Case for diagnosis

Caso para diagnóstico

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CASE REPORT

A 27-year-old mixed-raced (pardo) female patient presented with flat ovate hypochromic plaques with a rough surface on the back and upper limbs, with an aspect resembling pityriasis versicolor, which she reported having had for three years (Figure 1). She reports family history involving a brother with similar lesions.

The material for histopathology was obtained by punch biopsy, fixed in 10% formaldehyde and stained with hematoxylin-eosin (HE). The test results showed hyperkeratosis without papillomatosis, hypergranulosis and enlarged keratinocytes with basophilic and microvacuolated cytoplasm occupying the upper portion of the spinous layer and granular layer (Figure 2). They also showed mononuclear perivascular infiltrate in the papillary dermis.

After anatomo-clinical correlation, the diagnosis of epidermodysplasia verruciformis pityriasis versicolor like was confirmed. The patient was counseled about the disease, possible treatment and sent for HIV testing.

The patient received treatment with retinoic acid for six months without satisfactory results.

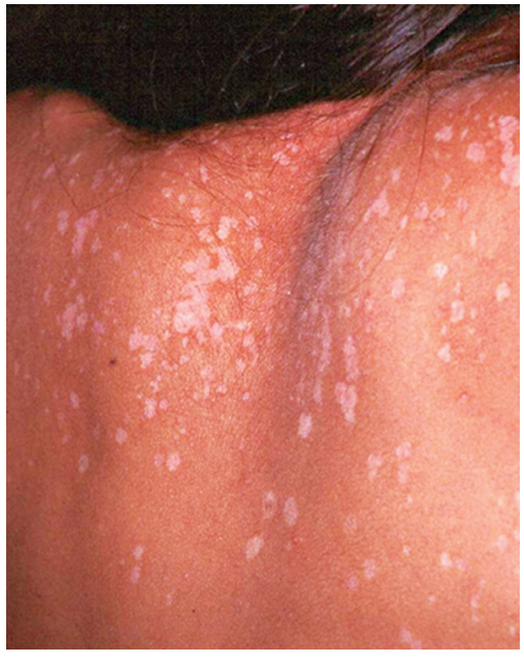

FIGURE 1: Flat ovate hypochromic plaques with a rough surface on the back

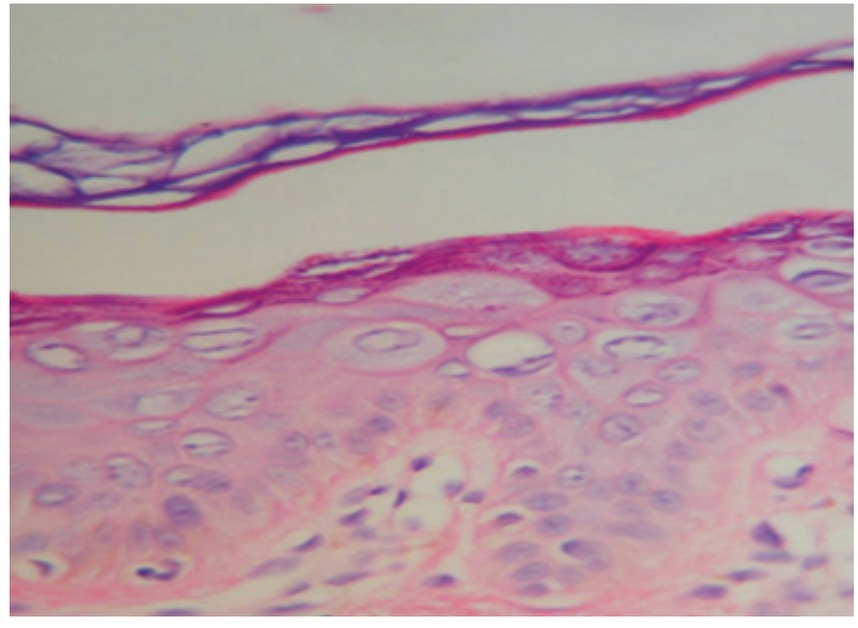

FIGURE 2: Enlarged keratinocytes with basophilic and microvacuolated cytoplasm (HE - 40X)

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What is your diagnosis?

Caso para diagnóstico

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DISCUSSION

Epidermodysplasia verruciformis (EV), originally described by Lewandowsky and Lutz in 1922, is a rare autosomal recessive genodermatosis attributed to infection by specific types of HPV in immunologically deficient individuals. It presents a high familial incidence, especially in children of consanguineous marriages.[3]

Initial manifestations of the disorder most frequently occur in childhood, and clinical manifestation of EV may present only flat warts, associated with non-oncogenic HPV 3 and / or 10, called "benign form". It may also be expressed in a polymorphic way, with a tendency to malignancy associated with multiple Epidermodysplasia Verruciformis Human Papillomavirus (EVHPV), some oncogenic, such as 5 and / or 8, being called "malignant form", which is the most frequently observed. [4,5]

The clinical expression of pityriasis versicolor like, characteristic of the benign form of EV, develops some years after the first lesion. They are initially erythematous macules, but hypochromic in later stages of the disease. [5,6]

When EV is associated with a state of immunosuppression, with HIV being the most prevalent, the main manifestations are hypopigmented lesions of pityriasis versicolor like, with history of antifungal therapy not being rare. Given the rare occurrence of EV in HIV patients, some authors support the hypothesis that EV arises only in those who also have a genetic susceptibility which has not been defined yet. [7]

A totally effective treatment has not been described yet, and EV is considered an extremely resistant disease. Acitretin (oral, 0.5-1 mg / kg / day for 6 months) is the substance that brings the best results. However, the treatment is less successful when associated with HIV, and anti-viral therapy does not appear to significantly affect the course of the disease. [7,8,9] ❑

Abstract: A 27-year-old mixed-raced (pardo) female patient presented with flat ovate hypochromic plaques with a rough surface on the back and upper limbs, with an aspect resembling pityriasis versicolor. She reports family history involving a brother with similar lesions. Lab tests, including anti-HIV, showed no alterations and a histopathological examination showed enlarged keratinocytes with basophilic and microvacuolated cytoplasm occupying the upper portion of the spinous layer and granular layer. After anatomo-clinical correlation, the diagnosis of epidermodysplasia verruciformis pityriasis versicolor like was confirmed.
Keywords: Epidermodysplasia verruciformis; Papillomavirus infections; Pityriasis

Resumo: Paciente do sexo feminino, parda, de 27 anos de idade, apresentando há três anos, placas hipocrômicas planas, ovaladas, de superfície áspera, no dorso e membros superiores com aspecto que lembrava pitiríase versicolor. Refere história familiar de um irmão com lesões semelhantes. Exames laboratoriais, incluindo anti-hiv, sem alterações e com histopatológico evidenciando ceratinócitos aumentados de volume com citoplasma basofílico e microvacuolado ocupando a porção superior da camada espinhosa e a granulosa. Após correlação anatomo-clinica confirmou-se o diagnóstico de epidermodisplasia verruciforme pitiríase versicolor like.
Palavras-chave: Epidermodisplasia verruciforme; Infecções por papillomavirus; Pitiríase

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