

Dyschromatosis universalis hereditaria with involvement of palms

Kikkeri Narayanshetty Naveen, U. S. Dinesh¹

Department of Dermatology,
¹Department of Pathology, SDM College of Medical Sciences and Hospital, Sattur, Dharwad, Karnataka, India

ABSTRACT

Dyschromatosis universalis hereditaria is a rare genodermatosis characterized by hyper- and hypopigmented macules in a reticulate pattern. Here, we present a case of DUH with involvement of the palms.

Key words: Dyschromatosis universalis hereditaria, palms, reticular

INTRODUCTION

Dyschromatosis universalis hereditaria (DUH) is a rare genodermatosis characterized by hyper- and hypopigmented macules forming a reticulate pattern. Pigmentation appears in a generalized distribution. Although initial and subsequent reports are from Japan, later it has been reported from other countries.^[1] Both autosomal dominant and autosomal recessive patterns of inheritance have been documented.^[2] We herein report a case of autosomal dominant inherited DUH with involvement of the palms.

CASE REPORT

A 38-year-old married male presented to the outpatient department with generalized, asymptomatic symmetrical and spotty hyper- and hypopigmented macules over the body. The lesions appeared in infancy and progressed with age. Lesions started over the extremities and then gradually spread to the trunk over a period of six years. The face was spared. There was no history of photosensitivity or photophobia. There was no history of preceding dermatosis or associated systemic involvement.

He was born to healthy non-consanguineous parents following an uneventful pregnancy. There was history of similar lesions in his brother, sister, and paternal grandmother's sister. The patient did not have any children.

Cutaneous examination revealed multiple discrete circular and irregular-shaped mottled

hyperpigmented and hypopigmented macules measuring 2-3 mm in size distributed symmetrically over the trunk and upper limbs [Figures 1 and 2]. There was a symmetric, diffuse hypopigmentation with spotty hyperpigmented macules on the lower limb sparing knees and dorsum of the foot [Figure 3]. Symmetric hypopigmented patch with spotty hyperpigmentary macules were seen on the palms [Figure 4]. Soles were not affected. There was no atrophy or telangiectasia of the affected skin. The face was spared. Mucous membrane, hair, and nails were uninvolved. Systemic examination did not reveal any abnormality.

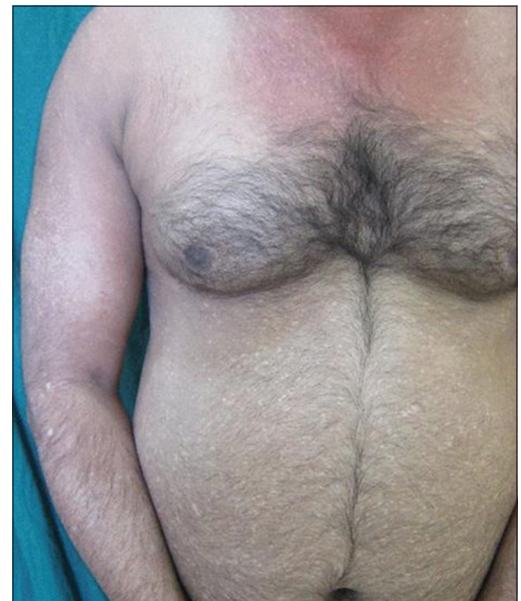


Figure 1: Multiple discrete, circular, and irregular-shaped mottled hyperpigmented and hypopigmented macules distributed symmetrically on the trunk and upper limbs

Access this article online

Website: www.idoj.in

DOI: 10.4103/2229-5178.137781

Quick Response Code:



Address for correspondence:

Dr. Naveen KN,
 No 10 Skin Opd,
 Sdmcms&h, Sattur,
 Dharwad - 580 009,
 India.

E-mail:
naveenkn80@yahoo.com



Figure 2: Hyper- and hypopigmented macules arranged in reticular pattern on the shoulder



Figure 4: Symmetric hypopigmented patch with spotty hyperpigmentary macules seen on the palms

Laboratory examination including complete hemogram, renal and liver function tests, blood HIV, and HbsAg were nonreactive or within normal limits. Ultrasonogram of the abdomen showed no detectable abnormality. Based on the clinical features, a diagnosis of DUH was made.

