CASE REPORTS

DYSCHROMATOSIS UNIVERSALIS HEREDITARIA

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A 50-year-old man had dyschromatosis universalis hereditaria manifesting as asymptomatic macular pigmentation and depigmentation present all over the body almost since birth. There was no atrophy or telangiectasia. In addition, the patient had associated features of solar elastotic syndrome.

Key words : Dyschromatosis universalis hereditaria, Solar elastotic syndrome.

Dyschromatosis universalis hereditaria is a rare disorder of pigmentation described mostly among the Japanese and less often in other races. It is characterised clinically by asymptomatic pigmented macules of various sizes and shapes arranged irregularly and admixed with depigmented macules. The disease manifests at birth or during the first few months of life and affects both sexes equally. The lesions are distributed all over the body with the exception of the palms and soles and the face is involved less severely than other areas.[1] The lesions remain stationary and there is no specific treatment for the disorder. We are reporting a case seen by us recently.

Case Report

A 50-year-old man had asymptomatic pigmentary lesions of the skin present almost right from birth. The patient was born to consanguinous parents and gave history of similar skin lesions in his paternal uncle and one of his daughters. The hyperpigmented macules were 2-3 mm in size, varied in shape and were distributed all over the body except the palms and soles. Face was relatively spared. In addition, depigmented macules were also present in the midst of the hyperpigmented lesions (Figs. 1, 2 and 3). There was no evidence of

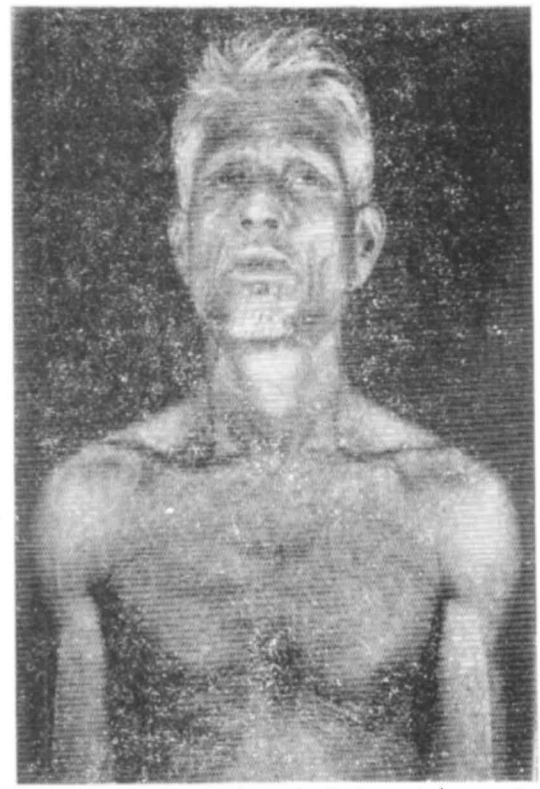

Fig. 1. Hyperpigmented and depigmented macules diffusely spread over the trunk with relative sparing of the face.

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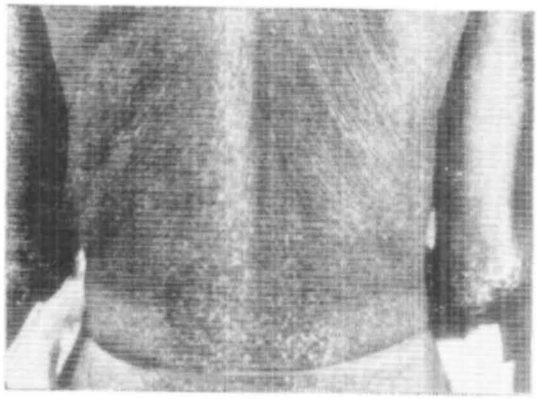


Fig. 2. Lesions on the back.

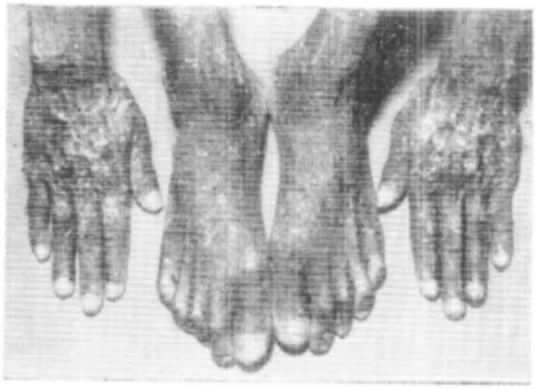
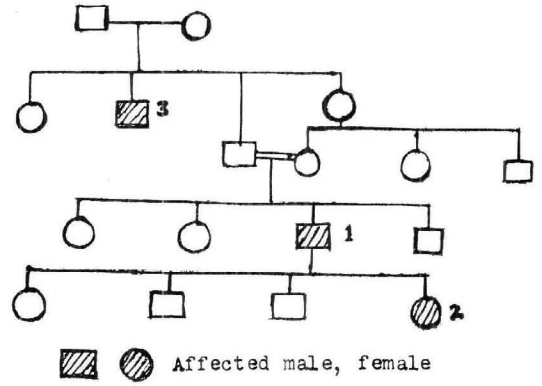


Fig. 3. Lesions on the hands and feet.

atrophy or telangiectasia of the affected skin. Elastotic nodules of the ears, rhomboidalis nuchae and acrokeratoelastoidosis were present representing the syndrome of solar elastoidosis. Ophthalmological examination revealed bilateral glaucoma and immature cataract of the left eye. Rest of the dermatological and systemic examinations were normal. Routine blood and urine analysis were within normal limits. Biopsy of the affected skin showed a non-specific histopathology with varying degrees of pigmentation of the epidermis.

Comments

The exact cause of this disorder is not known but it is reported to be inherited autosomally in more than one way.[2] In the present instance, as shown in the family pedigree, (Fig. 4) the



1 Patient 2 Daughter 3 Paternal uncle

Fig. 4. Family Pedigree of the patient

patient's parents were of consanguinous marriage and one of his daughters was also affected implying thereby that both autosomal recessive and autosomal dominant modes of inheritance could be operative in the transmission of this disorder. However, sporadic cases have also been seen.[3] Most reported cases are females[4] but males have also been affected[3] just as in the present report. The disease once thought to be confined to the Japanese is being reported with increasing frequency in other races as well. Findlay and Whiting[2] have described the disorder in two Bantu females and Rycroft et al[5] in an Iraqi girl.

Case reports of this condition in association with other abnormalities are few. Rycroft et al[5] have reported the disorder in a short-statured girl with high-tone deafness. Our patient had features of solar elastotic syndrome and glaucoma of both eyes with immature cataract of the left eye which may be co-incidental.

A localised variant of this disorder is also known and is called dyschromatosis symmetrica hereditaria. It is inherited as an autosomal dominant condition,[6] is believed to be relatively common among the Japanese[7] and characterised

by pigmented and depigmented macules forming a reticulate pattern affecting the dorsa of hands and feet symmetrically. No associated abnormalities have been reported with this condition.[4]

The disorder of dyschromatosis universalis hereditaria closely resembles xeroderma pigmentosum clinically but is easily differentiated from it by the lack of atrophy, lack of telangiectasia, the stationary course of the disease and the benign prognosis.[1]

References

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