



A Case of Enterocolic Lymphocytic Phlebitis Mimicking Surgical Abdomen

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Vasculitis that involves the gastrointestinal (GI) tract often occurs as part of a systemic inflammatory process. It is a well-recognized manifestation of the small and medium sized vessel vasculitides. Vasculitis of the GI tract may occur in isolation; although it can progress to a systemic illness. It usually involves the arterioles, venules, and capillaries; however, it is very rare for only the venules to be affected. Enterocolic lymphocytic phlebitis is a localized vasculitis, typically affecting the small and medium-sized intramural and mesenteric veins of the intestines. We report a case of enterocolic lymphocytic phlebitis of the colon. A 38-year-old woman was presented with hema-

tochezia and severe abdominal pain on the day of admission. She had no history of intestinal disease or systemic disease. Computed tomography showed an extremely thickened wall of the colon, along with several air bubbles in the colon with diffuse subcutaneous emphysema in the abdominal wall. An emergency exploration laparotomy and extended right hemicolectomy was performed. The patient recovered completely after surgery and remains well without further therapy.

Key Words. Enterocolic lymphocytic phlebitis, Localized gastrointestinal vasculitis, Single-organ vasculitis

Introduction

The term acute abdomen refers to signs and symptoms of abdominal pain with tenderness and a clinical presentation that often requires emergency surgical therapy (1). One cause of acute abdomen is intestinal ischemia that can result from vasculitis.

Various vasculitides affect the gastrointestinal (GI) tract and are classified by type of affected vessel, etiology and underlying disease. Cancer, infection, drug, and systemic autoimmune disease can present as GI vasculitis (2). Primary systemic vasculitis also affect the GI tract, especially polyarteritis nodosa, granulomatosis with polyangiitis (Wegener's granulomatosis), eosinophilic granulomatosis with polyangiitis (Churg-Strauss syndrome) and microscopic polyangiitis (3). Localized vasculitis of the GI tract is very rare.

Enterocolic lymphocytic phlebitis (ELP) is a localized vasculitis typically affecting small and medium-sized intramural and

mesenteric veins of the intestines (4,5). We report a case of ELP of the colon that was clinically and radiologically suspected as intestinal ischemia accompanied by necrosis and perforation of the colon.

Case Report

A 38-year-old woman was admitted to the hospital because of hematochezia on the day of admission and severe abdominal pain with diarrhea of 36 hours' duration. She had previously been healthy and had no history of intestinal or systemic disease. She was a non-smoker and not taking any medications. Blood pressure was 110/60 mmHg with a regular heart rate of 96/min; body temperature at the time of admission was 37.0°C. On physical examination, the abdomen was remarkable for tenderness in the right upper and lower with rebound tenderness. A digital rectal examination revealed fresh and clotted blood in the rectum.

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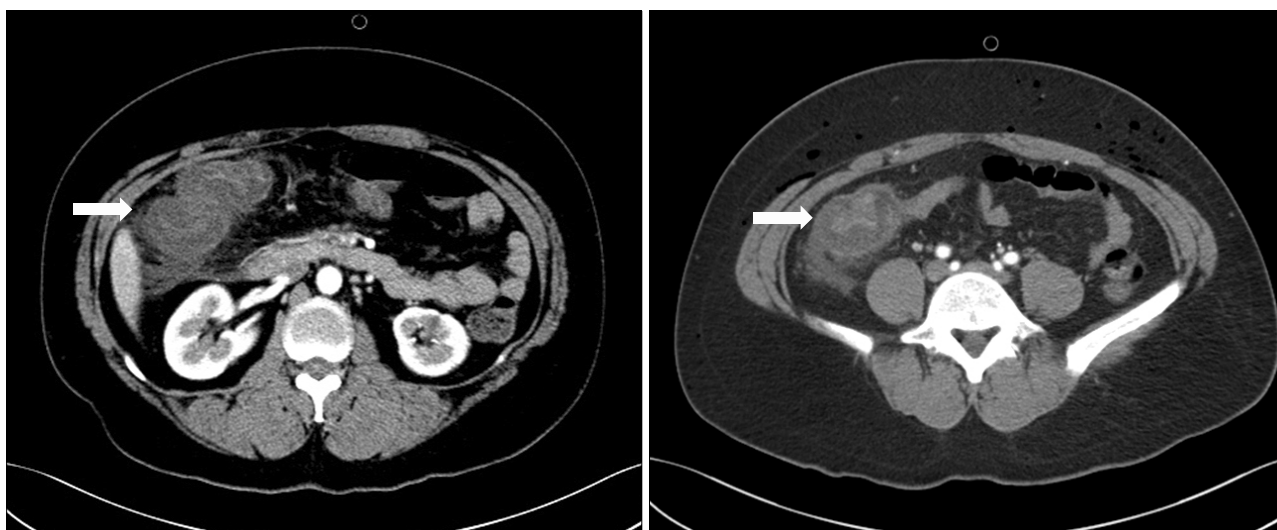