Biopsy was taken from both the hyperpigmented and hypopigmented lesions. The section studied showed keratinized thinned out epidermis. The dermis showed a few pilosebaceous units surrounded by sparse lymphocytic infiltrate. There was no evidence of increased epidermal pigmentation or pigment incontinence [Figures 5-7].

DISCUSSION

Dyschromatoses are a group of disorders characterized by



Figure 3: Symmetric, diffuse hypopigmentation with spotty hyperpigmented macules on the lower limb sparing knees and dorsum of the foot

the presence of both hyperpigmented and hypopigmented macules. It is a spectrum of diseases, which includes DUH, dyschromatosis symmetrica hereditaria (DSH) or acropigmentation of Dohi, and a segmental form called unilateral dermatomal pigmentary dermatosis. In 1929, Toyama described DSH as a distinct entity, later in 1933, Ichikawa and Hiraga described DUH, which had similar features of DSH but occurring in generalized as opposed to acral distribution.^[2] DSH has been considered as localized form of DUH.^[3]

The etiology of this disorder is not known. A novel mutation in RNA-specific adenosine deaminase gene (ADAR1, DSRAD) was reported in DSH, without identifying such mutation in DUH.^[4] Recently, ABCB6 has been identified as the first pathogenic gene associated with DUH.^[5] It has been suggested in the past that DUH is a disorder of melanocyte numbers. In a recent ultrastructural skin investigation, Nuber *et al.*

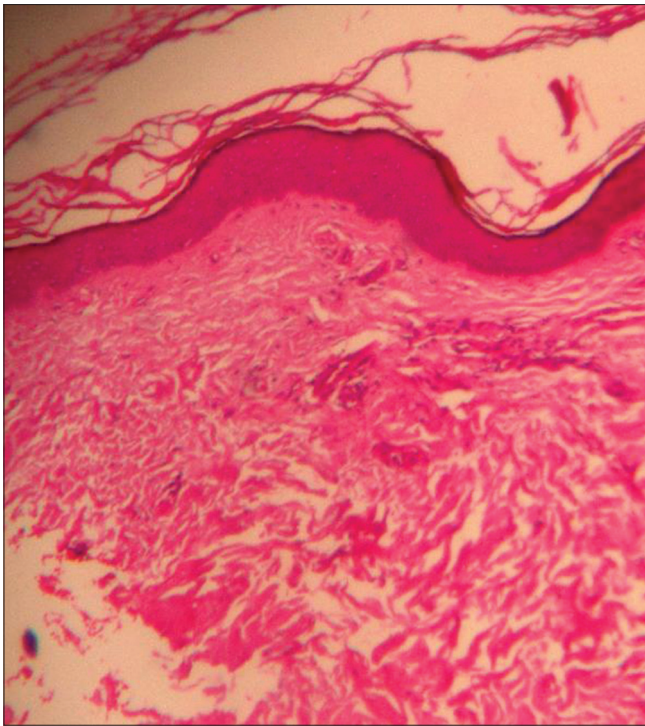


Figure 5: Histopathology showing no evidence of increased epidermal pigmentation or pigmentary incontinence (H and E, ×10)

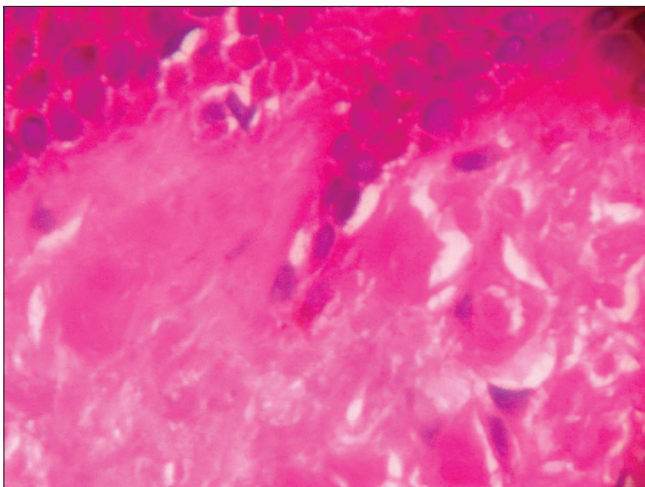


Figure 7: Closer view not revealing any pigmentary incontinence (H and E, ×100)

