

Dohi's Reticulated Acropigmentation: Case of a Girl

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Abstract

We present the case of a 7-year-old female patient with hyperpigmented and hypopigmented skin lesions on the back of both hands, asymptomatic, without any relevant history, non-consanguineous parents from Guatemala. Due to the clinical manifestations, the diagnosis of Dohi's reticulated acropigmentation is made, a rare pathology that occurs mainly in people of Japanese descent, although cases have been seen in South America and Europe, despite having autosomal dominant inheritance in our case the patients were a new case; For the year 2015, 47 cases have been reported in the literature.

Keywords: Reticular Pigmentation; Macules; Hyperpigmentation; Hypopigmentation; Girl

Abbreviations

DM: Diabetes Mellitus Type 2; LE: Lupus Erythematosus; ARD: Dohi Reticulate Acropigmentation

Introduction

Dohi's reticular pigmentation, also called hereditary symmetric dyschromia, is an entity that belongs to the skin disorders with hereditary reticular pattern pigmentation. It is characterized by presenting hypopigmented and hyperpigmented macules in acral regions in patients of Japanese descent; unlike other types of reticular pigmentation disorders in which only hyperpigmented macules occur in generalized locations. Cases of autosomal dominant, recessive and sporadic inheritance have been reported [1-3].

Materials and Methods

A 7-year-old female patient, native and resident of Guatemala with a history of presenting dark brown spots on the back of both hands at 8 months of age, punctiform, which increase in number; however, at 3 years old, whitish spots begin to appear in the same

place, affecting arms for what he decides to consult, he does not apply any cream.

Does not suffer from any disease, within the important family history, non-consanguineous parents, third daughter of 3 pregnancies, sister with presence of linear nevus in left arm and brother with joint hypermobility in elbows and arms, mother with rosacea and father with acne, paternal aunt with Lupus erythematosus.

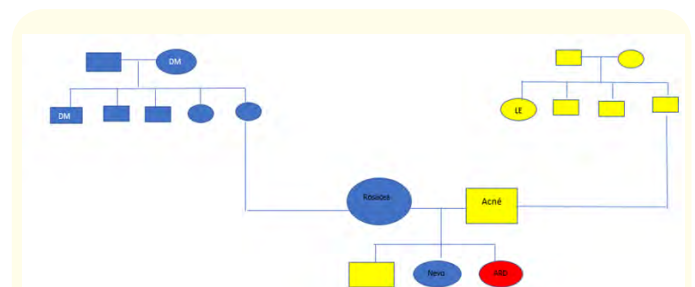


Figure 1: Family tree.

On evaluation, the patient was in good general condition, presenting a chronic dermatosis located on the back of both hands characterized by multiple hypochromic and hyperchromic macules of variable sizes (approximately 0.3 x 0.5 cm), round, smooth surface, defined contours on the basis of normal skin.

On dermoscopic evaluation, mild reticular pigmentation with hyperpigmented points is observed in hyperchromic macules; in hypochromic macules some areas with diffuse light brown pigmentation and others without pigment or adjacent structures.



