

Case Report

Possible Contribution of a Diverticulum to the Development and Rupture of Colonic Lymphangioma

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ABSTRACT

A 55-year-old Japanese man with a history of diverticulitis underwent colonoscopy for careful evaluation of progressive anemia. A 5-mm depressed lesion oozing spontaneously was observed at the hepatic flexure. On suspicion of depressed-type of cancer, right-sided hemicolectomy was performed. Histopathological examination indicated a collapsed lymphangioma exactly over a diverticulum, which had previously been complicated diverticulitis. The colonic mucosa and lymphangioma prolapsed beyond the subserosal layer via the muscularis propria defect, resulting in a depressed lesion and mucosal laceration with hemorrhage. This case suggests the contribution of a colonic diverticulum to the development and rupture of lymphangioma, which needed to be distinguished from depressed-type colon cancer.

Key words: *Lymphangioma, Colonic diverticulum, Anemia, Depressed type of colon cancer*

Lymphangioma is a benign tumor characterized by the proliferation of dilated lymph vessels with a lining of a single layer of endothelial cells^{2,5,7-9}. Although lymphangioma may occur anywhere in the body^{2,5,9}, it is most often found in head, neck, and axillar regions during childhood^{3,7,9}. Although the intestinal tract is one of the least common sites of lymphangiomata^{2,3,5,7,9}, colonic lymphangioma is being reported more frequently with the widespread use of endoscopy^{2,3,5,8,9}. In this case report, we describe what is presumably the first presentation that a colonic diverticulum contributed to both the development and rupture of lymphangioma to cause intestinal blood loss. We also discuss the need of differentiation of a ruptured lymphangioma from depressed-type colon cancer on the basis of endoscopic appearance.

CASE REPORT

A 55-year-old Japanese man was referred to our hospital for careful examination of asymptomatic anemia, detected during a medical health checkup. The patient did not report a history of obvious

bleeding such as the occurrence of bloody stools. However, he had a history of colonic diverticulitis at the hepatic flexure, treated conservatively over the preceding 27 months (Fig. 1). Total colonoscopy performed approximately 1 month after the diverticulitis demonstrated nothing but a diverticulum at the site of hepatic flexure. The physical examination findings were unremarkable, except for anemic conjunctiva. A peripheral blood cell count revealed microcytic hypochromic anemia with a red blood cell count of 446/mm³, a hemoglobin level of 7.4 g/dl, and a hematocrit of 26.7%. These values had been within the normal range 27 months prior to his most recent visit. Blood biochemistry findings were normal, except for observations of mild diabetes with a hemoglobin A1c level of 6.4%. The findings of esophagogastroduodenoscopy were unremarkable. However, colonoscopy showed a depressed lesion, 5 mm in diameter, with an elevated edge located at the hepatic flexure, from which the blood was spontaneously oozing (Fig. 2). A biopsy sample from the depressed area did not have malignant cells. No other lesions that could possibly be responsible for the patient's anemia were detected in

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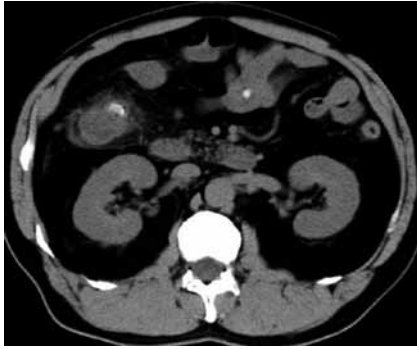


Fig. 1. Abdominal computed tomography (CT) scan demonstrating acute colonic diverticulitis at the hepatic flexure. Diffuse colonic wall thickening, a fecal stone and increasing density of the surrounding adipose tissue were found on CT, but no cystic lesion suggestive of a lymphangioma was detected.



Fig. 2. Conventional colonoscopy revealed a 5-mm-diameter superficial depressed lesion surrounded by a circulatory elevated edge at the hepatic flexure. Spontaneous oozing from the central depression was identified as the cause of anemia.

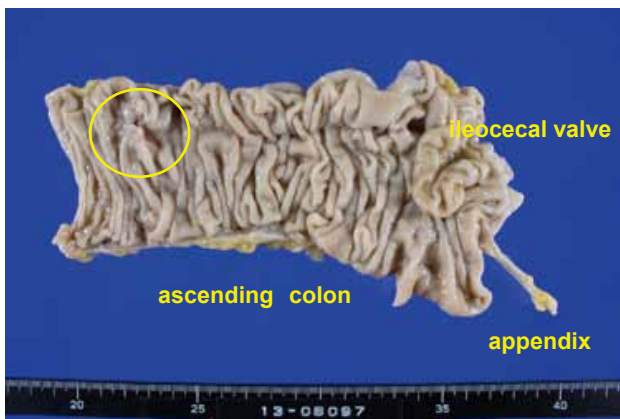


Fig. 3. Careful examination of the resected right-sided colon did not show evidence of any other diverticulum except the small depressed lesion. The circle indicates the site of the ruptured lymphangioma developing exactly over the diverticulum.

