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An uncommon form of localized colonic eosinophilic vasculitis with extensive thrombosis of the spleen and liver: A case report and literature review

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ABSTRACT

INTRODUCTION: Localized vasculitis of the gastrointestinal tract is an uncommon disease that mainly presents as polyarteritis nodosa and is mainly located on small bowel and gall bladder. Localized eosinophilic vasculitis of the colon, which needs surgical intervention, has never been reported before. **CASE PRESENTATION:** A 40-year-old man was diagnosed with localized eosinophilic vasculitis of the colon with an initial presentation of necrotizing colitis of ascending colon. After right hemicolectomy, extensive thrombosis of the liver and spleen occurred with the presentation of abdominal pain. The histopathological analysis revealed ischemic colitis with eosinophilic vasculitis in medium-sized vessels throughout the colon. The thrombosis was improved after prednisolone and azathioprine were given. The results of all autoimmune tests, including those for anti-neutrophil cytoplasmic antibodies, were all negative except the elevation of serum immunoglobulin E (680 kU/L [normal, <25 kU/L]).

CONCLUSION: Although the patient failed to meet the criteria of the Churg-Strauss syndrome, this case may represent an atypical localized variant of eosinophilic vasculitis of the gastrointestinal tract. Immunosuppressant therapy should be considered after surgery.

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1. Introduction

Localized vasculitis of the gastrointestinal tract (LVGT) is an uncommon type of vasculitis that affects the entire gastrointestinal system, including the esophagus, stomach, small and large intestines, appendix, omentum, gallbladder, and pancreas [1]. Most LVGT cases present as polyarteritis nodosa, and eosinophilic vasculitis has been rarely reported so far [1,2]. Although some LVGT cases can be treated through surgical resection alone, many patients still require following immunosuppressant therapy [1–6].

The work of this case has been reported in line with the SCARE 2018 criteria [7].

2. Patient information

A 40-year-old man presented to our emergency department with progressive periumbilical pain and bloating for three days.

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He denied nausea, vomiting, diarrhea, bloody stool, or fever. He was generally healthy before, denied any substance using (Tabaco, alcohol or illicit substances). He also denied any systemic diseases, including asthma or allergic rhinitis. He reported no any surgery received before nor hereditary diseases in his family.

2.1. Clinical findings and diagnostic assessment

Physical examination revealed lower abdominal tenderness without any signs of peritonitis. Abdominal computed tomography (CT) revealed right side colitis with pericolic fat stranding. Laboratory tests revealed leukocytosis (white blood cell [WBC] count, 24,200/ μ L) with left-shift granulocyte (segment 68%, band 3%), elevated C-reactive protein levels (50.78 mg/dL), and absolute eosinophil count (AEC) of 968/ μ L. The patient was initially admitted with the diagnosis of colitis. Bowel rest and intravenous antibiotics were given initially. However, progressive abdominal pain and diffuse peritoneal signs were presented three days after admission. Subsequent laboratory tests showed progressively elevated C-reactive protein (153.67 mg/L), leukocytosis (WBC count, 24,200/ μ L), and marked eosinophilia (AEC, 3316/ μ L). In contrast, serum lipase (24 U/L) and aspartate transaminase (22 U/L) were normal.

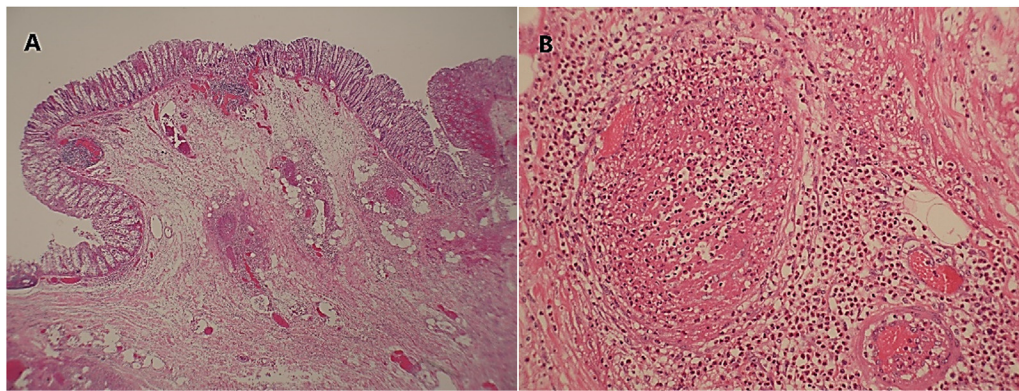


Fig. 1. Histopathology of the resected colon. (a) Low-power magnification: thrombosis of blood vessels and multiple foci of necrosis. (b) High-power magnification: thrombus formation and dense eosinophilic infiltration of the vessels.

2.2. Therapeutic intervention

The patient underwent laparoscopic right colectomy on the third day of admission. Ascending colon inflammation with omentum shifting and mild turbid ascites were observed intraoperatively. The postoperative course was smooth, except for persistent eosinophilia with an AEC of 8277/ μ L on a postoperative day 9, the date of discharge. Histopathological analysis revealed ischemic colitis with eosinophilic vasculitis in medium-sized vessels throughout the colon, including the resection margin (Fig. 1).

