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Case Report

Eosinophilic Myenteric Ganglionitis as a Cause of Digestive Tract Perforation

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Abstract

The etiology of eosinophilic myenteric ganglionitis (EMG) remains unclear. We present the case of a 62-year-old man who underwent right hemicolectomy with ileostomy and transverse colon mucous fistula due to ascending colon perforation. Pathological examination revealed severe eosinophilic infiltration in Auerbach's plexus and fibrosis extending from the external longitudinal muscle layer to the subserosal layer, suggesting that the perforation resulted from pseudo-obstruction and EMG-related increased intestinal pressure. Eosinophilic infiltration was observed not only near the perforation site but throughout the entire length of the resected intestine. Four months postoperatively, the patient underwent ileostomy closure, during which the ileal and colonic tracts left external to the wound were resected. Notably, no eosinophilic infiltration in Auerbach's plexus was found in the new specimen, unlike that in the previous surgical specimen, despite the patient receiving no postoperative medication. The patient has remained symptom-free for over 2 years. This is the first report to document histological time-course changes in eosinophil infiltration in Auerbach's plexus and demonstrate the efficacy of surgical treatment in a patient with EMG.

Keywords

Auerbach's plexus, chronic intestinal pseudo-obstruction, eosinophilic myenteric ganglionitis, perforation

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Introduction

Eosinophilic myenteric ganglionitis (EMG) is characterized by marked eosinophilic infiltration of the enteric plexus and is associated with inflammatory neuropathy that mainly occurs in children. Inflammation of the enteric ganglia is recognized as a major cause of functional intestinal obstruction[1-5]. The main diagnostic feature of EMG is eosinophilic infiltration in Auerbach's plexus, manifesting as an intestinal motility disorder and constipation. However, owing to the lack of simple screening tests and diagnostic imaging, EMG has been an under-detected disease, and no standard treatment for EMG has yet been established. Furthermore, histopathological changes that occur over time for EMG are not well understood.

Here, we report a surgical case of a patient with ascending colon perforation resulting from an EMG-derived pseudo-obstruction. To the best of our knowledge, this report is the first to show histological time-course changes in eosinophil infiltration in Auerbach's plexus in a patient with EMG.

Case Report

A 62-year-old man with a history of atopic dermatitis was referred to our hospital with abdominal pain. His height was 180.7 cm, weight was 68.6 kg, and body mass index was 21.0 kg/m^2. On arrival, his body temperature was 37.0°C,

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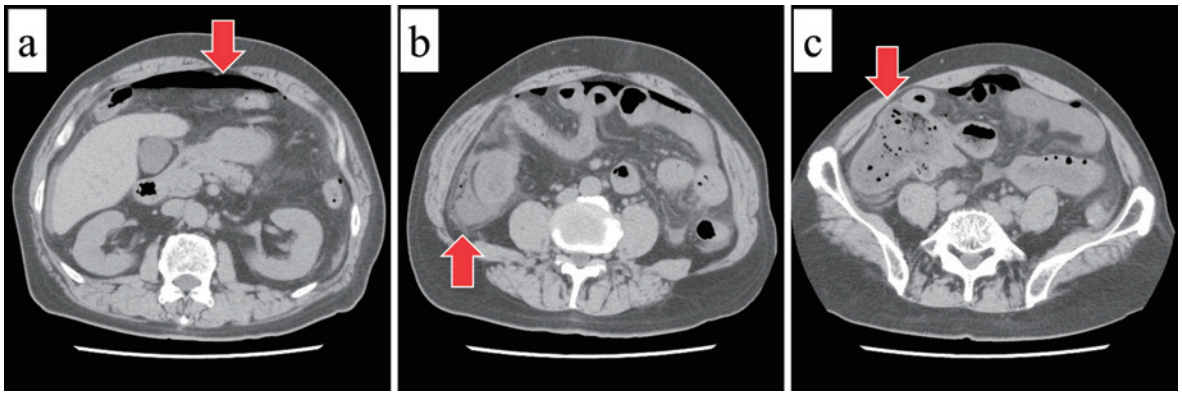

Figure 1. Findings of contrast-enhanced computed tomography.
a. Intra-abdominal free air (arrow indicating free air).
b. Ascitic fluid in the right paracolic gutter (arrow indicating ascitic fluid).
c. Gut distention from the cecum to the ascending colon with fat stranding (arrow indicating cecum).

