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Case Report

Ectodermal Dysplasia with Partial Anodontia: A Case Report and Review

Tarun Vyas[1*], Satish Bhosale[2]

[1]Department of Oral Medicine and Radiology, RR Dental College and Hospital, Udaipur, India

[2]Department of Oral Pathology and Microbiology, RR Dental College and Hospital, Udaipur, India

[*]Correspondence should be addressed to Tarun Vyas, Department of Oral Medicine and Radiology, RR Dental College and Hospital, Udaipur, India; Tel: 7568348887; E-mail: tarunvyas14@gmail.com

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ABSTRACT

Ectodermal dysplasia is an X-Linked, recessive hereditary disease characterized by dysplasia of tissues of ectodermal origin. In Hypohidrotic Ectodermal Dysplasia (HED) males are more affected than female .The incidence of ectodermal dysplasia is rare (1 in 100,000 birth). It is characterized by hypotrichosis, hypohidirosis with frontal bossing. Some they have partial anodontia or anodontia. This treatment plan would be able to provide psychological, aesthetic and functional boost to suffer.

Keywords: Midface hypoplasia, Hypohydrotic ectodermal dysplasia (HED), Partial anodontia.

INTRODUCTION

Ectodermal dysplasia, was initial illustrated by Thurman [1,2] may be a X-linked recessive, hereditary disorder going down as an impact of disturbances within the exoderm of the developing embryo. The triad of nail dystrophy (onchodysplasia), phalacrosis or hypotrichosis (scanty, fine light-weight hair on the scalp and eyebrows), and palmo plantar hyperkeratosis is typically accompanied by a scarcity of sweat glands (hypohidrosis) and a partial or complete absence of primary and/or permanent dentition [2-5].

Ectodermal dysplasia represents an out sized and sophisticated cluster of diseases comprise over a hundred and seventy completely different clinical conditions [3]. The incidence of this condition is 1:100,000, with a death rate of twenty eighth in males up to three years ancient [1] it's primarily present in male as compare to female. When a minimum of a pair of sorts of abnormal ectodermic options occur, like distorted teeth and very thin hair, the patient is diagnosed with ectodermic dysplasia syndrome [3,5].

There are two major styles of this condition counting on the quantity and practicality of the sweat glands: (1) sex chromosome anhidrotic or hypohidrotic, wherever sweat glands are either absent or considerably reduced in variety (Christ-Siemens-Touraine syndrome), and (2) secretion, wherever sweat glands are traditional and therefore the condition is inheritable as chromosome dominant (Clouston's syndrome) [1,3,5]. The dentition and hair are affected equally in each variety, however the hereditary patterns and nail and sudoriferous gland manifestations tend to dissent [2].

Christ-Siemens-Touraine syndrome, with sex chromosome recessive inheritance, is that the most often according manifestation of ectodermic abnormality [2,6,7] counting on the severity of clinical manifestations, Christ-Siemens-Touraine syndrome will be classified as either hypohidrotic or anhidrotic ectodermic abnormality [7]. Ectodermic dysplasia additionally referred to as Clouston syndrome. Two novel and six known mutations were identified in the EDA, EDAR, and WNT10A genes. New evidence ascertaining WNT10A as a candidate gene for HED was provided [8].

CASE REPORT

A 14-year-old boy given with a complaint of missing teeth to the Department of Oral medicine and radiology, R.R Dental college and Hospital, Udaipur. He was in the midst of his father who gave a history of the boy's missing teeth since infancy, aside from 2 teeth within the jaw that erupted once the boy was 6-years previous. He conjointly gave a history of frequent bouts of fever throughout infancy and childhood, and mentioned that he was intolerant to heat and takes frequent dips in summer to stay cool. There was no case history of missing teeth. The boy was moderately built and poorly nourished. His skin was dry and also the hair was scanty. He had the everyday face of an ectodermic abnormal condition patient with options like: frontal bossing, periorbital skin pigmentation, depressed nasal bridge, bulbous lips, low-set ears, and scanty scalp hair [9,10]. The nails weren't terribly brittle. It was fine in texture, truthful and short. Most of them were chromosome recessive inheritance. Eye brows are absent (**Figure 1**).

