

Dermatopathia Pigmentosa Reticularis with Addisonian Pigmentation: Atypical Presentation of a Rare Case

Abstract

Dermatopathia pigmentosa reticularis is a rare ectodermal dysplasia with mottled pigmentation. Here we report a case of 15-year-old boy with variable (reticulate as well as diffuse) pigmentary disorder and adermatoglyphia.

Keywords: *Adermatoglyphia, dermatopathia pigmentosa reticularis, reticulate pigmentary disorder*

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Introduction

Dermatopathia pigmentosa reticularis is a very rare disorder of reticulate pigmentary abnormality. It is an ectodermal dysplasia characterized by clinical triad, with widespread reticular pigmentation, non-scarring alopecia, and nail changes. We encountered a patient with classical features, along with addisonian pigmentation. Only less than 20 cases of dermatopathia pigmentosa reticularis have been reported so far.

Case Report

A 15-year-old boy presented with complaints of speckled pigmentation over face, dorsae of hand and palms, dorsae of feet, soles and pubic area since the age of 7 years. He also developed deformed nails around the same time. There has been occasional blistering of palms with heavy manual labor since 3 years and thinning of hair since 1 year. According to the patient, one elder male sibling has similar complaints which started around the same age as him. However, four sisters and a brother are normal. Antenatal and postnatal history was uneventful. There was no history of hyperhidrosis, plantar hyperkeratosis, photosensitivity, or any ocular complaints. General physical examination was normal.

On cutaneous examination, there were multiple freckles and lentigenes on face, predominantly in centofacial and periorbital location. There was sparsening of lateral third of eyebrows, lower lid lashes and scalp hair with bleaching of the color of scalp hair.

There was reticulate hyperpigmentation over neck extending onto V of chest anteriorly and nape of neck posteriorly [Figure 1]. Reticulate hyperpigmentation mottled with hypopigmentation was present over palms [Figure 2a], dorsae of hands extending to forearms, soles [Figure 2b] and dorsae of feet extending to the ankle joint. There was adermatoglyphia of distal phalynx of fingers [Figure 3a] while reduced dermatoglyphics were seen over the soles [Figure 3b]. Multiple discrete to coalesced hypopigmented macules of variable size over hyperpigmented background were present on lower abdomen and groins. There was diffuse pigmentation in axillae [Figure 4a], both groins [Figure 4b], cubital fossae [Figure 4c] and popliteal fossae. On mucosal examination, there were hyperpigmented macules on lips, palate, buccal mucosa and glans penis. Scalp showed bleached, light-colored, sparse hair. Thinning of all toe nails and loss of distal nail plates in both great toes [Figure 5a] along with finger nail dystrophy, melanonychia, onycholysis, thinning and ridging [Figure 5b] were found on nail examination.

Thus on the basis of history and examination, we kept differentials of dermatopathia pigmentosa reticularis and dyskeratosis congenita along with addisonian pigmentation. We worked up the patient for evidence of bone marrow suppression and to evaluate the cause of Addisonian pigmentation.

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Figure 1: Reticulate pigmentation over anterior chest and neck

On investigations, his hemoglobin was 9.8 gm/dL with megaloblastic anemia and eosinophilia on peripheral smear. His vitamin B12 levels {78 pg/ml (normal: 180–914 pg/mL)} and folic acid {3.89 nmol/L (normal: 6.5–35 nmol/L)} levels were below reference range. Other investigations and evaluations including sweat test, dental, otorhinolaryngeal and ophthalmological evaluations were within normal limits. Vitamin B12, folate, and iron supplementation were done. The patient was kept under strict follow-up and after 3 months, there was near complete resolution of flexural pigmentation and complete resolution on examination at 6 months.

While the reticulate pigmentation over the chest was in favor of dyskeratosis congenita, the absence of leukoplakia and bone marrow suppression went against this differential. On the other hand, the patient fulfilled the diagnostic triad of dermatopathia pigmentosa reticularis. Thus, we made a diagnosis of dermatopathia pigmentosa reticularis with addisonian pigmentation. For the pigmentary and hair changes, we started the patient on topical azelaic acid 20% cream to be applied at night on the lesions on face and neck and topical minoxidil 5% to be applied 1 ml twice a day on scalp, respectively. While the reticulate pigmentation has not responded, patient reports significant improvement in hair density after 6 months of use of minoxidil.