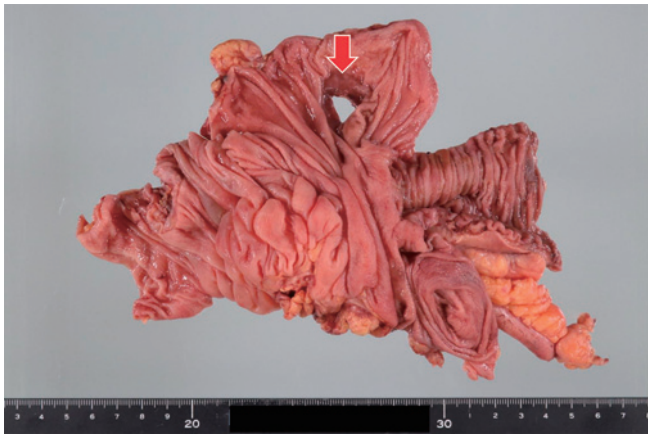

Figure 2. Macroscopic findings of the surgically resected specimen.
Resected specimen showing marked dilatation from the cecum to the ascending colon with perforation of the ascending colon (arrow indicating perforation site).

blood pressure was 150/70 mmHg, and pulse rate was 116 beats/min. Physical examination revealed abdominal swelling, tenderness, and peritoneal signs. Laboratory data indicated a white blood cell count of 1,670/μL (basophils, 0.3%; eosinophils, 0.5%; neutrophils, 68.4%; lymphocytes, 24.5%; monocytes, 6.3%), a C-reactive protein level of 2.78 mg/dL, and a base excess of −1.4 mEq/L. Contrast-enhanced computed tomography showed intra-abdominal free air, gut distention from the cecum to the ascending colon with fat stranding, and ascitic fluid in the Douglas pouch and the right paracolic gutter (Figure 1). Therefore, the patient was preoperatively diagnosed with acute generalized peritonitis secondary to ascending colon perforation. Emergency surgery was performed, and a perforation site with a diameter of 1.5 cm was confirmed in the ascending colon. A one-stage anastomosis was considered to carry a risk of leakage; therefore, right hemicolectomy with ileostomy and transverse colon mucous fistula was performed. Four months postoperatively, the patient underwent ileostomy closure. The ileum and colonic tracts that had been left out of the wound and were located next to the proximal and distal margin of the previously resected intestinal tract were also resected. During the 2-year postoperative period, no abnormalities in intestinal peristalsis have been observed.

Pathological findings

First operation specimen

The resected specimen showed marked dilatation from the cecum to the ascending portion. Histopathological examination revealed no tumor, significant fixed obstruction, or diverticulum in the resected specimen (Figure 2). However, a full-layer wall defect with fibrin deposition and neutrophil infiltration was noted at the perforation site. In addition, diffuse eosinophilic infiltration was observed in Auerbach's plexus, partly within the proper muscular layer, extending beyond the serositis associated with the perforation, including the distal resection margin and Bauhin's valve (Figure 3 a). However, minimal eosinophilic infiltration was noted from the mucosal to the submucosal layer (Figure 3b). The ganglionic structure was maintained, and multifocal fibrosis with vascular proliferation, along with macrophage and lymphocyte infiltration, was observed from the longitudinal muscle layer to the subserosal layer (Figure 3c), indicating chronic changes. The extent of chronic changes exceeded that of secondary serositis due to perforation.

Second operation specimen

Pathological examination revealed no eosinophilic infiltration of Auerbach's plexus, as observed in the previous surgical specimen (Figure 4).

