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Ectodermal Dysplasia with Case Study & Current Scenario in India

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Abstract- Ectodermal dysplasia (ED) is not a single disorder but a group of genetic Syndromes all deriving from abnormalities of the Ectodermal structures More than 150 different syndromes have been identified. The triad of nail dystrophy, alopecia or hypotrichosis and palm plantar hyperkeratosis is usually accompanied by a lack of sweat glands and a partial or complete absence of primary and/or permanent dentition. One Case reports illustrating from state of Maharashtra in India suffering from ED. The treatment for such patient is quite difficult. So the patient treated with Dentistry at local level Doctor.

Index Terms- Ectodermal Dysplasia

History Review

According to Perabo-et -al ectodermal dysplasia may have been recorded as early as 1792 by Danz. In 1838, Wedderburn documented ectodermal dysplasia in a letter to Charles Darwin, describing a case of 10 Hindu male family members. Thurnam in 1848 reported 2 cases of hypohidrotic form. Similar cases were reported by Guilford and Hutchinson in 1883 and 1886 respectively. Weech, in 1929 introduced the term hereditary ectodermal dysplasia and suggested the term anhidrotic for those with inability to perspire. Felsher, in 1944, changed the adjective anhidrotic to hypohidrotic because the author agreed that person with hypohidrotic form are not truly devoid of sweat glands.

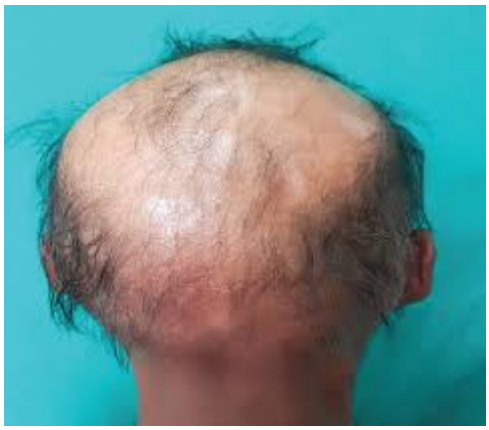
Introduction

Ectodermal dysplasia, as first described by Thurman,1,2 is a hereditary disorder occurring as a consequence of disturbances in the ectoderm of the developing embryo. The triad of nail dystrophy (onchodysplasia), alopecia or hypotrichosis (scanty, fine light hair on the scalp and eyebrows), and palm plantar hyperkeratosis is usually

accompanied by a lack of sweat glands (hypohidrosis) and a partial or complete absence of primary and/or permanent dentition.

Hair

Individuals affected by an ED syndrome frequently have abnormalities of the hair follicles. Scalp and body hair may be thin, sparse, and very light in color, even though beard growth in affected males may be normal. The hair may grow very slowly or sporadically and it may be Excessively fragile, curly, or even twisted. Kinky hair is also a possibility.



Nails

Fingernails and toenails may be thick, abnormally shaped, discolored, ridged, slow-growing, or brittle. The cuticles may be prone to infections.

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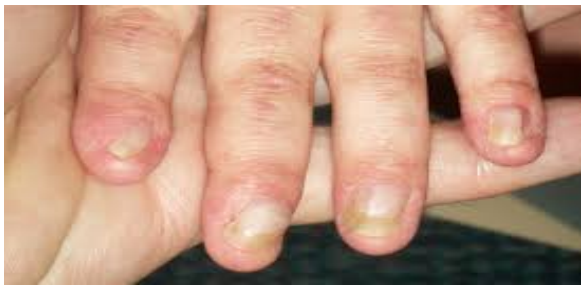
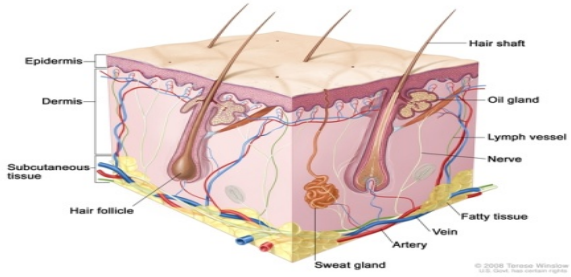


Skin

The skin may be lightly pigmented. Skin sustaining injury may grow back permanently hypo-pigmented. In some cases, red or brown pigmentation may be present. Skin can be prone to rashes or infections and can be thick over the palms and soles. Care must be taken to prevent cracking, bleeding, and infection.

Sweat glands

Individuals affected by certain ED syndromes cannot perspire. Their sweat glands may function abnormally or may not have developed at all because of inactive proteins in the sweat glands. Without normal sweat production, the body cannot regulate temperature properly. Therefore, overheating is a common problem, especially during hot weather. Access to cool environments is important.



