CASE REPORT

Ectodermal Dysplasia: Features and Dental Management

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Abstract
The current case report presents a rare clinical case of ectodermal dysplasia for a three years old Libyan child. The child's parents were concerned about the missing dentition for their child at his age, and his malnutrition as consequence of the inability to eat healthy food. Clinical and radiographic examinations were carried to confirm the diagnosis and to make the proper treatment plan. A prosthodontic treatment offered to the child and he kept under observation.

Key Words: Ectodermal dysplasia, Hypodontia

Introduction

Developmental anomalies of primary teeth might occur individually or associated with general defects in the developmental processes of other organs from the same embryological layer (1). These developmental anomalies can have a spectrum of clinical manifestations, ranging from anomalies in the size, shape and number of teeth to a more wide-spread disfigurement in the dentofacial region. A deficiency in the normal number of teeth is termed as hypodontia, and it is considered as the most common developmental defect among humans (2). Although the congenital absence of teeth (tooth agenesis) may occur solitarily as a result of local disruption of the ectomesenchyme, it could also be associated with different systemic manifestations systemic, which constitute a group of diseases or a syndrome. This case report presents a Libyan child with total clinical absence of his teeth and the treatment was provided to the child in the Pediatric Department.

Case Report

A three-year-old male child presented to the pediatric dentistry clinic accompanied by his parents who were concerned with their son's missing teeth and difficulty in chewing food which lead to less consumption of nutritional

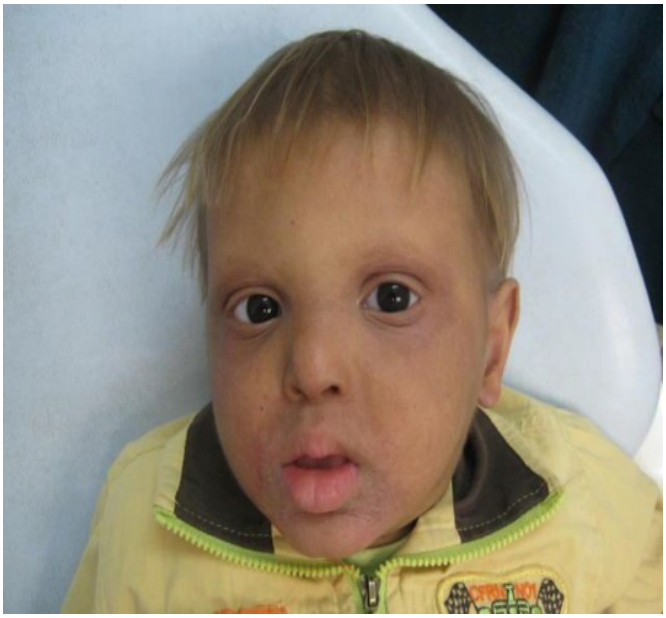

Figure 1. shows the 3-year-old child with the clinical features of ectodermal dysplasia i.e., scanty hair, saddle nose, absence of eyelashes.

needs with subsequent bad effect on general health. A pediatric specialist had already informed his parents that he had ectodermal dysplasia. The past medical and dental histories of the child were of no significance to be mentioned. As of the social history, it was non-contributory other than the child's parents were cousins, i.e., they had a consanguineous marriage. Additionally, the child's maternal uncle is also affected with the same condition. The child was not on any systemic medication. His family lives in a remote area from Benghazi, which made it difficult to communicate and come in time for treatment sessions. In the first visit, the extra-oral examination revealed that the scalp hair of the child was sparse and there were no eyelashes (Figure 1), saddle nose and other features suggestive of ectodermal dysplasia. Intra-orally, both of the upper and lower ridges of the mouth were edentulous (Figure 2). Radiographic examination showed the existence of only five teeth, which were not fully formed; only the crowns could be seen in their follicles (Figure 3). They appeared to be two canines (one in each side of the anterior area), and three molars in the posterior region (one in the right side and two in the left). Having conducted a thorough examination of the patient, it was explained to the father that a removable denture could not be provided at this age; rather, he should reach his fifth year in order to carry on with the prosthetic treatment. However, under the insistence of his father, the treatment was decided to commence at his child's fourth year of age. The child was brought again by his father when he was four-year-old. Having explained to the father the necessity of the replacement of both upper and lower dentures due to the continuous growth of the orofacial tissues of his child, a written consent was obtained and an appointment was fixed to start the prosthetic management. Both upper and lower impressions were taken and sent to the lab for constructing a primary cast upon which a special tray was fabricated. On the next visit, a final impression was taken of both upper and lower ridges, followed by pouring and preparing the master casts. Subsequently, the patient was given an appointment for the final denture (Figure 4). Upon trial in the patient's mouth, few corrections in the flanges of the denture were undertaken and finally the patient was satisfied with the shape and shade of the artificial teeth (Figure 5).

