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Case Report

# Dermatopathia pigmentosa reticularis and atopic dermatitis: A case report of two siblings

**Aisha Ghias**

Department of Dermatology, Unit I, King Edward Medical University/Mayo Hospital, Lahore

**Abstract** Dermatopathia pigmentosa reticularis is a rare reticulate pigmentary disorder which starts during infancy or childhood and consists of a triad of generalized reticulate hyperpigmentation, nonscarring alopecia, and nail dystrophy. We report 2 sisters with this syndrome with an additional association of atopic dermatitis in the elder one.

***Key words***
Dermatopathia pigmentosa reticularis, Naegeli-Franceschetti-Jadassohn syndrome.

## Introduction

Dermatopathia pigmentosa reticularis (DPR) is a rare disorder with the diagnostic triad of generalized reticulate hyperpigmentation, nonscarring alopecia, and nail changes. It was first described by Hauss and Oberste-Lehn in 1958.[1] Approximately 13 cases of DPR have been reported worldwide; however, there is no case report from Pakistan so far.

## Case report

We report case of two siblings. The elder sister, 8-year old, presented with reticulate pigmentation on almost entire skin. It started on the neck at the age of 6 months and later involved progressively whole of the body within a year. There was history of short-lived blistering episodes around 2 years of age but these healed without scarring or pigmentation. She had a xerotic skin since always and gave positive history of atopy. Palms and soles were hyperkeratotic, as well as, pigmented. Trunk showed diffuse, as well as, reticulate hyperpigmentation but there were no areas of hypopigmentation, atrophy or telangiectasias (**Figures 1** and **2**). Lingual and buccal mucosa also showed pigmentary spots but teeth were normal. The dermatoglyphics of the hands were poorly developed and absent at places (**Figures 3** and **4**).

She had diffuse nonscarring alopecia with dry lusterless, short hair. Hair shaft microscopy however did not reveal any abnormality. There was history of drenching sweats around 3 years of age followed by decreased sweating later in life. Ichthyotic scaling was there on lower legs along with xerosis and reticulate pigmentation (**Figure 5**). All the fingernails were white and atrophic and there was brachyonychia of most toenails along with atrophy. Hearing and vision were normal and she had a normal height for age. Histopathology of involved skin showed increase in number of melanocytes along with basal layer degeneration and mild infiltrate at dermoepidermal junction (**Figure 6**).

Her younger sister was 8-month old with history of similar involvement of skin and nails. There was no family history of such pigmentation among parents. Both sisters were born by normal vaginal delivery.

**Address for correspondence**
Dr. Aisha Ghias
Department of Dermatology,
King Edward Medical University/
Mayo Hospital, Lahore
Email: draishaghias@gmail.com

