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Case Report

Hypohidrotic Ectodermal Dysplasia: A Case Report

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ABSTRACT

Ectodermal dysplasia is a heterogenous group of hereditary disorders affecting development of two or more ectodermally derived anatomic structures. The Incidence of the syndrome is considered to be around 1 case per 100,000 live births. The two most common types of ectodermal dysplasias are hypohidrotic ectodermal dysplasia (Christ-Siemens-Touraine syndrome), and hidrotic ectodermal dysplasia (Clouston syndrome). Hypohidrotic ectodermal dysplasia is usually inherited as X-linked recessive pattern and has a full expression in males, whereas females show little to no signs of the disorder. The triad of nail dystrophy, alopecia or hypotrichosis and palmoplantar hyperkeratosis is usually accompanied by a lack of sweat glands and a partial or complete absence of primary and/or permanent dentition. In this case report prosthetic rehabilitation of a young boy with Hypohidrotic ectodermal dysplasia is presented.

Keywords- hypodontia, hypohidrotic ectodermal dysplasia, removable overdentures, prosthetic rehabilitation

INTRODUCTION

Ectodermal dysplasia(ED) is a genetic disorder affecting development of two or more ectodermally derived anatomic structures. There is hypoplasia or aplasia of structures such as skin, hair, nails, teeth, nerve cells, sweat glands, parts of the eye, ear and other organs. [1] Ectodermal dysplasia can be inherited in any form of several genetic patterns including autosomal-dominant, autosomal-recessive, and X-linked modes. [2] Incidence of the syndrome is considered to be around 1 case per 100,000 live births. [3]

Currently more than 170 different types of ectodermal dysplasias have been identified. They can be classified based on the presence or absence of four primary ectodermal dysplasia defects:

ED1: Trichodysplasia (hair dysplasia)
ED2: Dental dysplasia
ED3: Onychodysplasia (Nail dysplasia)
ED4: Dyshidrosis (Sweat gland dysplasia)

According to the presence or absence of sweat glands defect, two major groups are distinguished: (1) Hypo-hidrotic or anhydrotic (Christ-Siemens-Touriane syndrome) in which sweat glands are either absent or significantly reduced in number and seems to show an X-linked inheritance pattern; (2) Hydrotic (Cloustone syndrome) in which sweat glands are normal, which is inherited in an autosomal dominant pattern.

In general, the skin of affected children appears thin and lightly pigmented; surface blood vessels are easily visible. Other manifestations include fine sparse hair, reduced density of eyebrow and eyelash hair. When hair is present, it may be dry and fragile. Nails may appear small,

thick or thin, brittle, discoloured, cracked, and/or ridged. [4] Sweat glands may be absent or reduced in number or non-functioning due to which the affected individuals may display heat intolerance and elevated body temperature.

The teeth are markedly reduced in number (oligodontia or hypodontia) and often manifest abnormality in size and/or shape. The alveolar processes of the jaws remain underdeveloped due to lack of tooth bud formation. The affected individuals have old age appearance with protuberant lips due to reduced vertical dimension of occlusion. Xerostomia is also seen when salivary glands are affected.

This article presents the early prosthetic rehabilitation of a child with anhidrotic ectodermal dysplasia associated with oligodontia in mixed dentition.

CASE REPORT

A 6 year old boy reported to the Department of Pedodontics and Preventive Dentistry with the chief complaint of several missing teeth due to which he had difficulty in chewing food. He was intolerant to heat and had medical history of recurrent chest infection. According to his family history, his maternal uncle also had oligodontia and intolerance to heat.

Extraoral examination revealed large head size with sparse hair, lack of hair in eyebrows and midface deficiency with protruded lips. Apart from this purulent discharge from right ear was evident during his visit to hospital, which was in coordination with the history of frequent visit to ENT specialist due to recurrent ear infection.

The intra oral examination revealed 3 teeth in maxillary arch and 1 tooth in mandibular arch, all of them was peg shaped. The alveolar ridge appeared very thin and short in height, due to which the depth of vestibule also appeared to be shallow.

