ACG CASE REPORTS JOURNAL

CASE REPORT | INFLAMMATORY BOWEL DISEASE

Isolated Celiac Artery Vasculitis Presenting as Ileus in a Patient With Ulcerative Colitis

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ABSTRACT

There is a known association between inflammatory bowel disease (IBD) and vasculitis, which can present with mesenteric ischemia or cutaneous manifestations. Infliximab, an anti-tumor necrosis factor (anti-TNF) used to treat IBD, has been implicated as a potential culprit. We present a unique case of a patient with ulcerative colitis who developed isolated celiac artery vasculitis presenting with abdominal pain and ileus after decreasing his dosage of azathioprine. Vasculitis resolved with steroids and increasing azathioprine dosage, while continuing anti-TNF therapy, suggesting that anti-TNF agents are not the only cause of vasculitis in patients with IBD or that thiopurines may be protective.

KEYWORDS: Ulcerative Colitis; Vasculitis; Inflammatory bowel disease

INTRODUCTION

Vasculitis is typically grouped by vessel size or by presence of specific immune markers.[1] Systemic vasculitides can affect multiorgan systems, with gastrointestinal (GI) tract involvement more commonly noted with polyarteritis nodosa, granulomatosis with polyangiitis, or IgA vasculitis. Inflammatory bowel disease (IBD) has been associated with large-vessel vasculitis, such as Takayasu arteritis, ANCA-associated vasculitis, and cutaneous vasculitides. Clinical manifestations include abdominal pain, diarrhea, fever, ileus, or GI bleeding, and complications can include intestinal ischemia or perforation.[2] Isolated GI tract vasculopathy has been reported, and patients may present with GI ischemia without other systemic findings.[3] Of these, isolated celiac artery vasculitis has been rarely reported in the literature. We describe the first case of isolated celiac artery vasculitis in a patient with ulcerative colitis (UC) presenting with abdominal pain and ileus after decreasing his dosage of azathioprine (AZA).

CASE REPORT

A 69-year-old man with hypertension and an 11-year history of UC in clinical and endoscopic remission (for at least 2 years) presented with lower abdominal pain, vomiting, and constipation. He was on infliximab 5 mg/kg every 8 weeks and AZA 200 mg daily for the past 8 years, but had decreased AZA to 100 mg daily 1 month before presentation. He initially presented to 2 different emergency departments where he was diagnosed with constipation. Computed tomography (CT) scan of the abdomen/pelvis with contrast revealed no explanation for his symptoms. He presented to our institution the next day because worsening symptoms.

Abdominal/pelvic CT scan with contrast showed stranding surrounding the celiac artery, fluid-filled small bowel loops, and proximal colon distention. Barium lower GI series did not show stricture or obstruction. Mesenteric duplex was inconclusive. He developed progressive small bowel and right colonic distention, and colonoscopy and esophagogastroduodenoscopy (EGD) with nasogastric and colonic decompression tube placement were performed. There was no evidence of active UC. Dilation of the transverse and ascending colon was seen. EGD revealed dusky-appearing mucosa at the gastroesophageal junction and ulceration in the gastric cardia and body (Figure 1). Biopsy of the gastroesophageal junction showed ischemic-type changes and extensive ischemic necrosis (Figure 1). Gastric

ACG Case Rep J 2024;11:e01335. doi:10.14309/crj.0000000000001335. Published online: April 18, 2024

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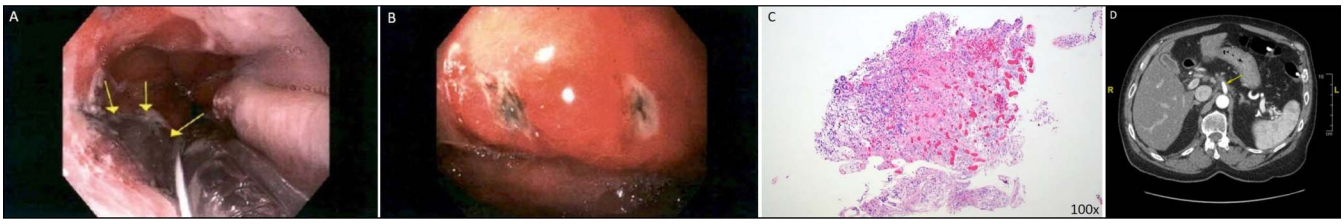