Figure 1. Abdominal CT shows an extremely thickened wall of the ascending colon (arrow) with diffuse scattered subcutaneous emphysema in the abdominal wall.

A complete blood count revealed white cell count of 12,670 cells/mm³ (neutrophil 80.6%, lymphocyte 11.4%), hemoglobin of 13.0 g/dL and platelet count of 255,000 cells/mm³. CRP was 6.19 mg/dL (normal range 0~0.5 mg/dL). Blood chemistry revealed total protein 6.2 g/dL, albumin 3.8 g/dL, BUN 11.3 mg/dL, creatinine 0.8 mg/dL, AST 8 IU/L and ALT 9 IU/L. Urinalysis showed neither microscopic hematuria nor proteinuria. Hepatitis B surface antigen and hepatitis C antibodies were negative.

The plain chest and abdomen radiographs were normal. Computed tomography (CT) of the abdomen showed extremely thickened wall of the ascending colon and proximal portion of the transverse colon. There was an extreme inflammatory process in the pericolic and mesenteric area, several small air bubbles in the ascending colon, and diffuse scattered subcutaneous emphysema in the abdominal wall. There was also a small amount of ascites in the right paracolic gutter. Those findings were most likely due to underlying ischemic colitis, possible necrosis and perforation of the ascending colon (Figure 1).

An emergency exploratory laparotomy was done. The surgical findings were dilatation of the cecum, wall thickening in the mid portion of the ascending colon, intact ileocolic and middle colic artery, but no perforation. Because multiple lesions with necrotic change were found, an extended right hemicolectomy was performed.

Histopathologic findings: The resected tissue consisted of 30.5 cm length of the terminal ileum and 41.4 cm length of the colon. There were no specific findings in the serosa. A necrotic lesion 17 cm in length was visible in the mucosa of

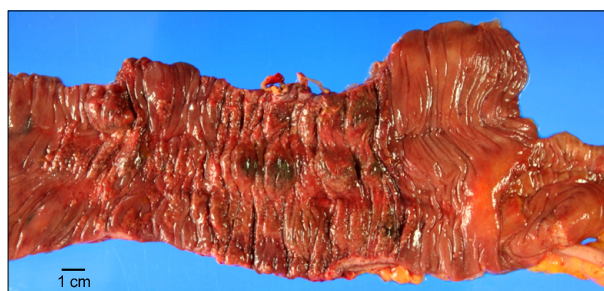


Figure 2. Gross features of the specimen. An ill defined lesion with irregular surface is localized in ascending colon. The mucosa of the lesion is necrotic, stiff and reddish.

the ascending colon, focally covered by a hemorrhagic exudate. Marked edematous mucosa surrounded the necrotic lesion. The necrotic lesion was firmer and thicker than surrounding tissue. There were no pathologic findings in the terminal ileum (Figure 2).

Histologically, in sections from the necrotic lesion, ischemic necrosis was found in the mucosa. Acute inflammation, congestion and edema were found in the submucosa. In particular, many small veins of the submucosa, muscularis propria and subserosa presented with inflammation infiltrate of the vessel walls, composed mostly of lymphocytes. In some affected vessels, thrombosis was found. The arteries were normal, not affected by inflammation (Figure 3).