Orthopantomogram (OPG) revealed one more tooth present distal to the tooth present on the left side of maxillary arch. It also showed reduced height of alveolar bone in maxillary and mandibular arch.

Tooth supported overdenture was considered for the treatment to improve the mastication and aesthetics. Primary impression was recorded with alginate impression material. Custom made trays were prepared for the final impression which was recorded with elastomeric impression material. Jaw relation was recorded and teeth arrangement was done. After laboratory processing, overdentures were delivered and instructions for oral hygiene maintenance were given. Continuous follow-ups every six months were planned for adjustment or replacement of old denture.

Figure 1: Facial Profile Of The Ectodermal Dysplasia Patient: Front View

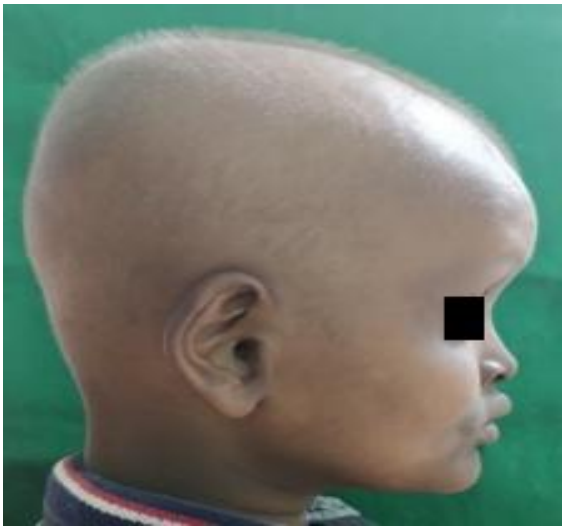

Figure 2: facial profile of the ectodermal dysplasia patient: lateral view

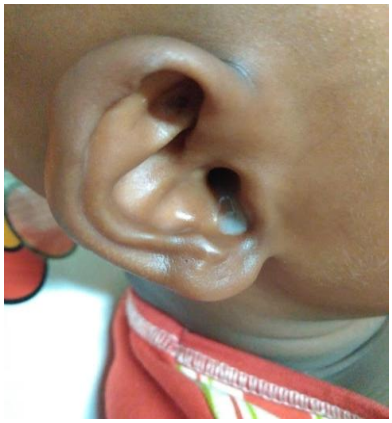

Figure 3: Purulent Discharge from Ear

