

## Palpable and Isolated Purpura Revealing Granulomatosis with Polyangiitis

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### Abstract

### Case Report

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a rare autoimmune vasculitis that can follow a rapidly progressive and potentially fatal course. Cutaneous signs complete the symptomatic presentation of the disease. We report a case of recurrent cutaneous leukocytoclastic (vascular) purpura that led to the diagnosis of the disease.

**Keywords:** Purpura, granulomatosis, vasculitis, ANCA.

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## INTRODUCTION

Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitides are necrotizing vasculitides affecting small-caliber vessels, encompassing granulomatosis with polyangiitis, microscopic polyangiitis, and eosinophilic granulomatosis with polyangiitis. Cutaneous involvement is frequent and may be the presenting manifestation of the disease. We report an original observation of vascular purpura occurring as the inaugural clinical feature of granulomatosis with polyangiitis [1-3].

## CASE REPORT

A 50-year-old male patient with no significant past medical history presented with a 4-month history of

diffuse petechial purpuric lesions without other associated clinical signs. Skin biopsy revealed evidence of purpura without identifiable vasculitis lesions. ANCA testing was positive for c-ANCA. Otolaryngologic (ENT) examination was unremarkable. Laboratory workup revealed a systemic inflammatory syndrome, urinalysis showed microscopic hematuria, and chest X-ray demonstrated diffuse reticulonodular infiltrates. The diagnosis of GPA was retained based on the presence of two ACR criteria: hematuria and abnormal chest radiograph. Treatment with systemic corticosteroids combined with intravenous cyclophosphamide pulses was initiated. Outcome was marked by complete resolution of the purpura after three weeks.



Figures 1 and 2: Palpable purpura of the trunk and lower limbs

## DISCUSSION

Granulomatosis with polyangiitis is a granulomatous vasculitis distinguished from other vasculitides by predominant ear, nose, and throat (ENT) and pulmonary involvement (present in 80% to 100% of patients), as well as the presence of cytoplasmic antineutrophil cytoplasmic antibodies (c-ANCA) targeting proteinase 3 (PR3). Cutaneous manifestations are highly variable, occurring in 10% to 50% of cases, and may present at onset or during the course of the disease. Diagnostic delay ranges from 9 months to 18 years. Vascular purpura is the most common cutaneous feature and can occasionally reveal the underlying disease. The combination of glucocorticoids with an immunosuppressive or immunomodulatory agent remains the standard of care for GPA. Disease course is characterized by frequent relapses, with half of patients relapsing within 5 years of the initial flare [1] [2] [3].

## CONCLUSION

Granulomatosis with polyangiitis (GPA) is a rare systemic vasculitis characterized by involvement of the upper respiratory tract, lungs, and kidneys; however,

cutaneous lesions may serve as the presenting sign of the disease.

**Conflicts of Interest:** The authors declare no conflicts of interest regarding this work.

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