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Dyschromatosis Universalis Hereditaria - Case Report of a Rare Genodermatosis in India.

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ABSTRACT

Dyschromatosis universalis hereditaria (DUH) is a rare genodermatosis, clinically manifesting as hyperpigmented and hypopigmented macules forming a reticulate pattern in a generalized distribution. We report a case of DUH in a child with involvement of oral mucosa, palm and soles.

Keywords: Dyschromatosis Universalis Hereditaria, Genodermatosis, Macules.

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. It is a spectrum of diseases which includes dyschromatosis universalis hereditaria (DUH), dyschromatosis symmetrica hereditaria (DSH) or acropigmentation of Dohi 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] It is characterised by a symmetrical distribution of hyperpigmented and hypopigmented macules on the extremities especially over dorsa of the hands and feet. In 1933, Ichikawa and Hiraga described dyschromatosis universalis hereditaria (DUH) which was basically the same disorder but with distribution pattern all over the body.[2] The etiology is unknown. Most of the cases are reported from Japan.[3] Here, we report this rare pigmentary disorder in an eight year old male child.

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CASE REPORT

Eight year old male presented with asymptomatic generalized hyperpigmentation all over the body after 7 months of age. There was no history of consanguinity among the parents and none of the family members were affected. Patient was apparently normal prior to 7 months of age, skin darkening started after 7 months with progression of lesions all over body. Hyper and hypopigmented macules were present over the whole body. The lesions had started over face, both the legs and gradually spread upwards towards the thighs. Few lesions had appeared during this time over the hands subsequently spreading upwards to the elbows. Later, lesions developed over the buttocks and trunk also. Palms and soles were also involved. His face showed mild involvement with hyperpigmentation on forehead and bilateral temporal regions. There was no erythema or telangiectasia. Buccal mucosa, palatal mucosa and tongue were involved. Eyes and ear examination was normal. The hair, nails and teeth appeared normal.

There was no history of any drug intake, systemic illness, or exposure to chemicals. There was no history of photosensitivity or photophobia. Developmental milestones were normal.

Systemic examination did not reveal any abnormality. Routine hematological investigations, serum electrolytes, liver and kidney function test and urine and stool routine showed no abnormality. Serum vitamin B12 and cortisol were normal.

Histopathology of biopsy from sole and upper back showed a focal increase or decrease in melanin content of the basal layer (depending on the type of the lesion biopsied) and occasional melanin incontinence. Deep dermis and subcutaneous tissue were unremarkable. There was no evidence of atypical/ vacuolated keratinocytes.

