skin, (B) Midface hypoplasia, (C) Partial Anodontia, (D and F) Upper and Lower Arch with a reduced vertical height of alveolar bone respectively.

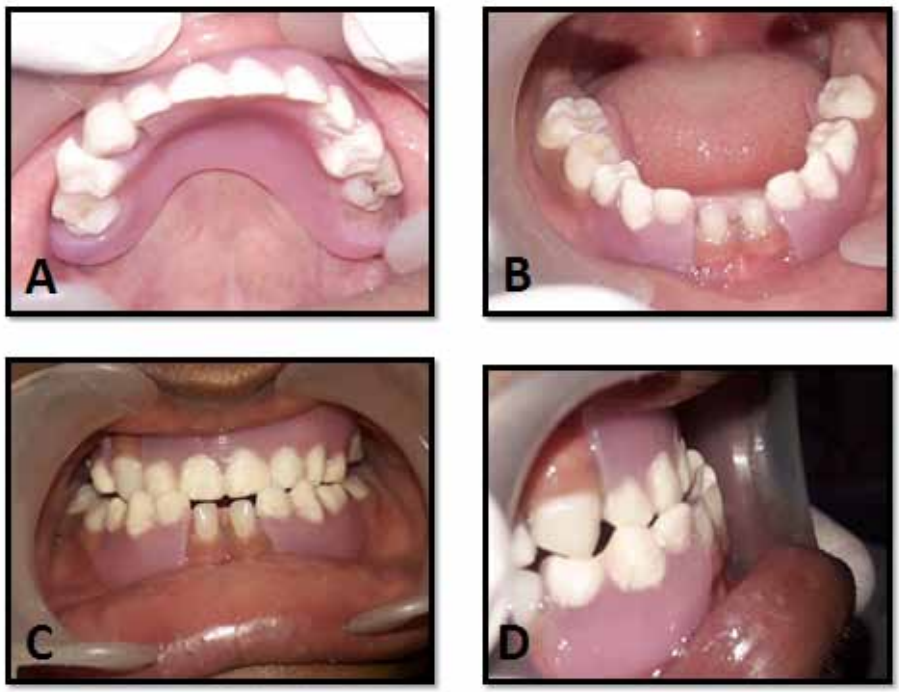

Figure 2A to D: Post-Operative Photographs

Discussion

Freire-Maia and Pinheiro proposed the first classification system of ectodermal dysplasia in 1982.[1] They classified ectodermal dysplasia into different subgroups according to the presence or absence of (1) anomalies of Hair(2) anomalies of Teeth (3) abnormalities of Nails (4) eccrine gland dysfunction.[6] As in the above-mentioned case report, all features of ectodermal dysplasia classification were present; like scanty hair, partial anodontia, thin-brittle nails, dry mouth as well as dry cornea.

Medical care of ectodermal dysplasia depends on which ectodermal structure is involved. In this case report, the temperature was raised,So it was advised for frequent intake of liquids to maintain hydration patient having dental issues are advised for early dental evaluation and treatment and encouragement for routine dental hygiene is done.[7] In the above case report, upper partial denture and lower complete denture was advised. After the construction of dentures, the patient was educated for adjustments and reconstruction of dentures at different stages of growth and advised that dental implants may eventually be required. Patients with xerostomia and reduced lacrimation may benefit from artificial saliva and tears respectively. The defect of the lip can be corrected by cheiloplasty to improve esthetics. The patient is advised to take consultation with a genetic counselor to find out the diagnosis and genetic analysis.[8]

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Ethical Approval: Approved

Conflict of Interest: None

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