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Authors

Sardar, Swapan Kumar
Das, Anupam
Bandyopadhyay, Debabrata Bandyopadhyay

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Case presentation

Dyschromatosis universalis hereditaria: report of six cases from a family

Swapan Kumar Sardar¹, Anupam Das², Debabrata Bandyopadhyay³

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¹ **Department of Dermatology, North Bengal Medical College, Sushurtanagar, Darjeeling, India**

² **Department of Dermatology, KPC Medical College and Hospital, Kolkata, India**

³ **Department of Dermatology, Medical College and Hospital, Kolkata, India**

Correspondence:

Anupam Das
Email: anupamdasdr@gmail.com

Abstract

Dyschromatosis universalis hereditaria (DUH) is a rare pigmentary disorder characterized by the presence of mottled hyperpigmented and hypopigmented macules over the trunk, extremities, and face. We have presented a case series comprised of six members of a family who had numerous hyperpigmented and hypopigmented macules distributed all over the body. Histological findings were suggestive of dyschromatosis universalis hereditaria.

Keywords: Dyschromatosis universalis hereditaria; pigmentation disorder

Introduction

Dyschromatosis universalis hereditaria (DUH) was described by Ichikawa and Higara in 1933 [1]. Since then, most of the cases have been reported from Japan. Additional cases have been reported from Europe, South America, India, China, Iraq, South Africa, Saudi Arabia, and Tunisia [2-4]. The disease may be inherited in an autosomal dominant or recessive pattern [5].

Case synopsis

Case 1

A 20-year-old otherwise healthy man, born of non-consanguineous parentage, presented with multiple hypopigmented and hyperpigmented macules over the thighs, buttocks, and lower abdomen since the age of 14 years. The lesions first appeared over the thighs and gradually spread to involve the lower abdomen and buttocks. He was prescribed topical steroids and antifungal creams without any appreciable improvement. There was no history of exposure to any chemical or any significant history of drug intake. Cutaneous examination showed multiple hyperpigmented as well as hypopigmented macules over the lower abdomen, thighs, and buttocks in a reticulate pattern (Figure 1).



Figure 1. Multiple hyperpigmented and hypopigmented macules over the lower abdomen and thighs.

Mucosae, hair, nails, palms, and soles were spared. There was no feature suggestive of chronic arsenicosis. Systemic examination was normal. His father and sister reportedly had similar lesions. Skin biopsy from a hypopigmented macule showed a normal epidermis, absent melanization of basal cells, and few melanocytes. However, a biopsy of a hyperpigmented lesion revealed mild acanthosis, intense hypermelanization of basal keratinocytes and suprabasal cells along with increased vacuolated melanocytes (Figure 2).

