



## Case Report

# Splenic Flexure Aneurysm Secondary to Granulomatosis with Polyangiitis and Mayhegglin Anomaly

Jessica Cox<sup>1</sup>, Devin B Haney<sup>2\*</sup>, Varalaxmi Nannaka<sup>2</sup>

<sup>1</sup>School of Medicine, University of Texas Medical Branch, Galveston, TX, US

<sup>2</sup>Department of Internal Medicine, University of Texas Health Tyler, Tyler, TX, US

\*Corresponding author: Devin B Haney, Department of Internal Medicine, University of Texas Health Tyler, Tyler, TX, US.

**Citation:** Cox J, Haney DB, Nannaka V (2024) Splenic Flexure Aneurysm Secondary to Granulomatosis with Polyangiitis and Mayhegglin Anomaly. Ann Case Report 9: 1781. DOI: 10.29011/2574-7754.101781

**Received:** 24 April 2024; **Accepted:** 29 April 2024; **Published:** 01 May 2024

### Abstract

This case details a 64-year-old white male diagnosed with Granulomatosis with polyangiitis, complicated by the May-Hegglin anomaly and presenting with a severe gastrointestinal bleed. Hypotension and anemia prompted packed red blood cell and fresh frozen plasma administration, culminating in ICU admission for intubation and norepinephrine support. Despite spontaneous resolution at the splenic flexure, ongoing bleeding necessitated 11 units of packed red blood cells and an endovascular intervention. The patient's unique combination of Granulomatosis with polyangiitis and May-Hegglin anomaly, accentuated by a splenic artery aneurysm, represents a rare clinical entity. Granulomatosis with polyangiitis typically targets lungs and kidneys, with aneurysms seldom documented. This case is the first to report a splenic artery aneurysm in Granulomatosis with polyangiitis, further complicated by May-Hegglin anomaly.

The success of the endovascular approach, involving coiling the splenic artery aneurysm and a selective region of the inferior mesenteric artery, underscores its viability as a primary treatment for associated aneurysms in Granulomatosis with polyangiitis. This case emphasizes the importance of tailored interventions in managing complex medical scenarios, shedding light on the intricate interplay between May-Hegglin anomaly, Granulomatosis with polyangiitis, and splenic artery aneurysm.

**Keywords:** Granulomatosis with polyangiitis; May-Hegglin Anomaly; Splenic Flexure Aneurysm

### Introduction

Granulomatosis with polyangiitis (GPA) is a rare and debilitating disease requiring immediate intervention, characterized by an immunologically mediated systemic small vessel vasculitis. [1] GPA exhibits an inflammatory reaction pattern presenting with necrosis, granulomatous inflammation, and vasculitis, typically localized to the lower respiratory tract and kidneys. [1] The prevalence of GPA in the United States is estimated to be 3 cases per 100,000 people with 70-100% of cases involving the upper respiratory tract. [2] The exact etiology of GPA is unknown.

Current hypotheses include: 1) environmental exposures to dust inhalation or silica (10% of cases); 2) autoimmunity via molecular mimicry; and 3) genetic predispositions. [3]

GPA is a clinical diagnosis made from clinical presentation with anti-neutrophil cytoplasmic antibody (ANCA) serology, necrotizing vasculitis, and granulomatous inflammation on histology providing support. [1] Other manifestations include oral ulcers, purulent or bloody nasal discharge, pulmonary nodules, fixed pulmonary infiltrates, pulmonary cavitory lesions, and microscopic hematuria with or without RBC casts.[1] The classic triad of vasculitis, necrosis and granulomatous inflammation can be observed in up to 16% of cases.[1]

Nonspecific constitutional symptoms of fever, weakness, and weight loss are observed in 50% of the population. [3] Other manifestations occur in various systems including ear, nose, and throat (ENT) with 70% of cases presenting with rhinorrhea, sinusitis, otitis media, or damage to facial cartilage. [3] Lung involvement can be observed in 50-90% of the cases with alveolar hemorrhage or parenchymal nodules. Common renal manifestations include focal segmental necrotizing glomerulonephritis associated with pauci-immune crescent formation observed in 40% of the cases. [3] Other systemic manifestations include ocular involvement (60%), the peripheral nervous system (33%), cardiac involvement (less than 10%), and gastrointestinal involvement (5-10%). [3] Gastrointestinal involvement typically presents with ulcerative lesions. [3]