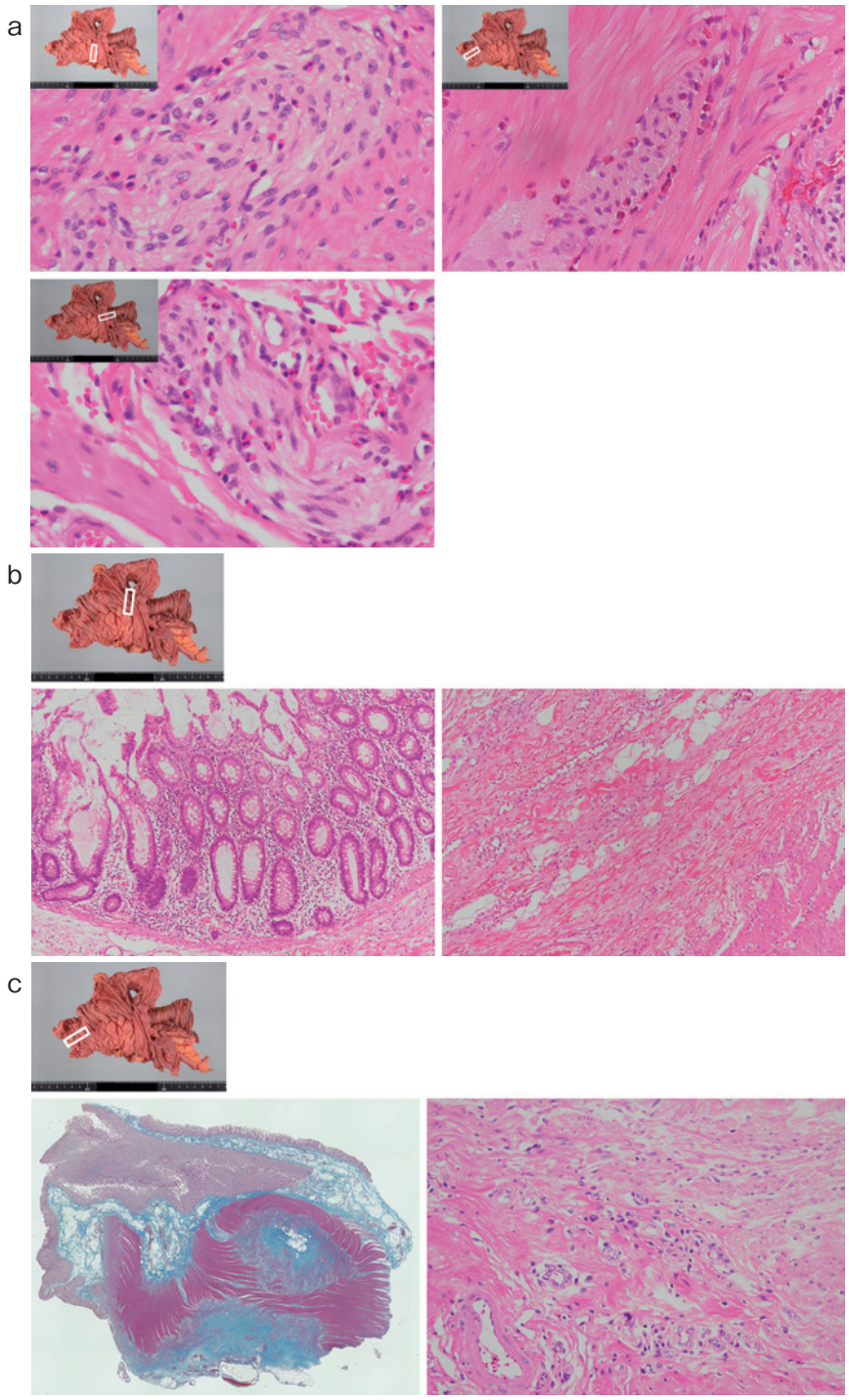

Figure 3. Microscopic findings of the surgically resected specimen.
a. There is marked eosinophilic infiltration in the myenteric plexus at the perforated site, at the distal resection margin, and in Bauhin's valve. Ganglion cells are recognized and appear normal in morphology.
b. There is little eosinophilic infiltration from the mucosal layer to the submucosal layer.
c. Fibrosis extends from the external longitudinal muscle layer to the subserosal layer (Masson trichrome stain). There was vascular proliferation, along with macrophage and lymphocyte infiltration.

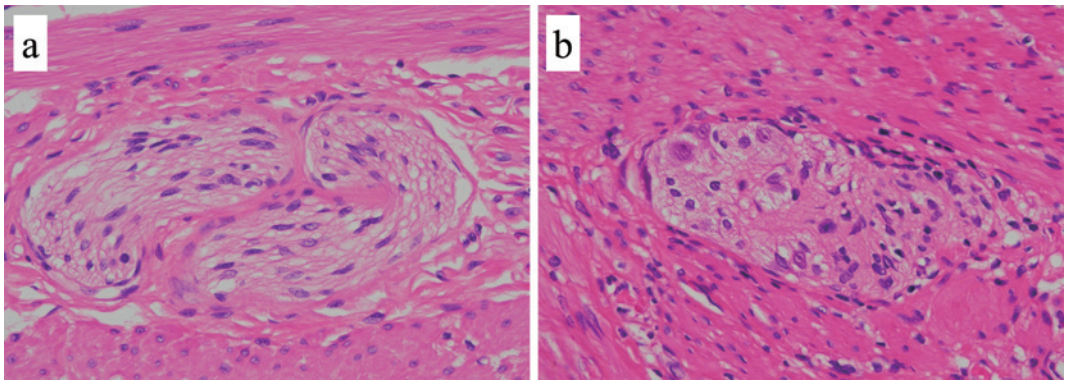

Figure 4. Histopathological appearance of Auerbach's plexus at closure of ileostomy.
No apparent abnormalities are identified in the myenteric plexus (a. resection stump of the ileum, b. resection stump of the colon).

