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An Unusual Presentation of Granulomatosis with Polyangiitis (GPA)

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Conflict of interest: None declared

Patient: Female, 72
Final Diagnosis: Granulomatosis with polyangiitis
Symptoms: Finger pain • gangrene
Medication: —
Clinical Procedure: —
Specialty: Rheumatology

Objective: Unusual clinical course

Background: This is a very interesting case of Granulomatosis with Polyangiitis (GPA) presenting with hand ischemia which rapidly evolved into dry gangrene from the involvement of digital arteries. GPA usually affects the small and medium sized blood vessels. GPA affecting muscular artery causing limb ischemia is a rare manifestation.

Case Report: A 72 years old Caucasian female was sent to our hospital for the evaluation of left hand pain and bluish discoloration, which she noticed for few days. Physical examination was notable for bluish discoloration of left 2nd to 5th fingers, which later evolved, into dry gangrene at the tips. Angiogram revealed ischemia with no evidence of thrombosis. Administration of intra-arterial nitroglycerin resulted in improved blood flow through the radial artery. Labs revealed ESR of 142 mm/hr. C-ANCA titer was 5120, (normal <20) and antiproteinase 3 was 1117 (normal <20) consistent with GPA.

Conclusions: GPA usually affects the small and medium sized blood vessels. This case highlights an unusual presentation of GPA manifesting as critical limb ischemia. Given the rarity of this circumstance, a high index of suspicion is necessary in order to initiate proper treatment and limb salvaging intervention.

MeSH Keywords: Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis • Gangrene • Ischemia

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/909718

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Background

Granulomatosis with polyangiitis (formerly Wegner's granulomatosis) is a small-to-medium vessel vasculitis which affects multiple organs. Pathologically, there is a focal necrotizing vasculitis involving the small arteries and veins of the ear, nose, and throat, the lungs, the kidneys, the heart, the nervous system, the eyes, the skin, and, infrequently, other organs [1–3]. Involvement of digital arteries causing ischemia or gangrene is very rare. This paper describes a patient with GPA who initially presented with multiple ischemic fingers. The diagnosis was supported with a positive cANCA (anti-proteinase 3) and kidney biopsy. The patient responded well to therapy, with improvement of all organs involved and had no further digital ischemia or gangrene on follow-up. The involvement of digital arteries in this condition is very unusual and only a few cases have been reported so far.

Case Report

A 72-year-old white woman was sent to our hospital for the evaluation of left hand pain and bluish discoloration which she first noticed a few days before. She had recently developed bilateral uveitis, for which she was started on prednisone eye drops. She claimed to be healthy until a few months ago when she developed bilateral hearing loss followed by left foot drop. A physical examination was remarkable for bilateral sensorineural hearing loss, left foot drop, and bluish discoloration of the left 2nd to 5th fingers, which later evolved into dry gangrene at the tips (Figure 1). An angiogram revealed a diminutive left brachial artery distal to the elbow, with sluggish flow in the radial and ulnar arteries, terminating in the proximal forearm. There was no evidence of thrombosis. An intra-arterial nitroglycerin resulted in improved blood flow through the radial artery (Figure 2).

Routine blood work revealed normal complete blood count, with elevated creatinine to 2.3 mg/dl. Urinalysis showed mild proteinuria with a few red blood cell casts. ESR was markedly elevated to 142 mm/h. cANCA (titer: 5120, Ref. <20) and antiproteinase 3 (titer: 1117, Ref. <20) were significantly elevated, suggesting the diagnosis of GPA. The other rheumatologic panels, including P- ANCA, were negative.

Computed tomography (CT) of the chest showed extensive consolidation, with areas of cavitation and pseudo-cavitation (Figure 3). Bronchial lavage was negative for malignancy and infection. A subsequent kidney biopsy demonstrated pauci-immune crescentic glomerulonephritis, confirming the diagnosis (Figure 4). She was started on pulse methylprednisolone followed by oral prednisone and IV Cytoxan with some clinical improvement of hand ischemia proximally. The distal digits

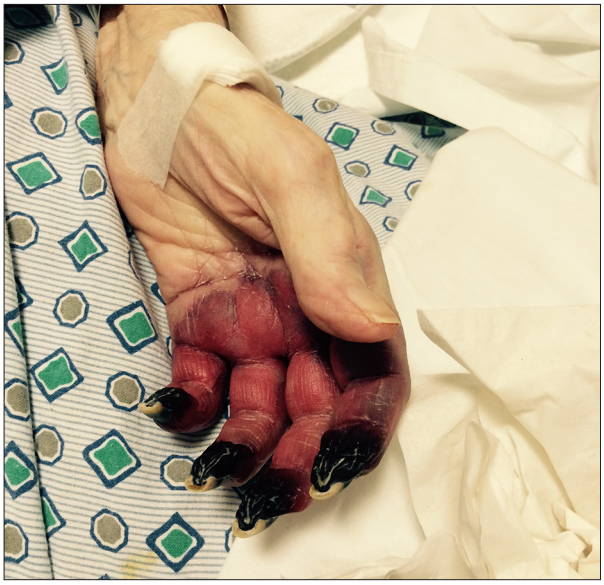

Figure 1. Dry gangrene involving finger tips.