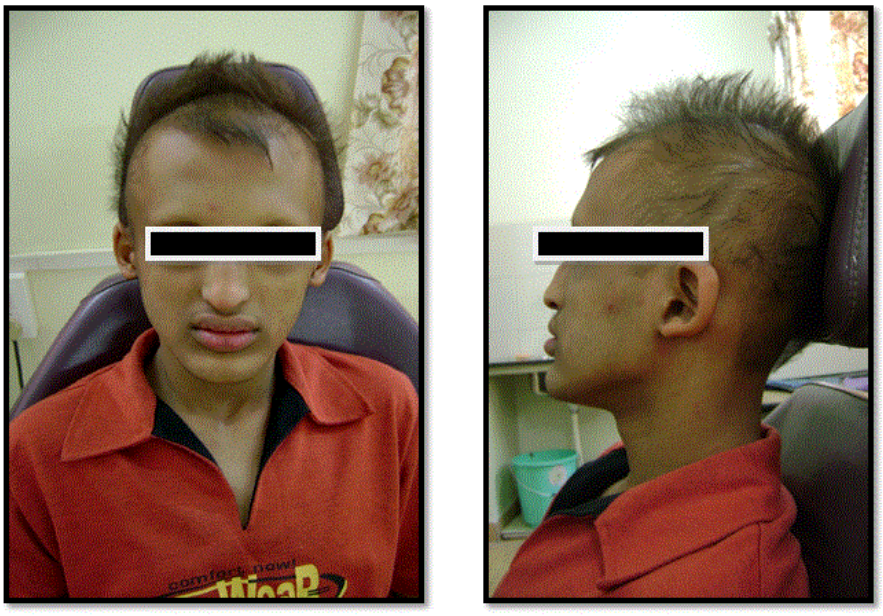

Figure 1: Extraoral examination of the patient.

Intraoral examination disclosed a comparatively dry mucosa, articulator arch was fully toothless with poorly developed alveolus and also the jaw arch had 2 standing molars (**Figure 2**). Picture taking investigations (OPG and lateral cephalogram) showed unshaped incisors compact within the jaw arch (**Figures 3 and 4**). Since hypohidrosis, hypotrichosis, and hypodontia were terribly evident on physical examination, the boy was diagnosed with Hypohidrotic germ layer abnormal condition (HED) with partial anodontia. The treatment of all patients with craniofacial anomalies includes not only the management of the defect or anomaly itself, however conjointly the psychological management of the person as a full [11,12]. Radiographically they need round shape angular teeth. During this we tend to see the generalized biological process of bone. Due to absence of teeth there was a scale back in vertical dimension with protrusive lip. This could be a case Hypohydrotic ectodermal dysplasia.

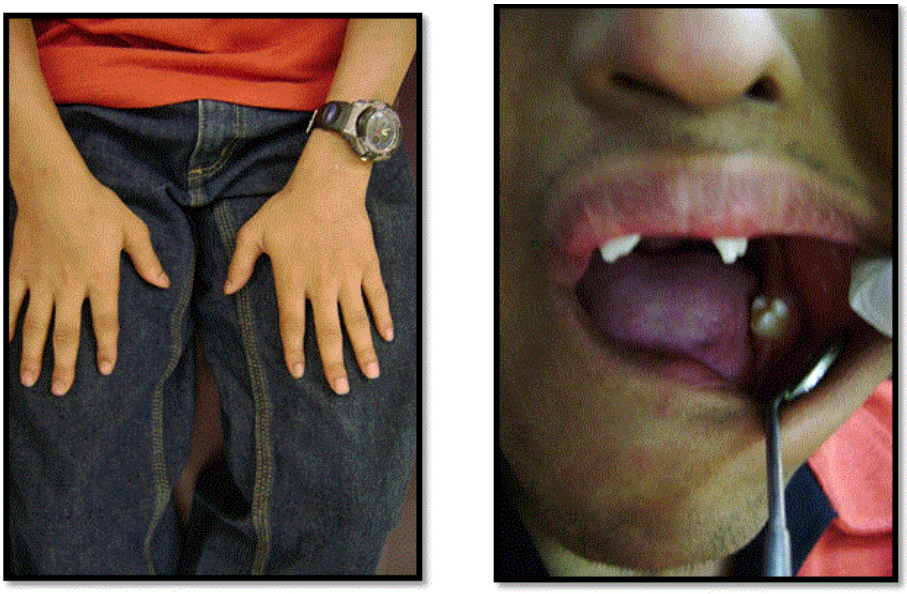

Figure 2: Intraoral examination of the patient.

