Enteroscopic appearance of vasculitis presented with melena and signs of nephrotic syndrome

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SUMMARY
A 69 year-old man was admitted to our hospital with melena, and clinical signs of nephrotic syndrome. The site of hemorrhage was not revealed by upper gastrointestinal (GI) endoscopy and colonoscopy. Combined surgical and enteroscopic interventions were employed to investigate the small intestine and the bleeding was located in the ileum, part of which was resected and examined histologically. Histology showed the presence of vasculitis. The literature of such systemic causes of gastrointestinal bleeding is reviewed.

Key words: vasculitis, melena, nephrotic syndrome, endoscopy, enteroscopy, Commercial interests & grant support: None

INTRODUCTION
Approximately 5% of all patients with gastrointestinal hemorrhage, and up to 38% of subjects with iron deficiency anemia without overt gastrointestinal bleeding, do not have a bleeding site identified after routine evaluation by esophagogastroduodenoscopy and colonoscopy. The source of bleeding in these subjects is often the small intestine. Most vascular lesions of the small bowel present as chronic gastrointestinal bleeding, which may be severe in some cases. Depending on the underlying disease, other systemic signs and symptoms may be present. The diagnosis and often therapy are heavily dependent upon endoscopic techniques, and in some cases upon nonendoscopic methods.1

In this paper we report successful exploration and treatment of hemorrhage in the small intestine, caused by vasculitis, by employment of combined surgical and enteroscopic interventions.

CASE REPORT
We report a case of 69 year-old man who was admitted to our hospital with the first episode of melena in his life, and clinical signs of nephrotic syndrome. Upper GI endoscopy and colonoscopy did not reveal the location of hemorrhage. Similarly, arteriography and scintigraphy by use of autologous-labelled red blood cells did not focus the source of GI bleeding. As the hematocrit declined rapidly from 25% to 19%, we decided to explore a potential source of hemorrhage in the small intestine by introduction of combined surgical and enteroscopic interventions. Through the enterotomy section we introduced the enteroscope from the duodenum to ileum, where we found the source of the GI bleeding (Figure 1). Histological examination of the resected segment of the ileum (Figure 2) showed findings of vasculitis (mainly in mucosa or submucosa) with eosinophilic infiltration, as the cause of the blood loss.

DISCUSSION
The mesenteric circulation is acutely sensitive to processes that affect the entire body. Such systemic diseases and syndromes are reviewed with particular
emphasis on the mechanisms by which they influence the mesenteric vasculature and blood flow.\textsuperscript{2}

Vasculitis can affect the bowel in several ways.\textsuperscript{7} A patient’s presentation depends on the caliber of the vessels involved. Although large artery vasculitis, as with aortitis, can lead to arterial occlusion and subsequent bowel gangrene and perforation, venulitis, with obstruction of the venous return, leads to mucosal edema and malabsorption. Vasculitis of medium and small arteries generally causes gastrointestinal bleeding. Medium-sized arteries develop aneurysms, which can rupture and massively bleed, as occurs in polyarteritis nodosa.\textsuperscript{3} This condition is a systemic necrotizing vasculitis affecting small and medium-sized arteries of nearly every organ. Approximately half of the cases are associated with hepatitis B infection.\textsuperscript{4} Gastrointestinal complaints are present in about two thirds of patients, and are usually secondary to visceral ischemia.\textsuperscript{5}

Necrotizing vasculitides of the bowel also occur in rheumatoid arthritis (although usually affecting the colon) and systemic lupus erythematosus (SLE).\textsuperscript{5} Gastrointestinal (GI) symptoms occur in about 30\% of SLE patients,\textsuperscript{5} of whom approximately 75\% have findings suggestive of vasculitis.\textsuperscript{2} Acute, severe abdominal pain was the presenting symptom in an adolescent with SLE. There have been reports of patients with SLE who underwent exploratory laparotomy because of acute abdominal pain with peritoneal irritation and had intraabdominal vasculitis.\textsuperscript{8}

Vasculitis of small arteries (intramural arterioles) presents with pain, fever, and occult bleeding. Infectious and inflammatory processes usually affecting these vessels occur in tuberculosis, amyloidosis, sarcoidosis, and multiple myeloma.\textsuperscript{1}

Hypersensitivity vasculitis, a vasculitis usually affecting the smaller vessels in response to a specific antigen, is seen in Henoch-Schönlein purpura. It has a peak age of onset in early childhood, but may occur at any age. The gastrointestinal tract is involved in as many as 50\% of patients. Any part of the GI tract may be affected and can precede the onset of the characteristic purpuric rush.\textsuperscript{4} Immune complex vasculitis of small vessels (leucocytoclastic vasculitis) appears on the skin of dependent sites as crops of palpable purpura and is mediated by deposition of immunoglobulin G in most cases in postcapillary venules.\textsuperscript{10}

Radiation injury to the bowel can also cause vasculitis and subsequent bleeding. Bowel irradiation initially affects the intestinal mucosa directly, causing ulceration and bleeding. Late injury usually occurs 6 to 24 months after radiation treatment from a progressive occlusive vasculitis.\textsuperscript{3}

Churg-Strauss disease is a systemic vasculitis that resembles polyarteritis nodosa but has such characteristics as granuloma formation, eosinophilia, and pulmonary clinical presentation. Gastrointestinal manifestations are related to mesenteric vasculitis.\textsuperscript{9}

The clinical presentation of Wegener’s granulomatosis is the triad of focal glomerulonephritis vasculitis that may result in intestinal or colonic ischemia, bleeding and perforation. The finding of anti-neutrophil cytoplasmic

![Figure 1](image1.png) Figure 1. Endoscopic appearance of the erosions in the terminal ileum due to vasculitis.

![Figure 2](image2.png) Figure 2. Macroscopic appearance of the resected portion of the small intestine.
antibodies may be helpful in establishing the diagnosis.4

In the United States, four diseases account for the vast majority of cases of lower intestinal bleeding: arteriovenous malformation, diverticulosis, neoplasms, and internal hemorrhoids. “Common” less frequent causes of gastrointestinal bleeding include solitary rectal ulcer syndrome, colonic varices, mesenteric vascular insufficiency, small bowel diverticula, Meckel’s diverticulum, aortoenteric fistula, vasculitis as in our case, small intestinal ulceration, endometriosis, radiation-induced injury, and intussusception. Less frequent causes of gastrointestinal bleeding that have been recently described include portal colopathy, diversion colitis, and gastrointestinal bleeding in runners.3

This case should raise awareness among clinicians, that although rare, severe GI vasculitis may be the only presenting symptom of many conditions. Making the diagnosis may be difficult, even with the aid of imaging studies. Early diagnosis is essential to prevent complications and unnecessary surgery. In our case we decided to explore a potential source of hemorrhage in the small intestine by introduction of combined surgical and enteroscopic interventions. Through the enterotomy section we introduced the enteroscope from the duodenum to ileum, where we found the source of the GI bleeding.

REFERENCES