Progress: After surgery, the diarrhea and hematochezia disappeared and acute inflammatory markers normalized. The surgeon referred this patient to a rheumatologist to rule out systemic autoimmune disease because she was relatively young and had no risk factors for bowel ischemia. The authors took

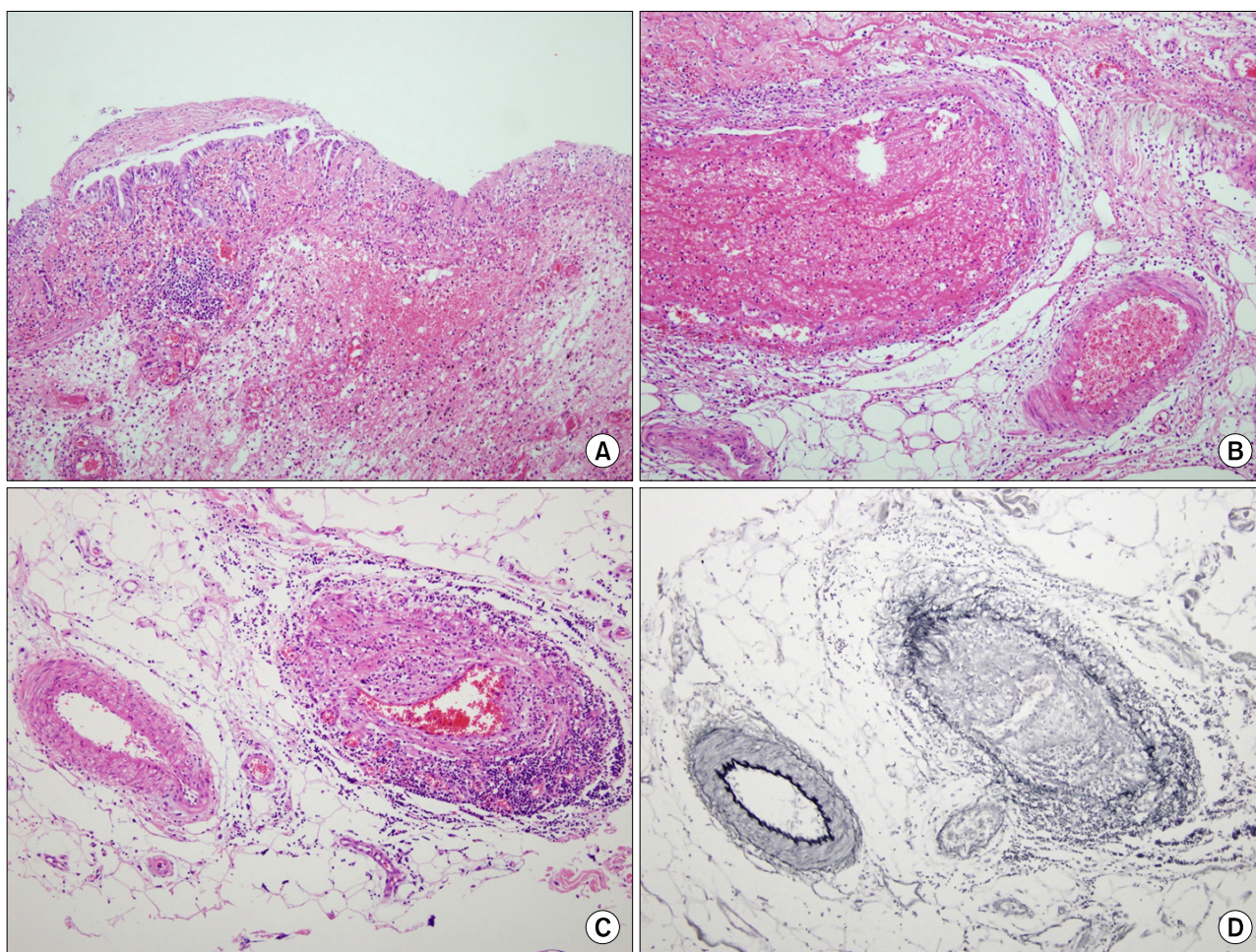


Figure 3. Microscopic findings of enterocolic phlebitis. The lesional mucosa and submucosa are shown in (A). The mucosa reveals ischemic necrosis and the submucosa is hemorrhagic and edematous. The submucosal veins of the lesion (B) are filled with thrombi, and show necrotizing vasculitis with fibrinoid degeneration. The vessels at the periphery (C and D) show lymphocytic phlebitis, relatively. Special stain for elastin (D) demonstrated that arteries are spared. (A-C, Hematoxylin-eosin stain $\times 200$; D, Elastic stain $\times 200$).