Figure 1. EGD findings show inflammation with a dusky-appearing mucosa (arrows) at the gastroesophageal junction (GEJ) (A). and nonbleeding gastric ulcers with overlying pigment (Forrest class IIc) (B). (C) An H&E stain image showing the GEJ biopsy with extensive ischemic necrosis and hemorrhage. (D) The initial abdominal/pelvic CTA with fat stranding surrounding the celiac artery (arrow showing celiac artery). EGD, esophagogastroduodenoscopy.

biopsies showed chronic gastritis, focal hemorrhage, mild subacute ischemic type changes, and *Helicobacter pylori* gastritis. There was no previous EGD for comparison and no history of ischemic events. Celiac artery vasculitis was presumed to be the etiology.

CT angiography (CTA) was performed and demonstrated perivascular inflammation of a patent celiac artery without evidence for dissection, suggestive of acute vasculitis (Figure 1). Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were 50 mm/hr and 7.8 mg/dL, respectively. Myeloperoxidase (MPO, tests for p-ANCA), serine-proteinase 3 (tests for c-ANCA), rheumatoid factor, and antinuclear antibodies were negative. Rheumatology was consulted and concluded; this was not a systemic vasculitis.

The patient received intravenous methylprednisolone 1 g daily for 3 days, with resolution of symptoms. He was discharged on oral prednisone 20 mg 3 times a day for 2 weeks, with a tapering schedule. AZA was increased back to 200 mg daily. Infliximab was continued. Oral proton pump inhibitor treatment for *H. pylori* was recommended. Abdominal/pelvic follow-up CTA 1 month later showed minimal residual inflammation. ESR and CRP had normalized. Repeat EGD was not performed, but the patient remains asymptomatic 1 year later.

DISCUSSION

The systemic vasculitides cause inflammation and fibrinoid necrosis of blood vessel walls and may lead to tissue ischemia or necrosis. Vasculitis may be primary or secondary to infection, malignancy, or autoimmune diseases.[1,4]

Systemic vasculitides affecting the GI tract are categorized as small- or medium-sized vessel disorders, including polyarteritis nodosa, granulomatosis with polyangiitis, microscopic polyangiitis, and Henoch-Schönlein purpura. In patients with systemic vasculitis, GI manifestations usually indicate a poor prognosis.[2] Even localized vasculitis in the GI tract can be associated with significant morbidity and mortality.[5] Endoscopic findings such as erosions, petechiae, ulcers, submucosal hemorrhage, and strictures strengthen the suspicion for vasculitis.[6] In our patient, the dusky-appearing gastroesophageal junction and ulcers on endoscopy put vasculitis high on our differential.

Small case series have reported an association between IBD and large-vessel vasculitis, ANCA-associated vasculitides, and cutaneous vasculitides. There are roughly 100 case reports that report an association between Takayasu arteritis and IBD. It is unclear whether these patients have a primary vasculitis with GI involvement or IBD with secondary vasculitis. In a case series and literature review, IBD preceded the diagnosis of vasculitis in 12/13 patients with large-vessel vasculitis and in all 8 cases of ANCA-associated vasculitis. There was no case of isolated celiac artery vasculitis noted.[7] Previous case reports raised suspicion that the vasculitis was drug-induced by anti-TNF agents.[8–12] One case series showed the mean duration of treatment before the development of leukocytoclastic vasculitis as 30.8 months, whereas another showed it to be 34.5 months for cutaneous small-vessel vasculitis.[8] The treatment in some cases was to use AZA, but overall included stopping anti-TNF therapy, changing to a different anti-TNF drug, and starting steroids. Some suggested changing to a different biologic class.