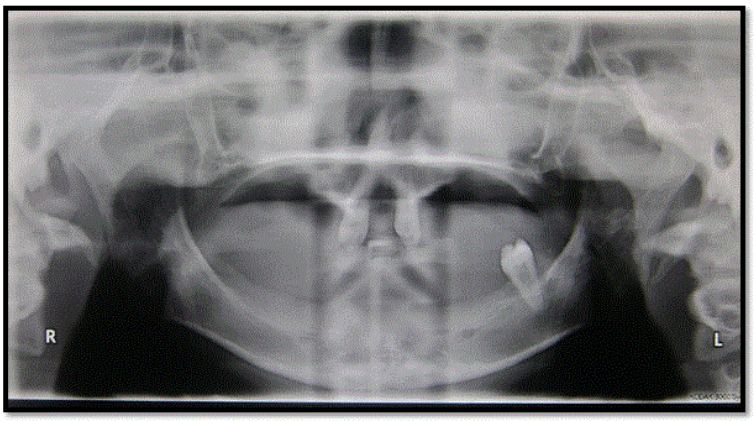

Figure 3: OPG of the patient.

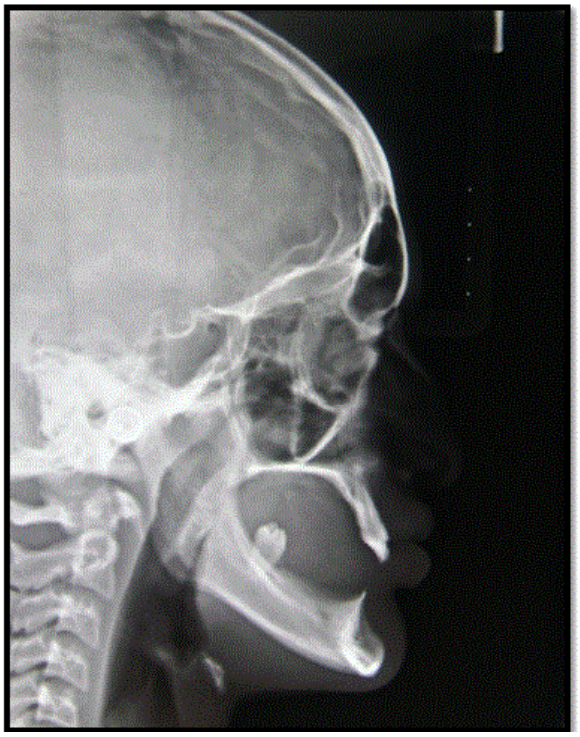

Figure 4: Lateral cephalogram of the patient.

DISCUSSION

Oral rehabilitation of ectodermal dysplasia is required to boost sagittal and vertical skeleton relationship throughout natural growth and development further on improve aesthetic, speech and masticatory efficiency. So, the treatment set up for a child full of HED should embrace a prosthodontics side and a psychological side [13]. But although removable complete/ partial dentures still stay the most keep of treatment in several cases, FPDs and implant-supported prostheses are to be thought-about once deemed viable. Implants appear to be significantly indicated in cases with complete anodontia. In youngsters wherever severe midface dysplasia is anticipated, early growth modification and implants is also useful. Though dentures are poor alternatives to healthy dentition, they produce conditions for maintenance of a normal, satisfactory daily diet, therefore serving to ascertain a long dietary pattern at associate early age. Also, within the absence of occlusal stops (or dentures), the associate rotation of the lower jaw causes an upward and forward displacement of the chin, with a reduction within the height of the lower-third of the face; a bent to C1 III disorder. Dentures facilitate positioning of the chin in situ [2]. Histopathology shows flattened epidermis, reduced number of sebaceous glands and hair follicles [14]. The management of children and adults with HED is a challenge because of their heat intolerance (especially during febrile illness or physical activities and in warm climate) and because of their susceptibility to pulmonary infections. During hot weather, affected individuals must have access to an adequate supply of water and a cool environment, which may mean "cooling vests," air conditioning, a wet t-shirt, and/or a spray bottle of water. However, external cooling is less effective in these patients because their heat transfer from the core to the skin is also reduced, presumably due to poor capillary dilatation [15]. Future treatment may involve gene correction administration. This patient is counsel regarding the condition and referred to dental and advised regarding the lifestyle changes including restriction of physical exertion, choice of occupation to ensure a disease free life. Dental prosthesis was offered and is advised regarding regular follow up and to be extra careful during febrile episodes. This presentation of case is done due to its infrequency and the need for the dermatologist to diagnose such cases at the earliest. It is to be stressed that the counselling of the patients empowers them to get adjusted to the society and helps them live a better life by simple lifestyle modifications. In above patient complete denture is advised or implant is advisable to improve aesthetic and psychological condition.

CONCLUSION

Hypohydrotic ectodermal dysplasia is a rare heterogeneous group of disorders and is non-progressive in nature. Therefore dentist must be properly aware of clinical symptoms diagnosis and then follow the treatment.

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