Journal of Pakistan Association of Dermatologists. 2017;**27 (1)**:80-82.

Case Report

Dyschromatosis universalis hereditaria: first case report from Northern Pakistan

Inam Ullah Khan, Muhammad Riaz Khan*

Department of Dermatology, Kabir Medical College, Gandhara University, Peshawar, KPK, Pakistan
* Khwazakhela Medical Center, Swat, KPK, Pakistan.

Abstract Dyschromatosis universalis hereditaria is a rare genodermatosis, first reported from Japan. Later on many cases have been reported from other countries of the world. It is characterized by mottled pigmentation. We report first case of this disorder from Northern Pakistan with positive family history.

Key words
Dyschromatosis universalis hereditaria, dyschromatosis symmetrica hereditaria.

Introduction

Dyschromatosis universalis hereditaria (DHU) was first reported from Japan.[1] Later on it was reported from other countries.[2,3] Dyschromatoses are a group of disorders characterized by mottled pigmentation. It includes dyschromatosis universalis hereditaria, dyschromatosis symmetrica hereditaria and a segmental form called unilateral dermatomal pigmentary dermatosis.[4]

Case Report

A ten-year-old male child presented to us with generalized asymptomatic mottled pigmentation involving trunk, arms, legs, hands and feet with relative sparing of the face. According to his father he noticed these changes at the age of two years and have been progressively increasing since then. The disease first appeared on hands arms and legs slowly becoming generalized. Child was born to parents who were first cousins. According to the father his other male child had similar problem. There was no history of photosensitivity or photophobia. His systemic history was unremarkable and IQ was normal.

On examination there was generalized mottled pigmentation with hyperpigmented and hypopigmented macules present in a reticulate pattern **(Figure 1)**. His palms and soles had similar findings **(Figure 2)**. Tongue showed cobblestone hypopigmented papules with hyperpigmented patches **(Figure 3)**. His face showed mild involvement. Nails, hair and teeth were normal. Routine investigations and chest X-ray were normal.

Skin biopsy showed basal layer hyperpigmentation and some melanophages in the upper dermis.

Based on clinical history, examination and pathological correlation a diagnosis of dyschromatosis universalis hereditaria was made.

Discussion

DUH is rare genodermatosis first reported from japan.[1] Later on it was reported from other countries.[2-6] To the best of our knowledge this is the first case report from Pakistan.

Address for correspondence
Professor Inam Ullah Khan
Department of Dermatology,
Kabir Medical College, Gandhara University,
Peshawar, KPK, Pakistan
Email: dr.inam57@hotmail.com

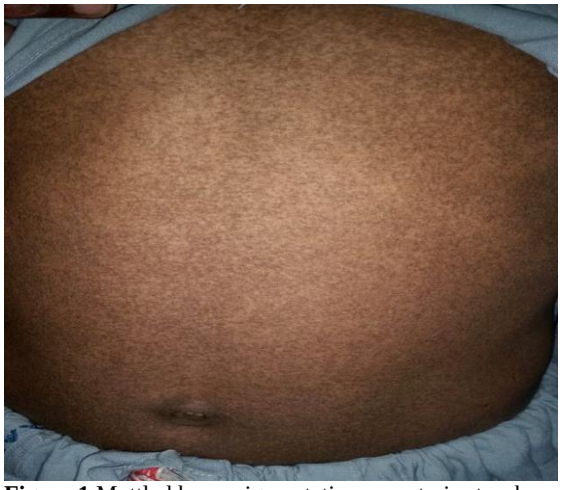

Figure 1 Mottled hyperpigmentation on anterior trunk.

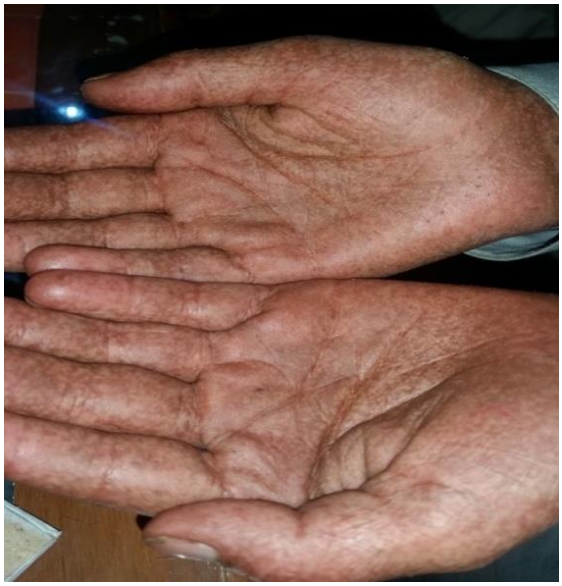

Figure 2 Mottled hyperpigmentation affecting palms.