Figure 2

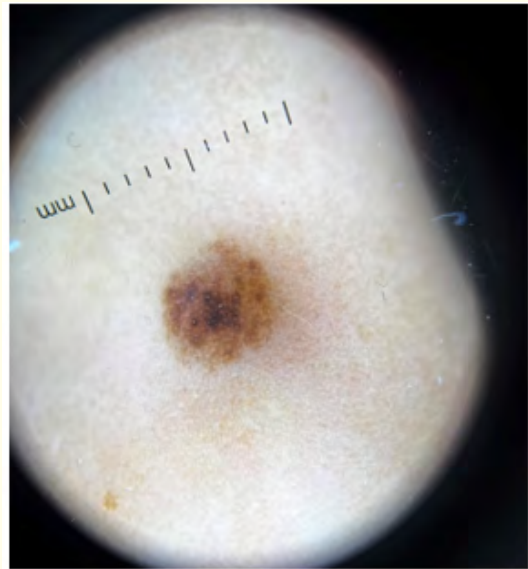


Figure 4



Figure 3

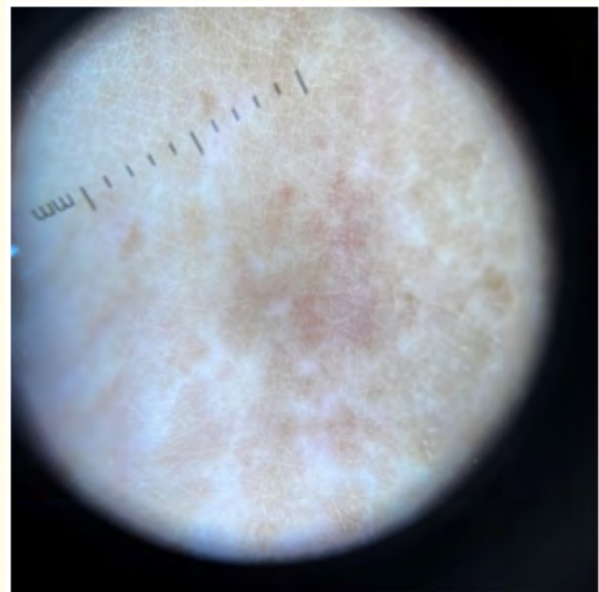


Figure 5

Skin biopsy of a hypopigmented macula reports basket-net stratum corneum, rough epidermis with an adequate number of basal melanocytes without hyperpigmentation, with a small area of spongiosis and mild exocytosis of lymphocytes. In superficial reticular dermis with mild perivascular inflammatory infiltrate by lymphocytes without loss of pigment.

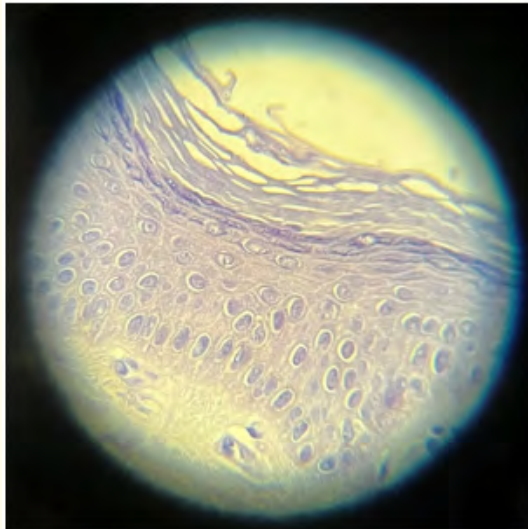


Figure 6

Results and Discussion

Dohi's reticulated acropigmentation is a rare entity, presenting during childhood or early childhood. Most of the cases that have been reported come from Japan, China and Korea, although they have also been described in Asia, Europe and South America with autosomal dominant, recessive and sporadic inheritance. Our patient was not of Asian descent, nor had a family history, which is why it is considered a *de novo* presentation [3,4].

This pathology is due to mutations in the gene that encodes RNA-Adenosine deaminase (ADAR1 or DSRAD) on chromosome 1q21, causing multiple mutations that cause less activity in the enzyme, causing irregular migration of melanoblasts from the neural crest to the skin and irregular in the differentiation of melanosomes during the embryonic period [3].

Clinical manifestations occur before the age of 6 with hyperpigmented and hypopigmented macules that mix to give a reticular pattern, with symmetrical and acral distribution on the

back of the hands and feet. These lesions can extend to the elbows, knees, but usually stop by adolescence. The face is respected although ephelides can be seen. The patient was compatible with the symptoms, however she did not present involvement on the back of her feet and at the facial level ephelides were observed as well as slightly bluish ocular sclera [1,2,4].

The histology of the hyperpigmented areas presents fibroform proliferation of interpapillary ridges with increased melanin in the basal layer with a preserved number of melanocytes, and the hypopigmented areas present a decrease or absence of melanin pigment, without observing differences in the number of melanocytes. The diagnosis is made by clinic; however, a biopsy was performed to rule out other possible entities that affected the number of melanocytes or that presented pigment loss, acantholysis of the subbasal epidermis, epidermal atrophy, pseudocysts corresponding to differential diagnoses such as postinflammatory hyperpigmentation, vitiligo, *incontinentia pigmenti*, *poikiloderma* and other disorders with hereditary reticular pigmentation [3].

There is no specific treatment. The use of PUVA and topical corticosteroids in hyperpigmented macules without success. Partial autografts and laser treatments can improve the cosmetic appearance of some patients [2].

Conclusion

The first case of Dohi reticular acropigmentation in Guatemala was presented, with no family history or Asian descent, presenting the characteristic clinical manifestations of hyperpigmented and hypopigmented macules only on the back of both hands, it is important to make the proper diagnosis, in this way They avoid unnecessary treatments since there is no specific one.

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Conflict of Interest

No conflict of interest exists.

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