SKIN

BRIEF ARTICLE

# An Atypical Case of Granulomatosis with Polyangiitis with Cutaneous Features in a 19-year-old Female

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## INTRODUCTION

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a rare ANCA-associated, necrotizing vasculitis of the medium and small vessels. The annual incidence is 10-20 cases per million globally and 3 per million in the U.S. with an average age of onset of 64-75 years. It is most common among Northern European Caucasians.[1] Although the pulmonary and renal systems are most commonly impacted, GPA manifests with cutaneous findings in approximately 34% of patients. Skin lesions may be the initial presenting finding in about 13% of patients, however they do not typically manifest until 12-15 months of disease onset.[2] The most common skin finding is palpable purpura involving the lower extremities (16%), followed by painful subcutaneous nodules(9.4%), maculopapular rash (6.7%), and papulonecrotic lesions that typically affect the face, scalp, and extremities(4.76%).[3,4]

## CASE REPORT

Here we discuss a case of a 19-year-old hispanic female who presented to our consult service with painful sanguineous skin lesions on the head, neck, arms, axilla, and back that had begun 3 weeks prior to admission. Associated symptoms included fevers, palpitations, excessive sweating, arthralgias, productive cough with hemoptysis, nasal discharge, ocular pain, and a 20-lb weight loss over 6 months. The patient had been recently diagnosed with hyperthyroidism and managed on propranolol prior to admission. On examination, the patient presented with 1-3mm punched-out-appearing, tender and hemorrhagic ulcerations and subcutaneous nodules on the cheeks, jaw, neck, upper arms, axilla, and back (**Figure 1**). A 4mm punch biopsy of a lesion on the upper back was sent for H&E, and lab work, imaging studies, and serologies were ordered.

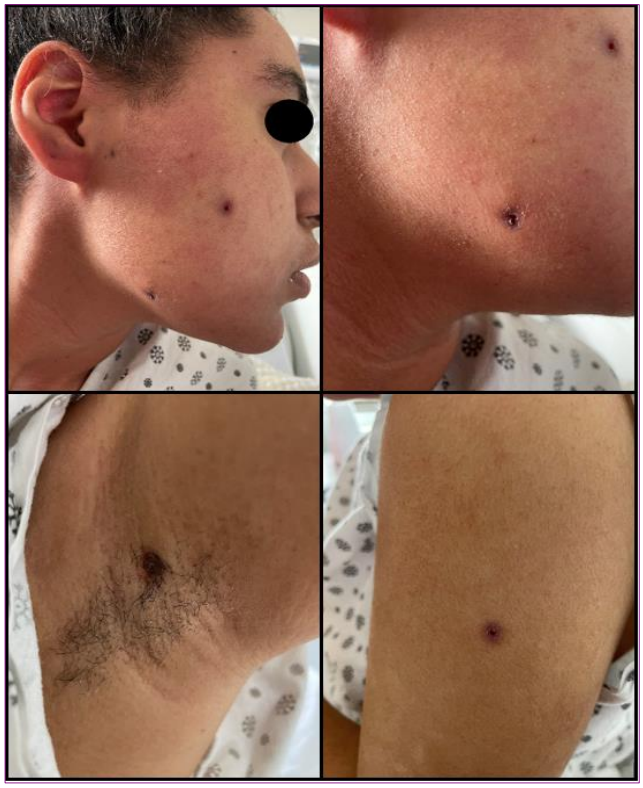

**Figure 1.** Punched out ulcerations on face (top left and right), axilla (bottom left), and arm (bottom right).

Histopathology demonstrated granulomatous dermatitis with extravasated erythrocytes, fibrin, and suppurative inflammation. C-ANCA/anti-PR3 were positive and p-ANCA, myeloperoxidase, and anti-GBM were negative (**Figure 2**). H&E of a biopsy performed on the intranasal wall demonstrated granulation tissue with an admixture of neutrophils, plasma cells, lymphocytes, eosinophils, and multinucleated giant cells (Figure 2). Chest CT demonstrated pulmonary cavitations and sinus CT showed signs of multiple resections, however the patient had not had any prior sinus procedures. In addition, the patient had a low TSH and free T4, high TSH receptor antibody, and a hypervascular thyroid on ultrasound.