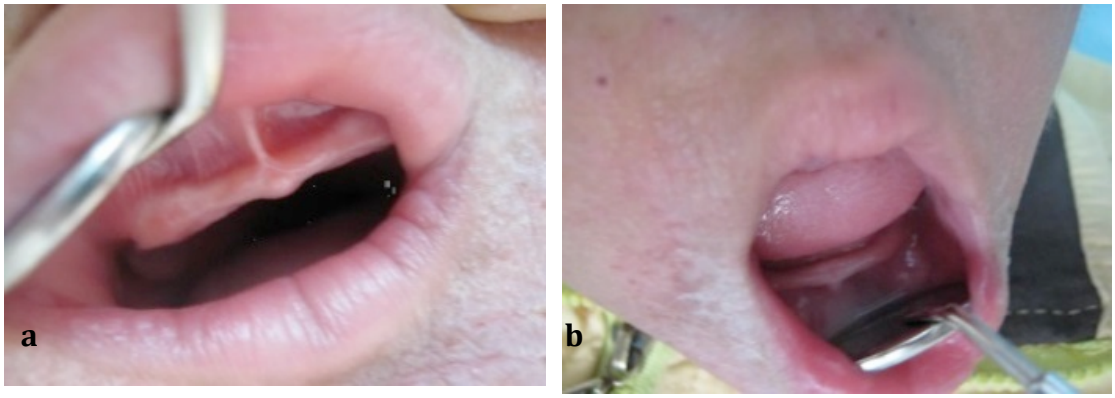

Figure 2. shows the edentulous upper (a) and lower (b) arches.

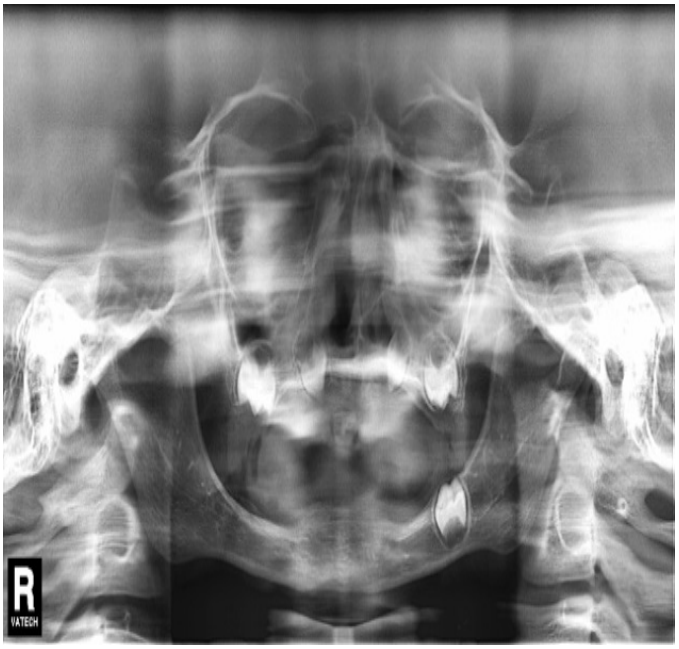

Figure 3. showing the crown follicles of the partially developed teeth (arrows) but failed to erupt.

