# Small Bowel Involvement of Microscopic Polyangiitis Presenting as a Massive Gastrointestinal Haemorrhage

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## ABSTRACT

**Microscopic polyangiitis is an Anti-Neutrophilic Cystoplasmic Antibody (ANCA) vasculitis disorder, mainly affecting the kidneys and respiratory tract, but also having the capacity to affect other organ systems, including the gastrointestinal (GI) tract. This is a case of microscopic polyangiitis presenting with massive life-threatening GI haemorrhage, with the bleeding source originating in the small intestine, and improving dramatically with immunosuppression. Massive GI haemorrhage is an unusual presentation of such cases, with involvement usually being with abdominal pain, obstruction, or evidence of ischaemia. Such patients usually present a diagnostic dilemma and require urgent treatment not only of the bleed itself, but of the underlying vasculitic process, as the condition can be life-threatening.**

## INTRODUCTION

Systemic vasculitic disorders, including those that are ANCA-associated, can infrequently involve the gastrointestinal system, which can present in a variety of ways including ischaemia, obstruction, perforation, and haemorrhage. Clinical presentation is often dramatic and severe, and can be life-threatening. Diagnosis depends on imaging and serology, which are not very specific, and most importantly, on a high level of clinical suspicion. Immunosuppression is the mainstay of management and should be prompt. This case presents a case of microscopic polyangiitis complicated by massive gastrointestinal haemorrhage, with the bleeding source originating in the small intestine, and discusses the approach to, and difficulties faced, in diagnosis of such cases[1-5].

## CASE

A 62-year-old male who was known to have microscopic polyangiitis with resultant stage V chronic kidney disease requiring hemodialysis presented with a one-day history of large-volume bleeding per rectum. The patient had received steroids at disease diagnosis four years earlier, which had been tapered off and discontinued. His disease course had remained stable once dialysis had been initiated. On presentation, there was fresh bleeding per rectum with large clots. There was no associated abdominal pain, vomiting, diarrhoea, constipation, or weight loss prior to this. There were no prior similar episodes.

The patient was hypotensive on arrival at 99/50 mmHg and tachycardic at 103 beats per minute. On physical examination, he was pale. Abdomen was soft and nontender with no palpable masses or organomegaly. Rectal examination showed fresh blood around the anal verge but was otherwise unremarkable.

Haemodynamic resuscitation was initiated with 0.9% saline and blood transfusion was commenced. Blood investigations showed a Haemoglobin of 5.2 g/dL, a White Blood Cell Count of 7.3 x $10^9$/L, and a Platelet count of 284 x $10^9$/L. Serum creatinine was 980 umol/L. A stool infection screen came back negative. A CT mesenteric angiogram showed no extravasation of contrast into the bowel lumen, indicating no significant active bleeding at the time of the scan. It also showed a short segment of distal small bowel with increased mucosal vascularity and mural thickening, appearing inflamed, and being suggestive of enteritis. He subsequently underwent a colonoscopy which was unremarkable apart from showing blood residue throughout the bowel with no evidence of a focal bleeding source. The patient's bleeding resolved spontaneously, and he was discharged with close follow-up. He presented again one week later with recurrence of his bleeding per rectum, this time with some melaena as well. Haemoglobin level at the time was 6.3 g/dL. A gastroscopy was unremarkable, and an ileocolonoscopy with 20 cm of terminal ileum intubated was also normal. A capsule endoscopy was considered, however in view of his known vasculitis and finding of small bowel thickening on initial CT, the patient would have required a patency test prior to this. A Magnetic Resonance Enterography (MRE) was requested to further assess the small bowel and revealed a short segment of stricturing in the mid-ileum. The previously noted terminal ileum thickening had resolved. In view of the presence of this stricture, a capsule endoscopy was definitively contraindicated and was therefore not performed (Figure 1).