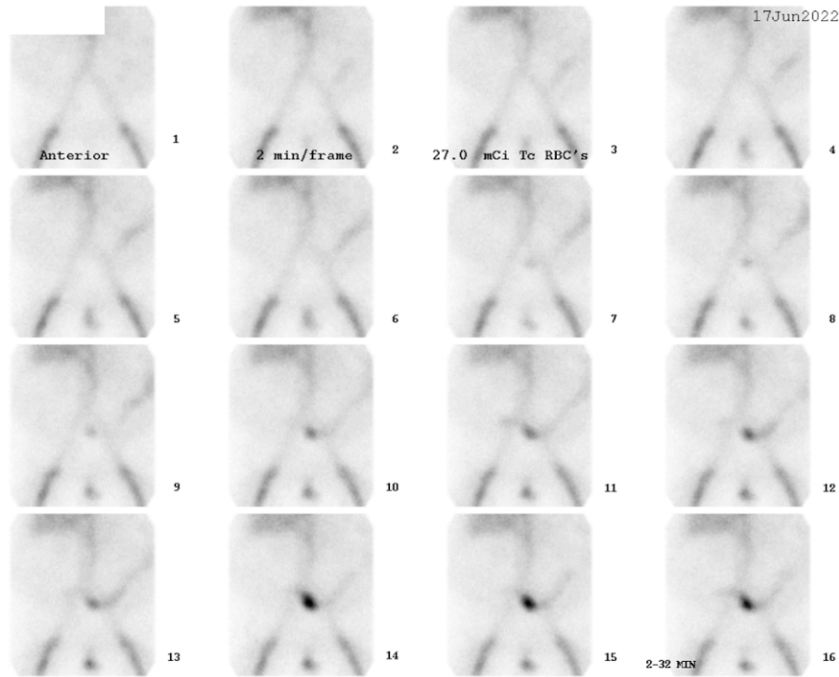
Early diagnosis is vital as 10-year survival rate is estimated to be 60-70% but can be as low as 40% with renal involvement. If left untreated, GPA has an 82% mortality rate at 1 year. [1] Treatment for GPA can be subdivided into induction and maintenance phases, with the induction phase involving systemic corticosteroids, cyclophosphamide, and glucocorticoids for 3-6 months followed by a maintenance phase with oral corticosteroids, azathioprine or methotrexate, and cotrimoxazole for 18-24 months. [3]

May-Hegglin anomaly is a rare autosomal dominant disease due to a MYH9 gene mutation which is characterized by neutrophils with abnormal cytoplasmic inclusions, large platelets, and variable thrombocytopenia. [4] The mutation occurs on chromosome 22q12-13 and encodes the non-muscle myosin heavy chain class IIA, a cytoplasmic protein present on many tissues including platelets. [4] This can cause macrothrombocytopenia secondary to defective megakaryocyte maturation and fragmentation. [4] Most patients are asymptomatic and do not experience bleeding complications. In rare cases, severe bleeding may be present and require platelet transfusion. [4] There are less than 100 cases reported in literature, and the exact etiology of this mutation is unknown. [4] Here, we present a case where granulomatosis with polyangiitis and May-Hegglin anomaly occur simultaneously leading to significant, uncontrollable blood loss.

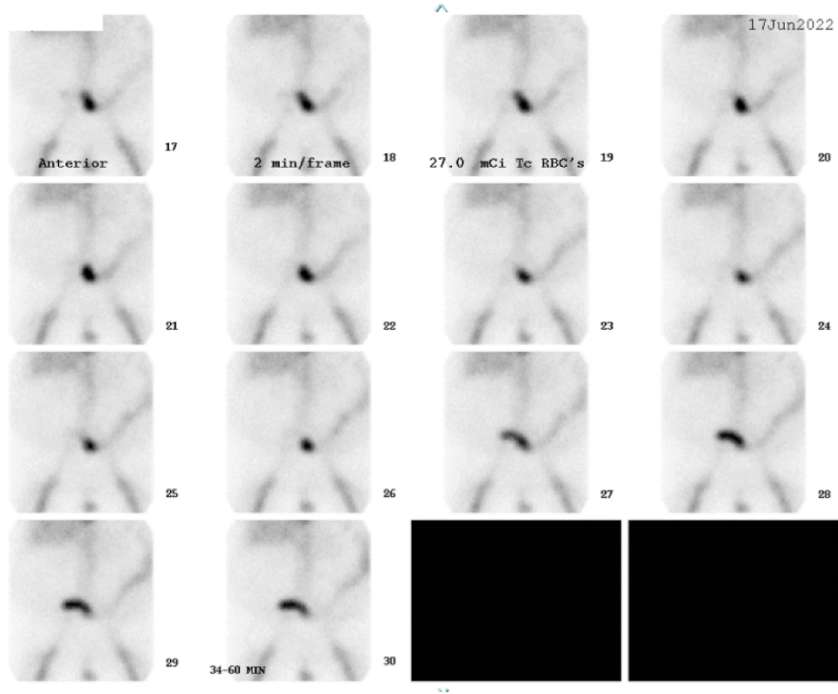
We present the case of a 64-year-old white male diagnosed one month prior with granulomatosis with polyangiitis in the setting