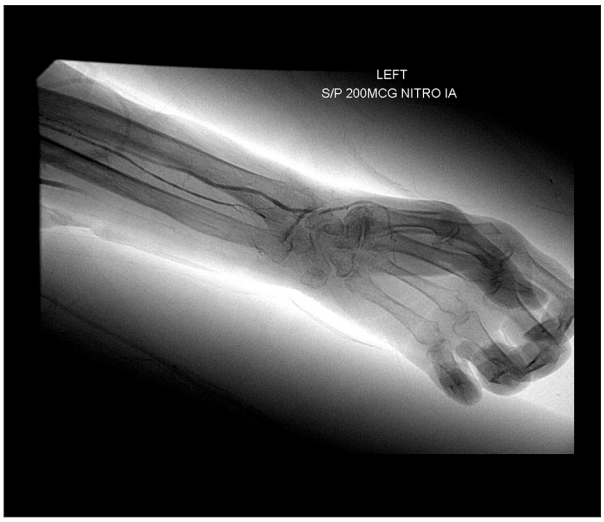

Figure 2. Angiogram showing slight improvement in blood flow, ruling out thrombosis.

were already gangrenous with no change. Her serum creatinine improved and stabilized at 1.5 mg/dl. Later, Cytoxan was transitioned to Rituximab for profound pancytopenia from former infusion.

Discussion

Cutaneous manifestations occur in 20% of patients with GPA, which usually includes papulonecrotic, pyoderma gangrenosum and sometimes generalized urticarial lesions [1–3]. There have been only a few cases reported of GPA presenting with digital ischemia as an initial manifestation [4–6]. Given the paucity of cases reported, the prevalence of digital

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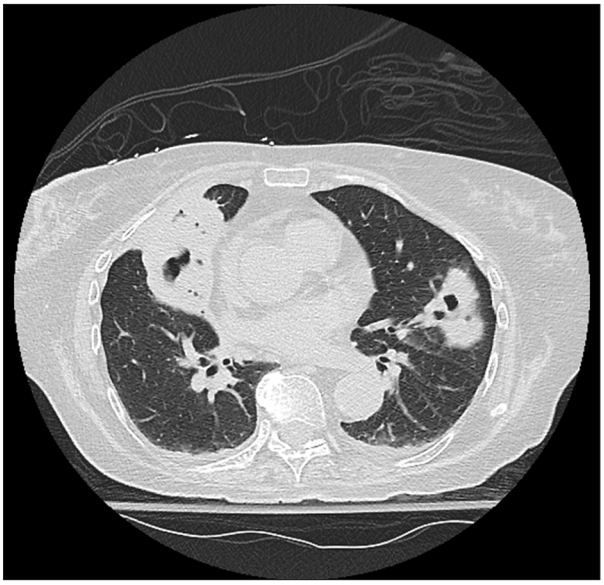

Figure 3. CT chest showing extensive consolidation, with areas of cavitation and pseudo-cavitation.

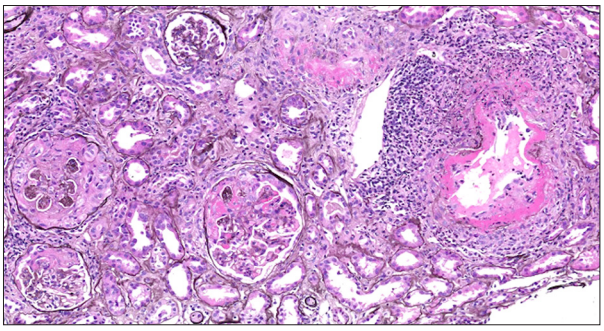

Figure 4. Kidney biopsy demonstrated pauci-immune crescentic glomerulonephritis.

ischemia and gangrene in the GPA population is suspected to be <1% [7–10]. The pathophysiology of digital ischemia and gangrene is thought to be from destruction of medium-sized vessels due to active vasculitis [1,2]. However, there is also evidence that *in situ* thrombosis, as a result of active vasculitis, can lead to ischemia and gangrene. The diagnosis is usually made based on clinical grounds, but angiographic examination can support vasculitic or thrombotic lesions [8–10]. There is no consensus on treatment, however, cyclophosphamide and steroids are the most commonly used treatment in the literature [8–10]. Without treatment, ischemia and gangrene can progress and lead to significant disability. Therefore, it is important to be aware of this rare manifestation and institute early treatment as indicated.

Conclusions

This case is unique as GPA usually affects the small- and medium-sized blood vessels. GPA affecting a muscular artery causing hand ischemia and gangrene is unheard of. Had we been vigilant of this rare manifestation, the patient's fingers could have been salvaged. All primary care physicians, including rheumatologists, should be aware of this rare manifestation so as to avoid delay in diagnosis and treatment.

Conflict of interest

None.

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