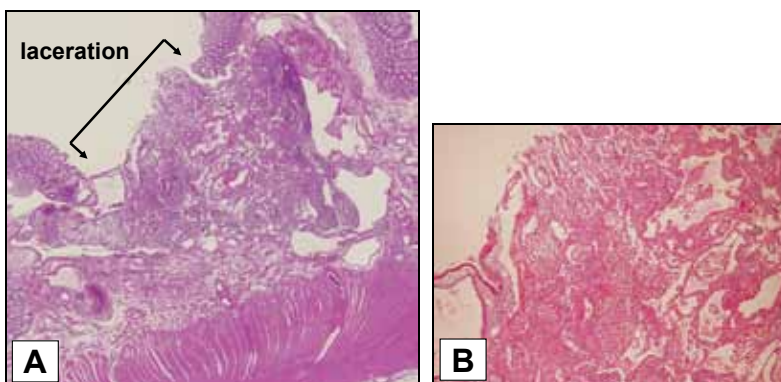


Fig. 4. Part of the colonic mucosa covering the depressed lesion was lacerated (A). The mucosal laceration exposed vasculatures with hemorrhage and collapsed lymph vessels (B). (hematoxylin and eosin staining at original magnification (A) 50 ×, (B) 200 ×).

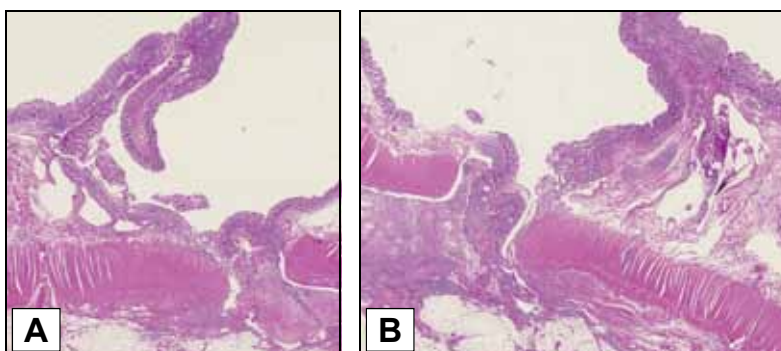


Fig. 5. Histopathological examination revealed a deformed and collapsed lymphangioma in a region of disrupted muscularis propria. The normal colonic mucosa and the lymphangioma had prolapsed through this defect beyond the subserosal layer, resulting in a central depression (A, B). The raised edge surrounding the central depression comprised an outgrowth of dilated lymph vessels lined by flat endothelial cells, with fibrosis, lymphocyte infiltration, and follicle formation in the submucosal layer (A, B). (hematoxylin and eosin staining at original magnification 10 ×)

the colon. Computed tomography (CT) performed immediately after colonoscopy, the findings of which were almost the same as those of CT colonography⁴⁾, merely revealed a localized subtle thickening of the haustra at the hepatic flexure. No evidence of lymph node swelling, ascites, or extra-colonic tumorous lesions was noted in the abdominal or pelvic cavity. Since the possibility of a depressed-type (0-IIc type)¹¹⁾ or small Borrmann type 3 colon cancer could not be completely ruled out and because the bleeding from the lesion had to be controlled, the patient underwent right-sided hemicolectomy. Careful macroscopic examination of the resected colon did not show evidence of any other diverticulum except a small hemorrhagic depressed lesion (Fig. 3). The postoperative clinical course was uneventful. Histopathological examination showed a collapsed and highly deformed lymphangioma in a region with disrupted muscularis propria, indicative of the diverticulum. The colonic mucosa and dilated lymph vessels had prolapsed beyond the subserosal layer, through the defect in the muscularis propria, resulting in the formation of a central depression. A part of the mucosa in the central depression was lacerated and collapsed lymph vessels and hemorrhaging vasculature were exposed (Fig. 4). The vasculature did not involve penetrating arteries that are often observed with colonic diverticulum. The raised edge around the central depression comprised an outgrowth of dilated lymph vessels lined by flat endothelial cells, with fibrosis, and lymphocyte infiltration, and follicle formation in the submucosal layer (Fig. 5).

DISCUSSION

Most intestinal lymphangiomas remain asymptomatic and are detected incidentally^{2,3,7-9)}. They rarely induce intussusception^{2,5,7,8,9)}, protein-losing enteropathy^{2,7-9)}, and hematochezia and anemia^{1,2,5,7,9,13)}. Although ulcerated mucosa⁹⁾, mucosal patchy defects¹³⁾, petechiae⁷⁾ or hyperemia⁵⁾, and hemorrhage of cystic lesions²⁾ have been identified as causes of gastrointestinal bleeding or anemia, we believe this is the first report on a ruptured lymphangioma where the source of bleeding was clearly demonstrated by endoscopy and/or pathology. In addition, this case was unique in that a colonic diverticulum presumably contributed to the development and rupture of lymphangioma.

