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CASE REPORT

Acquired reactive perforating collagenosis in association with hepatorenal failure: a case report and literature review

Colagenose perfurante reativa adquirida em associação com falência hepatorrenal: um relato de caso e revisão de literatura

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Abstract

Reactive perforating collagenosis is a rare perforating dermatosis characterized by transepidermal elimination of collagen. We report a 63-year-old male with diabetes, hypertension, chronic kidney disease, and cirrhosis from hepatitis B, presenting with keratotic, pruritic papules on the back. Histopathological examination showed a crater filled with a keratin plug and transepidermal elimination of collagen, confirming the diagnosis of acquired perforating dermatosis (APD). APD is associated with systemic diseases, mainly diabetes, and diagnosis depends on the clinicopathological correlation of the lesions. This case highlights the importance of early diagnosis to improve the patient's quality of life and demonstrates an atypical distribution/site of involvement.

Keywords: Perforating dermatosis. Pruritus. Systemic diseases.

Resumo

A colagenose perfurante reativa (CPR) é uma dermatose perfurante rara, marcada pela eliminação transepidérmica de colágeno. Relatamos um homem de 63 anos com diabetes, hipertensão, insuficiência renal crônica e cirrose por hepatite B, com pápulas ceratósicas e pruriginosas no dorso. O exame histopatológico evidenciou cratera preenchida por tampão córneo e feixes de colágeno em extrusão, confirmando o diagnóstico de dermatose perfurante adquirida (DPA). A DPA associa-se a doenças sistêmicas, principalmente ao diabetes, e o diagnóstico depende da correlação clínico-histopatológica das lesões. Este caso ressalta a importância do diagnóstico precoce para melhorar a qualidade de vida do paciente e evidencia uma localização atípica da doença.

Palavras-chave: Dermatose perfurante. Prurido. Doenças sistêmicas.

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Introduction

Reactive perforating collagenosis (RPC) is a rare cutaneous condition within the group of perforating dermatoses, characterized by transepidermal elimination of altered dermal material, such as collagen or follicular components.[1,2] Historically, RPC has been classified into a rare hereditary form presenting during childhood and an acquired form predominantly associated with diabetes mellitus and chronic kidney disease. However, owing to the considerable clinicopathological overlap among the perforating dermatoses observed in adults, Rapini et al. proposed the term acquired perforating dermatosis (APD) in 1984 to encompass acquired perforating disorders associated with systemic diseases. Currently, many authors reserve the term RPC for the hereditary form and use APD to describe adult-onset acquired cases. However, the terminology remains inconsistent throughout the literature.[1-3] The acquired form affects both sexes equally and shows a higher prevalence, occurring in approximately 10% of patients undergoing hemodialysis for renal failure.[1,2]

Case report

A 63-year-old male patient with a history of diabetes mellitus, arterial hypertension, chronic kidney disease secondary to diabetic nephropathy, and liver cirrhosis due to hepatitis B virus infection was evaluated. During hospitalization for clinical decompensation of his comorbidities, a dermatological consultation was requested because of the appearance, approximately 1 year earlier, of pruritic lesions on the back, whose onset coincided with the diagnosis of liver cirrhosis.

On dermatological examination, hyperpigmented, violaceous, keratotic, polygonal papules were observed, some of which were ulcerated and occasionally coalescing to form linear plaques on the upper back region (Fig. 1).

A skin biopsy was performed, and the main differential diagnoses included lichen planus, uremic prurigo, Besnier's prurigo, drug eruption, and acneiform eruption. Histopathological examination revealed marginal epithelium with mild acanthosis and a large area of continuity of the papillary dermis, forming a crateriform depression filled with a keratin plug, cellular debris, and neutrophils. At the base of the crusted ulceration, vertically oriented perforating collagen bundles in extrusion were identified, confirming the diagnosis of RPC, which was classified as the acquired form, in correlation with the clinical findings (Figs. 2 and 3).

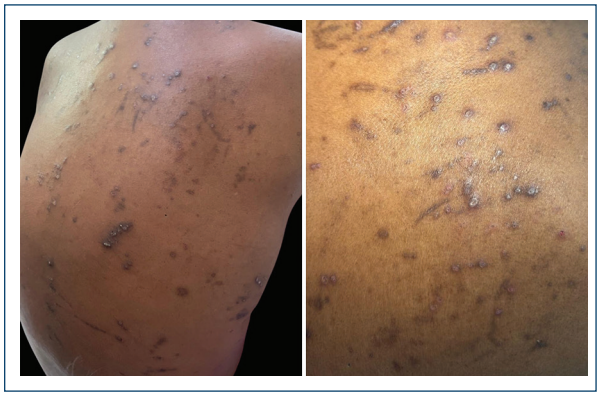

Figure 1. Clinical presentation of the lesion on the dorsal region.

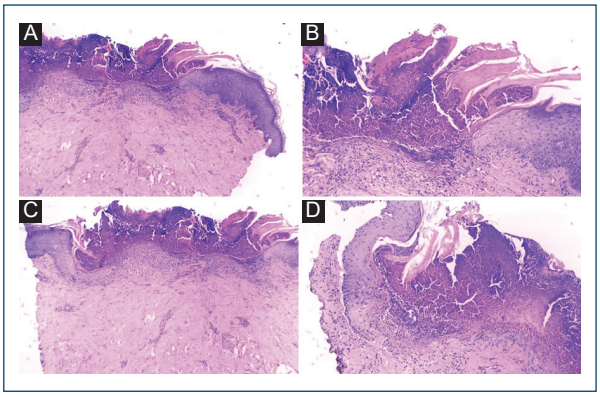

Figure 2. Histopathological findings with hematoxylin and eosin staining, shown at **A**: ×100, **B**: ×200, **C**: ×100 and **D**: ×200 magnification, demonstrating a crateriform depression filled with a keratin plug and, at the base of the crusted ulceration, vertically oriented perforating collagen bundles in extrusion.