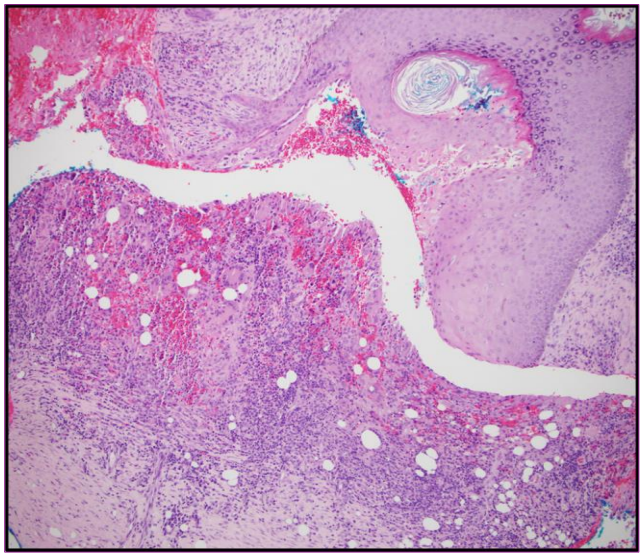

**Figure 2.** H&E shows granulomatous dermatitis with extravasated erythrocytes, associated fibrin, and suppurative inflammation.

## DISCUSSION

Although GPA is a rare condition, cutaneous lesions present in only a minority of cases and typically on the lower extremities. This case demonstrated further atypicality as cutaneous findings were present above the trunk only and materialized fairly early in the disease process. In addition, the patient was 19 years old, significantly below peak incidence, non-Caucasian, and had an accompanying thyroiditis. Hyperthyroidism is an extremely rare concomitant finding in patients with GPA, described only in few case reports, with an estimated prevalence of about 1%. In addition, thyroid disease is more common in p-ANCA vasculitides rather than c-ANCA.[5] In this case, the patient presented with concomitant thyrotoxicosis.

## CONCLUSION

Although uncommon, cutaneous lesions can be among the initial prominent findings in patients with atypical presentations of GPA, and thus may serve as a crucial tool in

diagnosing this systemic and often fatal disease.

**Conflict of Interest Disclosures:** None

**Funding:** None

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**References:**

1. E. Ntatsaki, R. A. Watts, and D. G. I. Scott, "Epidemiology of ANCA-associated vasculitis," *Rheum. Dis. Clin. North Am.*, vol. 36, no. 3, pp. 447–461, Aug. 2010.
2. L. E. Gibson, U. Specks, and H. Homburger, "Clinical utility of ANCA tests for the dermatologist," *Int. J. Dermatol.*, vol. 42, no. 11, Nov. 2003, doi: 10.1046/j.1365-4362.2003.01914.x.
3. G. Ragab, M. T. Hegazy, M. Ali, M. R. E. Abdel-Halim, and X. Puéchal, "Three Patterns of Cutaneous Involvement in Granulomatosis with Polyangiitis," *Journal of Advanced Research*, vol. 24, p. 311, Jul. 2020.
4. R. Micheletti, Z. Chiesa, A. Craven, R. Watts, R. Luqmani, and P. Merkel, "LB943 Cutaneous manifestations of ANCA-associated vasculitis are common, varied, and associated with systemic disease," *Journal of Investigative Dermatology*, vol. 137, no. 10. p. B3, 2017. doi: 10.1016/j.jid.2017.07.016.
5. S. Lionaki *et al.*, "Association between thyroid disease and its treatment with ANCA small-vessel vasculitis: a case–control study," *Nephrol. Dial. Transplant*, vol. 22, no. 12, pp. 3508–3515, Aug. 2007.