

Dyschromatosis Universalis Hereditaria: A Rare Case Report in Southern India

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ABSTRACT

Dyschromatosis Universalis Hereditaria (DUH) is a rare genodermatosis characterised by hyper and hypo pigmented macules in reticulate pattern. The clinical manifestations includes occurrence of lesions at the trunk and extremities as the dominants sites. The lesions may spread to the face, hands and feet. Hair, teeth, nails, palms and soles may also be involved. Here, we present a case of 7 year old male patient with DUH with the involvement of whole body surface. The treatment pattern in this case includes cortisol hormone replacement therapy with the use of hydrocortisone tablets for a long period of time.

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Introduction

Dyschromatosis Universalis Hereditaria (DUH) is a rare genodermatosis characterized by hyper and hypopigmented macules forming a reticulate pattern [1]. This abnormal pigmentation appears in a generalised distribution [2]. Both autosomal dominant and autosomal recessive forms have been reported [3]. This disorder is reported mainly from Japan. Only a few cases have been reported from other countries [4,5]. Hence, we are reporting a case of DUH from southern part of India.

Case Report

A 7 year old male patient who was presented with full body hyperpigmentation from the past 6 years was observed with diffused hyperpigmentation on the whole body involving arms, plantar of feet, buccal mucosa and nails. There was no family history of similar complaints and also no history of any chemical or drug intake and no history of photosensitivity & photophobia. Systemic examination was found to be normal. His ACTH stimulation test reveals low level of cortisol was 1.12 mcg/dl (normal: 6.2 - 19.4 mcg/dl). Further skin biopsy from right forearm was performed which shows epidermis with diffuse hypermelanosis confined to basal cell layer and upto lower 1/3rd of epidermal thickness at places.

Deep dermis shows increased collagenization. Based on these findings, it was diagnosed as Dyschromatosis Universalis Hereditaria. The dermatologist has started the cortisol hormone replacement therapy which mainly includes the long term use of hydrocortisone tablet. The condition of patient has been improved but, no satisfactory treatment modality has ever been reported for this condition. Although hyper-pigmented macules can improve with the use of lasers, intense pulsed light or by chemical peeling.

Discussion

DUH is a rare genodermatosis which has been reported most often in Japan. Most of the cases show autosomal dominant pattern of inheritance and a few have inherited in an autosomal recessive pattern. The etiology of this disorder is unknown. In DUH, skin lesions are usually present in the first year of life which includes the occurrence of mixtures of hyper and hypo pigmented macules all over the body. This histopathology typically shows a focal increase or decrease in melanin content of the basal layer (in this case increase in melanin content has been observed) and occasionally pigmentary incontinence. In this condition, the lesions may occur in the exposed and unexposed sites. In a recent ultrastructural skin investigation, Nuber and his team stated that DUH is a disorder of melanosome synthesis rate or melanocyte

activity and not a disorder of melanocyte number. It does not progress or worsen with age.

Conclusion

Dyschromatosis Universalis Hereditaria is a rare genodermatosis condition which involves hyper or hypopigmentation of the skin. Patients often suffer from depression because of the cosmetic disfigurement and hence, psychological counselling is required. Role of NBUVB (Narrow Band Ultraviolet-B therapy) as a treatment modality should be evaluated and tried for better cosmetic appearance. Dermatologists should focus on the better management for improving the quality of life the patients with the help of other health care professionals.

Abbreviations

- ACTH - Adreno Cortico Tropic Hormone
- DUH - Dyschromatosis Universalis Hereditaria
- NBUVB - Narrow Band Ultra Violet - B

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