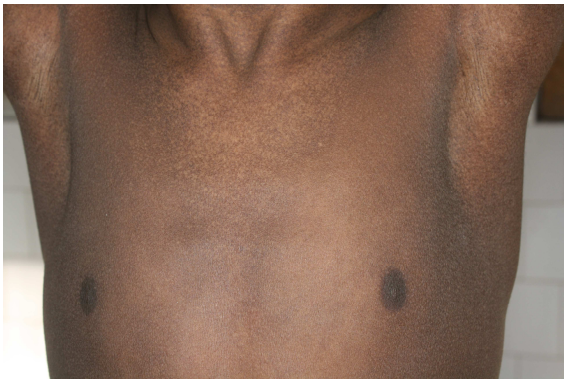

**Figure 1** Generalised reticulate pigmentation not sparing flexures.

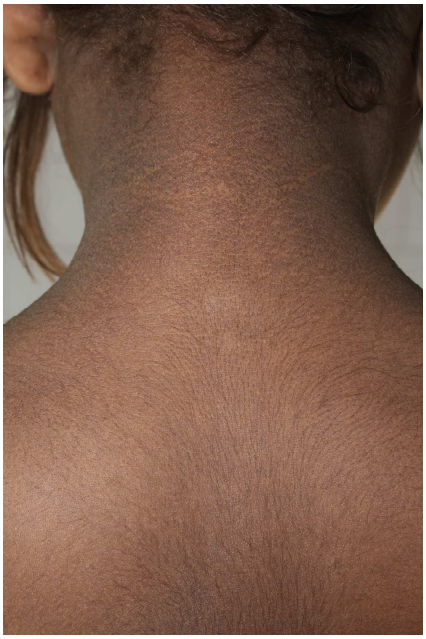

**Figure 2** Mottled pigmentation of back of trunk.

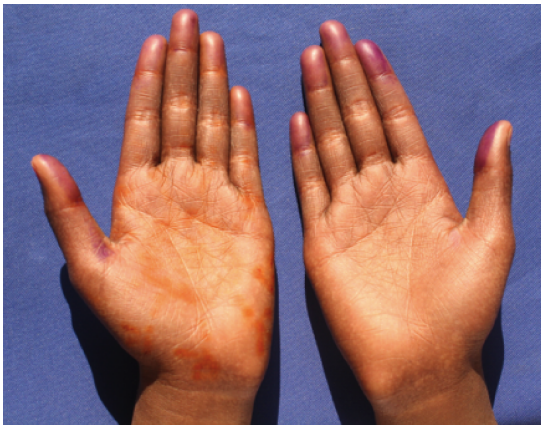

**Figure 3** Partial loss of dermatoglyphics and mottled pigmentation of palmer surface of both hands.

**Figure 4** Loss of fingerprints.

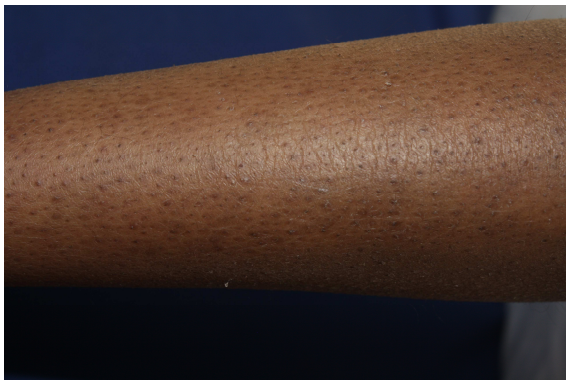

**Figure 5** Mild ichthyosis on legs.

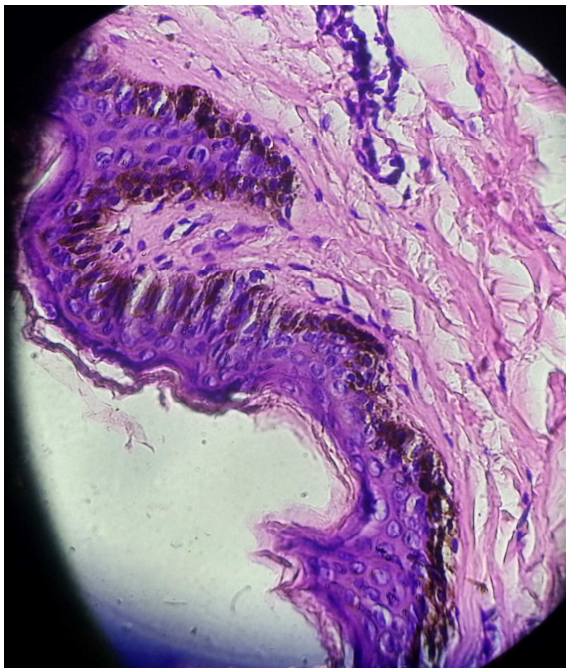

**Figure 6** Abundance of melanocytes in the basal layer with basal layer degeneration and mild inflammatory infilterate.

Routine investigations including complete blood picture, liver function tests, renal function tests and chest radiography were all normal however serum IgE levels were markedly raised.

A diagnosis of dermatopathia pigmentosa reticularis (DPR) with a differential of Naegeli-Franceschetti-Jadassohn syndrome (NFJS) was made. Normal teeth enamel and the presence of diffuse nonscarring alopecia favoured DPR.

**Discussion**

DPR is an autosomal dominant disorder characterized by a triad of widespread reticulate hyperpigmentation, nonscarring alopecia and nail changes.[2] Other associated findings include dermatoglyphic changes, hypohidrosis or hyperhidrosis, palmoplantar hyperkeratosis, and acral dorsal nonscarring blisters.[3] The reticular pigmentation of DPR occurs at birth or during infancy and persists throughout life.[4]

Reticulate pigmentation of early onset can be either localized or generalized. Localized reticulate skin pigmentation along with involvement of hair, nail, teeth and CNS is seen incontinentia pigmenti.[5]

Widespread or generalized reticulate skin pigmentation is seen in epidermolysis bullosa simplex with mottled pigmentation. Reticulate pigmentation in association with involvement of hair, nail, teeth and eccrine glands is seen in dermatopathia pigmentosa reticularis and Naegeli-Franceschetti-Jadassohn syndrome. Widespread pigmentation of skin with involvement of hair, nail, teeth and systemic involvement is seen in dyskeratosis congenita, Fanconi′s anemia and certain mitochondrial diseases.[5]

Causes of adult-onset reticulate pigmentation are different and include dirty neck in atopic dermatitis, reticulate pigmentary anomaly of flexures, reticulate acropigmentation of Kitamura, reticulate acropigmentation of Dohi, LP pigmentosus, drug-induced reticulate pigmentation, ashy dermatosis, pigmented contact dermatitis, prurigo pigmentosa, systemic sclerosis, Galli-Galli disease and erythema ab igne.[5,8]

Reticulate hyperpigmentation, oral leukoplakia, bone marrow dysfunction and a predisposition to malignancy are characteristic of dyskeratosis congenita. These patients can have dental abnormalities, reticulate hyperpigmentation, palmoplantar hyperkeratosis, and nail anomalies similar to NFJS and DPR patients.[6]

Palmoplantar keratoderma, nail dystrophy, and enamel defects are common in NFJS, whereas diffuse alopecia is only seen in DPR. Teeth are always severely affected, leading to early total loss in NFJS. In some NFJS pedigrees, the reticulate pigmentation fades after puberty and may disappear completely in old age. In DPR the hyperpigmentation persists throughout life, showing no tendency of spontaneous fading. The reticulate network of hyperpigmented macules occurs particularly on the trunk, neck, and proximal areas of the limbs.[5,9]

Genetic studies have recently confirmed that DPR and NFJS are allelic with dominant mutations in KRT14 at chromosome 17q11.2-q21. Severe keratin 5 and 14 mutations induce downregulation of junction proteins in keratinocytes, which likely underlies these diseases.[7]

Atopic eczema has not yet been reported in association with DPR.

No specific laboratory anomalies have been detected in DPR. There is no specific treatment for DPR, except for symptomatic management of cutaneous problems e.g. palmoplantar keratoderma for which topical retinoids and keratolytics may be useful.[10]

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