of May-Hegglin anomaly. The patient presents after experiencing four episodes of frank hematochezia. Vital signs on arrival showed a blood pressure of 73/61 mmHg and a pulse of 96 bpm with no other disturbances. Laboratory evaluation revealed a serum BUN of 49, serum creatinine of 1.6, normocytic, normochromic anemia with a hemoglobin of 7.4 mg/dl, and coagulation studies revealing PT of 12.8 and INR of 1.2. While in the emergency department, the patient had voluminous bowel movements consisting of clots and blood. He then received 3 units of packed red blood cells (PRBC) which raised his Hgb to 7.8. Due to resistant hypotension, intensive care was consulted for admission. Diagnosis on admission was hypovolemic shock secondary to gastrointestinal hemorrhage in the setting of Wegener's granulomatosis with May-Hegglin anomaly. Upon arrival to ICU, patient was started on norepinephrine for pressure support. The patient then passed two more voluminous, bloody stools and was given 2 more units of PRBC, 1 unit of platelets, and 2 units of fresh frozen plasma. The patient's blood pressure was poorly controlled despite continuous norepinephrine. DDAVP (desmopressin) was given upon admission, after which vasopressin was added as a secondary pressure agent. There was continued difficulty managing the patient's bleeding and the patient was intubated in an elective fashion due to impending respiratory failure. Gastroenterology and interventional radiology were consulted and recommended a bleeding scan.

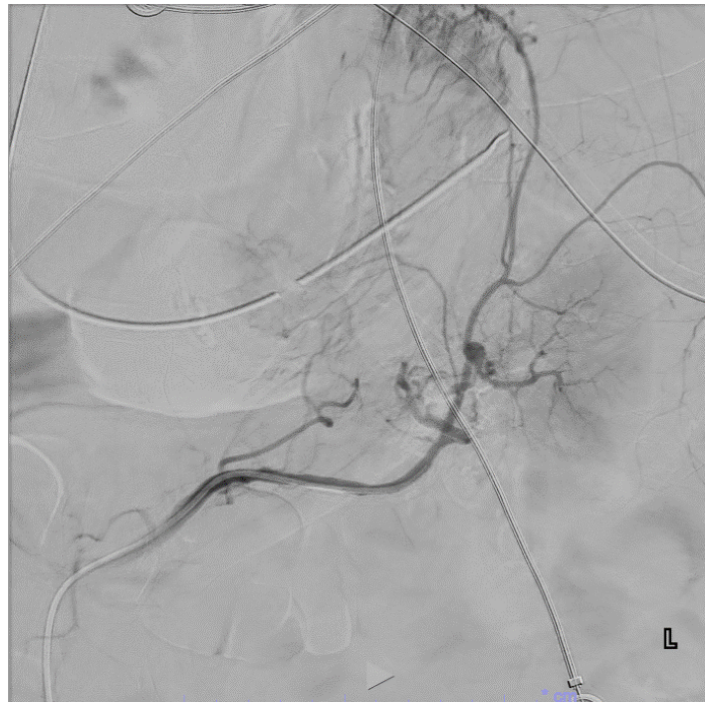
The nuclear bleeding scan revealed aneurysmal changes to the distal splenic artery with evidence of bleeding requiring coil embolization (figure 1,2). Selective catheterization of the branches of the superior mesenteric artery and inferior mesenteric artery revealed a bleeding diverticulum supplied by a distal branch of the inferior mesenteric artery which was subsequently embolized (figure 3,4). Post-procedural hemoglobin stabilized at 10.2, as did blood pressure. At this point in the hospital course, hematology was consulted to help manage this patient. The patient was then successfully extubated and blood pressure stabilized, holding vasopressin at time of extubation. Upon extubating, the patient had episodes of bloody bowel movements requiring an additional 2 units of PRBC and 1 unit of platelets; DDAVP was not given due to his factor VIII depletion.