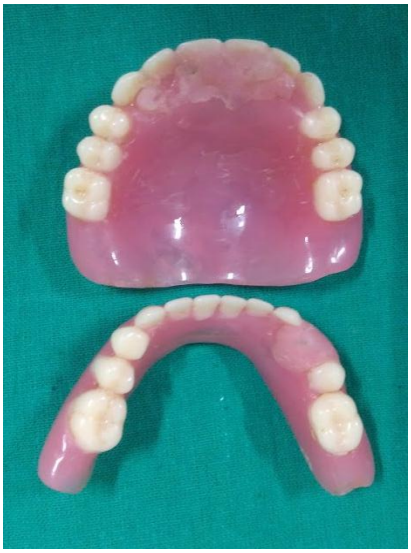

Figure 6: Removable Complete Denture

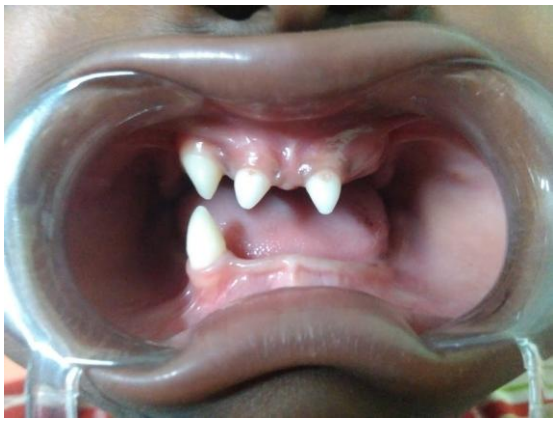

Figure 4: Intraoral Examination

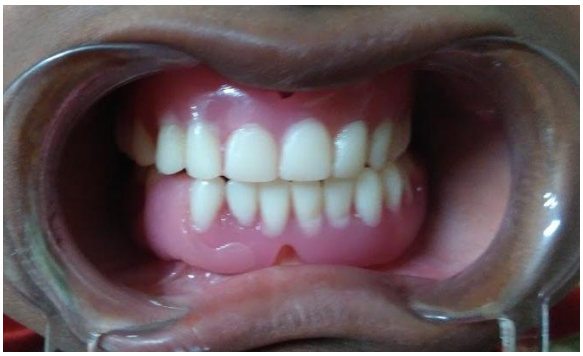

Figure 7: Removable Complete Denture Inserted

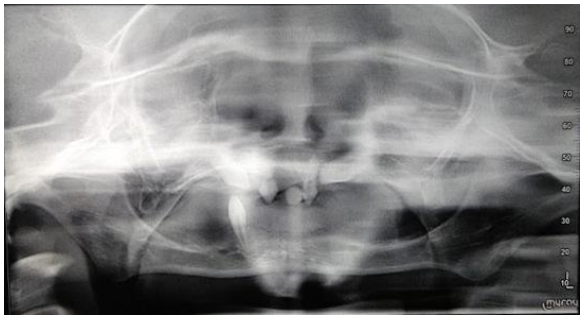

Figure 5: Radiographic Examination: OPG

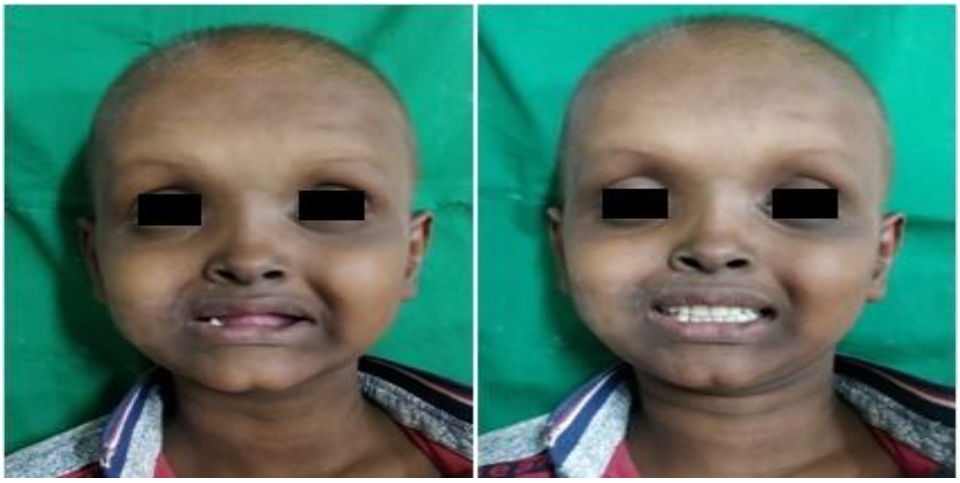

Figure 8: Improved Appearance after Insertion of Complete Denture

DISCUSSION

Ectodermal dysplasia is a rare congenital disorder resulting from disturbances in ectodermal development, first described by Thurman. [5] The clinical features may include nail dystrophy, hypotrichosis, palmoplantar hypokeratosis, odontodysplasia and dysplasia of the other structures of ectodermal origin. Among the manifestations, hair abnormalities occur with the highest frequency (>90% of patients); this is followed by defects of teeth (80%), nails (75%), and sweat glands (42%). [6] Dental abnormalities may be in the form of enamel hypoplasia, microdontia or partial to complete absence of teeth.