Discussion

Inflammation of the neuromuscular tissue occurs in some patients with chronic intestinal pseudo-obstruction and can occasionally lead to severe intestinal motility disorders. When the inflammatory reaction is confined to the enteric plexus and the submucosal plexus is normal, the condition is termed myenteric ganglionitis[6]. Two major histopathologic types of myenteric ganglionitis have been reported[1-7]. The common form is lymphocytic ganglionitis, which is characterized by the infiltration of the myenteric plexus by CD4 and CD8-positive lymphocytes. Lymphocytes are believed to target antigens/proteins on the surface of enteric neurons, leading to their destruction. The inflammatory infiltrate is often associated with neuronal degeneration that may progress to a complete loss of neurons. The other type is eosinophilic ganglionitis, which has a much lower incidence than the lymphocytic type. A potent chemotactic factor for eosinophil interleukin 5 is expressed in the enteric ganglia, resulting in eosinophil infiltration in Auerbach's plexus[5]. Unlike lymphocytic ganglionitis, eosinophilic ganglionitis is often reported not to involve neuronal degeneration or loss[2,4,5,8-10]. Eosinophil infiltration or fluid factors released by eosinophils may secondarily cause enteric neuron dysfunction and result in pseudo-enteric obstruction[5].

It is important to differentiate EMG from eosinophilic enterocolitis, which causes intestinal motility disorders and pseudo-intestinal obstruction similar to EMG. In eosinophilic enteritis, eosinophilic infiltrate is prominent in the mucosal, proper muscular, and submucosal layers of the intestine, whereas in EMG, eosinophilic infiltrate is characteristically localized around the nerve plexus and ganglion cells.

In our patient, no degeneration or loss of neurons was noted, with minimal eosinophilic infiltration from the mucosal layer to the submucosal layer; therefore, the patient was ultimately diagnosed with EMG. Multifocal fibrosis was observed from the longitudinal muscle layer to the subserosal layer, indicating chronic changes. The extent of fibrosis exceeded that of secondary serositis due to perforation, suggesting that the intestinal perforation may have resulted from EMG-derived pseudo-intestinal obstruction.

Previously, most cases of EMG were reported from infancy to 15 years of age[4,5,8], but in recent years, many adult cases, including the present one, have been reported (Table 1)[1,2,9-12]. Owing to the rarity of this disease, its exact prevalence in the general population remains unknown. Peripheral eosinophilia often occurs in patients with EMG; however, it does not occur in all patients[1,2,11]; it was not observed in the patient reported herein. While the exact etiology of EMG is unknown, it is particularly prevalent in children, occasionally with peripheral eosinophilia, and allergic or infectious mechanisms have been suggested. The patient's medical and medication history was examined for the possibility of allergic reactions to drugs or other substances; however, an apparent cause for the EMG could not be identified.

To date, most cases of EMG have been reported in the left colon, including the sigmoid colon and rectum[1,2,5,8-10,12], and our study reports the first case of EMG in the ascending colon. Patients with EMG often present with chronic constipation, abdominal distention, and recurrent bowel obstruction of unknown cause. However, our patient did not present with these symptoms and developed sudden abdominal pain. This might be attributed to the site of the EMG lesion in the right-sided colon, where stool passing through the lesion as a liquid was less likely to cause obstructive symptoms. Most of the reported cases of EMG to date have been limited to the left-sided colon, possibly due to the nature of the stool at this site.

EMG has been reported to indicate progressive disease. Although no standard treatment has been established, several studies have reported that steroid therapy is effective for EMG[4,5]. However, certain case reports have noted that

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Table 1. Summary of Case Reports.

