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PARENTAL COUNSELING A PREREQUISITE FOR PEDIATRIC HYPOHYDROTIC ECTODERMAL DYSPLASIA - A CASE REPORT

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ABSTRACT: Ectodermal dysplasia's are inherited diseases that have developmental defects in at least two major structures derived from the ectoderm. The most characteristic findings are sparse scalp and body hair, reduced number of sweat glands, heat intolerance and brittle nails. Some infants and children have premature look referred to as an 'old man' facies. Characteristic dental findings include hypodontia, abnormal shaped teeth, delayed teething. Early dental treatment promotes self confidence improves individuals psychosocial condition. The parents must be educated and motivated about the importance of dental treatment in such patients. The present case is a report of 6 year old boy having defects in all the Ectodermal derivatives with significant dental findings wherein the patient's mother was reluctant to accept the proposed treatment plan. Hence we felt that parental motivation is a prerequisite in such cases.

KEYWORDS: Hypohydrotic Ectodermal Dysplasia (HED), oral rehabilitation, parent counseling.

INTRODUCTION

Ectodermal dysplasia (ED) affects males and is inherited through female as carriers. The incidence of ED in males is estimated at 1 in 1, 00,000 births, and carrier incidence in females is around 17.3 in 1, 00,000 births.[1] Genetic studies reveal that mutations in the ectodysplasin A and ectodysplasin A receptor genes are responsible for X-linked and autosomal HED. Oral rehabilitation is important as this would help in improving saggital and vertical skeletal relationship during craniofacial growth & development.[2] There is no literature stating the importance of parental counseling as a prerequisite for proposed treatment in HED patients. The aim of this case report is to describe the importance of parental counseling for HED patients.

Case report

A six year old boy reported with his mother to Aimst reception and primary care unit, with a chief complaint of unerupted teeth. History revealed that since his birth there are only few milk teeth erupted and erupted teeth appears sharp and pointed. History revealed that this was the child's first dental visit, child was getting chastened in the school and nick named as "old man" hence refused to go to school and this has led the parent to approach the dentist. Upon eliciting the Medical history it was revealed that child experienced frequent episodes of fever since his birth and has undergone multiple consultations, but no systemic abnormality has been detected. Child could not withstand hot climate and he failed to sweat. Family history revealed absence of consanguity and no similar dental manifestations been identified among the siblings. On extra oral examination patient had receded hair line, with prominent supraorbital ridges, depressed nasal bridge was noticed **(Fig.1)**.Her nails were brittle and are associated with frank bleeding.**(Fig.2.).** On intraoral examination there were multiple missing teeth in the upper and lower jaw and the teeth in the upper incisal region appeared conical in shape**(Fig.3).** orthopantomograph (OPG) revealed congenitally missing teeth in the clinically missing tooth region with few unerupted tooth buds**(Fig.4).**. Hence with the above history and clinical findings we arrived at a provisional diagnosis of Hypohydrotic Ectodermal Dysplasia. We explained the mother about the child's developmental condition and advised for partial denture fabrication with the existing teeth. The parent was worried that the child was too young to undergo the proposed dental treatment plan and psychologically unwilling. We have explained the consequences of long term edentulism and for better psychosocial development of the child early dental intervention would be the correct treatment option.

Discussion

ED syndromes have been described as a group of disorders of morphogenesis displaying two or more of the following signs and symptoms Trichondysplasia (abnormal

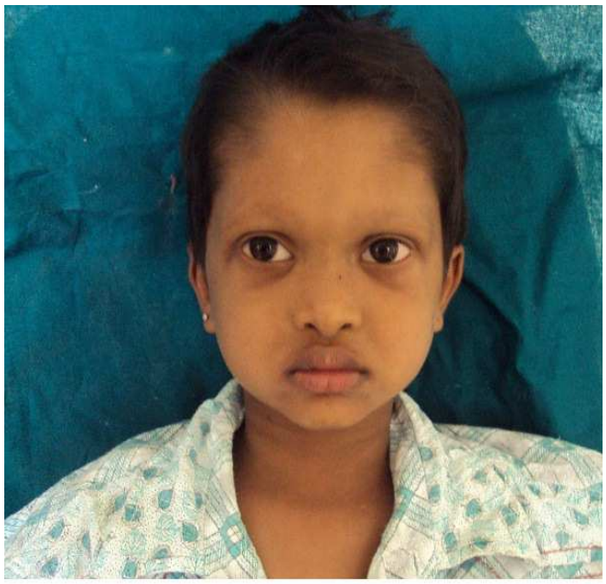

Fig.1. Extra Oral Photographs- receded hair line, with prominent supraorbital ridges, depressed nasal bridge