Discussion

Dermatopathia pigmentosa reticularis (DPR) was originally documented by Hauss and Oberste-Lehn^[1] in 1958, and is an uncommon reticulate pigmentary disorder. The mode of inheritance is autosomal dominant.^[2] DPR is characterized by a triad of widespread reticular pigmentation, non-scarring alopecia, and nail changes. Other associated findings include adermatoglyphia, hypohidrosis or hyperhidrosis, palmoplantar hyperkeratosis, and acral dorsal non-scarring blisters. The reticular pigmentation of DPR occurs at birth or during early childhood and persists throughout life.^[3]



Figure 2: Mottled pigmentation over palms (a) and soles (b)

Few extracutaneous manifestations have been reported in the literature, which include fine punctate superficial spots in the cornea, Salzmann's nodular degeneration of the cornea, and early-onset gastric carcinoma.^[4] 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.^[4] Various differential diagnoses of DPR may include overlapping features i.e., acromelanosis progressiva, hereditary symmetric dyschromatosis of Dohi, reticular acropigmentation of Kitamura, heterochromia extremitarium, reticulate pigmented dermatosis of the flexures, congenital diffuse mottling of the skin, hereditary universal dyschromatosis, dyskeratosis congenita syndrome, Naegeli-Franceschetti-Jadassohn (NFJ) syndrome. NFJ

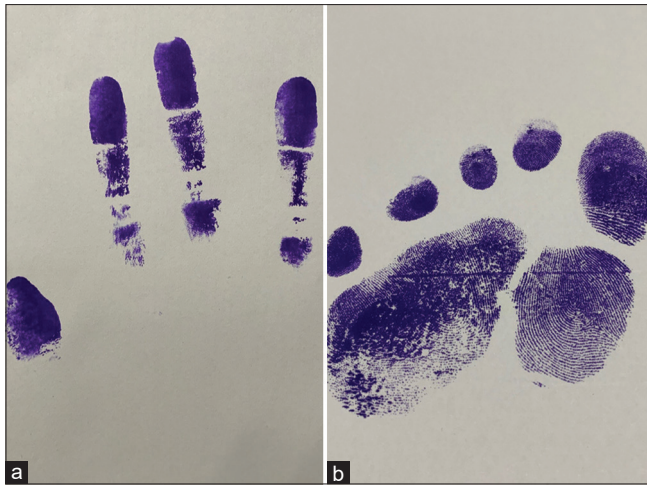


Figure 3: Dermatoglyphics of palms (a) and soles (b)

syndrome is autosomal dominant and has been described as a gray-brown reticulate pigmentation of the trunk and neck; however, the pigmentation is classically described as improving in adulthood, which has not been documented in cases of DPR.^[5] Hypoplasia of the dermatoglyphics has been reported in one family diagnosed as having NFJ syndrome by Sparrow *et al.*^[6] in 1975. In contrast, patients affected by DPR have a total absence of dermatoglyphics. Dental anomalies are a primary feature of NFJ syndrome, but they have not been described in cases of DPR. Congenital diffuse mottling of the skin can be excluded based on histologic grounds; there were no elongated club-shaped rete ridges but rather pigmentary incontinence. Dyskeratosis congenita is almost always confined to males and is believed to be inherited as an X-linked recessive condition; leukokeratosis and blood dyscrasias occur frequently.^[2]

Our closest differential in this patient was dyskeratosis congenita. We have tabulated the clinical features of the two differentials with features in our patient in Table 1.

Addisonian pigmentation, which presents as diffuse hyperpigmentation of skin with more pronounced pigmentation in photo exposed sites, flexures, frictional sites, palmar creases, may be caused by tuberculosis, autoimmune adrenalitis, adrenal hemorrhage, congenital hypoplasia, sarcoidosis, amyloidosis, malignancy, and vitamin B12 deficiency. In our patient, concomitant vitamin B12 deficiency led to Addisonian pigmentation, which resolved on replenishment of vitamin B12.

No specific treatment is available for DPR. Symptomatic management of hyperkeratosis and other secondary changes can be done with keratolytics, topical steroids, and emollients.^[7]

This is a rare case of dermatopathia pigmentosa reticularis with addisonian pigmentation.



Figure 4: Diffuse pigmentation in axilla (a), both groins (b), cubital fossa (c)

Table 1: Comparison of clinical features in the patient with classical cases of DPR and DKC

	DPR	DKC	PATIENT
PIGMENTATION			
Neck, palms, soles	+	+	+
Flexures	-	+	+
Atrophy	-	+	+
Mottled pigmentation	+	+	+
Nail dystrophy	+	+	+
Scalp hair sparsening	+	+/-	+
Madarosis	+	+/-	+
Acral blistering	+	+/-	+
Leukoplakia	-	+	-
Adermatoglyphia	+	+	+
Dental abnormalities	-	+	-
Bone marrow dysfunction	-	+	-

DPR-Dermatopathia pigmentosa reticularis, DKC- Dyskeratosis congenita

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.



Figure 5: Nail deformities of toe nails (a) and finger nails (b)

Conflicts of interest

There are no conflicts of interest.

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