

Spontaneous Gastrointestinal Perforation as a Rare Complication of Granulomatosis with Polyangiitis (Wegener's Granulomatosis): A Case Report

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Publication Date: 2026/09/17

Abstract: Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a systemic necrotizing vasculitis that primarily affects the respiratory tract and kidneys, while gastrointestinal involvement is uncommon. We report a rare case of gastrointestinal perforation as a complication of GPA. The patient presented with acute abdominal pain and features of generalized peritonitis, and imaging revealed hollow-viscus perforation. Emergency exploratory laparotomy was performed, which identified intestinal perforation requiring resection and appropriate surgical management. Histopathological examination demonstrated features consistent with vasculitic involvement. Gastrointestinal manifestations of GPA may include abdominal pain, ulceration, bleeding, ischemia, obstruction, and perforation, with perforation being a rare but potentially fatal complication. Early recognition of gastrointestinal involvement, prompt surgical intervention when indicated, and appropriate immunosuppressive therapy are essential to reduce morbidity and mortality. This case highlights the importance of considering GPA as an underlying cause of gastrointestinal perforation, particularly in patients with known or suspected systemic vasculitis.

Keywords: Granulomatosis with Polyangiitis; Wegener's Granulomatosis; Gastrointestinal Perforation; Intestinal Perforation; Systemic Vasculitis; ANCA-Associated Vasculitis; Acute Abdomen; Bowel Ischemia.

How to Cite: Dr. J. G. Vagadiya; Dr. Bhargav Trivedi; Dr. Disha Padhi; Dr. Harpal Gadhvi (2026) Spontaneous Gastrointestinal Perforation as a Rare Complication of Granulomatosis with Polyangiitis (Wegener's Granulomatosis): A Case Report. *International Journal of Innovative Science and Research Technology*, 11(8), 3682-3685. <https://doi.org/10.38124/ijisrt/26aug1300>

I. INTRODUCTION

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a rare systemic necrotizing granulomatous vasculitis affecting predominantly small- and medium-sized blood vessels. It most commonly involves the upper respiratory tract, lungs, and kidneys, with gastrointestinal manifestations occurring infrequently. Gastrointestinal involvement may result from vasculitis-induced ischemia, ulceration, bleeding, obstruction, or perforation. Intestinal perforation is an exceptionally rare but potentially life-threatening complication of GPA and may present as an acute abdomen requiring urgent surgical intervention. Due to its rarity and nonspecific clinical presentation, gastrointestinal involvement can be challenging to diagnose, particularly when it occurs without previously established systemic disease. Early recognition of this complication, prompt surgical management, and appropriate

immunosuppressive therapy are essential for improving patient outcomes. We present a rare case of gastrointestinal perforation associated with granulomatosis with polyangiitis, highlighting the diagnostic and therapeutic challenges of this unusual manifestation.

II. CASE REPORT

A 35yrs/Male patient presented to Emergency Room on 18/07/2026 with complaints of bleeding per rectally since 10 days and right hand swelling since 4 days. Op/c/o Stapler haemorrhoidectomy done on 9/7/2026.

Routine blood investigations were within normal limits. Ultrasound- Abdomen +pelvis was suggestive of ileocolitic changes. Chest X-ray was suggestive of diffuse ground glass opacities on right upper lobe.

Patient was then managed conservatively as a case of ileocolitis with injectable antibiotic.

However within 1 week, patient developed petechial purpura, periorbital edema, swelling, conjunctival congestion, painful oral aphthous ulcers. There was marked bilateral hand swelling, acral edema, and progressive blackening/ischemic discoloration of multiple digits.



Fig 1 Left Hand Progressive Blackening



Fig 2 Right Hand Progressive Blackening

Following the onset of vasculitic symptoms, a multidisciplinary approach was initiated.

- CECT (Abdo+pelvis) revealed distal ileum, terminal ileum, IC junction appear inflamed with thickened wall with surrounding mesenteric fat stranding noted. *p/o* – infective/ inflammatory etiology .

- HRCT (Throat)- A well defined soft tissue density lesion (70*46mm) with few internal areas of cavitation is seen in posterior segment of right upper lobe inferiorly abutting the right major fissure.
- 2D ECHO – Normal
- Sputum CBNAAT & KOH- Negative
- S. Procalcitonin – 2.61 ng/ml
- CRP – 324 mg/L
- P-ANCA, C-ANCA, ANA, MPO IgG- NEGATIVE
- Anti PR3 IgG – 102.5 (positive)
- Skin biopsy (from back of Right leg)- revealed leucocytoclastic changes.

Thereafter, patient was diagnosed as a case of GRANULOMATOSIS WITH POLYANGITIS (WEGNER'S GRANULOMATOSIS).

Patient was then treated as per advice of Rheumatologist. We started with 1 cycle of cyclophosphamide. Along with it, he was kept on steroid coverage and higher antibiotics. Later, 3 cycles of phasmapheresis were also initiated.

However, approx. 2 weeks after being admitted, patient had complaints of abdominal pain * 1day (since 30 July 2026) and abdominal distention morning of 31st July 2026. There was no h/o vomiting, diarrhoea, GI bleed, constipation.

• Vitals

- ✓ P -116/min
- ✓ RR – 20
- ✓ BP– 130/90mmhg

Per Abdomen Examination – Abdomen was distended with generalized tenderness.

✓ URGENT CECT SCAN WAS DONE

• CECT(31/07/2026)-

Evidence of thinning of terminal ileal wall with suspicious focal gap defect with adjacent air foci are noted.

Multiple dilated bowel loops noted with most of them showing air –fluid levels, with transition point noted at distal jejunal bowel loops at umbilical region.

