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Ectodermal Dysplasia- A Case Report

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KEYWORDS

Ectodermal Dysplasia, Sebaceous Glands, Partial Anodontia, X linked recessive hypohydrotic

ABSTRACT:

Ectodermal dysplasia is a hereditary genetic disorder that manifests various in one or more ectodermal derivatives such as skin ,nails, hair ,teeth ,exocrine glands and sebaceous glands ,the incidence of ectodermal dysplasia is rare 1 in 100000 birth, this case depicts the ectodermal dysplasia type 2 (dental dysplasia) revealing negligible hair or sebaceous glands with slightly chubby cheeks, saddle nose with absence of clubbing and partial anodontia in the maxillary and mandibular arches ,the most common type is X-linked recessive hypohydrotic ectodermal dysplasia and hydrotic ectodermal dysplasia ,the current case is x-linked recessive type since the father has history of delayed eruption ,saddle nose with sparse distribution of hair .

Introduction

Ectodermal dysplasia (ED) is rare hereditary disorder which is congenital(1) Patients with ectodermal dysplasia are characterized by hypoplasia or aplasia of structures such as skin, hair, nails, teeth, nerve cells, sweat glands, parts of the eye and ear and other organs. Ectodermal dysplasia might be inherited in any form of several genetic patterns including autosomal-dominant, autosomal-recessive, and X-linked modes.(2) 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(3) Facial appearance is typical which is characterized by frontal bossing, sunken cheeks, a saddle nose, and hyperpigmented skin around the eyes and low-set ears. Dental manifestations include conical or peg shape teeth, hypodontia or anodontia, and delayed eruption of permanent teeth. In hypohydrotic ED eccrine sweat glands may be absent or sparse and rudimentary.(1) The most common ED is hypohidrotic and hidrotic ED which is inherited as an X-linked recessive trait.(1)

Case Report

A 14 year old female patient visited to the OPD of Department of pediatric and preventive dentistry, St. Joseph Dental College, Eluru with the chief

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complaint of missing teeth in the upper and lower front teeth region since few years, pt father gave history of delayed eruption for him, no reported complications during the delivery, upon extra-oral examination her facial features are as follows with slightly chubby face and saddle nose with competent and thick lips, absence of clubbing for nails and even distribution of hair was observed, no history of decreased lacrimation or sweating. The institutional advisory committee/RAC has provided clearance letter for submission/publication of this manuscript vide letter number SJDC/RAC/008/2024 Dated 24/07/2024. On clinical examination partially edentulous teeth irt 12,15 ,23 ,24 ,31 ,41,42 with midline diastema and peg shaped lateral incisors are seen irt 22,32 ,On radiolographic examination retained deciduous teeth irt 54,55,53 are evident, Prelimnary treatment plan involves a multi -disciplinary approach of pedodntist, orthodontist, periodontist. Pediatric rehabilitation with extraction of retained deciduous teeth along with fabrication of removable partial dentures followed by orthodontic correction and once the dentofacial development is achieved following relining of dentures and correction of jaw discrepancies, jaw line and cephalometrics gives way for implants in oral health and wellbeing

Discussion

ED is a group of hereditary disorders characterized by developmental abnormalities of ectodermal derivative structure.[1] According to them ED is classified into different subgroups:

- ED1: Presence of hair anomalies or Trichodysplasia
- ED2: Dental anomalies
- ED3: Nail abnormalities or onychodysplasia
- ED4: Eccrine dysfunction or dyshidrosis