The pathogenesis of lymphangioma in children seems to be related to inherent errors in the development of lymphatic channels, such as the proliferation and dilatation of a blind-ended lymphatic sac^{3,8)}. In adult cases, the pathogenesis of lymphangioma is suspected related to acquired conditions that affect local lymphatic circulation, such as abdominal trauma, localized lymphatic degeneration or obstruction, cancer, surgery, and radiation^{3,7,8)}. In the present case, pathological evaluation showed that

lymphangioma developed on a diverticulum that had previously complicated with inflammation (diverticulitis). The development of lymphangioma after diverticulitis, probably due to impeded local lymphatic flow associated with local inflammation, is a more plausible speculation. In addition, we believe that the simultaneous presence of the diverticulum, represented by a disruption of the muscularis propria, contributed to the rupture of lymphangioma, as noted on the pathological examination. Because of its soft-tumor-like properties represented by change in the tumor outline on compression with endoscopic forceps (cushion or pillow sign), peristalsis, or postural alterations^{2,5,8)} reflecting the submucosal localization of dilated lymphatic vessels filled with serous fluid^{5,7,8)}, the lymphangioma was probably pushed beyond the subserosal layer via the muscularis propria defect due to the increase in intra-luminal pressure or peristalsis of the colon. This retraction of lymphangioma and the overlying mucosa might have induced central depression and laceration to the mucosa to cause bleeding. Since no cellular or structural atypia was found in the lymphangioma, the possibility of tumor invasion to the subserosal layer with disruption of the muscularis propria was disregarded. Thus, this case is intriguing because it suggests that an acquired factor such as diverticulum may contribute to both the development and rupture of lymphangiomas.

In the present case, differentiation of the ruptured lymphangioma from colon cancer was another critical issue. Macroscopically, the central depression of the ruptured lymphangioma resembled 0-IIc depressed-type cancer^{10,11)}. If the raised edge around the central depression—pathologically proved to be due to an overgrowth of the residual dilated lymph vessels in the submucosal layer—is interpreted as submucosal cancer invasion, the ruptured lymphangioma may be misjudged as 0-IIc + IIa type^{10,11)} or small Borrmann type 3 cancer. Further, 0-IIc and 0-IIa + IIc type lesions are reported to be usually located in the right colon and are at the highest risk for submucosal invasion¹⁰⁾. With regard to imaging studies for the diagnosis of lymphangioma, were it not ruptured, the lymphangioma could have been detected by CT colonography as a submucosal cystic lesion⁹⁾ with the same attenuation as water⁴⁾; however, CT colonography in this case merely showed the collapsed lymphangioma as a local subtle thickening of colonic haustra. The patient had the typical characteristics of colonic lymphangioma: male gender, age in the fifth to seventh decades of life, Oriental ethnicity, presence of a right colon lesion, and presence of a solitary lesion^{3,7,8,13)}. However, these factors are not conclusive in distinguishing lymphangioma from colon cancer. Furthermore, it is reported that colon cancers including the depressed type do arise within a diverticulum⁶⁾, although the pathogenic

linkage is still controversial⁶). Thus, despite the biopsy result being negative for malignancy, we believe that surgery was justified in this case because macroscopic differentiation of ruptured lymphangioma from depressed-type colonic cancer was difficult and also because the depressed-type colon cancer often exhibits greater biological aggressiveness, irrespective of its size^{10,12}, and is more prevalent than previously thought¹². Complete regression of colonic lymphangioma either spontaneously⁷ or after incomplete polypectomy (e.g., upper half of resection) has been intriguingly reported^{5,8}. Regression in the latter case may be explained by the collapse of the lymphangioma⁷ and its absorption by lymphatic fluid outflow from the dilated lymph vessels through the cutting plane after polypectomy. In the present case, the disappearance of the lacerated and partially collapsed lymphangioma may have been spontaneous.

In conclusion, this present case suggests that a colon diverticulum possibly contributes to both the development and rupture of lymphangioma. A colonic diverticulum should be added to the list of the acquired causes of colonic lymphangioma. Although the presented condition may be rare, awareness that a ruptured lymphangioma related to a diverticulum resembles depressed-type cancer on the basis of endoscopic appearance may help clinicians to provide appropriate diagnosis and treatment, in addition to careful evaluation of mucosal surface pits patterns using magnifying and/or chemoendoscopic techniques¹¹.

(Received January 16, 2015)

(Accepted March 5, 2015)

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