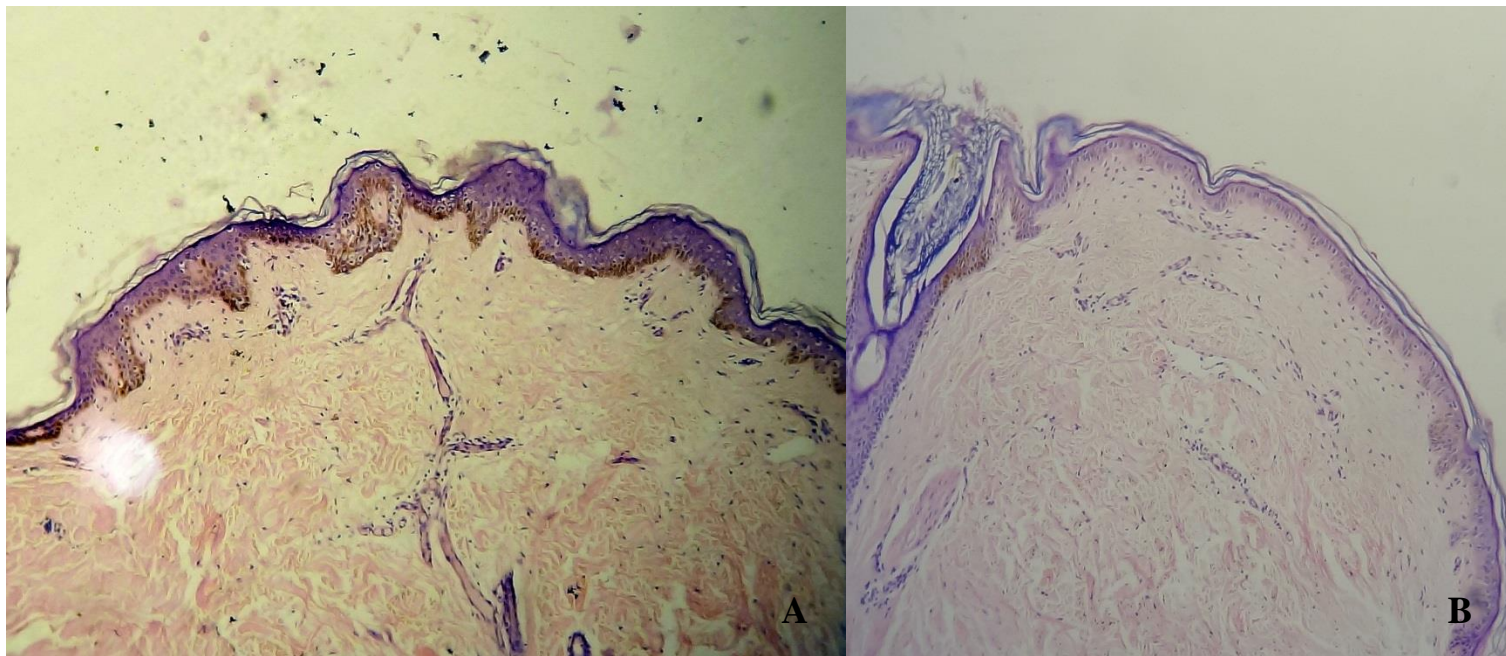


Figure 2. Photomicrographs of (A) hyperpigmented lesion showing mild acanthosis, intense hypermelanization of basal keratinocytes and suprabasal cells along with increased vacuolated cells and a (B) hypopigmented lesion showing absent melanization of basal cells and few melanocytes.

Based on clinicopathological correlation, a diagnosis of dyschromatosis universalis hereditaria was made. We subsequently examined the other affected members of the family. The pedigree tree has been provided (Figure 3).

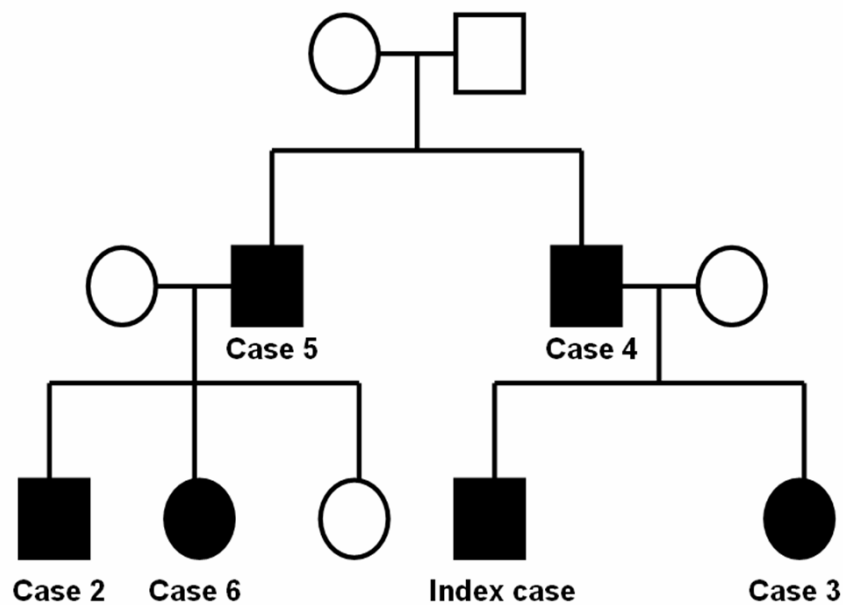


Figure 3. Pedigree chart for the family.

