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DYSCHROMATOSIS UNIVERSALIS

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Hyperpigmented macules all over the body intermingled with mottled depigmentation were observed in two brothers from Varanasi. Hyperpigmented macules were present since birth and increased in number and size during childhood. Depigmentation developed at the age of eight years and increased during adolescence. This entity described as dyschromatosis universalis in the Japanese and the Europeans has not been described in the Indians so far.

Key words : Dyschromatosis universalis, Hyperpigmentation, Depigmentation.

Dyschromatosis, universalis, characterized by generalized hyperpigmented macules intermingled with mottled depigmentation has been reported only in the Japanese,[1-3] and the Europeans.[4] It is a genetic disorder transmitted as an autosomal dominant trait.

Case Report

A 19-year-old male student presented with hyperpigmented macules all over the body intermingled with mottled depigmentation on bony prominences, neck and upper trunk for the preceding eleven years. The black dots on the face and a large cafe-au-lait spot were present at birth. Hyperpigmented macules increased in number and size in early childhood and involved other areas too. At the age of eight years, the patient noticed many small depigmented macules on the extensor aspects of limbs and bony prominences. Depigmentation increased in size and shape upto adolescence and intermingled with pigmented macules. There was no history of any symptom preceding it.

Physical examination revealed a large number of small (1-5 mm) hyperpigmented macules varying in size, shape and in depth of colour, all over the body, but mainly concentrated over the face, neck, upper trunk, buttocks and upper limbs without any tendency for grouping. The hyperpigmented macules were also present on the lower limbs, genitalia, axillae, groins, palms, soles and scalp though less in number. There was a large cafe-au-lait spot on the middle of right forearm with a serrated margin. Mucous membranes were free from such lesions. There was depigmented mottling with hyperpigmented macules, more conspicuous over the neck, upper trunk, upper arms, dorsa of hands and

Fig. 1. General distribution of lesions.

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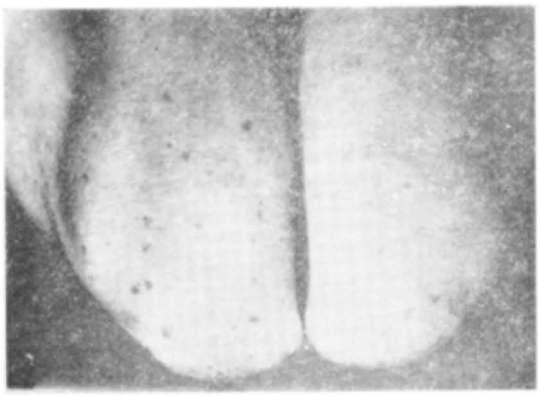

Fig. 2. Depigmented mottling on the elbows.

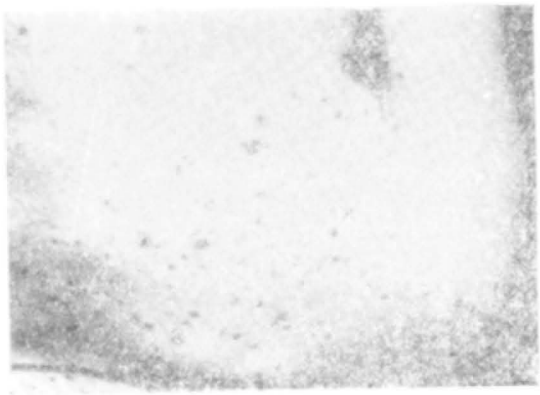

Fig. 3. Mottled depigmentation with hyperpigmented macules on the chest.

feet, bony prominences such as shoulders, elbows, knuckles, knees and malleoli etc. Nails, hair, palms and soles were normal. The vision, hearing, cardio-vascular system, central nervous system, chest, abdomen and uro-genital system were also normal.

Family history revealed that the patient had two brothers and two sisters. The elder brother had similar type of hyperpigmented and depigmented macules since his childhood. Parents and other family members were not affected. There was no history of consanguinity among parents.

The histopathological findings from hyperpigmented and depigmented macules were non-contributory.

Comments

Dyschromatosis has been reported in the Japanese[1-3] and the Europeans,[4] but not in the Indians. We are herewith presenting two Indian cases. It is transmitted as an autosomal dominant gene,[4] in our cases, however, only two brothers of one family were affected. Age of onset of hyperpigmented macules is reported to be the first two years of life.[1] In our cases, however, hyperpigmented macules were present at birth or appeared shortly after that and gradually increased in number and size during early childhood followed by the appearance of mottled depigmented macules at the age of eight years increasing upto adolescence. Hyperpigmented macules were present more or less all over the body except mucosa. Sparing of the face has, however, been described by Rook et al.[1] In our cases, mottled depigmentation was heavily concentrated on the neck, upper trunk, upper arms, dorsa of hands and feet and bony prominences, while Rook et al[1] have described it to be more conspicuous on abdomen. Involvement of acral parts has been reported by other workers.[3,4]

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