CASE REPORT

Dermatopathia pigmentosa reticularis with beard alopecia: first report from Syria

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Abstract

Dermatopathia pigmentosa reticularis (DPR) is a rare disorder with characteristic triad of generalized reticulate hyperpigmentation, noncicatricial alopecia and onychodystrophy. We report the first case from Syria with classical features of the triad along with hyperhidrosis and adermatoglyphia. Based on previous studies this case is distinguished by the location of alopecia on the beard which is not reported before and distinctive histopathological features were found on the biopsy.

INTRODUCTION

Dermatopathia pigmentosa reticularis (DPR) is a very rare autosomal dominant congenital ectodermal dysplasia mainly affects the skin, nails and hair with a characteristic diagnostic triad of widespread reticulate hyperpigmentation, noncicatricial alopecia and onychodystrophy. Patients with this syndrome may also have adermatoglyphia, palmoplantar hyperkeratosis, hyperhidrosis or hypohidrosis, and acral dorsal nonscarring blisters [1,2].

Very few occurrences have been reported in the literature but none of the cases presented with beard alopecia, and to the best of our knowledge this is the first case of this extremely rare syndrome reported from Syria.

CASE REPORT

A 38-year-old Syrian man presented to the Dermatology department at Tishreen University Hospital with main complaint of multiple nonscarring alopecia on the beard associated with diffuse brown reticulate hyperpigmentation. The patient was born to Syrian second-degree relatives with no family history of similar disorder. However, he had a cousin who was born with the same symptoms. The patient had normal physical, social and mental development without prominent medical, surgical or drug history.

On examination, the reticulate hyperpigmentation involved the neck, trunk, extremities (Fig. 1) and was more obvious on axilla (Fig. 2) and palms (Fig. 3). It began at birth and became severe in adulthood; the intensity of hyperpigmentation slightly decreased at the age of 30 but remained present. The oral mucosa was not affected, no oral leukoplakia or pallor. No dental abnormalities were found. Scalp and body hair was thin since childhood. No eyebrow, axillary or pubic hair loss was noted. Nine months ago he developed multiple nonscarring alopecia on the beard (Fig. 4). The hair pull test was negative. The morphology of the hair shaft was normal on clinical and microscopic examination. Onychodystrophy of many fingernails and toenails was found, it started at 13 years of age then the fingernails improved during the last 10 years while the dystrophy of toenails became worse (Fig. 5). Nail clipping for fungus and a fungal culture were negative. There was a history

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of hyperhidrosis along with hypoplastic dermatoglyphics of fingerprints over fingers and toes (Figs 6 and 7). There was not palmoplantar hyperkeratosis or acral dorsal nonscarring blisters.

Neurological, cardiovascular, respiratory, auditory and visual examination was normal. Abdominal ultrasound did not reveal any abnormality. Blood chemistry analysis, thyroid function tests, cortisol, total testosterone levels, tumor markers, hematology profile and blood film morphology, urinalysis and
24-h porphyrins urine test were all normal. We could not conduct genetic phenotyping because it is not available.

Histopathological examination of skin biopsy revealed unremarkable epidermis. Scattered variable numbers of melanophages in the papillary dermis (Fig. 8) and pigment incontinence with marked tendency of melanophages to be situated around blood vessels (perivascular melanophages) (Fig. 9). Melanophages showed both spindle and epithelioid morphology with some particularly large forms contain prominent coarse melanin granules (Fig. 10), adnexal structures were absent.

Topical minoxidil 5% has not been shown to be effective for beard alopecia.

DISCUSSION

DPR is a rare autosomal dominant pigmentary dermatosis caused by mutations in the KRT14 gene located on chromosome 17 [1,3]. Since first described by Hauss and Oberste-Lehn in 1958 [3,4], DPR has been acknowledged as a very rare disorder with approximately 21 reported cases among the regions of America, Europe and Asia [5]. It is characterized by a triad of diffused reticulate hyperpigmentation, nonscarring alopecia that may include the scalp, eyebrows or axillary hair and onychodystrophy [6]. The diagnosis of DPR can be made clinically based on the previous triad [7,8]. Other associated features include: hyperhidrosis or hypohidrosis, acral dorsal nonscarring blisters, adermatoglyphia and palmoplantar hyperkeratosis [2,9]. The reticular pigmentation of DPR occurs at birth or in early childhood on the trunk, neck and proximal limbs, it presents throughout life without spontaneous fading.

Other diseases fall under the category of reticulated pigmentary disorders that should be considered and excluded are: Naegeli-Franceschetti-Jadasson syndrome (NFJS), Dyskeratosis Congenita (DKC), Dyschromatosis Universalis Hereditaria (DUH), Dowling-Degos disease, reticulate acropigmentation of Kitamura, acromelanosis progressiva and hereditary symmetric dyschromatosis of Dohi.
NFJS is an autosomal dominant ectodermal dysplasia syndrome manifested with poorly developed dermoglyphics, onychodystrophy, abnormal sweating, and palmoplantar keratoderma. It is distinguished by reticulate hyperpigmentation that fades after puberty, and severe dental anomalies leading to early dental loss which does not correlate to the manifestations of our case [4,6].

DKC is an X-linked disease characterized by reticulate hyperpigmentation, nail dystrophy, adermatoglyphia and palmoplantar hyperkeratosis. Other features of DKC mainly include bone marrow failure, oral leukoplakia and abnormal dental findings. However, our patient did not show any of these features thus DKC was excluded [4,6].

Dyschromatosis Universalis Hereditaria is a rare genodermatosis characterized by small irregular hyper-and hypopigmented macules present in early childhood on the trunk and extremities but palms and soles usually are not involved [10]. This dyschromatosis is associated with high-tone deafness that was not present in this case.

Dowling-Degos disease or reticulate pigmented dermatosis of the flexures was also excluded as it presents in adult life where distinct distribution of pigmentation in the flexural areas (groin, axillae and inframammary), facial pits and perioral scars are found [6].

While in reticulate acropigmentation of Kitamura, acromelanosis progressiva and hereditary symmetric dyschromatosis of Dohi, hyperpigmented macules can be observed especially on the dorsal aspect of the hands and feet in a reticular pattern.

Based on previous studies, it is the first time that we report a DPR patient with beard alopecia which suggests that non-scarring alopecia in DPR may include other sites of the body not just the scalp, eyebrows or axillary hair.

Histologically, some distinctive features such as perivascular localization of spindle and epithelioid melanophages with some large forms containing prominent coarse melanin granules were noted in our case.

Unfortunately, there is no specific treatment for DPR but we can manage some cutaneous distress such as palmoplantar keratoderma by keratolytic and topical retinoids.

CONCLUSION

DPR is a very rare disorder usually misdiagnosed but it should be taken into account when seeing the triad of generalized reticulate hyperpigmentation, non-scarring alopecia and onychodystrophy. This case suggests that alopecia in DPR may include other sites of the body not just the scalp, eyebrows or axillary hair. Therefore, correlation with future reported cases will be required.

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Conflict of Interest statement. No conflicts of interest.

ETHICAL APPROVAL

No approval was required.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and the figures related to it.

GUARANTOR

Lama Elias.

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