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Dohi's Reticulated Acropigmentation

CASE REPORT

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Abstract

Background: Dohi's Reticulated Acropigmentation is a rare autosomal dominant genodermatosis with high penetrance, characterized by small, irregular, hypo and hyperpigmented macules on the dorsal surface of the distal extremities.

Case: The authors report a case of Dohi's Reticulated Acropigmentation in a 61 years old female patient. The patient complained of spots on hands, feet and posteriorly face involvement since she was 7 years old.

Conclusion: Dohi's Reticulated Acropigmentation is a rare clinical condition, which usually appear in childhood and commonly interrupt their onset before adolescence. The diagnosis is based on clinical data, physical examination and histopathological findings. The treatment is unsatisfactory and still no therapy is proposed.

Keywords
Acropigmentation; Dohi; Dermatology.

Introduction

Also called Hereditary Symmetric Dyschromatosis (DSH), Dohi's Reticulated Acropigmentation is a rare autosomal dominant genodermatosis with high penetrance reported initially in the Asian population. It is characterized by small, irregular, hypo and hyperpigmented macules on the dorsal surface of the distal extremities, especially hands and feet. The lesions usually appear in childhood and commonly interrupt their onset before adolescence [1].

About 20% of the patients do not have a family history, being presumed to have spontaneous mutations [2].

Current treatments are unsatisfactory, maintaining depigmentation throughout the patient's life [3].

Case report

Patient is female, 61 years old, from Ananideua-Pará, complains of spots on hands, feet and posteriorly face involvement since she was 7 years old. She reports a similar case presented by her father, paternal uncle, brother and son.

At the dermatological examination, the patient presented hypochromic macules associated with brownish hyperchromic macules with clear borders and irregular contours on the back of hands, feet and face. No other injury on her body.

She had previous diagnosis of vitiligo, which was treated with oral and topical Mamacadela, as well as tacrolimus 0.1% ointment, showing a small clinical response and no areas of re-pigmentation. **(Figure 1)**.

Figure 1: Hypochromic spots associated with brownish macules with a cross-linked pattern at the extremities.

Discussion

First described in 1929, DHS is a rare type of dyschromatosis, characterized by the development of macules on the extremities area and in face 50% of the cases [4]. The gene responsible for autosomal dominant inheritance was first located in the region of chromosome 6q24.2-q25.2 [5].

The diagnosis is based on clinical data, physical examination and histopathological findings [1]:

- **Clinical:** depigmenting lesions with onset in childhood, usually before 6 years old, with gradual increase in size and extension. Family history is present.
- **Physical examination:** hyperchromic and hypochromic macules located at the extremities and face.
- **Histopathology:** hyperpigmentation of basal keratinocytes with a slightly increased or normal number of melanocytes. However, histopathological study is not mandatory for the diagnosis.

Other pigmentary disorders are included in the differential diagnosis and peculiar features help to distinguish these entities, such as [2]:

- **Vitiligo:** presence of only acromic macules, without areas of hyperpigmentation, except in situations of response to treatment.
- **Dowling-Degos disease:** begins in the third decade of life with reticulated hyperpigmentation in areas of flexure. Additional features present with comedo-like lesions on the back area, pitted facial scars and epidermal cysts.
- **Kitamura's reticulated pigmentation:** autosomal dominant inheritance with acral distribution, differentiating from DHS because of the presence of atrophic and depressed lesions interspersed with hyperchromic macules.
- **Hereditary universal discromatosis:** a similar case of hypo and hyperchromic macules, but of universal distribution.

- **Xeroderma pigmentosum:** genodermatosis with autosomal recessive inheritance presenting extreme sensitivity to UV radiation with varying degrees of atrophy, keratosis, hyperpigmentation, neoplasms in exposed areas, as well as ocular and neurological alterations.

Regarding to treatment, it is unsatisfactory and no therapy is proposed.

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Conflict of interest disclosures

The authors declare that there are no conflicts of interest in this case report.

Contribution

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Francisca Regina Oliveira Carneiro: Concept and orientation regarding the manuscript, data acquisition and methodological review of the manuscript.

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