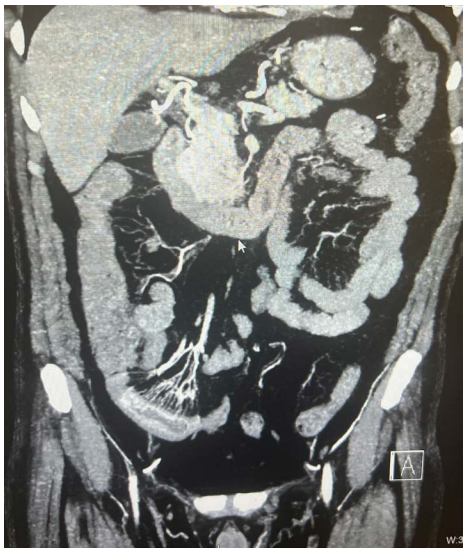

**Figure 1:** Thickened strictured ileum

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The patient was started on intravenous Methylprednisolone on presentation at his second admission. Bleeding ceased within a day of administering this. He re-bled, although less massively, on cessation of the steroids, and therefore they were re-introduced with a slow taper. The bleeding again recurred once a Prednisolone dose of 20 mg was reached, therefore the patient was considered for steroid-sparing therapy. He was subsequently started on Rituximab. He has had no recurrence of his bleeding since then.

## DISCUSSION

Microscopic polyangiitis is an ANCA-associated vasculitis with multisystem involvement, typically mainly affecting the kidneys and lungs. Other organ systems can potentially be involved, including less commonly the gastrointestinal tract. GI involvement can present with abdominal pain, evidence of obstruction, ischaemia, or perforation. These often present as abdominal pain. Patients can also be asymptomatic with findings only picked up on autopsy. Much less commonly, clinical presentation is with bleeding. This can be from the stomach, small intestine, or large intestine, and the bleeding source is often difficult to localize, particularly if the bleeding site is in the small intestine.

Investigation begins with an oesophagogastroduodenoscopy (OGD) and an ileocolonoscopy. Should those be negative, a small bowel capsule endoscopy would be the next line of investigation. The sooner after presentation this is done, the higher the diagnostic yield. However, it is important to remember that in this subset of patients, ongoing vasculitis and inflammation can often lead to stricturing. Therefore, a patency test is usually indicated prior to small bowel capsule endoscopy. Radiologic investigation in the CT scan, with mesenteric angiography if bleeding is active, can show features of inflammation but is nonspecific. These include mucosal oedema and narrowing as was present in our case. Angiography will only detect an active bleed if this is ongoing at a rate of at least 0.5 ml per minute. In diagnosing a small bowel source, MR is helpful in marking sites of pathology however and can show thickening or stricturing of the small intestine. Should MRE or a small bowel capsule endoscopy confirm evidence of small bowel disease, a push enteroscopy, in either an anterograde or a retrograde manner can be performed for tissue sampling to confirm the diagnosis of vasculitic involvement of the GI tract.

Since the diagnosis is difficult and time-consuming to confirm, timely treatment of GI involvement in ANCA vasculitis is often difficult. Immunosuppression is central to the management of these cases, and is in the form of steroids initially, bridged with Rituximab or Cyclophosphamide, with the former having a more favourable side effect profile. Since GI presentations of ANCA vasculitis can be life-threatening, particularly if they present in the form of a massive haemorrhage or ischaemia, a high index of suspicion is required, especially in those known to have vasculitic disease or those with concomitant involvement of other organ systems. Importantly, it is often not practical to wait for a tissue diagnosis prior to starting immunosuppressive therapy as patients' condition can be critical.

This case is unique in that GI involvement of vasculitis itself is not very common, even more so if it presents as a massive bleed. Patients with gastrointestinal ANCA vasculitis typically present with abdominal pain rather than massive bleeding. There have been previous case reports of ANCA vasculitis presenting with melaena, however only one other to our knowledge with fresh bleeding per rectum. A push enteroscopy and biopsies in this case would have further confirmed the diagnosis, however the patient declined further endoscopy, and his background history of vasculitis, imaging findings, and prompt response to immunosuppression made the diagnosis instead.

## CONCLUSION

Large-volume gastrointestinal hemorrhage is an atypical feature of microscopic polyangiitis, which usually favours the kidneys and respiratory tract. A high index of suspicion and prompt treatment are essential, given that a tissue diagnosis can sometimes be difficult and/or time-consuming to obtain, and more crucially, as the bleed may be fatal if not treated urgently with immunosuppression.

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