Dental management of a patient with ectodermal dysplasia is a multidisciplinary team approach with the clinical knowledge of growth, development and behavior management. [7,8] Treatment modalities for dental management of patients with ectodermal dysplasia includes: 1) Complete dentures, 2) Removable partial dentures, 3) Fixed partial dentures, and 4) Endosseous implants. [4] Prosthodontic treatment of each patient should be customized according to the patient's age, growth and development. [9]

Conventional removable prosthesis is a good option for young patients with ectodermal dysplasia as they require repeated change of prosthesis as the patients grow. [7] But the over-denture is advantageous as it helps in preserving the alveolar ridge for future implant placement and utilizes the natural undercuts in the teeth for retention. [7] Removable prosthesis requires regular adjustments and should be replaced when a decreased vertical dimension of occlusion and an abnormal mandibular posture are detected due to growth. [10] Long term follow ups are necessary to match up to the developing jaws. A broad guideline recommends relining/rebasing intraoral prostheses in a growing patient every 2-4 years and remaking a new prosthesis after 4-6 years. [11]

Conventional fixed prosthodontic treatment should not be used in children with hypohidrotic ectodermal dysplasia as they have a minimal number of teeth. The fixed partial dentures (FPDs) with rigid connectors should not be used in young, growing patients. [12] Individual crown restorations can be used, but larger pulp sizes and shorter clinical crown heights will be of concern. [13]

Implant retained prosthesis: In the last decades oral rehabilitation of partially or totally edentulous patients with dental implants has become common practice. [14] But implants should not be placed in growing children before completion of craniofacial growth as it will lead to several problems in routine practice. [15] This is especially crucial in the maxilla, where implants can be submerged by the downward growth of investing tissues. [16]

Cronin et al, stated that transverse growth of the maxilla occurs mostly at the midpalatal suture. [17] Thus, fixed implant constructions crossing the midpalatal suture will result in a transversal growth restriction of the maxilla. The insertion of implants in the growing maxilla should be avoided until early adulthood. Implants are indicated only in the anterior mandible of ectodermal dysplasia patients who are older than 12 years and exhibit anodontia. [18]

Provisional implants are an alternative option because they do not osseointegrate, they are retrievable and do not interfere with the growing bone. [9] Bergendal et al. demonstrated a frequent dental implant loss among patients with ectodermal dysplasia (64.3%), unlike patients with dental trauma and agenesis treated with dental implants. [19] Children with ED should be rehabilitated with smaller implants due to the reduced dimension of the maxillary and cortical bones. Sfeir et al. presented three cases of children with ED where mini-implants were used for prostheses retention. [20] The authors affirmed that the use of mini-implants should be part of the treatment in cases of ED and that their use reduces the

number of surgeries required for conventional implants. [20] Further clinical evaluations must be performed each six months to observe the need of mini-implant replacements and/or substitution of prostheses due to the growth and development processes.

Clinical studies with long-term follow-up are needed to test the mini-implants as an alternative for oral rehabilitation of children with ectodermal dysplasia.

In the present case, the patient had consistently raised body temperature along with frequent nasal and throat infections. His parents were advised to keep him hydrated with cool liquids consumption and bathing frequently to help in thermoregulation. He was advised to consult an ENT specialist for recurrent ear, nose, and throat infections. No diet restrictions were given.

The patient was advised to undergo genetic counseling to determine the underlying molecular basis of the disease. The life expectancy of patient with ectodermal dysplasia is usually normal. Murdock et al in 2005 carried out a study addressing cost analysis of dental treatment of patients suffering from ectodermal dysplasia and reported US$2038 to US $3298 for those who received prosthetic treatment alone to US$12,632 to US$41,146 for those who received a combination of orthodontic, prosthodontic and implant placement. [21] Keeping the financial condition of the patient in mind the best possible treatment plan was decided.

CONCLUSION

A proper family history of patient with ectodermal dysplasia along with assessment of the socioeconomic status of the family is very essential for making a proper treatment plan. Children with ectodermal dysplasia may present a shy and self withdrawn behavior due to their esthetic and functional limitations. Oral rehabilitation of such children at the earliest with removable overdentures is important to help him improve his esthetics, masticatory function and phonetics. This further helps to develop a positive self esteem and goes a long way to restore ones social life.

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