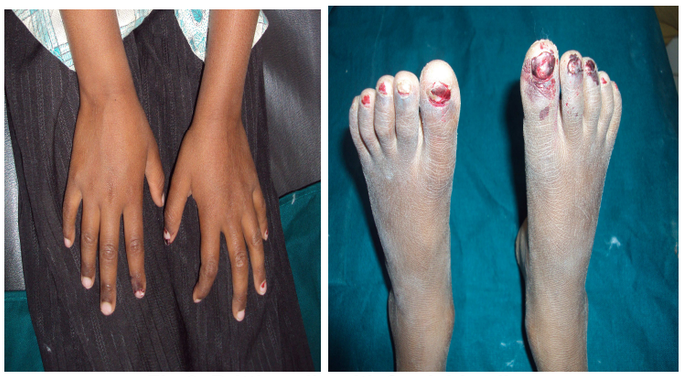

Fig.2. Brittle nails with frank bleeding

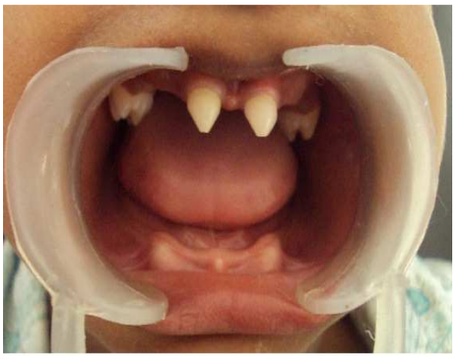

Fig.3. IntraOral photograph-Multiple missing teeth and conical shaped teeth

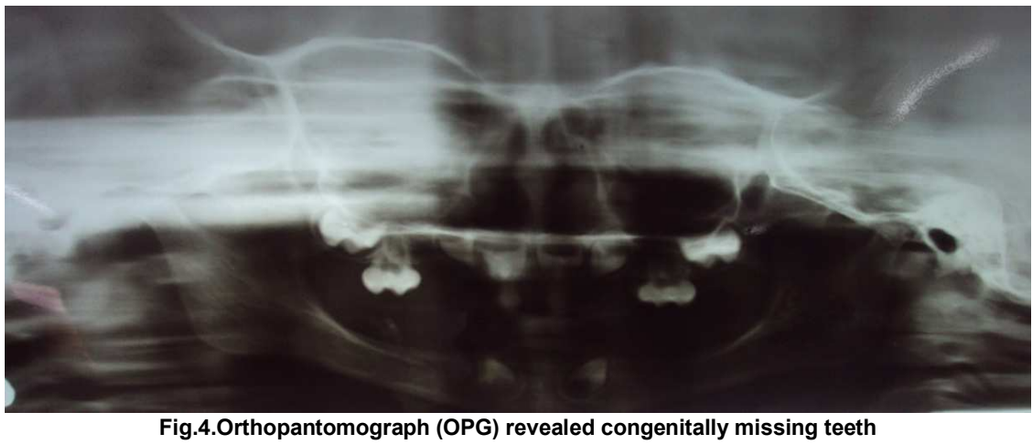

Fig.4.Orthopantomograph (OPG) revealed congenitally missing teeth

hair) , Abnormal dentition, Onchondysplasia (abnormal nails), Dyshidrosis (abnormal or missing sweat glands)[3] Freire-Maia , Pinkeiro [4] and Giansanti gave a classification of ectodermal dysplasia, and have reviewed extensively, the associated syndromes. In their studies, hereditary hypohidrotic ectodermal dysplasia was the most common.[5] Hypohydrotic form is characterized by either absent or significantly reduced sweat glands this in turn causes decreased sweating and patients may show heat intolerance, and experience frequent episodes of hyperthermia, sparse scalp and body hair (hypotrichosis). The hair is often light-colored, brittle, and slow-growing, facial features include a prominent forehead, thick lips, and a flattened bridge of the nose. Half the affected individuals exhibit mild fingernail abnormalities & nail dystrophy split nails with longitudinal ridging thinning and superficial peeling is also often noted[6,7]

The present case exhibited all the above mentioned extra oral finding hence was diagnosed as the HED. Oral findings are of particular interest, since patients with hyphohidrotic Ectodermal dysplasia manifest anodontia, or oligodontia, complete or partial absence of teeth, seen both in permanent and deciduous dentition. Even the tooth erupted are conical or peg shaped with delayed eruption. [8] The present case shared all the oral findings mentioned in the above literature. Diagnosis of HED is most commonly based on medical history, family history, intra oral clinical and radiographic examination .[9] The present case was diagnosed as Hypohydrotic Ectodermal dysplasia based on the above mentioned diagnostic criteria. However in the family history there was aabsence of consanguinity between the parents, and no similar case of HED identified among sibling suggest that the condition might be fresh mutation in the present case.