more history and ran tests to evaluate systemic autoimmune disease, especially systemic vasculitis, and hypercoagulable states. She had no history of hypersensitivity to sunlight, malar rash, arthritis, oral ulcerations, genital ulcerations and eye disease. She was unmarried with no history of pregnancy. Rheumatoid factor, antinuclear antibodies, antiphospholipid antibodies (lupus anticoagulant, anticardiolipin IgG/IgA/IgM, and anti-beta2-glycoprotein I IgG/IgM) and antineutrophil cytoplasmic antibodies were negative. Complement levels of C3 and C4 were within a normal range. Hypercoagulable workup showed no abnormalities (prothrombin time, activated partial thromboplastin time, protein C, protein S, Factor V Leiden, antithrombin, homocysteine, cryoglobulin). Stool bacteriological and parasitological examinations were negative. In mesenteric CT angiography and echocardiography, there were no abnormal findings. There were no other manifestations of autoimmune diseases such as systemic lupus erythematosus,

Behcet's disease or systemic vasculitides.

Therefore, we diagnosed her as ELP by clinical manifestations and histologic findings. The patient has recovered completely after the surgery without other therapy and was symptom-free as of six months post-surgery.

Discussion

The vasculitides are a heterogeneous group of disorders characterized by inflammation of blood vessel walls. The etiology is usually unknown. Therefore, the vasculitides are mainly classified by the type of vessels predominantly involved. Another practical approach is to divide them into systemic or localized vasculitides. The 2012 revised Chapel Hill Consensus Conference defined the single-organ vasculitis as vasculitis in arteries or veins of any size in a single organ, with no features indicating that it is a limited expression of a systemic vasculitis (2). Therefore, the term "single-organ

vasculitis" is synonymous with localized vasculitis (6).

Vasculitis of the GI tract is a rare condition. The manifestations of GI symptoms are wide, ranging from mild transient abdominal pain to surgical abdomen. Systemic disorders such as systemic lupus erythematosus, Behcet's disease, mixed connective tissue disease and rheumatoid arthritis can manifest as vasculitis of GI tract (7). Indeed, primary systemic vasculitides, especially medium and small vessel vasculitides, may involve the GI tract. GI tract involvement of systemic vasculitides has been reported at rates of 40~60% for polyarteritis nodosa, 30~56% for microscopic polyangiitis, 20~50% for eosinophilic granulomatosis with polyangiitis, and 5~11% for granulomatosis with polyangiitis (3). GI involvement occurs in IgA vasculitis (Henoch-Schönlein purpura), Kawasaki disease, Takayasu arteritis and giant cell arteritis (4,5,8). The GI manifestations could be the first signs of those diseases, and progress to systemic disorders. It is important to be aware that isolated vasculitis of GI tract carries a risk for progression to a systemic vasculitis. Therefore, isolated vasculitis should be followed for at least six months before completing the diagnosis (9).

Localized vasculitis of the GI tract is an extremely rare disease. Pagnoux and colleagues reviewed a series of small and medium-sized vessel vasculitides with GI tract involvement; only one of 62 patients presented with isolated vasculitis of GI tract (3). Endoscopic biopsies have a low sensitivity to diagnose GI vasculitis due to unable to obtain affected vessels. In general, localized vasculitis of the GI tract is diagnosed after surgery (4,5,8,10). Burke and colleagues classified isolated vasculitis of the GI tract into six groups histologically. Those were polyarteritis, ELP, eosinophilic necrotizing inflammation of both arteries and veins, small-vessel vasculitis, thromboangiitis obliterans, and giant cell arteritis (10).

The one cause of localized vasculitis of the GI tract is ELP. This term is synonymous with mesenteric inflammatory veno-occlusive disease, intramural mesenteric vasculitis, isolated granulomatous phlebitis, lymphocytic venulitis, idiopathic myointimal hyperplasia of mesenteric veins, and necrotizing and giant cell granulomatous phlebitis (11). ELP is a rare type of venulitis involving only the intramural and mesenteric veins and venules of bowel wall. It spared the arteries and arterioles as well as the systemic circulation. The inflammation can lead to thrombotic obstruction and fibrointimal proliferation with subsequent venous occlusion, causing edema and ischemia of the involved intestinal segment. It can involve all sites of the intestine, but the commonly affected sites are in the large bowel, predominantly right colon (7,11). Clinically, the presenting feature of ELP is subacute to acute intestinal ischemia

manifested as abdominal pain, hematochezia, and bloody diarrhea. The endoscopic or imaging findings are not diagnostic, but computed tomography scanning often showed thickened and edematous bowel walls. Our patient presented with acute abdomen and remarkably thickened and edematous right colon wall in CT. The etiology and pathogenesis of the disease are undetermined, although the associations with some medications, diversion colitis and lymphocytic colitis have been proposed (12-14). But our patient was not taking any medications and reported no history of surgery or previous GI symptoms. The diagnosis of ELP depends on histopathology. ELP was refractory to medical treatment and surgical resection of the affected bowel is usually curative without any relapse. Of reported ELP, none progressed to systemic vasculitis to our knowledge (11-14).