Management was primarily directed toward symptomatic relief of pruritus and optimization of the patient's underlying systemic comorbidities through a multidisciplinary approach. Treatment included topical betamethasone dipropionate 0.1% cream applied twice daily for 7 days, and the patient was referred for evaluation for narrowband ultraviolet B (UVB) phototherapy. However, owing to the severity of the underlying systemic diseases, particularly advanced chronic kidney disease, the patient died before initiation of phototherapy and before the prescribed treatment could be fully implemented.

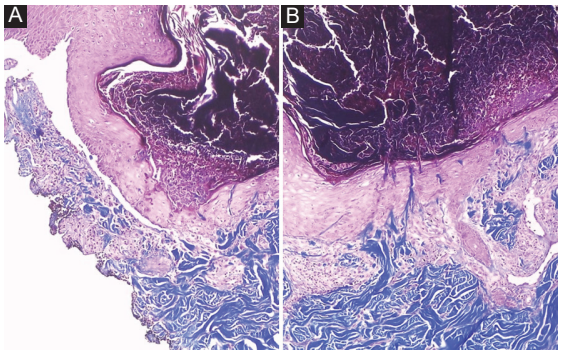

Figure 3. Histopathological examination with Masson's trichrome staining, highlighting transepidermal elimination of collagen (**A** and **B:** ×200).

Discussion

RPC can be classified into hereditary or acquired variants. The hereditary form is rarer, inherited in an autosomal recessive pattern, and typically manifests in early childhood. In contrast, the acquired form is more commonly associated with systemic diseases and, unlike the hereditary variant, usually has its onset in adulthood.[2,4]

In APD, lesions are generally widespread but may present as papules or nodules that evolve into crateriform ulcerations covered by a keratotic plug, accompanied by intense pruritus.[4,5] According to the literature, the lower extremities and the trunk are the most commonly affected sites; however, in the present case, lesions were confined exclusively to the patient's back.[4-6] In addition, new lesions are prone to develop due to the Koebner phenomenon following excoriations caused by intense pruritus.[4]

It is virtually impossible to clinically distinguish APD from other perforating dermatoses solely on lesion morphology; however, histopathological examination reveals characteristic findings that aid in diagnosing APD.[4,5] The main histopathological feature of APD is transepidermal elimination of degenerated collagen, frequently associated with a chronic inflammatory infiltrate and local vascular alterations.[5] Masson's trichrome staining is used to highlight the eliminated collagen fibers.[5]

Studies such as those by Yazdi et al. suggest considering a possible association between APD and other cutaneous neoplasms, proposing this dermatosis as a paraneoplastic syndrome.[4,7] However, although most reported cases of APD have been preceded by the diagnosis of a malignant neoplasm, such as Hodgkin lymphoma or leukemia, the patient described in the present case showed no evidence of oncologic disease.[4,7] In contrast, the association between APD, diabetes mellitus, and chronic kidney disease is well established, with the trunk and lower extremities representing the most commonly affected anatomical sites. However, in the present case, lesions were exclusively distributed on the back.

There is no standardized treatment or therapeutic consensus for APD, and management varies according to the extent and severity of the lesions and the patient's overall clinical condition.[4] The current literature consists predominantly of case reports and small case series, with a lack of controlled clinical trials and established management guidelines. Reported therapeutic options include topical corticosteroids, emollients, UVB phototherapy, systemic retinoids, allopurinol, and cryotherapy for the treatment of perforating dermatoses and associated severe pruritus.[4,8,9] Nevertheless, the available evidence remains limited, and no standard treatment has been established to date.

APD is a rare cutaneous condition frequently associated with systemic diseases such as diabetes mellitus and chronic renal failure, and more rarely with hepatic disorders. The absence of associated neoplasms in this case and the finding of lesions restricted to the back underscore the heterogeneity of clinical presentations and the importance of clinicopathological correlation for confirmation of the diagnosis of APD. Therefore, the present case reinforces the notion that, although rare, APD should be considered in patients with multiple metabolic comorbidities, as early diagnosis and appropriate management of associated factors may contribute to pruritus control and improve quality of life.

Conclusion

This case report describes a perforating dermatosis in an unusual location, associated with hepatic failure, an association that is poorly documented in the literature. This case highlights the importance of early recognition of this entity to establish optimal lesion management and improve patients' quality of life. Furthermore, it suggests that acquired RPC should be considered among the differential diagnoses of lesions affecting the dorsal region, particularly in the presence of hepatorenal failure or other hepatic disorders.

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Conflicts of interest

None.

Ethical considerations

Protection of human subjects and animals. The authors declare that no experiments on humans or animals were performed for this research.

Confidentiality, informed consent, and ethical approval. The authors have followed their institution's confidentiality protocols, obtained informed consent from all patients, and secured approval from the Ethics Committee. SAGER guidelines have been followed as applicable to the nature of the study.

Declaration on the use of artificial intelligence. The authors declare that no generative artificial intelligence was used in the writing or creation of the content of this manuscript.

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