CASE REPORT
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A CASE OF DERMATOPATHIA PIGMENTOSA RETICULARIS

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Abstract

Dermatopathia pigmentosa reticularis is a rare ectodermal dysplasia with a triad of generalized reticulate hyperpigmentation, noncicatricial alopecia, and onychodystrophy. We report a case of a 3 year old female with reticulate hyperpigmentation present over whole body including oral mucosa and sclera. Diffuse thinning of hair on scalp was present. Poorly developed dermatoglyphics were there. There was onychodystrophy. Histopathology revealed superficial perivascular lymphocytic infilterate which also supported the diagnosis. There was no evidence of involvement of other ectodermally derived organ.

Key words: Ectodermal dysplasia, Reticulate pigmentation, Dermatoglyphics

Introduction

First described by Hauss and Oberste-Lehn [1] in 1958, Dermatopathia Pigmentosa Reticularis (DPR) is a rare ectodermal disorder. The diagnostic triad includes generalized reticulate hyperpigmentation, noncicatricial alopecia and onychodystrophy.

Case Report

A 3 year old female child, born out of non consanguineous marriage presented with darkening of skin since 6 months of age, which increased progressively to whole of the body including oral mucosa and sclera within a span of 1 month and diffuse thinning of hair on scalp. There was no history of photophobia. Hearing and sweating were normal. There was no history of similar illness in the family. She was born to a primigravida by normal vaginal delivery. She had one male sibling who was normal. The morphology of the hair shaft was normal on clinical and microscopic examination. Her developmental milestones were below 3rd centile. On examination, generalized reticulate hyperpigmentation was present (Figure No.1). Scalp hair was short and there was diffuse thinning. Nails were dystrophic. Poorly developed dermatoglyphics were there (Figure No.2).

Figure 1 : Diffuse reticulate hyperpigmentation of skin.

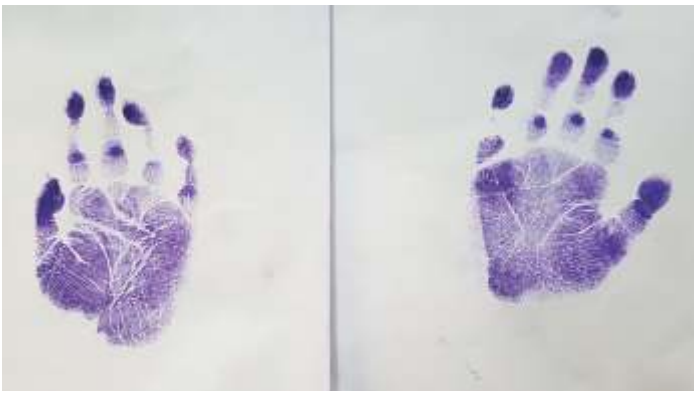

Figure 2 : Poorly developed dermatoglyphics.

Oral mucosa and sclera showed reticular pigmentation and teeth showed mineralization defect (Figure No.3). Her intelligence quotient was estimated to be in the normal range. Routine investigations in the form of complete hemogram, liver function test, renal function test and chest radiography were normal. Her thyroid profile was deranged and she was diagnosed with juvenile hypothyroidism. Her adrenal function was normal.

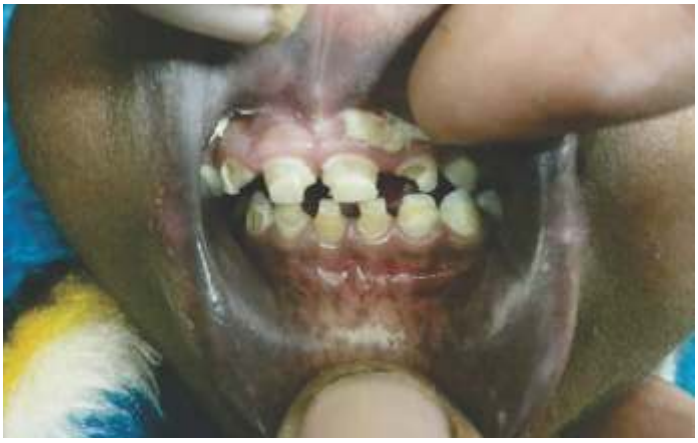

Figure 3 : Teeth mineralization defect.