However, the patient returned to the emergency department three days after discharge due to diffuse, dull pain in the abdomen. Abdominal CT revealed multiple tiny hypodense lesions over the spleen and liver. Laboratory test results revealed a WBC count of 12,200/ μ L and AEC of 3720/ μ L; the serum C-reactive protein level was elevated (119 mg/L), and erythrocyte sedimentation rate (ESR) was 59 mm/h. The symptoms of abdominal pain improved after methylprednisolone (80 mg per day) and enoxaparin administration. Subsequent tests revealed elevated serum total immunoglobulin (Ig) E levels (680 kU/L [normal, <25 kU/L]). The test results for hepatitis B and C viruses were negative. Autoimmune test results revealed anti-neutrophil cytoplasmic antibody, antinuclear antibody, rheumatoid factor IgM, anti- β 2 glycoprotein I IgG, anti-cardiolipin IgG, anti-SSA/B antibodies, protein-S, and protein-C levels to be either negative or within normal limits. Under stable conditions, immunosuppressants (prednisolone, 50 mg, and azathioprine, 50 mg per day) and anticoagulants were provided to the patients orally.

2.3. Follow-up and outcomes

The patient was discharged and followed up in the outpatient department. Oral prednisolone dosage was gradually tapered to 2.5 mg/day, whereas azathioprine was discontinued 1 month after discharge. ESR and AEC gradually returned to normal limits after 2 and 6 months of follow-up, respectively.

3. Discussion

This case report illustrates that LVGT can initially present as eosinophilic vasculitis of the colon with symptoms of acute abdomen due to necrotizing colitis. The patient in the current study did not present with paranasal sinusitis, transient pulmonary opacities or a history of asthma, and therefore, failed to meet the criteria of eosinophilic granulomatosis with polyangiitis (EGPA). Furthermore, the histology of the resected colon demonstrated eosinophilic vasculitis instead of diffuse layers of infiltration by

eosinophils, thus making the diagnosis of eosinophilic gastroenteritis unlikely [8,9]. Most LVGT cases present as polyarteritis nodosa, and localized eosinophilic vasculitis in the gastrointestinal tract has been reported in only two cases [2,3,6]. In the largest case series of the Mayo Clinic, which included 18 cases over a 12-year period, only one case of large bowel infarction was observed, and no eosinophilic vasculitis was reported [1]. Another case series focusing on incidental LVGT over a 12-year period included nine cases, of which one presented with eosinophilic venulitis of the appendix; the patient recovered well after appendectomy [2]. To the best of our knowledge, this is the first report of eosinophilic vasculitis (i.e., LVGT) of the colon, which initially presented as necrotizing colitis and required surgical intervention.

The laboratory findings in patients with LVGT are mostly non-specific, and the autoantibody profiles are mostly negative [1–3]. In addition to eosinophilia and elevated C-reactive protein level and ESR, the autoimmune tests in the current report were negative with IgE elevation as the only positive finding. Although the course of eosinophilic vasculitis with end-organ damage was similar to that of EGPA, the absence of a history of asthma failed to meet the criteria. Similarly, another case (a 13-year-old boy) with isolated eosinophilic mesenteric vasculitis and extensive thrombosis presented with negative serological test results and no history of asthma, which implies the possibility of an atypical presentation of EGPA [3].

LVGT is mainly treated through surgery initially, but following immunosuppressive therapy are often required in many cases [1–6]. Of the two cases with localized eosinophilic vasculitis in the gastrointestinal tract, the one with incidental localized eosinophilic venulitis of the appendix that initially manifested as appendicitis was resolved through appendectomy [2]. By contrast, another 13-year-old boy with eosinophilic mesenteric vasculitis who underwent small bowel resection initially due to infarction received oral prednisolone, cyclophosphamide, and warfarin due to extensive infarction after surgery [3]. In addition, the current patient presented with vasculitis on the surgical margin, which indicated that the condition could extend beyond the infarcted segment. Furthermore, multiple intraabdominal thromboses immediately after the discharge of the patient confirmed the need for follow-up therapy. Hence, immunosuppressive therapy should be provided to patients immediately after surgery to prevent subsequent episodes of thrombosis.

In conclusion, this was the first report of eosinophilic vasculitis, presenting as LVGT of the large bowel with symptoms of acute abdomen followed by spleen and liver thrombosis. In addition to surgical resection, subsequent immunosuppressive therapy should be considered if the patient presents with eosinophilia or vasculitis that extends up to the surgical margin. Although the patient failed

to meet the EGPA criteria, atypical types of LVGT with localized variants should be considered.

Declaration of Competing Interest

The authors report no declarations of interest.

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None.

Ethical approval

Ethical approval has been exempted by my institution for reporting this case.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Huang SH is the first author of the above case report who wrote the case report and completed the literature review.

Liao CK is an attending surgeon took the consent from the patient for publishing this case.

Wu RC is an attending pathologist who did the pathological analysis.

All authors have read and approved the final manuscript.

Registration of research studies

This work is case report and there is no need of registration.

Guarantor

Dr. Chun-Kai Liao is the guarantor for the work.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Patient's perspective

It's never come to my life as a patient of autoimmune disease. I should keep regular follow up with my doctor.

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