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Ectodermal Dysplasia : A Case Report

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Abstract
Ectodermal Dysplasia Causes Deformities In One Or More Ectodermal Derivatives And Is A Hereditary Disease.The Ectodermal Dysplasia Shows Various Sign And Symptoms Like Scanty Hair,Dry Skin ,Dystrophic Finger Nails Etc. There Are Many Syndromes That Shows Oligodontia Or Anodontia And Ectodermal Dysplasia Is One Of Them. The Report Herein Is Of A 8yr Old Female Child Who Visited The Peditaric Department Of Dental Hospital Who Provided With Prosthetic Rehabilitation.

Key Words*: Ectodermal Dysplasia, Dystrophic Finger Nails, Oligodontia*

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I. INTRODUCTION

In 1929 weech coined ectodermal dysplasia and in 1848 thurman published a paper on ectodermal dysplasia.[1] .It is a group of inherited disorder characterized by hypodontia, hypotricosis and hypohidrosis etc.Depending on the presence of hair(1), nail(2), the tooth(3) or sweat gland(4) abnormality freire-maia subclassified the ectodermal dysplasia into various groups .It is broadely classified into hypohydrotic and anhidrotic type.[2]

Sarnat et al concluded that apart from defective alveolar growth, jaw and facial development is essentially normal.[3] The article present one such case in detail:

II. CASE DETAILS

A 8 yrs old female patient along with her parents reported in our department of pedodontics and preventive dentistry with the complaint of missing teeth and difficulty of mastication ,speech and have esthetic concern .She has history of mild hyperthermia but no history of any medical evaluation. There was no family history.On general physical examination the patient showed increase in body temperature and dry skin.On extraoral examination patient showed presence of sparse and scanty hairs ,everted and dry lips,sparse eyelashes and frontal bossing.Intraorally patient showed oligodontia with teeth missing 12,22,54,64,31,32,33,74,41,42,43,84.

Teeth appear in the upper arch conical shaped with spacing in upper anterior region and thin edentulous area in lower anterior region.

III. TREATMENT

Prosthodontic rehabilitation of the mandibular arch replacing the missing teeth done. After taking impression of both the arches followed by bite registration porcelain teeth were placed. After final trial removal partial denture for the mandibular arch was fabricated and delivered to the patient.

Fig1. Extraoral clinical picture

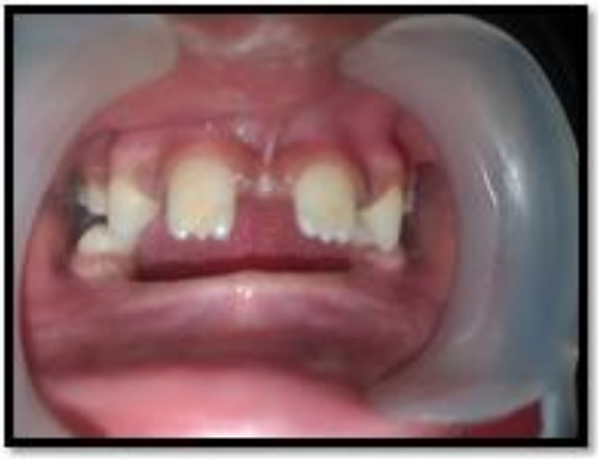

fig.2 intraoral picture showing missing 12,22,54,64,31,32,33,74,41,42,43,84

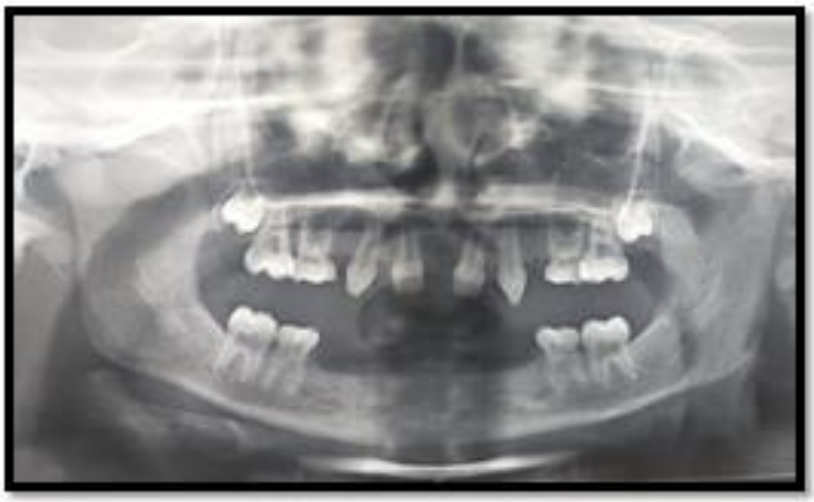

Fig 3.Panoramic view showing oligodontia

Fig 4. Prosthetic rehabilitation of mandibular arch done

IV. DISCUSSION

With a variable mode of inheritance ectodermal dysplasias are single-gene disorders. Mutations in cell involved in the induction and development of ectodermal structures are central to the clinical manifestations of ectodermal dysplasias. [2]The common variant is hypohydrotic ED, which is commonly X-linked . The prevalence of ectodermal dysplasia is approximately 7 per 10,000 births. Hypohidrotic Ectodermal Dysplasia shows constellation of hair and tooth anomalies along with an inability to sweat. The hair in scalp and eyebrows are sparse or absent with light-brown pigmentation. Affected infants clinically present with hyperthermia as early as the first few hours of life. This happens due to an inability to sweat , which leads to elevation of core body temperature. Teeth are usually reduced number and peg-shaped .[4]

Hydrotic Ectodermal Dysplasia is also known as Clouston syndrome. The condition mainly affects the hair and nails with sparing of teeth and eccrine glands.[5]

In Wiktop Tooth and Nail Syndrome both deciduous and permanent dentition is affected. The primary teeth may be normal or conical and showing prolonged retention. Permanent dentition may be partially or totally absent, especially the mandibular incisors, maxillary canines, and second molars. [6]

The treatment mainly consists of temperature maintenance by frequent drinking of cold liquids, wearing special cooling vests and caps.In case of teeth early dental management may lead to improved function and esthetic outcome. Bone grafting, dental implants, and dental prosthesis are recomended.[7]In our case the patient showed protruded lip ,scanty hair, dry skin and frontal bossing.The radiographic examination showed conical shaped ,spaced and missing teeth with elongation of the pulp chamber more common in molar teeth. Roots are also in conical shape.Depending on the clinical and radiographical examination we diagnosed our case as a case of ectodermal dysplasia. The patient was provided prosthetic rehabilitation and the follow up showed patient with improved speech,mastication and esthetic appearance.

V. CONCLUSION

An early detection of ectodermal dysplasias (EDs) have a good prognosis with a normal life span. Proper timely management of related dental and skeletal problems and maintainance of body temperature considerably improves these patient's quality of life.

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