Case 2

A 24-year-old man (paternal cousin of case 1) was brought to us on the first follow-up visit of the index case. He had multiple hyper- and hypo-pigmented macules (size ranging from 4 to 30 mm in diameter) over the lower part of abdomen, thighs, buttocks, hands and shoulders present for the preceding 11 years. (Figure 4) There was no apparent atrophy, erythema or telangiectasia.

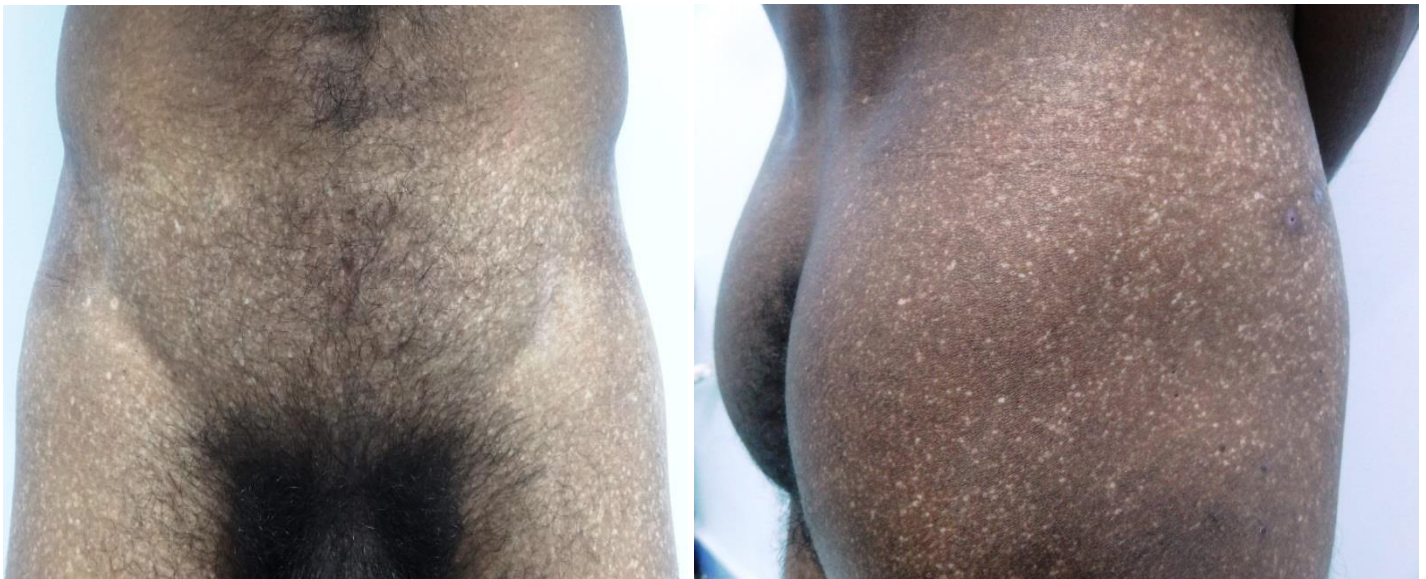


Figure 4. Numerous hyperpigmented and hypopigmented macules over the lower abdomen, thighs and buttocks

Case 3

The 16-year-old sister of the index case had multiple hyper- and hypo-pigmented macules in a reticulate pattern over the lower part of abdomen and legs sparing the acral regions, since the age of 13 years (Figure 5). Palms and soles and mucous membranes were normal.



Figure 5. Note the presence of dyschromatosis over the lower extremity and abdomen.