• *p/o* Acute GI Obstruction with Hollow Viscous Perforation

Patient was then taken for Emergency Exploratory Laprotomy on 1/08/2026 at 2:30 am. Ot findings were –

- ✓ Approximately 50cm and 40cm proximal to ICJ, impending perforation noted at antimesenteric side.
- ✓ Approx 5*3cm perforations noted at 20cm and 10cm proximal to ICJ noted at antimesenteric side.
- ✓ Sloughed off distal ileum with necrosed IC Jn, with necrosed caecum, ascending colon noted
- ✓ Complete transection of part of proximal transverse colon noted at hepatic flexure.

- ✓ Hence. Resection of Sloughed Off Distal Ileum, Caecum, Ascending Colon, Transverse Colon with Double – Barrel Ileal-Transverse Stomy with Tension Closure of Abdomen was done.



Fig 3 Resected Bowel Segment Containing Multiple Perforation and Necrosis

Thereafter, patient was shifted to SICU with ET in situ. Patient was kept on higher antibiotics and injectable steroids were resumed. Liquids was started on POD 2.

5th cycle of plasmapheresis was completed on 10/08/2026

- ✓ HPE report of resected bowel segment – Nonspecific changes

III. DISCUSSION

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a rare systemic necrotizing granulomatous vasculitis predominantly involving small- and medium-sized vessels.

It most commonly affects the upper respiratory tract, lungs, and kidneys, although any organ system can be involved.

The estimated annual incidence is approximately 10–20 cases per million population, with variation according to geographic region.

The clinical presentation is highly variable and depends on the organs affected. Common manifestations include constitutional symptoms such as fever, fatigue and weight loss, chronic sinusitis, nasal crusting or epistaxis, otitis media, pulmonary nodules or infiltrates, hemoptysis, arthralgia or arthritis, skin lesions, peripheral neuropathy, and renal involvement in the form of hematuria, proteinuria, or rapidly progressive glomerulonephritis. Gastrointestinal involvement is uncommon but is clinically important because it may be

associated with significant morbidity and mortality. Reported gastrointestinal manifestations include abdominal pain, nausea, vomiting, diarrhea, gastrointestinal bleeding, mucosal ulceration, intestinal ischemia, obstruction, and, rarely, intestinal perforation.

The mechanism is believed to involve necrotizing vasculitis of the mesenteric and intestinal vessels, resulting in impaired blood supply, ischemia, mucosal ulceration, transmural necrosis, and eventual perforation. Gastrointestinal perforation may occur as an initial manifestation or during the course of established GPA and can present as an acute surgical abdomen.

The diagnosis of GPA is based on a combination of clinical findings, laboratory investigations, imaging, and histopathology, as no single investigation is diagnostic. Initial laboratory evaluation includes complete blood count, ESR, CRP, renal and liver function tests, urinalysis, and assessment for proteinuria and hematuria. ANCA testing is an important component of the diagnostic workup, with PR3-ANCA being particularly associated with GPA, although ANCA may be negative in a proportion of patients. Other investigations are directed toward excluding infectious, malignant, and other autoimmune causes of systemic inflammation. Histopathological confirmation, when feasible, may demonstrate necrotizing granulomatous inflammation and vasculitis. Renal biopsy may show pauci-immune necrotizing and crescentic glomerulonephritis. Imaging plays an important role in assessing the extent of disease. CT of the chest may reveal pulmonary nodules, cavitation, infiltrates, or alveolar hemorrhage, while CT of the paranasal sinuses can demonstrate chronic inflammatory changes. In patients with abdominal symptoms, contrast-enhanced CT of the abdomen is particularly useful for identifying bowel-wall thickening, ischemia, ulceration, necrosis, obstruction, or perforation. The nonspecific nature of gastrointestinal symptoms can make diagnosis challenging; therefore, the possibility of gastrointestinal vasculitis should be considered whenever a patient with suspected or established GPA develops new abdominal symptoms.

Management of GPA depends on the severity and extent of organ involvement. Patients with limited disease may be treated with less intensive immunosuppression, whereas organ-threatening or life-threatening disease requires prompt induction therapy. Current treatment strategies generally involve high-dose glucocorticoids combined with rituximab or cyclophosphamide. Rituximab is particularly useful in relapsing disease and is also commonly used for maintenance therapy. Avacopan may be considered in selected patients as a glucocorticoid-sparing option. Once remission is achieved, maintenance treatment with agents such as rituximab, azathioprine, or methotrexate may be used depending on the individual clinical situation.

Treatment requires careful monitoring because immunosuppressive therapy can increase the risk of serious infections, poor wound healing, metabolic complications, and disease-related relapse. In patients with gastrointestinal involvement, the decision to initiate

or intensify immunosuppression should be individualized and coordinated with the surgical team, particularly in the presence of bowel perforation, peritonitis, or an uncontrolled source of infection.

When gastrointestinal perforation occurs, surgical management becomes a critical component of treatment. Prompt resuscitation, correction of fluid and electrolyte abnormalities, broad-spectrum antibiotics, and urgent surgical assessment are required. Patients with generalized peritonitis, bowel necrosis, or free perforation usually require emergency exploration, resection of the affected bowel, and appropriate restoration of intestinal continuity or creation of a temporary stoma depending on the patient's clinical condition.

Histopathological examination of the resected specimen may demonstrate vasculitic, ischemic, or necrotizing changes and can provide important diagnostic information.

Although surgery provides definitive source control, it does not treat the underlying systemic vasculitis; therefore, immunosuppressive therapy remains essential after adequate control of the acute surgical problem and stabilization of the patient.

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