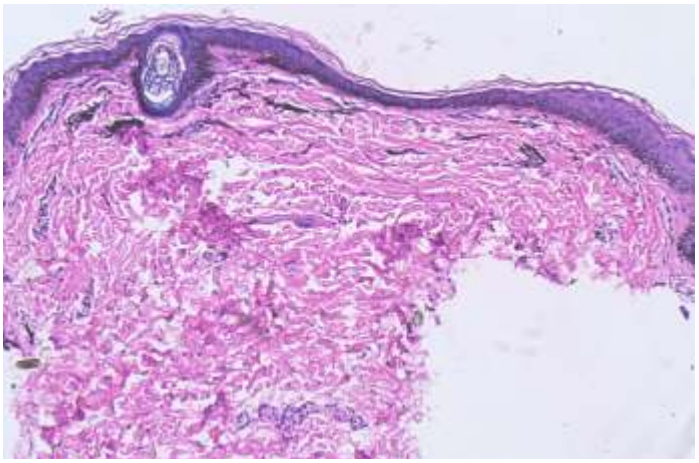

Figure 4 : superficial perivascular lymphocytic infilterate. The papillary dermis was slightly thickened with fibroblast and mucin.

Histopathology revealed superficial perivascular lymphocytic infilterate. The papillary dermis was slightly thickened with fibroblast and mucin. The epidermis was flattened at places (Figure No.4).

Discussion

DPR is an autosomal dominant ectodermal dysplasia. It usually presents as triad of reticular pigmentation, non-scarring alopecia and nail changes. The reticular pigmentation of DPR occurs at birth or during early childhood and persists throughout life.[2] Many other dermatologic findings have been associated with this triad, which include absent or decreased dermatoglyphia, hypohidrosis or hyperhidrosis, palmoplantar hyperkeratosis, acral nonscarring blisters, diffuse or punctate palmoplantar hyperkeratosis, darkly pigmented nipples, mucosal pigmentation, digital fibromatosis, neurofibromas, and wiry scalp hair.[3] Few extracutaneous manifestations that have been reported in the literature include fine punctate superficial spots in the cornea, Salzmann's nodular degeneration of the cornea, and early-onset gastric carcinoma.[4].

There are less than 20 cases of true DPR reported in the literature. Most of the cases were reported in Europe, and a few cases were reported in the USA and Asia; there was no race or sex predilection for DPR. The onset of the reticulate pigmentation of DPR usually occurs at birth or during early childhood, and the rest of its manifestations appear later. This condition should be differentiated from other genodermatosis associated with generalized reticulate pigmentation like dyskeratosis congenita (DKC), Naegeli-Franceschetti-Jadassohn Syndrome (NFJS), Dowling-Degos disease, reticulate acropigmentation of Kitamura and Haber's syndrome. Reticulate hyperpigmentation, mucosal leukoplakia, bone marrow dysfunction, cytogenetic instability, and a predisposition to malignancy are characteristic of DKC. These patients can have dental findings, reticulate hyperpigmentation, adermatoglyphia, palmoplantar hyperkeratosis, and nail anomalies similar to NFJS and DPR patients.[5] In Kitamura's disease, palmar pits and breakage in palmar ridges and acral hyperpigmentation, especially on the backs of hands and feet can be observed. Haber's syndrome is characterized by verruciform papular lesions of the trunk and a distinct facial erythema and telangiectasia, most commonly presenting in childhood. In X-linked reticulate pigmentary disorder in females, pigmentation occurs along the Blashcko's line.. Flexural reticulate hyperpigmentation occurs in Dowling-Degos disease. Additional findings, such as dark hyperkeratotic follicles, pitted perioral scars, and comedo-like lesions may occur. Galli-Galli disease also shows macular and papular reticulate pigmentation of flexures.The histopathology of the reticulate pigmentation of DPR is not diagnostic, and the reported histopathological features include mild orthokeratosis, papillomatosis, heavily pigmented epidermis, liquefaction degeneration of the basal layer, dermal pigmentary incontinence, melanophages, interface dermatitis, and sparse, superficial perivascular inflammations.[6] The microscopic examination of the hair shaft showed normal hair shafts [7] as observed in our patients.Although DPR and NFJS have poorly developed dermatoglyphics, specifically reticulate hyperpigmentation of the skin, DPR has been distinguished from NFJS by the lifelong persistence of the skin hyperpigmentation, partial alopecia, and absence of dental anomalies.[8]

No specific laboratory changes are seen in DPR. The typical histopathologic picture of DPR shows liquefaction degeneration of the basal layer and dermal pigmentary incontinence. No specific treatment exists for DP, except for symptomatic management of some of the associated conditions, such as palmoplantar hyperkeratosis. For hyperkeratosis, topical retinoic acids and keratolytics may be tried.

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