

Vasculitis with granulomatosis: An atypical presentation of giant cell arteritis



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INTRODUCTION

Giant cell arteritis (GCA) is a large- and medium-vessel granulomatous vasculitis that affects the aorta and its major branches, particularly the extracranial branches of the carotid artery.¹⁻³ GCA occurs primarily in those over 50 years of age and classically manifests as fevers, malaise, headaches, a temporal artery abnormality, jaw claudication, and visual disturbance.¹ The American College of Rheumatology criteria for the diagnosis of GCA require at least 3 of the following: age >50 years, new-onset headaches, temporal artery tenderness, an erythrocyte sedimentation rate of ≥ 50 mm/hour, and an arterial biopsy demonstrating vasculitis with a mononuclear cell infiltrate or granulomatous inflammation. However, the American College of Rheumatology criteria have poor diagnostic sensitivity for vasculitides, which is likely because of the development of newer imaging techniques and antibody testing methods.⁴

Cutaneous manifestations occur infrequently in patients with GCA; however, the skin over the affected artery may be tender, erythematous, edematous, or purpuric, with induration of the artery and a diminished pulse.^{1,5} Other cutaneous manifestations are categorized as ischemic due to arterial occlusion or nonischemic because of a variety of mechanisms that are poorly understood.¹ We report a case of GCA presenting with atypical systemic and cutaneous manifestations.

Abbreviations used:

GCA:	giant cell arteritis
PET-CT:	positron emission tomography computer tomography
TA:	Takayasu arteritis

CASE REPORT

A 60-year-old woman was admitted to the hospital with a 10-day history of quotidian fevers, myalgias, and generalized weakness, which had started in the lower extremities. The severity of her weakness had resulted in the inability to ambulate. An asymptomatic rash developed in the patient 5 days after the onset of these symptoms, which consisted of violaceous, nonblanchable indurated papules and plaques on the anterior aspects of the scalp, chest, and upper extremities (Figs 1 and 2). The temporal artery was nontender to palpation; however, the patient complained of a brief bilateral headache, without jaw claudication or visual changes. On admission, she was febrile (38.2 °C), with tachycardia, leukocytosis ($20.83 \times 10^3/\mu\text{L}$; range, $3.7 \times 10^3/\mu\text{L}$ to $8.4 \times 10^3/\mu\text{L}$), an elevated erythrocyte sedimentation rate (65 mm/hour; range, 0-30 mm/hour), an elevated C-reactive protein level (65 mg/L; range, >3.0 mg/L), and a normal creatine kinase level (84 IU/L; range, 26-192 IU/L). Electromyography demonstrated type 2 muscle atrophy. Urinalysis; blood cultures; tests for antinuclear antibody, antineutrophilic cytoplasmic antibody, rapid plasma reagin, HIV, viral hepatitis, cytomegalovirus,

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Fig 1. The image shows giant cell arteritis: indurated violaceous plaques on the anterior aspect of the scalp.



Fig 2. The image shows giant cell arteritis: indurated violaceous papules on the upper part of the chest.

Epstein-Barr virus, COVID-19, and tuberculosis; and flow cytometry yielded unremarkable results.

Punch biopsy of a lesion on the right arm revealed a robust neutrophilic infiltrate with a granulomatous component involving the vessel walls of medium-sized vessels of the subcutaneous tissue (Fig 3). Skin tissue cultures were negative for aerobic, fungal, and acid-fast microorganisms.

Positron emission tomography-computed tomography (PET-CT) was performed, which demonstrated uptake and mild thickening of the infrarenal abdominal aorta and uptake along the course of the iliac vessels. The diagnosis of atypical GCA was made based on the patient's age, elevated erythrocyte sedimentation rate, and histologic and PET-CT findings. The patient rapidly improved after starting 40 mg of oral prednisone per day. She regained the