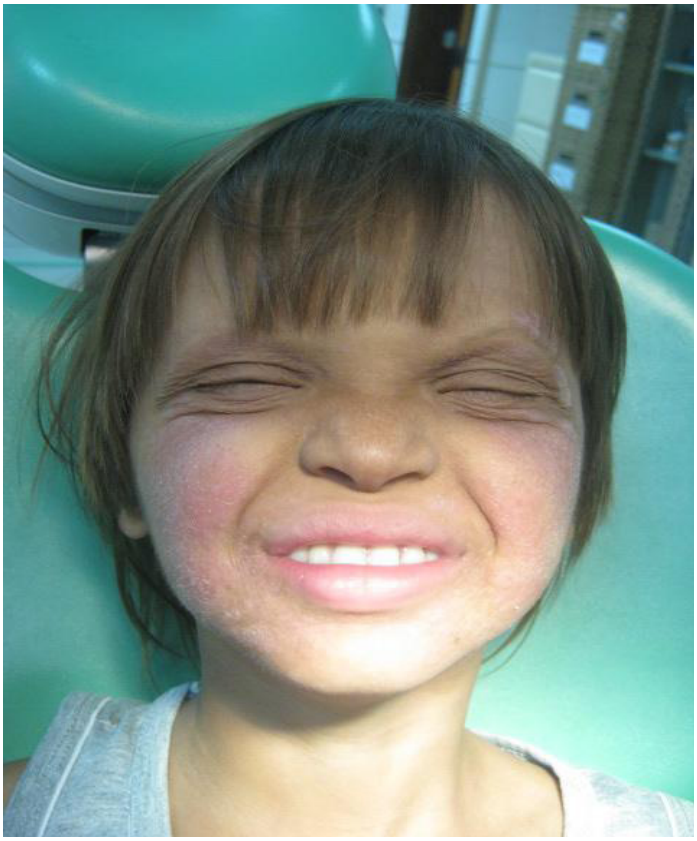

Figure 5. shows the child wearing the denture

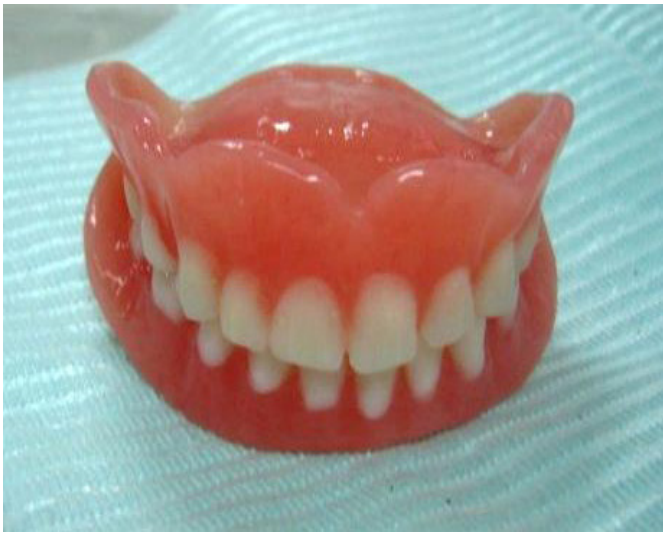

Figure 4. shows the delivered complete upper and lower denture

Discussion

Thurnam was the first who described ectodermal dysplasia as a group of disorders affecting two carrier patients as an x-linked recessive condition (3). Ectodermal dysplasia is a hereditary condition where there is a partial or complete absence of structures of the ectodermal layer, leading to hypoplasia of sweat glands, teeth and hair follicles. These hypoplastic defects will be manifested by hypohidrosis (and thus difficult in tolerating heat), hypodontia or anodontia,

and hypotrichosis. Intra-orally, the teeth that have erupted may show crown and root dysmorphism e.g. conical-shaped crowns, and also hyposalivation (4). The clinical type of ectodermal dysplasia encountered in our patient is also known as hypohidrotic ectodermal dysplasia and its most often subtype is mediated through x-linked mode of transmission, being either dominant or recessive in the way of inheritance (5). Recently, More et al. observed in a retrospective study of the effect of close-relatives marriage on the incidence of ectodermal dysplasia that two thirds of the affected child's parents had consanguineous marriage (6). Consanguineous marriages are fairly common in Libya and many areas in the Middle East, thus putting children at increased risks of ectodermal hypoplasia and other inherited conditions.

In the present case, the child was brought to the dental clinic because his parents were concerned about their child's deficient self-esteem in terms of psychosocial interaction with people due to his own appearance owing to loss of teeth. Interestingly, the child's uncle had the same condition, confirming the hereditary nature of this disorder. The removable denture provided to the patients critical for revitalizing the functional aspects i.e. masticatory and esthetic, of natural teeth, and that lead to improvement at the physiological and psychosocial levels. The parents were given the following points of advice for providing optimum care to their child: a) due to the absence of sweat glands, the child should be kept away from hot sunny weather, also use of hydration creams to the face and extremities several times a day, b) should be kept under soft diet c) should be followed by a dermatologist d) should be supervised to have his denture cleaned regularly and e) after 18 years of age the patient can receive an implant-supported complete denture. Further work is needed to find more cases among Libyan population to conduct relevant clinical investigations, research and provide appropriate genetic counseling.

Acknowledgement and Disclaimer: An informed consent was obtained from the patient's father to publish his son's condition as a case report in a scientific medical journal for educational purposes. We are grateful to the patient and his father for the permission. Reference to and use of this can only be made within this context.

References

1. Klein OD, Oberoi S, Huysseune A, Hovorakova M, Peterka M, Peterkova R. Developmental disorders of the dentition: an update. Am J Med Genet C Semin Med Genet 2013;163(4):318-32.
2. Chhabra N, Goswami M, Chhabra A. Genetic basis of dental agenesis - molecular genetics patterning clinical dentistry. Med Oral Patol Oral Cir Bucal 2014;19(2):e112-9.
3. Freire-Maia N, Pinheiro M. Ectodermal dysplasias - some recollections and a classification. Birth Defects Orig Artic Ser 1988;24(2):3-14.
4. Prager TM, Finke C, Miethke RR. Dental findings in patients with ectodermal dysplasia. J Orofac Orthop 2006;67(5):347-55.
5. van der Hout AH, Oudesluijs GG, Venema A, Verheij JB, Mol BG, Rump P, et al. Mutation screening of the Ectodysplasin - A receptor gene EDAR in hypohidrotic ectodermal dysplasia. Eur J Hum Genet 2008;16(6):673-9.
6. More CB1, Bhavsar K, Joshi J, Varma SN, Tailor M. Hereditary ectodermal dysplasia: A retrospective study. J Nat Sci Biol Med 2013;4(2):445-50.