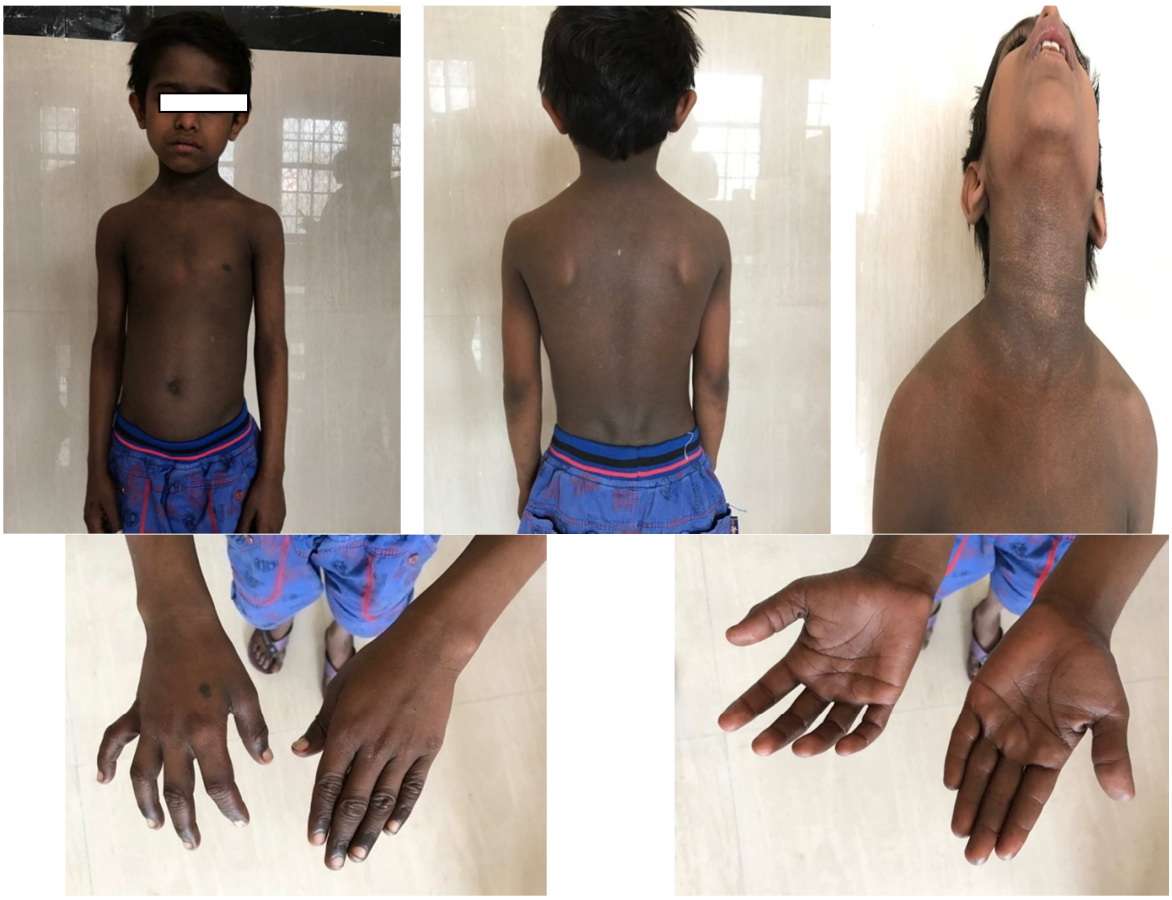

Figure 1: Characterstic, symmetrical rash involving face, neck back, palmar and dorsal surfaces of hands.

DISCUSSION

Dyschromatosis universalis hereditaria is a rare genodermatosis reported initially and mainly in Japan. Once thought to be confined to Japan, DUH is being increasingly reported from other countries including India.[4,5] Although majority of cases show autosomal dominant pattern of inheritance, but may be recessive or sporadically inherited.

In the past, it was considered to be a disorder of melanocyte number, but now, based on a recent electron microscopic study, it has been suggested that DUH may be a disorder of production of melanosomes in epidermal melanin unit.[6]

DUH is characterized by mixture of hyper- and hypopigmented macules occurring all over the body. Skin lesions are usually present in the first years of life. The trunk and extremities are the dominant sites. Facial lesions were seen in almost 50% of affected individuals, but involvement of palms and soles is unusual.[7, 8] In our case, face and palms and soles were also involved. Oral mucosa also was involved in our case. Abnormalities of hair and nails have also been reported. In our case, however, hair, nails were normal.

The histopathology typically 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. It can be associated with abnormalities of dermal connective tissue, nerve tissue, or with other systemic complications.[9,10] A large number of systemic associations have been reported with DUH including tuberous sclerosis, X linked ocular albinism, grand mal epilepsy, glaucoma, cataract, learning difficulties, insulin-dependent diabetes mellitus, teeth abnormalities, and small stature.[11] No such features, however, were present in our patient.

Lesions of DUH have to be differentiated from xeroderma pigmentosum, since in both the disorders patients clinically show lesions in the photoexposed areas. However, in DUH lesions occur in the unexposed sites as well. Moreover, the lesions show no atrophy or telangiectasia.

No treatment modality is available for DUH. In general, DUH does not progress or worsen with age. Spontaneous regression is not known.[12]

CONCLUSION

Dyschromatosis Universalis Hereditaria is a rare genodermatosis especially in India. Moreover involvement of palms and soles is also rare in DUH. This case is reported due to its rarity in India and also due to involvement of oral mucosa, palms and soles in our patient.

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