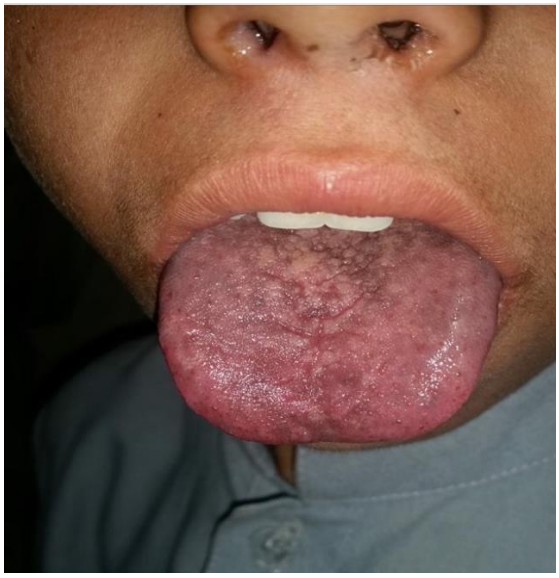

Figure 3 Cobblestone popular lesions on the tongue.

Exact etiology of this disorder is not known. Majority of cases show autosomal dominant pattern of inheritance. Very few have autosomal recessive inheritance.[4] Family history in our case was consistent with autosomal recessive mode of inheritance. In this disorder skin lesions are seen in the first year of life as in our case with trunk and limbs being the predominant sites.[4] Facial lesions are seen in about half of the affected patients but involvement of palms and soles is unusual.[6]

Out of 37 previously reported cases majority were women. In eighty two percent cases clinical symptoms started before the age of six years. Our patient was a male child and another affected sibling in the family who was also male. Age of onset was similar to the majority patients reported from Japan.[4]

Nail and hair involvement has been reported.[7] However, no such involvement was seen in our case.

The disease runs a benign course and no spontaneous regression has been reported. DUH must be differentiated from xeroderma pigmentosum an autosomal recessive disorder with severe photosensitivity, photophobia and a bad prognosis. Familial primary cutaneous amyloidosis may have similar skin changes but can easily be diagnosed by the presence of small foci of amyloid in the papillary dermis.[8]

Cosmetic concern in this disorder especially in females may be severe leading to severe depression.[3,4] There is no treatment for this disorder except for reassurance and genetic counselling. In one reported case narrowband UVB was tried with some improvement and is worth trying.[4]

References

1. Ichikawa T, Higara Y, Higara Y. About a pigmentary anomaly unprecedented. *Jpn Dermatol*. 1933;**34**:360-4
2. Bukhari IA, EI-Harith EA, Stuhrmann M. Dyschromatosis universalis hereditaria as

an autosomal recessive disease in five family members of one family. *J Eur Acad Dermatol Venereol*. 2006;**20**:628-9.
3. Kenani N, Ghariani N, Dengueszli M, Sriha B, Belajouza C, Nouira R. Dyschromatosis universalis hereditaria: Two cases. *Dermatol Online J*. 2008;**14**:16
4. Yadalla HHK, Pinninti S, Baboo AR. Dyschromatosis universalis hereditaria: Infrequent genodermatoses in India. *Indian J Hum Genet*. 2013;**19**:487-90.
5. Merino de Paz N, Roddriguez-Martin M, Contreras Ferrer P, Pestana-Eliche M, Martin-Herrera A. Dyschromatosis universalis hereditaria: An infrequently occurring entity in Europe. *J Dermatol Rep*. 2012;**3**:96-7.
6. Rai R, Kaur I, Handa S, Kumar B. Dyschromatosis universalis hereditaria. *Indian J Dermatol Venereol Leprol*. 2000;**66**:158-9.
7. Naik CL, Singh G, Rajashekar TS, Okade R. Dyschromatosis universalis hereditaria. *Indian J Dermatol*. 2009;**54**:74-5.
8. Morishima T. Amyloidosis cutis dyschromica. *Dermatol Clin*. 1981;**3**:627-30.