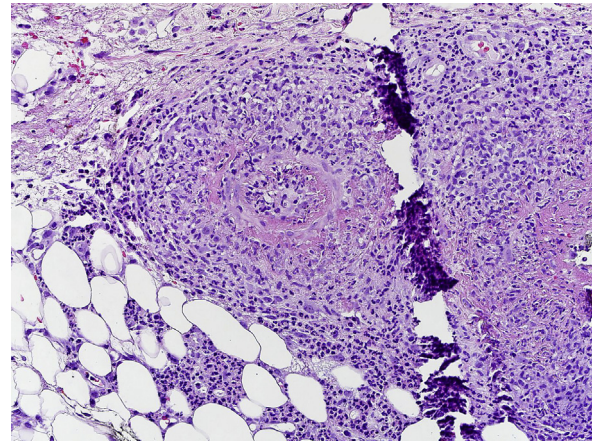


Fig 3. The image shows giant cell arteritis. Punch biopsy of a lesion on the right arm demonstrated an acute neutrophilic inflammatory infiltrate with a granulomatous component involving the vessel walls of medium-sized vessels (hematoxylin-eosin stain; original magnification: $\times 200$.) Specific stains for microorganisms and Epstein-Barr virus yielded negative results (not shown).

ability to ambulate within 24 hours of the first steroid dose and ultimately experienced complete resolution of all symptoms. Prednisone was gradually tapered off over several months, and she has remained off prednisone for 4 months, without recurrence of symptoms or elevation of inflammatory markers.

DISCUSSION

GCA rarely presents as cutaneous manifestations.¹ Our case highlights a unique presentation of GCA, with a few scattered indurated erythematous-to-violaceous plaques on the upper extremities, torso, and frontal aspect of the scalp. The cutaneous lesions were inflammatory, with vasculitis of medium-sized vessels.

The diagnosis of GCA can be challenging, particularly when patients present with atypical symptoms. If high-quality imaging is accessible, the European League Against Rheumatism prefers that high-quality imaging be performed early in the course of suspected GCA over temporal artery biopsy because of reduced morbidity, increased specificity, prompter results, and decreased rates of false-negative results.^{2,6}

In our case, cutaneous biopsy raised a concern for GCA and led to imaging being performed, which confirmed the diagnosis. The typical histologic findings of GCA include a granulomatous infiltrate centered at the inner media of arteries comprising prominent giant cells, leading to marked elastic layer fragmentation. However, giant cells may be absent, with the only histologic finding being granulomatous

involvement limited to the vessel wall. The latter is commonly referred to as vasculitis with granulomatosis based on the following pathologic differential diagnosis: polyangiitis; eosinophilic granulomatosis with polyangiitis; natural killer T-cell lymphoma; lymphomatoid granulomatosis; GCA; Takayasu arteritis (TA); drug-induced, inflammatory bowel disease; paraneoplastic syndrome; tuberculosis; tertiary syphilis; and previous herpetic infections.⁷ Some of these entities were easily dismissed based on our patient's clinical presentation. Her unremarkable workup for infectious and autoimmune diseases in combination with the PET-CT findings and her response to steroids was consistent with a large-vessel vasculitis.

TA is a large-vessel vasculitis with clinical features similar to those of GCA, which can make differentiating the 2 difficult. However, the patient did not demonstrate a diminished pulse, blood pressure difference of >10 mm Hg between the limbs, and bruits over the subclavian artery or aorta and was above 40 years of age, making the diagnosis of TA less likely. Additionally, patients with TA are more likely to present with involvement of the abdominal, renal, mesenteric, subclavian, and carotid arteries, detected via imaging, which was not seen in the PET scan in our patient. Patients with GCA can present with involvement of the aorta; aortic arch, axillary, or subclavian disease; and low burden of disease in the aorta.⁸ Our patient has been able to taper off steroids without disease recurrence, which is more consistent with GCA. TA typically requires chronic therapy to maintain disease control.⁸

Glucocorticoids are the mainstay of treatment for GCA.⁹ Immediate initiation of treatment is recommended as soon as the diagnosis is confirmed or suspected. There is a role of adjunctive therapy with methotrexate in those at high risk of adverse effects due to the use of systemic steroids.⁹

In this case, we highlight an atypical presentation of GCA. Initial diagnosis was challenging because of

nonspecific complaints and an atypical cutaneous eruption. Fortunately, the skin biopsy demonstrating vasculitis with granulomatosis prompted a workup for GCA. This case also emphasizes the role of imaging in the diagnosis of GCA, which is now recommended by the European League Against Rheumatism over temporal artery biopsy in suspected cases for which high-quality imaging is accessible.

Conflicts of interest

None disclosed.

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