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Case Report

ANCA-negative small vessel vasculitis: a case report in a 41-year-old male

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ABSTRACT

Non-leukocytoclastic small vessel vasculitis is an uncommon inflammatory disorder characterized by vascular wall damage without evidence of leukocytoclasia. We present the case of a 41-year-old male with ANCA-negative small vessel vasculitis limited to the skin. The patient developed pruritic violaceous macules on the lower limbs and showed histopathologic findings of small-vessel vasculitis with neutrophilic infiltrate. Treatment with systemic corticosteroids resulted in favorable clinical evolution without recurrence. This case highlights the importance of comprehensive clinical assessment and individualized management in cutaneous vasculitis, emphasizing clinical judgment over immunological markers.

Keywords: Small vessel vasculitis, Cutaneous vasculitis, Prednisone therapy

INTRODUCTION

Vasculitis is defined as damage and inflammation of the vascular wall, regardless of the caliber (small, medium, or large) and type of the affected vessel (arteries, veins, or both). It may be idiopathic or secondary to infections, drugs, neoplasms, or systemic inflammatory diseases.[1,2] Cutaneous involvement typically presents as purpuric macules or papules, most commonly on the lower limbs, probably due to hemodynamic factors.[3-5]

This case aims to emphasize the importance of comprehensive management, highlighting the value of clinical assessment over immunological markers in a case of non-leukocytoclastic vasculitis.

CASE REPORT

A 41-year-old male presented with dermatosis on the lower limbs. His mother had a history of rheumatoid arthritis. The patient was a chronic smoker, consuming 5–6 cigarettes per day. One month before medical evaluation, he developed pruritus accompanied by erythematous-violaceous pruritic macular lesions on the distal lower extremities, dorsum of the feet, and bilateral malleolar areas, initially attributed to pork ham consumption. Two weeks later, he experienced burning pain in the toes with intense pruritus on the contralateral limb, prompting consultation at our institution.

Physical examination

Lower limbs presented multiple residual violaceous lesions (<0.5 cm) that did not blanch with digital pressure, associated with hemorrhagic crusts over the bimaleolar region, dorsum of the feet, and anterior tibial surface, bilaterally. Mild edema was present without tenderness on palpation (Figures 1 and 2).

Histopathology

A punch biopsy revealed small-vessel vasculitis characterized by neutrophilic infiltrate without leukocytoclasia (Figure 3).

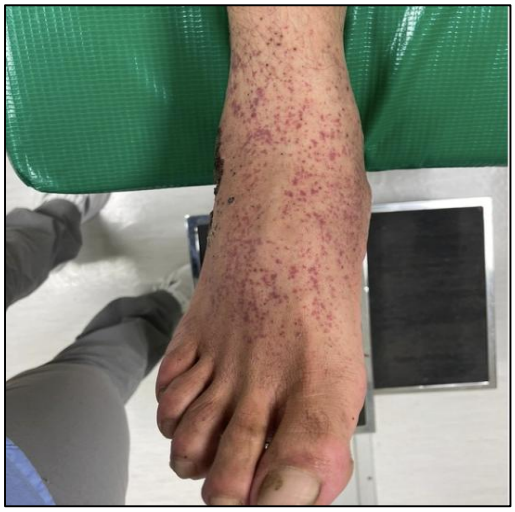

Figure 1: Violaceous lesions on the dorsal surface of the foot.

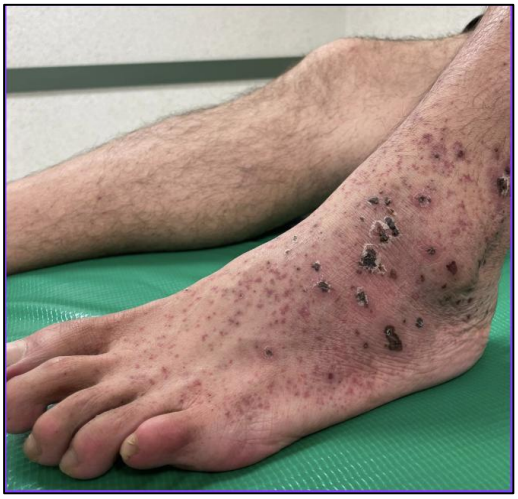

Figure 2: Violaceous lesions with haemorrhagic crusts on the bimalleolar region, dorsum of the feet, and anterior tibial surface.

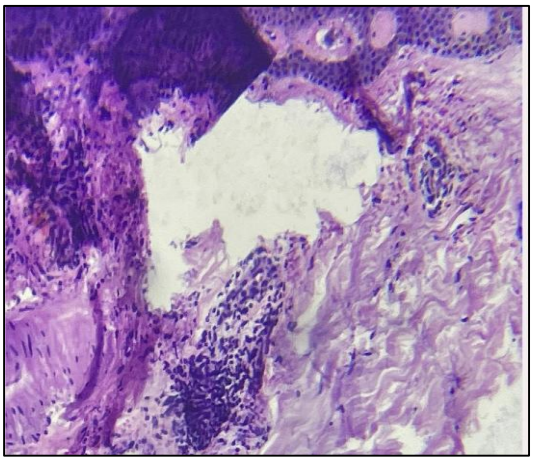

Figure 3: Skin biopsy.

Treatment and follow-up

Prednisone 0.5 mg/kg/day (45 mg) was initiated with a weekly taper of 5 mg until reaching 7.5 mg. Calcitriol 0.25 µg, two tablets every 24 hours, was also prescribed. On the fourth day of hospitalization, the patient was discharged with outpatient follow-up. Two months after discharge, prednisone was continued at 7.5 mg daily for one week, 5 mg daily for one week, and then half a tablet daily for one week; calcitriol was discontinued. The biopsy site healed without complications. Rheumatology follow-up confirmed clinical findings consistent with small-vessel vasculitis limited to the skin. At two-month reevaluation, the patient presented residual hyperpigmentation without active lesions.

Table 1: Laboratory results.

Test	Result
White blood cells	$8.94\times10^3/\mu l$
Erythrocyte sedimentation rate (ESR)	40 mm/hr
C-reactive protein (CRP)	23.3 mg/l
ANA	Negative
Anti-dsDNA	Negative
PR3-ANCA / MPO-ANCA	Negative

DISCUSSION

Cutaneous small-vessel vasculitis (CSVV) is a vasculitic disorder characterized by inflammation limited to the skin and histopathologic evidence of leukocytoclastic vasculitis involving capillaries, venules, and arterioles. Palpable purpura is the most frequent clinical manifestation.[6,7] Idiopathic CSVV is defined when no underlying cause can be identified.[8] Recent literature supports prednisone as the first-line systemic therapy over other immunomodulatory treatments. Prednisone at a dose of 0.5 mg/kg/day (based on ideal body weight) is recommended until new lesion formation ceases (typically within one to two weeks), followed by gradual tapering over three to six weeks. The potential for serious adverse effects limits the long-term use of systemic glucocorticoids. For patients who relapse during tapering or fail to respond adequately to prednisone, colchicine or dapsone are suggested as alternative treatments.[5,7-10]

CONCLUSION

This case underscores the relevance of clinical evaluation in diagnosing and managing non-leukocytoclastic small-vessel vasculitis. Comprehensive assessment, appropriate histopathologic correlation, and individualized corticosteroid therapy remain essential for optimal outcomes.

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