Differential diagnosis for Ectodermal dysplasia includes incontinentia pigmenti, ectrodactyly-ectodermal dysplasia clefting syndrome, Rapp-Hodgkin syndrome, and Ellis-vancreveld syndrome. Incontinentia pigmenti is a genodermatosis involving Ectodermal, Mesoderm, dermatome with dental and ocular features and some cases are associated with neurologic deficit. [10]

EEC is triad of ectrodactyly (development of anomalies of the structures derived from the embryonic ectodermal layer), ectrodactyly (extremities, hands and feet malformations), and cleft lip and/or palate. [11] Treatment should be initiated in children suffering from Ectodermal dysplasia by providing dentures as early as 2 years of age. [12]

ED is usually a difficult condition to treat with prosthodontic restorations because of the typical oral deficiencies and the young age when they are evaluated for treatment. Therefore, when treating a child with ED, it is important to motivate both the child as well as his parents prior to the treatment and to work with them to ensure their compliance. Oral rehabilitation is important this would help in improving saggital and vertical skeletal relationship during craniofacial growth & development thereby helps in improving the esthetics, speech and masticatory efficiency.[13] With regard to child cooperation denture can be fabricated as early as 3 to 4 years of age. This early dental intervention will in cooperate more self confidence improve psychosocial condition & provide positive outlook in the society. [14] The patient and the parents must be educated and motivated about the dental problems related to the genetic and psychological conditions. [15] In the present case patient is too young to be educated mother was educated about the existing condition but was psychologically unwilling to accept the fact about her child's dental problem however we have and also advised the mother that only under her cooperation it is possible to prepare the child to cooperate for the proposed treatment plan in near future.

CONCLUSION

Successful oral rehabilitation for young patients helps to overcome considerable social problems, helps to restore the normal function esthetics which is the main concern in these patients. This can be achieved only with parent's cooperation which can be accomplished only if the parents are educated and motivated about the dental problems and related psychological issues.

Hypohydrotic ED is a common variant. Clinical manifestations cause considerable social problems in individuals affected by this condition. Hence early diagnosis and oral rehabilitation will help in restoring the normal function esthetics which is the main concern in these patients. This can be achieved only when the parents are motivated regarding the consequences associated with early childhood edentulism.

References

1. Chugh A, Gupta N, Chugh VK. Ectodermal dysplasia with total anodontia: Case report with review.2016; 6(2)124-28.
2. Yenisey M, Guler A, Unal U. Orthodontic and prosthodontic treatment of ectodermal dysplasia-a case report. Br Dent J 2004; 196: 677-9.
3. El Tony et al. Hereditary Hypohidrotic Ectodermal Dysplasia with Anodontia: A Case Report. The Saudi Dental Journal. 1994; (6)1:31—34.
4. Freire-Maia N, Pinheiro M. Ectodermal dysplasia – some recollections and classification. Birth Defects 1988;24:3-14
5. Giansanti JS, Long SM, Rankin JL. The "tooth and nail" type of autosomal dominant ectodermal dysplasia. Oral Surg Oral Med Oral Pathol 1974; 37; 576.
6. RJ Patel, Rajesh S, GY Naveen, N Patil. Treatment considerations for a patient with ectodermal dysplasia: a case report. J. Int Oral Health, December 2010, volume 2 (issue 4)73-78.

7. Suri S, Carmichael RP, Thompson BD. Simultaneous functional and fixed applied therapy for growth modification and dental alignment prior to prosthetic habilitation in hypohidrotic ectodermal dysplasia: a clinical report. J Prosthet Dent 2004; 92: 428-33.
8. Jananee J, Satish kumar M, Sumathi Balaji. Ectodermal Dysplasia- A Case Report. Indian Journal of Multidisciplinary Dentistry. 2012; 2 (2): 465-67.
9. Patel A, Kshar A,Byakodi R, paranjpe A,Awale S. hypohydrotic Ectodermal dysplasia: A case report and review. International journal of advanced health sciences.2014; 1 (5):38-42.
10. X Li et al. Incontinentia Pigmenti: Case Report. Acta Dermatovenerol VENEROLOGICA CROATICA. 2013;21(3):193-197
11. Sharma D et al. Ectrodactyly, Ectodermal Dysplasia, Cleft Lip, and Palate (EEC Syndrome) with Tetralogy of Fallot: A Very Rare Combination. Frontiers in pediatrics.2015; 3(51):1-4.
12. Ohno K, Ohmori I. Anodontia with hypohidrotic ectodermal dysplasia in a young female: a case report. Pediatr Dent. 2000; 22:49-52.
13. Bani M,TM Ali, KNese, TTamer. Ectodermal dysplasia with anodontia: A report of two cases. European journal of dentistry.2010.4; 215-22.
14. Yavuz I., Baskan Z, Ulku R, Dulgergil T.C, Dari O, Ece A, Yavuz Ve Y, Dari O. “Ectodermal Dysplasia: Retrospective Study of 15 Cases,” Archives of Medical Research. 2006; 37:403-09.

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