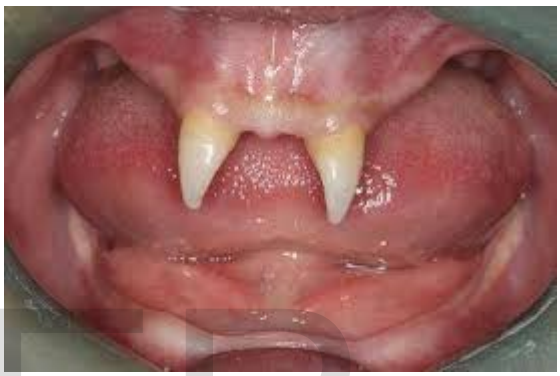
Salivary glands

Several studies have examined salivary flow rate in individuals and found parotid and submandibular salivary flow ranging from 5 to 15 times lower than average. This is consistent with the salivary glands being of ectodermal origin, although some findings have suggested that there is also mesodermal input

Teeth

The development of tooth buds frequently results in congenitally absent teeth (in many cases a lack of a permanent set) and/or in the growth of teeth that are peg-shaped or pointed. The enamel may also be defective. Cosmetic dental treatment is almost always necessary and children may need dentures as early as two years of age. Multiple denture replacements are often needed as the child grows, and dental implants may be an option in adolescence, once the jaw is fully grown. Nowadays the option of extracting the teeth and substituting them with dental implants is quite common. In other cases, teeth can be crowned. Orthodontic treatment also may be necessary.

Because dental treatment is complex, a multi-disciplinary approach is best.



Other features

People with ED often have certain cranial-facial features which can be distinctive: frontal bossing is common, longer or more pronounced chins are frequent, broader noses are also very common. In some types of ED, abnormal development of parts of the eye can result in dryness of the eye, cataracts, and vision defects. Professional eye care can help minimize the effects of ED on vision. Similarly, abnormalities in the development of the ear may cause hearing problems. Respiratory infections can be more common because the normal protective secretions of the mouth and nose are not present. Precautions must be taken to limit infection.

Classification of Ectodermal Dysplasia

Currently there are about 150 different types of ectodermal dysplasias. In an attempt to classify these, different subgroups are created according to the presence or absence of the

Four primary Ectodermal Dysplasia (ED) defects:

- ED1: Trichodysplasia (hair dysplasia)
- ED2: Dental dysplasia

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- ED3: Onychodysplasia (nail dysplasia)
- ED4: Dyshidrosis (sweat gland dysplasia)

Based on the above, the 150 different types of ectodermal dysplasias are categorized into one of the following subgroups made up from the primary ED defects:

- Subgroup 1-2-3-4
- Subgroup 1-2-3
- Subgroup 1-2-4
- Subgroup 1-2
- Subgroup 1-3
- Subgroup 1-4
- Subgroup 2-3-4
- Subgroup 2-3
- Subgroup 2-4
- Subgroup 3
- Subgroup 4

Case Report

A 30 Year Young Boy was referred at one Hospital for Dentistry treatment due to several missing teeth which called Hypodontia, and there was only 3 to 4 teeth had Peg liked shaped.
Parental history revealed that the patient was diagnosed with hypohidrosis ectodermal dysplasia after skin biopsy at J.J Hospital, Mumbai at Maharashtra state when he was a baby. (Near about 29 years ago)
During External examination of a patient the skin was soft, dry and without hair also there was absence of sweat Glands which called hypohidrosis. Sweating is a major way that the body controls its temperature; as sweat evaporates from the skin, it cools the body. Reduced sweating can lead to a dangerously high body temperature, Also loss of Hair and only few light coloured hairs on the head.

Discussion

In this case Dental Implant of the ectodermal dysplasia patient is necessary to improve the oral condition. In situations where implant therapy is indicated, the main problem is insufficient bone; if bone atrophy progresses in these already alveolar-deficient patients, implant placement may not be possible without bone grafting.
Also for lack of sweat glands, there was a hyperthermic condition in summer, for that there is not a proper and permanent treatment for hypohidrosis, so for that in summer season there will be use of a Air condition or Air cooler to control body heat. Some time the wet clothes use and wrapped on the body so it will helped out to maintain body temperature.

for hair loss, now a days there is new techniques using for hair that is hair transplantation, but due to thin hair this is not possible for the EC Patients, for that a hair wig can be use for less hair.

Current Scenario in India

In India Lots of patients are found who's suffered from Ectodermal Dysplasia disease. But the people are still unaware about this disease.
Patients of Ectodermal Dysplasia are rarely found in both Rural & Urban area of India, so the awareness is very low.
In most of the families the Ectodermal Dysplasic child was born and he looks like different at that time parents get confused & frightened.
The Indian health system divided many Levels like National Level; state Level, District Level, Sub District Level, and PHC Level. Also the different projects are running under Govt. of India like National Health Mission in which different Health Programs are to be running out.
So it will be needful to add a program for Ectodermal Dysplasia which helpful to know about the Disease to every people in Rural & Urban area.

Conclusion

The clinical manifestations of ectodermal dysplasia cause considerable social problems in individuals affected by the condition. In this case report, the patient suffered from Ectodermal Dysplasia associated with severe hypohidrosis & Hypodontia was described.
Also the awareness of the Ectodermal Dysplasia Disease is very less, for that its will important to add this Disease in our Health Programs so it will be helpful to know about the disease.

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