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Purpura annularis telangiectodes of Majocchi☆,☆☆

Dear Editor,

Purpura annularis telangiectodes of Majocchi is a rare subtype of pigmented purpuric dermatosis. It is more common in children and young females and predominantly affects the lower limbs, with symmetrical annular reddish-brown macules.[1,2] Little is known of its etiology, which may be associated with viral infections, chronic comorbidities, and use of medications. The diagnosis is clinical and histopathological. There is no consensus regarding treatment. Management is based on reports and case series, with variable response to the proposed treatments.[3,4]

A 6-year-old female patient, daughter of consanguineous parents, presented lesions since 2 years old. The patient had no history of systemic symptoms, allergies, or continued use of medications, except sporadic use of paracetamol. In the beginning, the lesions were erythematous, and subsequently evolved to annular and/or irregular hyperchromic macules, symmetrical in the legs, as well as in the right upper limb, and with an isolated lesion in the anterior cervical region (Fig. 1). A skin biopsy of the right thigh was performed, demonstrating a lichenoid lymphohistiocytic infiltrate in the upper dermis and red blood cell extravasation, as well as foci of lymphocyte exocytosis and perivascular mononuclear infiltrate, without pigmentary incontinence or vasculitis (Fig. 2). Perls' Prussian blue staining indicated the presence of hemosiderin deposition in the papillary dermis (Fig. 3). The patient was screened for hematological, infectious, and rheumatological diseases, all negative. The authors opted for treatment with colchicine orally, with no response after five months of medication use.

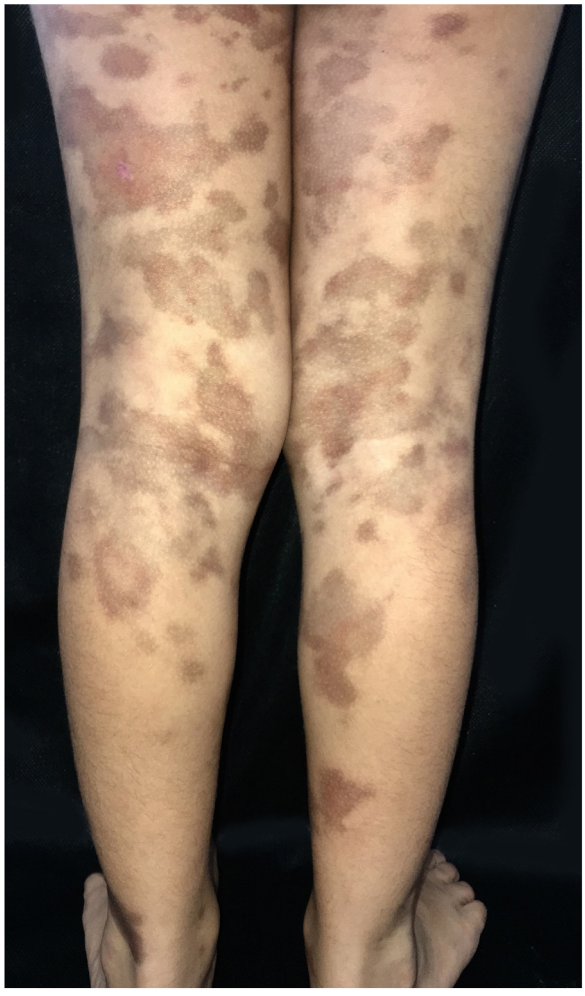

Figure 1 Multiple brownish macules on the legs and thighs.

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☆☆ Study conducted at the Universidade Federal da Bahia, Salvador, BA, Brazil.

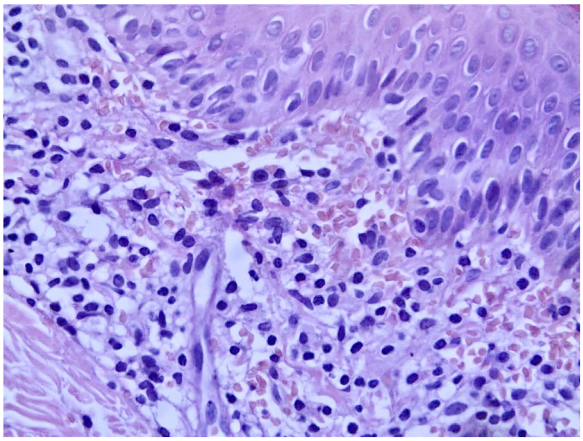

Figure 2 Lymphohistiocytic infiltrate in the upper dermis, as well as extravasation of red blood cells. It is possible to see some lymphocyte exocytosis (Hematoxylin & eosin, ×400).