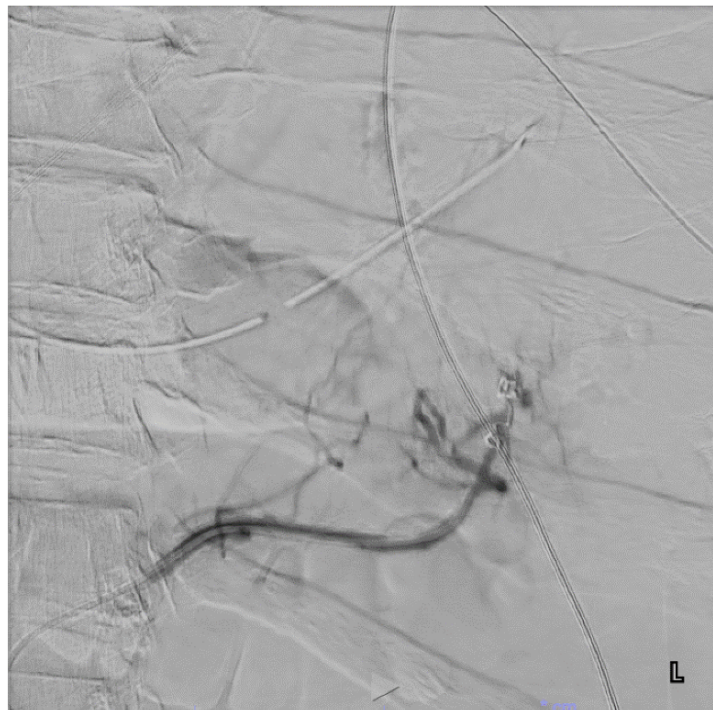
**Figure 1:** Nuclear medicine imaging: Abnormal uptake which appears to begin within LUQ on/about foramen to progress in more distal LUQ, finally centering over region of iliac bifurcation.



**Figure 2:** Nuclear medicine imaging: Abnormal exam with evidence of GI blood loss originating in the region of the splenic flexure.



**Figure 3:** Interventional radiology imaging: splenic flexure bleed.



**Figure 4:** Interventional radiology imaging: post coiling to control bleeding

Throughout the course of the significant blood loss, serum creatinine continued to rise from 1.6 to 2.6 leading to acute renal failure, secondary to acute tubular necrosis from hemorrhagic shock. Post-operative recovery was relatively uneventful. Over the course of his hospital stay, he received 11 units of PRBC. Hematology, nephrology, interventional radiology, and gastroenterology were involved in care. Upon discharge, the patient's Hgb and blood pressure was stabilized, and the patient was successfully discharged to an inpatient rehab facility.

This case represents a rare combination of notably difficult diseases to treat. To our knowledge, there has not been documented literature of patients presenting with granulomatosis with polyangiitis and May-Hegglin anomaly concurrently. In addition to the rarity of the patient's presentation, a large GI bleed located at the splenic flexure with granulomatosis with polyangiitis has also not been documented to our knowledge.

Our patient's hospital course was further complicated by continuous elevation in BUN and creatinine, leading to acute renal failure of pre-renal etiology, likely due to hypoperfusion from hypovolemic shock. With time, the patient's renal function returned back to his baseline at a serum creatinine of 1.3 at the time of discharge. The patient had multiple potential factors impacting renal function. Aside from the hypoperfusion and contrast insults, it is important to recognize the importance of renal function in overall mortality in patients with granulomatosis with polyangiitis. The literature also reports MYH9-associated nephropathy secondary to abnormalities of heavy chain myosin expression. These mutations can be expressed in podocytes and mesangial cells leading to proteinuria and chronic kidney disease in affected patients. [5]

The patient received two doses of DDAVP to mobilize his stores of Factor VIII. However, with the continuous rapid drop in hemoglobin, norepinephrine was started. Our patient did not receive a third dose of DDAVP due to the depleted Factor VIII stores, though he received continuous norepinephrine. With continuation of norepinephrine, his vitals eventually stabilized.

