Case Report

DYSCHROMATOSIS UNIVERSALIS HEREDITARIA

Chandra L Naik, Gurcharan Singh, T S Rajashekar, Rajendra Okade

Abstract

Dyschromatosis universalis hereditaria is an infrequently occurring genodermatosis with a peculiar pigmentary change, consisting of varying sized hyperpigmented macules mingled with hypopigmented lesions to give an overall impression of mottling. We hereby report a case of dyschromatosis universialis hereditaria in a child with no family history of the disorder.

Key Words: *Dyschromatosis universalis hereditaria, dyschromatosis symmetrica hereditaria, genodermatosis*

Indian J Dermatol 2009:54:S74-5

Introduction

A 3-year-old female child presented with generalized asymptomatic spotty hyper and hypo pigmented macules started at the age of one year. There was no history of photosensitivity, epilepsy or any other systemic complaints. The child had grade II mal nutrition and second degree consanguinity. There was no family history of similar skin lesions.

Examination revealed numerous, discrete and confluent circular and irregular shaped spotty hypo pigmented and hyper pigmented macules of various sizes, distributed bilaterally on trunk, limbs, neck and face [Figure 1]. Palms and soles were completely spared. There was no atrophy or telangiectasia of the affected skin. The hair was showing silvery discoloration and the nail, teeth and mucous membrane were within normal limit. Eye and ENT examination was normal, systemic examination was normal except for mild hepatosplenomegaly.

Investigation revealed elevated levels of SGOT and SGPT, ultrasound abdomen showing hepatosplenomegaly with mild ascitis. Other routine investigations and chest X-ray were within normal limits. VDRL and HIV was negative.

Histopathology of skin shows variable epidermal pigmentation with some pigmentary incontinence [Figure 2].

The dyschromatoses are a group of disorder characterized by the presence of both hyperpigmented and hypopigmented macules, with variable sizes and shapes. There are two major forms-dyschromatosis symmetrica hereditaria (DSH) and dyschromatosis universalis heriditaria (DUH). DUH, a rare autosomal dominant genodermatosis was first described by Ichikawa and Hiraga in 1933.[1] It was considered to be a disorder of melanocyte number in the past, however based

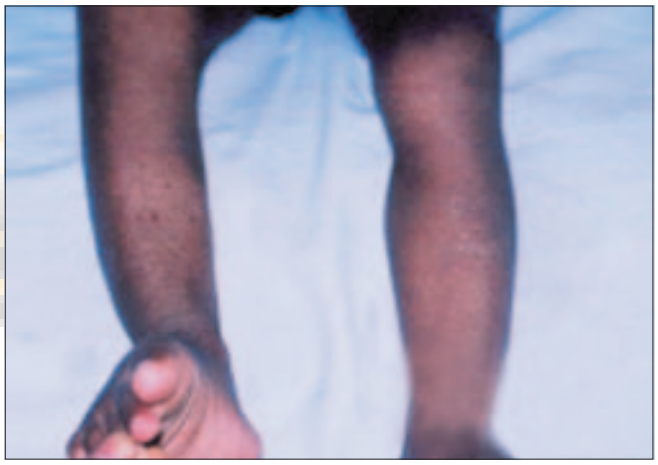

Figure 1: Variable sizes and shapes of hypopigmented and hyperpigmented macules over bilateral lower limb

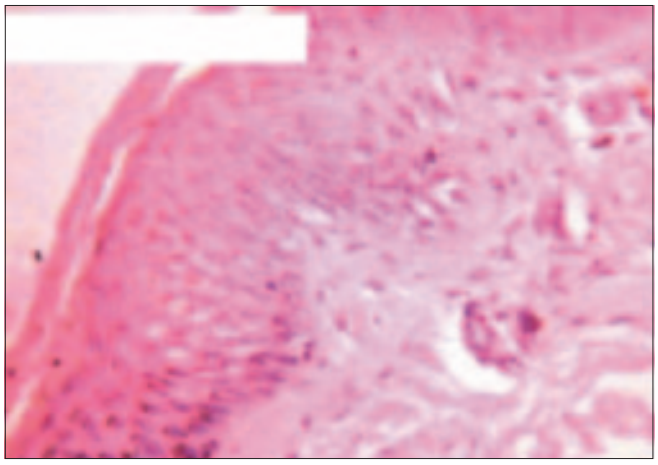

Figure 2: Histopathology showing variable epidermal pigmentation with some pigmentary incontinence (H and E, x100)

From the Department of Dermatology and STD, Sri Devraj Urs Medical College, Tamaka, Kolar - 563 101, Karnataka, India. ***Address for correspondence:*** *Dr. Chandra L Naik, Department of Dermatology and STD, Sri Devraj Urs Medical College, Tamaka, Kolar - 563 101, Karnataka, India. E-mail: chandra.naik@rediffmail.com*

on a recent electron microscopic study, it has been suggested that DUH, may be a disorder of melanosome production in epidermal melanin unit rather than a disorder of melanocyte number.[2] Lesions of DUH have to be differentiated from xeroderma pigmentosum, although in DUH lesions occurs in unexposed sites and does not show any atrophy or telangiectasia and DUH also differentiated from DSH by its presence in generalized form rather than acral distribution. Other differential diagnosis include dyschromic amyloidosis and exposure to chemicals such as diphenylcyclopropenone and monobenzyl ether of hydroquinone.[1,2]

Dyschromatosis universalis heriditaria is commonly encountered in Japan; however rare familial cases have been reported from Europe,[3] China[4] and India.[5,6]

Localized form,[7] involvement of oral mucosa, tongue and palms and soles[8] and association with high tone deafness and small stature,[3] solar elastosis[9] and grand mal epilepsy,[10] has been reported. This case is reported due its rarity and lack of familial involvement.

References

1. Urabe K, Hori Y. Dyschromatosis. Semin Cutan Med Surg 1997;16:81-5.
2. Rai R, Kaur I, Handa S, Kumar B. Dyschromatosis universalis hereditaria. Indian J Dermatol Venereal Leprol 2000;66:158-9.
3. Rycroft RJ, Calnan CD, Wells RS. Universal dyschromatosis with small stature and high tone deafness. Clin Exp Dermatol 1977;2:45-8.
4. Wang G, Li CY, Gao TW, Liu YF. Dyschromatosis universalis hereditaria: Two cases in a Chinese family. Clin Exp Dermatol 2005;30:494-6.
5. Gharpuray MB, Tolat SN, Patwardham SP. Dyschromatosis: Its occurrence in two Indian families with unusual features. Int J Dermatol 1994;33:391-2.
6. Kantharaj GR, Siddalingappa K, Chidambara Murthy S. Dyschromatosis universalis: Autosomal dominant pattern. Indian J Dermatol Venereol Leprol 2002;68:50-1.
7. Dhar S, Malakar S. Localized form of dyschromatosis universalis hereditaria in a 14-year-old girl. Pediatr Dermatol 1999;16:336.
8. Sethuraman G, Srinivas CR, D'Souza M, Thappa DM, Smiles L. Dyschromatosis universalis hereditaria. Clin Exp Dermatol 2002;27:477-9.
9. Sait MA, Garg BR. Dyschromatosis universalis hereditaria Indian J Dermatol Venereol Leprol 1985;51:277.
10. Pavithran K. Dyschromatosis universalis heriditaria with epilepsy. Indian J Dermatol Venereol Leprol 1991;57:102-3.

Received: July, 2006. **Accepted:** August, 2006.
Source of Support: Nil, **Conflict of Interest:** Nil.