Case Report

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Dyschromatosis universalis hereditaria: A rare case report

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ABSTRACT

Dyschromatosis universalis hereditaria is an autosomal dominant disorder but may be recessive or sporadically inherited disorder, infrequently occurring genodermatosis with peculiar pigmentary changes, consisting of varying sized, intermingled hyperpigmented and hypopigmented macules that give an overall impression of mottling. Herein, we report this extremely rare case of dyschromatosis universalis hereditaria in a young male with a family history of the same disorder in his younger brother.

KEYWORDS: Dyschromatosis universalis hereditaria, genodermatosis, dyschromatosis symmetrica hereditaria

Introduction

Dyschromatoses are a group of disorders characterized by the presence of both hyperpigmented and hypopigmented macules, many of which are small in size and irregular in shape. These are a spectrum of diseases, which includes dyschromatosis universalis hereditaria (DUH), dyschromatosis symmetrica hereditaria (DSH), acropigmentation of Dohi, Dowling-Degos disease, and a segmental form called unilateral dermatomal pigmentary dermatosis (UDPD). Dyschromatosis symmetrica hereditaria (DSH) was first reported as a clinical entity by Toyama in 1929.[1]

Case Report

A 22-year-old unmarried male presented to us with chief complaint of multiple hyperpigmented and hypopigmented macules over arms, legs, and trunk for the past 15 years. The lesions had started over both the forearms and legs and gradually spread upwards towards the thighs and arms respectively, and then the trunk over a period of 5 years [Figures 1-2]. There was no history of any photosensitivity. There was no history of handling any chemical directly or of any significant history of drug intake. On taking family history his younger brother had similar lesions in distribution as his elder brother. There was no history of consanguinity among the parents.

On dermatological examination multiple hyperpigmented and hypopigmented macules were present in a reticulate pattern on the trunk, arms, forearms, thighs, and legs. His face, palms, soles, and mucous membranes were spared. Systemic examination did not reveal any abnormality. Routine laboratory investigations, including blood count, urine analysis, liver function tests, renal function tests, electrolytes were within normal limits. HIV screening for both HIV-1&2 by CombiAIDS was negative. A skin biopsy was taken from both the hyperpigmented and hypopigmented lesions. The basal layer in the hyperpigmented lesions showed an increase in pigment

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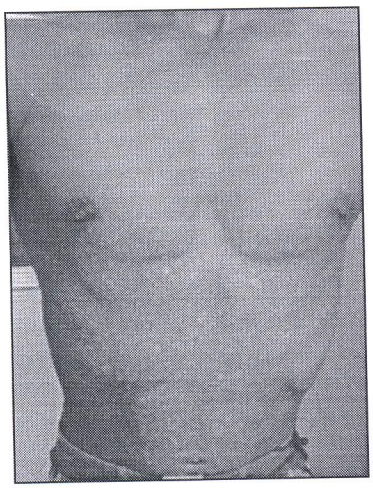

Figure 1: Lesions over chest and abdomen

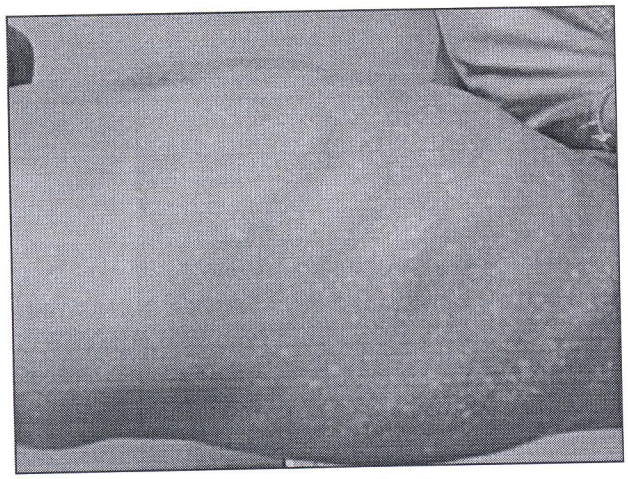

Figure 2: Lesions over lateral aspect of trunk

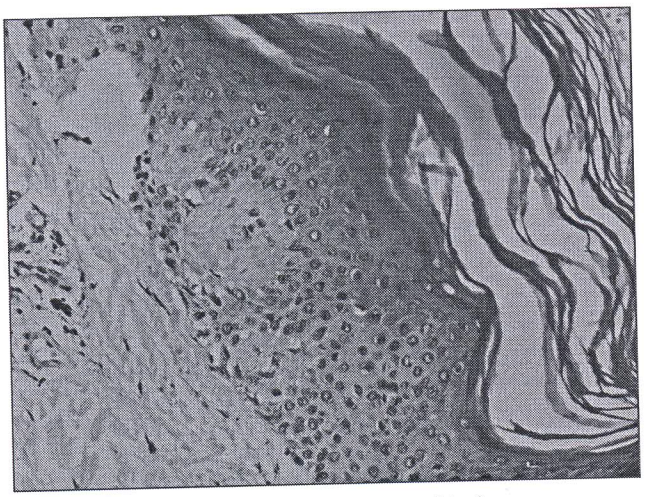

Figure 3: Histopathology of hyperpigmented lesion

along with melanin incontinence, [Figure 3] whereas in the hypopigmented lesions there was a decrease in pigment. The dermis in both the biopsies showed mild perivascular lympho-mononuclear infiltrate.

Discussion

DUH is an uncommon disorder characterised by hypo and hyperpigmented macules in reticulate pattern involving trunk and extremities but rarely the face, mucosa and palmo-plantar surfaces.[2] DUH was first described by Ichikawa and Hiraga in 1933.[3]

Previously, it has been suggested that DUH is a disorder of melanocyte number. Based on a recent electron microscopic study it has been suggested that DUH may be a disorder of melanosome production in epidermal melanin units rather than a disorder of melanocyte number.[4] Although, the precise etiology of this disorder is not yet known, the DUH locus has recently been mapped to chromosome 6q24.2-q25.2[5] and in chromosome 12 (12q21-q23).

However, DSH has been associated with different mutations in the gene ADAR1; this mutation is not present in patients with DUH and it is alternatively considered as two different entities.[6] In our patient, history of similar lesions in his younger brother was noted. No seasonal change or spontaneous regression with age was noted. Systemic abnormalities reported in isolated cases include short stature, deafness, erythrocyte, platelet and tryptophan metabolism abnormalities, cataract, and grandmal seizures[3] were conspicuous by their absence.

Lesions of dyschromatosis symmetrica hereditaria have to be differentiated from xeroderma pigmentosum because in both the disorders patients clinically show lesions in the photoexposed areas. However, in our patient, lesions were present in the unexposed sites as well. Moreover, the lesions did not show any atrophy or telangiectasia as seen in xeroderma pigmentosum. Other diseases in the differential diagnosis include dyskeratosis congenita, vitiligo, dyschromic amyloidosis, residual leukoderma, and exposure to chemicals such as diphenylcyclopropenone and monobenzyl ether of hydroquinone.[1,4] Yet, no such history of contact was present in our patient.

Conclusion

No treatment modality is available. Only genetic counselling is advised because of the recent reports of genetic etiology. This case of DUH is reported because of its rarity. Despite its rarity it assumes significance as it must be distinguished from xeroderma pigmentosum and other dyschromias.

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