Granulomatosis with polyangiitis is characterized as systemic necrosis of small to medium sized arteries usually within the lungs and kidneys, making aneurysms a rare but possible complication with few documented incidences. [6] Literature review revealed 20 cases of medium and large vessel aneurysms in patients with granulomatosis with polyangiitis. Within these cases, aneurysms were reported in the aorta, pancreaticoduodenal artery, renal artery, craniometrical artery, coronary artery, and superficial femoral artery. To the knowledge of the authors, this is the first case to present an aneurysm involving the splenic artery which was further complicated with the rare genetic disorder of May-Hegglin anomaly. [6]

To our knowledge, there have only been two reported cases of aneurysmal complications in patients with May-Hegglin anomaly. Our patient is the first reported case involving the peripheral vasculature. [7,8] Frequently, patients with May-Hegglin anomaly present with bleeding complications and are routinely diagnosed with idiopathic thrombocytopenia, as the thrombocytopenia occurs without obvious cause. [9] There have been previous reports of rectal bleeding likely due to May-Hegglin syndrome described in literature. While these patient presentations were not further complicated by aneurysm formation, they were treated similarly with platelet transfusions. [9]

In conclusion, we present a patient with a rare constellation of presentations involving MayHegglin anomaly, granulomatosis with polyangiitis, and splenic artery aneurysm. An endovascular approach should be considered as a first line treatment to address the aneurysms associated with granulomatosis with polyangiitis, and close attention should be paid to renal function to improve mortality outcomes.

### Take Home Message

A 64-year-old male with Granulomatosis with polyangiitis and May-Hegglin anomaly presented with severe gastrointestinal bleeding. Despite spontaneous resolution, ongoing bleeding necessitated 11 units of packed red blood cells and endovascular intervention, addressing a rare splenic artery aneurysm. This case highlights tailored interventions' importance, shedding light on the intricate interplay between May-Hegglin anomaly, Granulomatosis with polyangiitis, and splenic artery aneurysm.

**Conflicts of Interest:** The authors have no conflicts of interest to declare.

**Funding:** None

**Contributions:** (I): conception and design: all authors, (II): administrative support: Varalaxmi Nannaka, (III): Provision of patient: all authors; Manuscript writing: Jessica Cox, Devin Haney, (IV): final approval of manuscript: all authors.

### References

1. Greco A, Marinelli C, Fusconi M, Macri GF, Gallo A, et al. (2016) Clinic manifestations in granulomatosis with polyangiitis. *Int J Immunopathol Pharmacol* 29(2):151-9.
2. Tracy CL (2023) Granulomatosis with polyangiitis (GPA, formerly Wegener granulomatosis). *Practice Essentials, Background, Etiology*
3. Camarmond C, Cacoub (2014) Granulomatosis with polyangiitis (Wegner): Clinical aspects and treatment. *Autoimmunity Reviews* 13(11): 1121-1125.
4. Untanu RV, Vajpayee N (2023) May-Hegglin Anomaly. [Updated 2023 Mar 8]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing.

5. Furlano M, Arlandis R, Venegas M, Novellif S, Crespi J, et al. (2019) MYH9 Associated nephropathy. *Nefrologia* 39(2): 111-222.
6. Tomosugi T, Takahashi T, Kawase Y, Yoshida K (2017) Accessory left gastric artery aneurysms in granulomatosis with polyangiitis: a case report and literature review. *Nagoya J Med Sci* 79(1):75-83.
7. Doherty J, Millward C, Sarsam Z. (2021) Spontaneous and recurrent subdural hematoma in a patient with May-Hegglin anomaly. *BMJ Case Rep* 7(14).
8. Bejot Y, Barnay J, Chavent A, Daubail B, Jacquin A, et al. (2017) Subarachnoid Hemorrhage revealing Moyamoya syndrome in a patient with May-Hegglin Anomaly. *Neurologist* 22(5):204-205.
9. Reddy P, Kollipara R, Shammo J, Loew J (2021) The May-Hegglin anomaly: a rare cause of a common compliant. *BMJ Case Rep* 14(3).