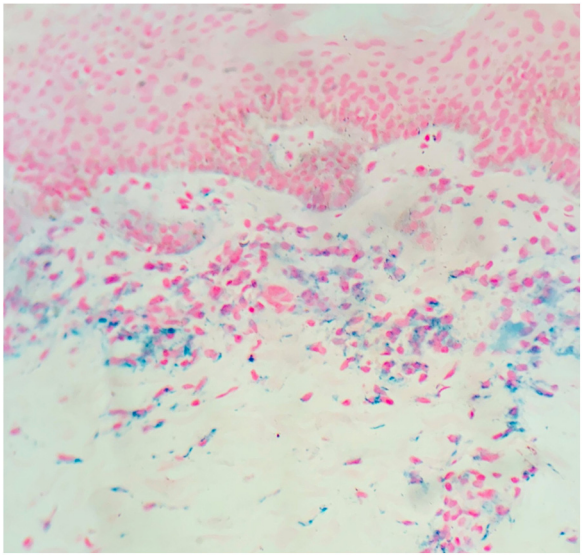

Figure 3 Presence of hemosiderin deposition in the papillary dermis (Perls' Prussian blue, ×200).

Purpura annularis telangiectodes of Majocchi is part of the pigmented purpuric dermatoses, which manifests as annular macules that are symmetrical, reddish-brown, and generally asymptomatic. It preferably affects children and young women, and there is no predominant ethnicity.[1–4] Commonly, it appears in the lower limbs; there appears to be an orthostatic component in the pathophysiology of the disease, as described in the present case. The etiology of pigmented purpuric dermatoses is not yet fully elucidated, and its triggers are not always detected; therefore, the etiology is idiopathic in most cases. An association with comorbidities such as diabetes mellitus, viral hepatitis, peripheral venous insufficiency, and use of certain medications has been reported, including the following: paracetamol, aspirin, carbamazepine, antihypertensives, infliximab, alpha-interferon, pseudoephedrine, raloxifene, and thiamine.[1,4] The present patient did not have any of the aforementioned comorbidities; the sporadic use of paracetamol was the only possible trigger identified in the clinical history. It is worth mentioning that most cases are idiopathic. The administration of paracetamol, even if occasional, may have been a trigger for the clinical picture as described by Kwon et al.[5] The diagnosis of purpura annularis telangiectodes of Majocchi is clinical and histopathological. The clinical characteristics are closely related to the anatomopathological findings. Lesion pigmentation is due to the extravasation of red blood cells and deposition of hemosiderin seen in the papillary dermis. It is also possible to observe perivascular inflammatory infiltrate composed of lymphocytes, histiocytes, and Langerhans cells. These findings corroborate the possible association of pigmented purpuric dermatoses with cellular immunity, contributing to capillary fragility. Humoral immunity also appears to play a role, which can be evidenced by direct immunofluorescence, which can detect perivascular deposits of immunoglobulins and complement. Foci of lymphocyte exocytosis may occur, as observed in this case. Leukocytoclastic vasculitis or epidermotropism are not observed.[3,4] The knowledge of this entity and its early diagnosis can allow the assessment of the presence of triggers and guide its management.

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Authors' contributions

Aline Soares Garcez: Elaboration and writing of the manuscript; critical review of the literature.

Vitória Regina Pedreira de Almeida Rego: Approval of the final version of the manuscript; critical review of the literature.

Thadeu Santos Silva: Approval of the final version of the manuscript; elaboration and writing of the manuscript; intellectual participation in propaedeutic and/or therapeutic conduct of studied cases; critical review of the literature, critical review of the manuscript.

Conflicts of interest

None declared.

