GENETICS AND GENODERMATOSES

HYPOHYDROTIC ECTODERMAL DYSPLASIA - CASE REPORT

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Background: The ectodermal dyplasias are a group of more than 150 hereditary disoders that affecet tissues and organs derived from the ectodermal germ layer. The most common variants of Ectodermal Dysplasia is the Hypohidrotic (HED) which causes dystrophy in the nails, alopecia and keratoderma in the hands and foot. Furthemore, HED can affect the teeth cusig hypodontia. This congetinal condition make the person seems older than his chronological age.

Observation: Male patient, six years old, born of natural child birth, without complications. By the age of two, his mother looked for medical assistance, beacuse her son presented warm and dry skin, he is not sweating, does have hair at all, as well. The mother also related episodes of bleedings by the nose, the ears and the mouth, when the kid was under high temperatures circunstances. In this scenario, received the Hypohydrotic Hectodermal Dysplasia diagnosis.
To the physical exam, we verify hypodontia, with just the presence of canine teeth; fragile and thin hair, with the thinning og hair mosto f the body; proeminente forehead; thin and wrinkled eyelids, with periorbital hyperpigmentation; proeminente lips; absence of sweat to the inspection; xerosis and normal nails. In addition, there is normal psycomotor development. The mother received orientations due to the needed cares with the patient's hair, who was directed to the dentist in order to prepare the dental prosthesis, besides him remaining under dermatological assistance.

Key Message: Hypohydrotic Ectodermal Dysplasia causes dystrophy in the skin's attachment