Case 4

The 45-year-old father of the index case had similar lesions over the entire body including the abdomen, thighs and distal parts of extremities, since the age of 10 years. (Figure 6)

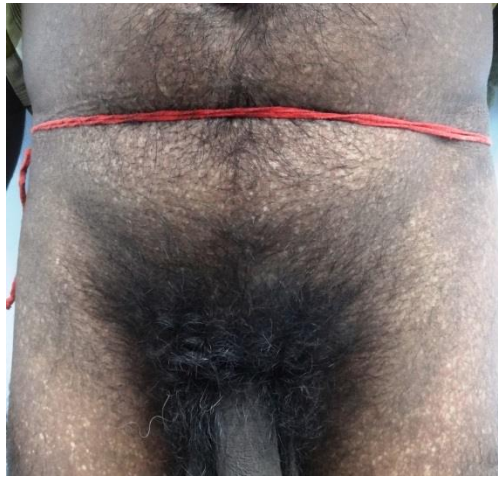


Figure 6. Numerous dyschromatotic macules over the lower abdomen and thighs.

Case 5

The 55-year-old father of case 2 had multiple hyper- and hypo-pigmented macules in a reticulate pattern over the trunk, thighs, buttocks, lower part of abdomen and hands since the age of 14 years. The lesions were dense on the trunk, thigh and buttock. (Figure 7) His palms and soles and mucous membrane were unaffected. There was no other significant positive history.



Figure 7. Numerous hyperpigmented and hypopigmented macules over the upper extremities, lower back, buttocks and thighs.

Case 6

The 27-year-old sister of patient number two had similar lesions over the abdomen and lower extremities, as reported by her brother. However, she was not available for clinical examination.

Discussion

Dyschromatosis is the presence of mixed hypo- and hyperpigmented macules over different regions of the body. DUH is a rare inherited cause of dyschromatosis present over the trunk, extremities and face. In rare cases, the lesions may be found over the palms, soles and mucous membranes [4,6]. Involvement of nails has been reported [7]. More than 80% of patients had presented before 6 years of age. However, late-onset disease may also occur [2].

Histopathologic features of DUH include a focal increase (hyperpigmented lesions) or decrease (hypopigmented lesions) in melanin content of the basal layer and pigmentary incontinence. It has been suggested that DUH is a disorder of melanosome production in epidermal melanin units rather than a disorder of melanocyte number [8]. Mutations in chromosome 6 (6q24.2-q25.2) and chromosome 12 (12q21-q23) have been described [9,10].

We considered and excluded the following differential diagnoses: xeroderma pigmentosum, an autosomal recessive condition characterized by photosensitivity, pigmentary changes, premature skin aging, and malignant tumor development; dyschromatosis symmetrica hereditaria, dyskeratosis congenita, Naegeli-Franceschetti-Jadassohn syndrome, and chronic arsenic toxicity.

DUH may be associated with abnormalities of dermal connective tissue, nerve tissue, short stature, photosensitivity, teeth abnormalities, deafness, cataract, epilepsy, insulin-dependent diabetes mellitus, erythrocyte and platelet abnormalities, tryptophan metabolism abnormalities, and adermatoglyphia [10-13]. However, none of our cases had any associated abnormality.

Familial cases, though rare, have been reported. Both autosomal dominant and autosomal recessive mode of inheritance have been suggested. Wang et. al. presented two cases of dyschromatosis universalis hereditaria (DUH) from a Chinese family [7]. Similar cases running in families have also been reported from Taiwan [9]. Bukhari et. al. reported an interesting series of five cases of DUH from a single Saudi Arabian family [14]. The pedigree tree of our patient indicated an autosomal dominant mode of inheritance.

DUH should be considered in the differentials of all cases presenting with mixed hyper- and hypopigmented macules and biopsy specimens should be obtained to corroborate the diagnosis. In view of the numerous reported associations of this condition, detailed history and thorough cutaneous and systemic examination is mandatory. To the best of our knowledge, this is the first report of six cases of dyschromatosis universalis hereditaria in a single family from India.

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