

Hypohidrotic Ectodermal Dysplasia : A Case Report

ABSTRACT

EDS- Ectodermal dysplasia syndrome is a large group of inherited disorders the manifestation of which can be seen in more than one ectodermal derivatives . these tissues primarily are skin , hair , nails , eccrine glands and teeth .

Hypohidrotic ED - Usually an inherited disorder due to X linked recessive trait.

Etiology - X linked hypohidrotic ED has been mapped in the proximal area of the long arm of band Xq-12-q13.1 . Decreased expression of epidermal growth factor receptor has been proposed as playing a cause role in this condition's phenotype.

Clinical features- Typical facies are characterized by frontal bossing; sunken cheeks ; saddle nose ; thick , everted lips ; wrinkled , hyperpigmented skin around the eyes ; ; and large , low set ears. include conical or pegged teeth , hypodontia or complete anodontia . Other common signs are short stature , eye abnormalities and photophobia.

Oral manifestations- Anodontia or oligodontia, with frequent malformation of any teeth present both deciduous or permanent dentitions . where some teeth are present they are commonly truncated or the cone shaped . Growth of the jaw is not impaired as the growth of jaws is independent of the presence of teeth .

PRESENT CASE

1. Partial anodontia , as some teeth were missing
2. Improper positioning of the teeth
3. Scarce hair or bald areas over the scalp
4. Eczema like skin with very less hair and dry as the sweat secretion was negligible



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5. The teeth present were not of the normal shape showing conical and pegged contours

Patient's history revealed that patient was not taking any drug or is not allergic to any drug .

Patient was sent to OPG and a radiograph was conducted and the radiograph of teeth showed that the erupted teeth had dysplastic enamel as the radiolucency was there in the areas of enamel over the dentine . jaw bone was found to be normal.

Histologic findings epidermis is thin , flattened , eccrine sweat glands are few or poorly developed.

Final diagnosis on the basis of symptoms and radiographic report , patient was diagnosed to be suffering from HYPOHYDROTIC ECTODERMAL DYSPLASIA or CHRIST-SIEMENS-TOURAINÉ SYNDROME

TREATMENT- There is no treatment for the condition . however the affected individuals with dental defects could be subjected to early dental evaluation and intervention beginning with dentures as early as two years.