Extraoral features include fine, sparse hair over the scalp along with extensive scaling of the skin and unexplained pyrexia along with heat intolerance is seen, most commonly occurs due to anhidrosis. Normal intelligence is observed. Other extraoral features are frontal bossing, sunken cheeks, depressed nasal bridge, thick everted protuberant lips, wrinkled or hyperpigmented periorbital skin, and a large low set of ears [1]. In the present case reported here didn't have involvement of sweat glands and lacrimal glands no heat tolerance levels with partially edentulous teeth with respect to upper and lower arches. 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.[3] 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. In youngsters wherever severe midface dysplasia is anticipated, early growth modification and implants is also useful.[3] Assessing the childs esthetic functional ,skeletal and dentofacial development ,the treatment aspect is designed to be multidisciplinary with the team paediatric dentist, orthodontist, periodontist and dietician, pediatric dentist role is significant in this case initiation of diagnosis based on case history ,familial history ,orthopantamogram. Although dentures are poor alternatives to healthy dentition, they create conditions for maintenance of a normal, satisfactory daily diet, thus helping to establish a lifelong dietary pattern at an early age. Also, in the absence of occlusal stops (or dentures), the anterorotation of the mandible causes an upward and forward displacement of the chin, with a reduction in the height of the lower-third of the face; a tendency to C1 III malocclusion. Dentures help positioning of the chin in place.[4] In differential diagnosis of the diseases, it is difficult to differential recessive autosomal hypohydrotic ectodermal dysplasia from x linked hypohydrotic form. Clinical characteristics are similar in both, but because of different inheritance, the autosomal recessive involves both men and women, and heterozygous condition does not show any symptoms.[5] treatment plan was carried out with initiation of extraction of retained deciduous tooth irt 53 and removable prosthesis for upper and lower arches followed by relining and rebasing in order to restore the vertical dimension and any futuristic

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arch discrepancies followed by osseus regeneration of bony defects irt 21 and implants or overdenture once the child reaches 18 years of age

Figure 1: Patient's Profile

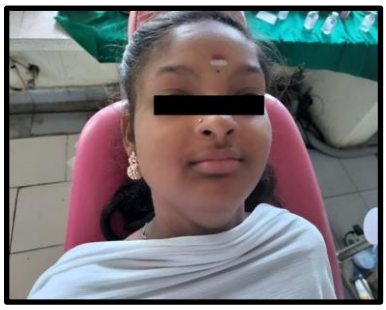

Figure 2: Extraoral Features

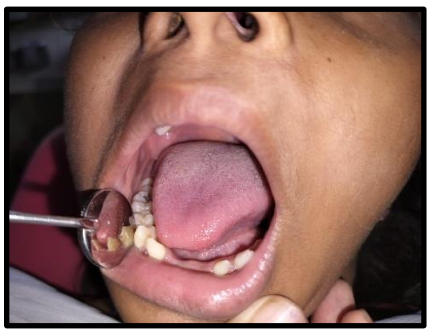

Figure 3: Intraoral Examination

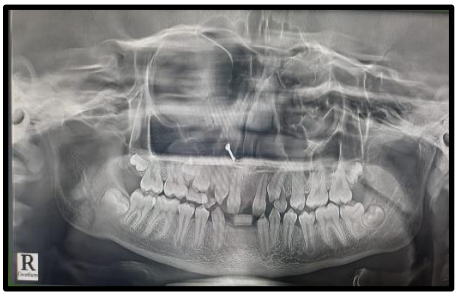

Figure 4: OPG

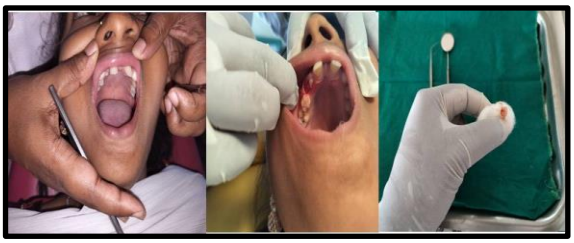

Figure 5: Extraction of 53

Conclusion

Ectodermal dysplasia is a rare inherited disease characterized by involvement of ectodermal structures like skin, nail and hair along with exocrine glands and lacrimal glands, careful assessment of Diagnosis and proper blue print of management and scope of ED needs further research and development of advances methods to treat in a comprehensive and time bound manner.

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