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Necrolytic migratory erythema associated with painful plantar keratoderma. A new diagnostic clue for this paraneoplastic syndrome?☆,☆☆

Dear Editor,

The presence of recurrent skin lesion outbreaks in intertriginous areas and lower extremities, during years of evolution, may be a presenting form of necrolytic migratory erythema (NME), which is a paraneoplastic skin disease that is associated with malignant glucagonoma in 90% of cases.[1]

This report presents the case of a 59-year-old woman with a history of keratoconjunctivitis, asthenia, and constipation. She was referred to evaluate recurrent skin lesions with four years of evolution. She was affected by erythematous, scaly plaques and hyperpigmentation in the legs, gluteal area, groin area, thighs, and elbows, with no associated systemic symptoms (Fig. 1). She provided several skin biopsies, which had been diagnosed as toxicoderma and eczema, but the patient denied taking drugs. A new biopsy showed an epidermis with a marked pale central area due to the presence of apoptotic keratinocytes of vacuolated appearance below a layer of extensive hyperkeratosis and parakeratosis (Fig. 2). This gave a ''tricolor flag'' image, which is suggestive of vitamin deficiency. In addition, a deficit of zinc and fatty acids was observed, and, after starting vitamin and zinc supplements, the patient remained asymptomatic for eight months. Nevertheless, she subsequently presented more severe outbreaks with blisters, edema, and scaling predominantly on the dorsum of the feet, associated with a very painful plantar keratoderma (Fig. 3).

With the suspicion of NME and high levels of glucagon, an abdominal CT scan was made showing a 4 cm mass in the pancreas, and the presence of a malignant glucagonoma was confirmed by pathological anatomy. The skin lesions completely resolved after tumor resection. However, one year later she developed liver metastasis with no recurrence of skin lesions.

Although malignant glucagonoma may be accompanied by systemic symptoms such as diarrhea, weight loss, newly developed diabetes, normocytic anemia, zinc deficiency, or fatty acid or amino acid deficiency, *etc.*, NME may be the first and only glucagonoma symptom.[2,3] The pathogenesis of NME can be explained due to the fact that hyperglucagonemia stimulates hepatocyte gluconeogenesis and lipolysis leading to hypoaminoacidemia. Liver dysfunction results in decreased albumin, which is the main carrier of zinc and fatty acids, and thus contributes to fatty acid and zinc deficiency. Glucagon also causes vitamin B and nutrient deficiencies, such as zinc deficiency, which may contribute to increased levels of arachidonic acid, prostaglandins, and leukotrienes, and thus predisposing individuals to develop inflammatory skin lesions, such as NME, and the classic epidermal necrolysis seen in the histology.[1] The evolution of skin lesions in outbreaks, which are sometimes self-healing, the nonspecific histology in some cases, and the long evolution of the lesions are the reason for the diagnostic delay.[3] It should be noted that the ''tricolor flag'' histological image is associated not only with skin lesions due to nutritional deficits and acral necrolytic erythema, but also with the advanced cutaneous lesions of NME.[4] It is also important to highlight the presence of a painful plantar keratoderma, which was not found to be associated with this syndrome in the literature, and which was completely resolved with the removal of the neoplasm. Even though malignant glucagonoma is a slow-growing tumor, more than 50% of the cases at diagnosis already have metastatic involvement.[5] For this reason, it is crucial to highlight the importance of an early diagnosis of this clinical presentation, which can lead to preventing the appearance of metastases leading to a worse prognosis. In conclusion, this report describes patient with recurrent skin lesions,

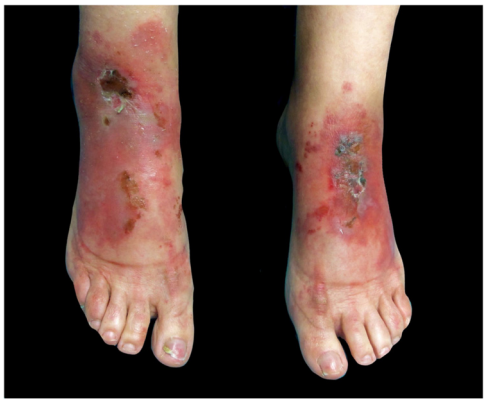

Figure 1 Erythematous, scaly, erosive, and crusty lesions on the dorsum of the feet.

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