indicated that DUH is a disorder of melanosome synthesis rate or melanocyte activity, and not a disorder of melanocyte numbers.^[4,6]

The disease manifests as generalized hypo- and hyperpigmented macules in first few years of life. The trunk and extremities are the dominant sites. The lesions spread to the face, hands, and feet. Hair, teeth, and nails may be involved. It usually spares palms and soles.^[3,7] In the present case, there were hypopigmented patches over the palms with spotty hyperpigmented macules in it. Pigmentary variation on the

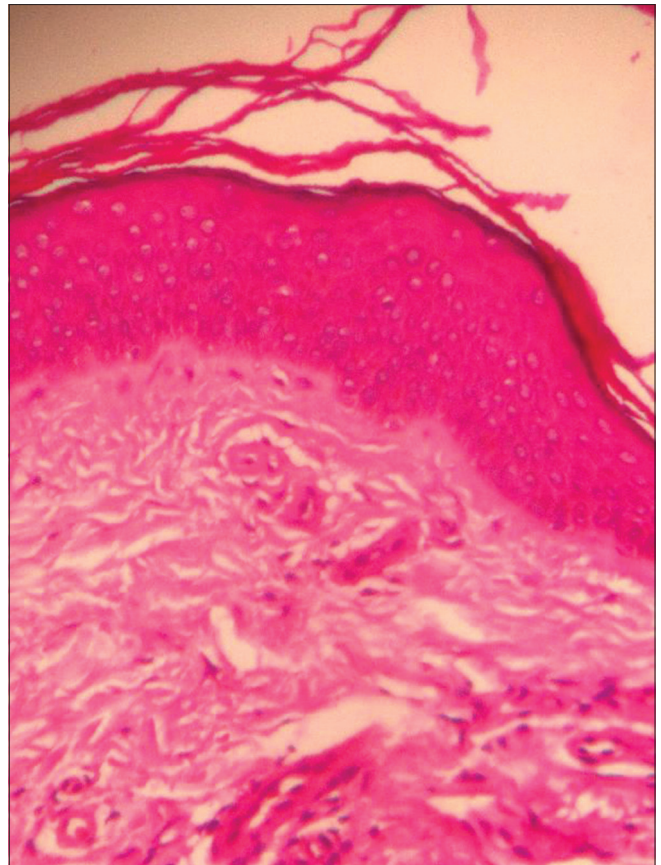


Figure 6: Histopathology showing no evidence of increased epidermal pigmentation or pigmentary incontinence (H and E, ×40)

palms and soles has been reported previously. Gharpuray *et al.*^[8] reported diffuse hypopigmentation of the palms, whereas Sethuraman *et al.*^[1] reported diffuse hyperpigmentation with spotty depigmented macules over the palms and soles. Kenani *et al.* described two unrelated Tunisian cases of DUH in a 3-year-old girl and a 3-month-old girl. Palms and soles were affected in the first child.^[4]

The histopathology shows a focal increase or decrease in melanin content of the basal layer (depending on the type of the lesion biopsied) and occasionally pigmentary incontinence.^[4] In our patient, there was no evidence of increased epidermal pigmentation or pigmentary incontinence.

In conclusion, we present here a rare case of DUH with the involvement of palms, which is sparingly reported in the literature.

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Cite this article as: Naveen KN, Dinesh US. Dyschromatosis universalis hereditaria with involvement of palms. *Indian Dermatol Online J* 2014;5:296-9.
Source of Support: Nil, **Conflict of Interest:** None declared.