Vasculitis of the GI tract can present as surgical abdomen. If acute intestinal infarction develops in a young patient without any risk factors of bowel ischemia, localized vasculitis such as ELP should be considered. The authors report a case of ELP that presented as an acute abdomen and recovered after surgery without other systemic treatment.

Summary

We report a 38-year-old woman presenting with acute abdominal pain and bloody diarrhea. We first suspected intestinal ischemia leading to necrosis and bowel perforation. The patient underwent emergency surgery. The pathologic diagnosis was ELP. She completely recovered after right hemicolectomy without further immunosuppressive treatment. GI vasculitides can be a rare but noteworthy cause of acute abdomen.

References

1. Ronald AS, Russell GP. Townsend: Sabiston Textbook of Surgery. 19th ed. p. 1141, Philadelphia, Saunders, An imprint of Elsevier, 2012.
2. Jennette JC, Falk RJ, Bacon PA, Basu N, Cid MC, Ferrario F, et al. 2012 revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. *Arthritis Rheum* 2013;65:1-11.
3. Pagnoux C, Mahr A, Cohen P, Guillevin L. Presentation and outcome of gastrointestinal involvement in systemic necrotizing vasculitides: analysis of 62 patients with polyarteritis nodosa, microscopic polyangiitis, Wegener granulomatosis, Churg-Strauss syndrome, or rheumatoid arthritis-associated vasculitis. *Medicine (Baltimore)* 2005; 84:115-28.
4. Gonzalez-Gay MA, Vazquez-Rodriguez TR, Miranda-Filloy JA, Pazos-Ferro A, Garcia-Rodeja E. Localized vasculitis of the gastrointestinal tract: a case report and literature review. *Clin Exp Rheumatol* 2008;26(3 Suppl 49):S101-4.

5. Salvarani C, Calamia KT, Crowson CS, Miller DV, Broadwell AW, Hunder GG, et al. Localized vasculitis of the gastrointestinal tract: a case series. *Rheumatology (Oxford)* 2010;49:1326-35.
6. Atisha-Fregoso Y, Hinojosa-Azaola A, Alcocer-Varela J. Localized, single-organ vasculitis: clinical presentation and management. *Clin Rheumatol* 2013;32:1-6.
7. Ahn E, Luk A, Chetty R, Butany J. Vasculitides of the gastrointestinal tract. *Semin Diagn Pathol* 2009;26:77-88.
8. Garcia-Porrúa C, Gutierrez-Duque O, Soto S, Garcia-Rodeja E, Gonzalez-Gay MA. Localized vasculitis of the gastrointestinal tract. *Semin Arthritis Rheum* 2006;35:403-6.
9. Hernández-Rodríguez J, Hoffman GS. Updating single-organ vasculitis. *Curr Opin Rheumatol* 2012;24:38-45.
10. Burke AP, Sobin LH, Virmani R. Localized vasculitis of the gastrointestinal tract. *Am J Surg Pathol* 1995;19:338-49.
11. Ngo N, Chang F. Enterocolic lymphocytic phlebitis: clinicopathologic features and review of the literature. *Arch Pathol Lab Med* 2007;131:1130-4.
12. Jain R, Chetty R. Enterocolic lymphocytic phlebitis and lymphocytic colitis: drug-related coexistent pathology. *Int J Colorectal Dis* 2009;24:473-4.
13. Saraga E, Bouzourenne H. Enterocolic (lymphocytic) phlebitis: a rare cause of intestinal ischemic necrosis: a series of six patients and review of the literature. *Am J Surg Pathol* 2000;24:824-9.
14. Chetty R, Hafezi S, Montgomery E. An incidental enterocolic lymphocytic phlebitis pattern is seen commonly in the rectal stump of patients with diversion colitis superimposed on inflammatory bowel disease. *J Clin Pathol* 2009;62:464-7.