ANCA-associated Leukocytoclastic Vasculitis with Mesenteric Involvement

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Introduction

Leukocytoclastic vasculitis (LCV) is a usually self-limiting, idiopathic small-vessel vasculitis with cutaneous symptoms. Extracutaneous manifestations should raise suspicion for underlying autoimmune conditions such as ANCA-associated vasculitis. Mesenteric involvement is uncommon and usually represents advanced disease. Delay in recognition of mesenteric vasculitis can lead to progression and perforation.

We present a case of gastrointestinal (GI) hemorrhage caused by ANCA-associated LCV.

Learning Objectives

- Identify the mesenteric involvement of ANCA-associated vasculitis and its clinical manifestations
- Recognize the endoscopic and histopathologic findings of mesenteric vasculitis

Case Presentation

A 69-year-old male with history of bilateral inguinal hernia repair was admitted for post-surgical groin wound infection. He was started on antibiotics and underwent mesh removal, washout, and debridement. He developed shock, renal failure, small vessel ischemic cerebrovascular infarction, and upper extremity papular eruptions.

- ↑ inflammatory markers
- ↓ C3
- (-) ANA
- (+) C-ANCA
- (-) P-ANCA
- Met Sapporo criteria for Antiphospholipid syndrome given small vessel arterial thrombosis and (+) Lupus Anticoagulant
- Shave biopsy of skin lesions: LCV

Imaging

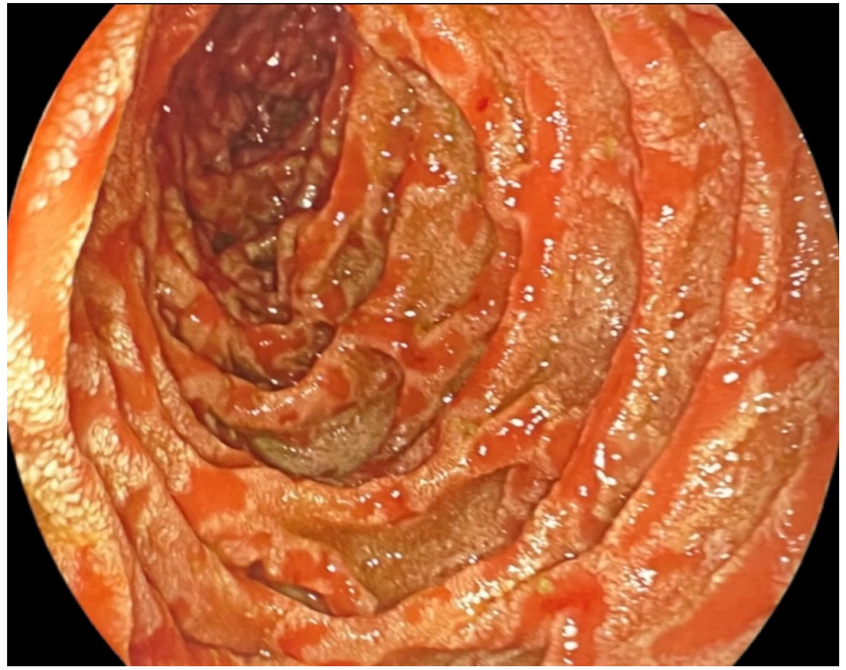
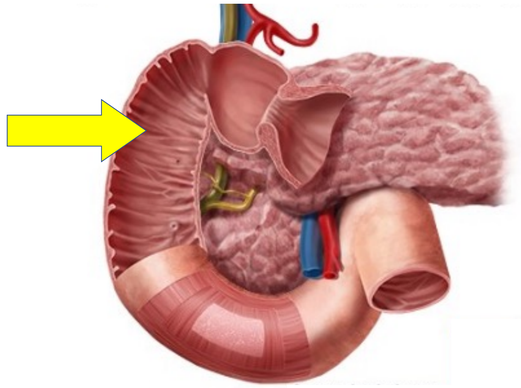


Figure 1. Endoscopic evidence of diffuse erosive duodenopathy interspersed between normal mucosa in the setting of mesenteric vasculitis

Hospital Course

He was started on IV methylprednisolone and heparin infusion. He subsequently developed melena and anemia. EGD showed grade C esophagitis, gastropathy, and diffuse erosive duodenopathy (Fig 1). He was diagnosed with **ANCA-associated LCV with mesenteric involvement.** Bleeding resolved after treatment with pantoprazole and holding heparin. He was transitioned to prednisone and started on rituximab with significant improvement.

Figure 2. Arrow indicates the second portion of the duodenum



Discussion

Mesenteric vasculitis merits consideration in patients with abdominal pain or GI bleeding and clinical evidence of systemic autoimmune disease.

Symptoms arise from intestinal ischemia which can progress to infarction and perforation. Endoscopy should be pursued with great caution due to increased perforation risk. Histopathology of superficial mucosal biopsies usually shows mucosal damage but does not yield a definitive diagnosis. Classic endoscopic findings include mucosal pallor, edema, or necrosis dispersed between normal mucosa. Ulcers and bleeding can be seen in severe cases.

Timely recognition of GI manifestations of ANCA-associated LCV and initiation of therapy are pivotal in ensuring reduction of morbidity, mortality, and the need for surgery. A high index of suspicion for infarction or perforation warrants early surgical intervention.