Study	Reporting year	Patient age, sex	Site of onset	Peripheral eosinophilia	Histopathological findings			Medical history			Results
					Gut distension	Peri/Intra ganglion eosinophilic infiltration	Neuronal degeneration/ loss	Surgery/Biopsy	Diet therapy	Steroid/ Immunosuppressive therapy	
Schäppi [5]	2003	1 month, F	Transverse colon~ Rectum	Yes	Avaluative (due to Biopsy)	Yes	No	Biopsy	Yes	Yes	Asymptomatic for over 1 year with diet therapy and sulphasalazine
		11, F	distal bowel	Unmentioned	Avaluative (due to Biopsy)	Yes	No	Biopsy	Yes	Yes	Asymptomatic for 1 year with azathioprine. Relapsed following its withdrawal
		15, F	Distal intestine	Unmentioned	Avaluative (due to Biopsy)	Yes	No	Biopsy	No	Yes	Asymptomatic for 1 year with azathioprine and prednisolone
Chander [2]	2011	93, F	Sigmoid	No	Yes	Yes	No	Hartmann's operation	No	No	Asymptomatic for 5 months
Ooms [4]	2012	4 months, M	Right bowel	Yes	No	Yes	No	Right bowel segment resection	Yes	Yes	Asymptomatic for over 18 months without therapy
Phillips [8]	2013	Term infant, M	Rectum	Yes	Avaluative (due to Biopsy)	Yes	No	Biopsy	Yes	No	Asymptomatic for over 6 months with diet therapy
Lee [11]	2017	53, F	Descending colon	No	Yes	Yes	Yes	Descending colon resectomy	No	No	Asymptomatic
Akazawa [1]	2019	34, F	Sigmoid	No	Yes	Yes	Yes	Sigmoidectomy	No	No	Lost to follow-up
		61, M	Sigmoid	No	Yes	Yes	Yes	Sigmoidectomy	No	No	Asymptomatic for 2 years
		76, M	Sigmoid	No	Yes	Yes	Yes	Sigmoidectomy	No	No	Asymptomatic for 1 year
D'Auria [9]	2020	3, M	Sigmoid~Rectum	Yes	Avaluative (due to Biopsy)	Yes	No	Biopsy	Yes	No	Asymptomatic for 15 months with diet therapy
Mitra [12]	2021	40, M	Sigmoid	Unmentioned	Yes	Yes	Yes	Sigmoidectomy	No	No	Single symptomatic recurrence for 36 months of follow-up
		85, M	Sigmoid	Unmentioned	Yes	Yes	Yes	Sigmoidectomy	No	No	Asymptomatic for 4 months
		67, F	Sigmoid	Unmentioned	Yes	Yes	Yes	Sigmoidectomy	No	No	Death for recurrence
		45, F	Sigmoid	Unmentioned	Yes	Yes	Yes	Sigmoidectomy	No	No	Lost to follow-up
Kim [10]	2023	72, M	Sigmoid	Unmentioned	Yes	Yes	No	Hartmann	No	No	Asymptomatic
Our case	2024	62, M	Ascending colon	No	Yes	Yes	No	Right hemicolectomy	No	No	Asymptomatic for over 2 years

symptoms sometimes remain stable clinically even when steroid treatment is not administered[1,2,9-12]. While these detailed case reports contain valuable information from a clinical aspect, they are limited in that they do not identify the time-course pathological changes that occur along the clinical course of EMG. In our report, the ileum and colon tract that were left out of the wound were resected at the time of ileostomy closure 4 months postoperatively. No eosinophilic infiltration was noted in Auerbach's plexus, which was observed in the previous surgical specimen, despite the patient not receiving steroid therapy postoperatively. Additionally, the patient had not experienced a relapse of symptoms after more than 2 years postoperatively. These findings suggest that patients with EMG can achieve symptom resolution through lesion resection without any medications. The pathogenesis of EMG has not yet been elucidated; however, the chronological and pathological findings identified in this patient may provide clues to its elucidation. Furthermore, the disease was alleviated through lesion resection, suggesting that surgical resection may be an effective treatment option. Early surgical intervention may be appropriate in cases with poor response to steroids or immunosuppressants, recurrent symptoms, or significant bowel dilatation.

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Conflicts of Interest

There are no conflicts of interest.

Author Contributions

TA wrote, edited, and reviewed the manuscript. ES revised and reviewed the manuscript. AI, SM, SF and MN reviewed the manuscript. AK and HT performed pathological evaluations and reviewed the manuscript. All authors approved the final version of the manuscript for publication. TA takes full responsibility for the work, including the decision to submit and publish the manuscript.

Approval by Institutional Review Board (IRB)

The information contained in, and preparation of, this manuscript complies with the journal's ethical standards.

Informed Consent

Written informed consent for publication was obtained from the patient.

Data Availability Statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

Disclaimer

Eiji Shinto is one of the Associate Editors of Journal of the Anus, Rectum and Colon and on the journal's Editorial Board. He was not involved in the editorial evaluation or decision to accept this article for publication at all.

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