Our case is unique because isolated celiac artery vasculitis occurred in our patient with IBD on anti-TNF therapy and did not recur while anti-TNF therapy was continued. Therefore, it seems that anti-TNF therapy was not the cause. Although infliximab levels and antibodies to infliximab were not checked, the UC was clearly in remission. A possible explanation is that decreasing the dose of AZA precipitated the episode. Our patient had not been on other drugs associated with vasculitis and had a large-vessel vasculitis, but not a systemic vasculitis. Our patient presented to 2 separate emergency departments before presenting at our hospital, highlighting the difficulty in making the diagnosis of vasculitis.

To the best of our knowledge, this is the first case of isolated celiac artery vasculitis in a patient with IBD on anti-TNF therapy. Long-term resolution was achieved with steroids and increased thiopurine dose while continuing anti-TNF therapy. This suggests that anti-TNF therapy may not be the cause of vasculitis in patients with IBD or that thiopurines may have a protective effect. In addition, there should be a high level of suspicion for vasculitis in patients with IBD who present with abdominal pain and ileus.

DISCLOSURES

Author contributions: S. Ali, M. Doniparthi, and A. Shapiro contributed to writing and editing the manuscript. N. Asado

and K. Borgen provided the pathology images. S. Ali is the article guarantor.

Financial disclosure: None to report.

Previous presentation: This case was presented as a poster presentation at the ACG conference; October 23, 2022; Charlotte, North Carolina.

Informed consent was obtained for this case report.

Received November 26, 2023; Accepted March 18, 2024

REFERENCES

1. Guillevin L, Dörner T. Vasculitis: Mechanisms involved and clinical manifestations. *Arthritis Res Ther.* 2007;9(Suppl 2):S9.
2. Chait J, Pavalonis A, Rajaee S, Hingorani A, Ascher E. Celiac artery vasculitis. *Ann Vasc Surg.* 2019;56:354.e17–e19.
3. Louie C, DiMaio M, Charville G, Berry G, Longacre T. Gastrointestinal tract vasculopathy: Clinicopathology and description of a possible "new entity" with protean features. *Am J Surg Pathol.* 2018;42(7):866–76.
4. Jarrot PA, Kaplanski G. Anti-TNF-alpha therapy and systemic vasculitis. *Mediators Inflamm.* 2014;2014:493593.
5. Salvarani C, Calamia KT, Crowson CS, et al. Localized vasculitis of the gastrointestinal tract: A case series. *Rheumatology (Oxford).* 2010;49(7):1326–35.
6. Hatemi I, Hatemi G, Çelik AF. Systemic vasculitis and the gut. *Curr Opin Rheumatol.* 2017;29(1):33–8.
7. Sy A, Khalidi N, Dehghan N, et al. Vasculitis in patients with inflammatory bowel diseases: A study of 32 patients and systematic review of the literature. *Semin Arthritis Rheum.* 2016;45(4):475–82.
8. Sokumbi O, Wetter DA, Makol A, Warrington KJ. Vasculitis associated with tumor necrosis factor-α inhibitors. *Mayo Clin Proc.* 2012;87(8):739–45.
9. Rogerio P, Julio C, Liliana C, et al. P007 anti-TNF-therapy-induced leukocytoclastic vasculitis: A rare complication in IBD patients. *Am J Gastroenterol.* 2020;115(Suppl 1):S2.
10. Bouhuys M, Armbrust W, van Rheenen PF. Case report: Systemic small-vessel vasculitis in an adolescent with active ulcerative colitis. *Front Pediatr.* 2021;9:617312.
11. Rasmussen C, Abitbol V, El Karoui K, et al. IgA vasculitis in patients with inflammatory bowel disease: New insights into the role of TNF-α blockers. *Rheumatology (Oxford).* 2022;61(5):1957–65.
12. Rivas S, Pandya AG, Dominguez AR. Drug-induced vasculitis. In: Hall J, Hall B (eds). *Cutaneous Drug Eruptions.* Springer: London, UK, 2015, pp 77–85.

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