CLOUSTON'S DISEASE IN THREE SISTERS

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In a family of four children, all females, three sisters presented with Clouston's disease or hidrotic ectodermal dysplasia. The case is reported for the rarity of presentation in a single generation with no history of other family members affected.

Key words : Clouston's disease, Hidrotic ectodermal dysplasia.

Clouston's disease or hidrotic ectodermal dysplasia is a dominantly inherited entity common in individuals of French-Canadian descent.[1] It is characterised by nail dystrophy, defects of the hair and keratoderma of palms and soles. The sweat glands, sebaceous glands and teeth are normal. Some patients are mentally retarded[2] and some have associated neural deafness.[3] There is some evidence that the disease is caused by a defect in keratinization due to an abnormality in the matrix polypeptide.[4]

Case Report

Three sisters aged 10 years, 5 years and 3 years were seen with nail discoloration, sparse scalp hair and palmo-plantar keratoderma. Another sister aged 8 years was normal. There were no other siblings in the family. Both parents were normal and no other family member had similar complaints.

Examination revealed yellowish discoloration of the nails in the three girls. All of them showed sparse, fine, pale and brittle hair. Diffuse hyperkeratosis with fissuring were seen on their palms and soles. Sweating was normal. No abnormality was seen in the general physical development, teeth, bones and hearing. Investigations showed no aminoaciduria.

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Comments

Clouston's disease in our three cases has probably presented as a forme fruste because true nail dystrophy was not seen. The sparing of one of the four children may be explained by variable penetrance of the causative dominant gene. The case is interesting for its occurrence in three out of four sisters, with normal parents. To the knowledge of the authors, this has not been reported so far.

References

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