

Lymphangioma of Colon Presenting as an Intramural Tumor

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Abstract: Lymphangiomas are rare and benign vascular malformations of the lymphatic system. They may arise in any location and at all ages and have variable presentation. These lesions in the intestinal wall are reported very rarely. In the case of colonic lymphangiomas, it is more common in late adulthood and old age, which, in this age group is thought to be associated with local disturbances of lymphatic circulation secondary to inflammation, degeneration, surgical procedure, trauma or radiation. The clinical presentation of colonic lymphangiomas varies from incidental findings on imaging to presenting with acute abdomen. The imaging features are usually multilocular cyst in intramural colon and submucosal mass on endoscopy. However, in the case of symptomatic lesions with atypical image findings, and the fact that the disease is rare, preoperative diagnosis is often difficult. On the other hand, although these cystic tumors do not transform into malignancy, they can be locally invasive or complicated, and often require resection. We report a 53-year-old male who had a cystic lymphangioma of the transverse colon illustrated by imaging modalities and recognized via postoperative histopathological examination.

Keywords: cystic lymphangioma, colonic lymphangioma, submucosal tumor

Introduction

Lymphangiomas are rare benign malformations of lymphatic vessels, therefore, they can occur in any organ of the body. The most common (95%) arise in the neck and axilla region; the rest (5%) occurs mainly in the abdomen and mediastinum.¹⁻³ It is worth noting that they are common in children and related to the congenital origin where lymphangiectasis arises from the failure to establish a patent communication with the lymphatic system; its discovery in adults is rare and the acquired origin has been suggested to be a lymphatic obstruction as a result of inflammation, trauma, or degeneration.³

Among the abdominal lymphangiomas, the majority are located in the mesentery and retroperitoneum.⁴⁻⁶ These lesions in the colonic wall have been reported even more rarely, accounting for about 0.7% of all abdominal lymphangiomas.⁶ Various imaging modalities including ultrasound (US), computed tomography (CT), magnetic resonance imaging (MRI), and endoscopy/endoscopic ultrasound (EUS) are increasingly important in the diagnosis and assessment of these lesions before therapeutic intervention. Herein, we present a case of a 53-year-old male presenting with acute abdominal pain, who was found to have an intramural cystic mass in the transverse colon on imaging, which was confirmed as a lymphangioma on postoperative histopathology.

Case Presentation

A 53-year-old male presented to the emergency room with acute pain in the umbilical region for two days with associated fever of 38°C and nausea without vomiting. He denied any melena, hematochezia or diarrhea, except for delayed bowel habit for a day. The patient reported no chronic abdominal pain or transit disorders. He did not have any significant personal history or family history. At the time of the initial visit, his vital signs were unremarkable. The abdomen was

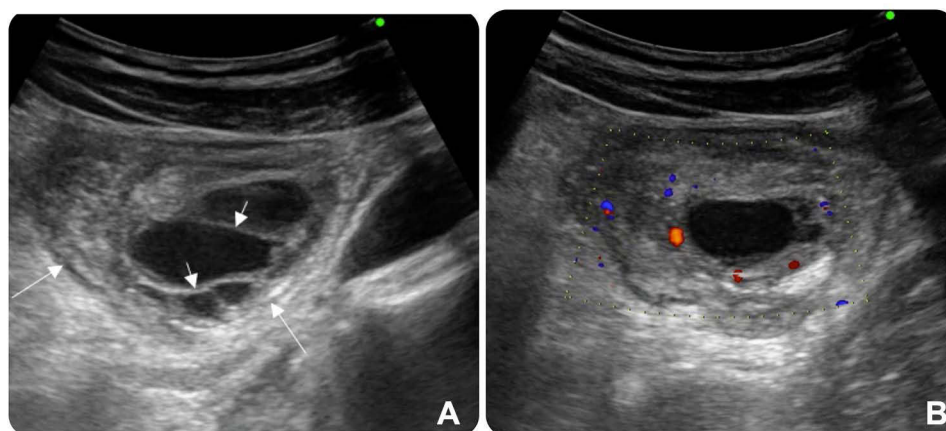


Figure 1 The longitudinal US of the supra-umbilical region (**A**) shows an anechoic cyst with multiple internal septa (short arrows), strongly demarcated by the intestinal wall structures (long arrows), suggestive of a cystic lesion located in the transverse colon. Color Doppler (**B**) demonstrated no hypervascularity of this multicellular cyst as well as of the colonic wall surrounding it.

soft and non-tender with no mass or organomegaly on palpation. Laboratory parameters upon admission showed a white blood cell count of 12.7 G/l (neutrophil 83.4%). The other laboratory tests, including those measuring amylase and tumor markers (CEA and CA19-9), were within normal limits.

For diagnosis, abdominal X-ray, US and CT were used. The abdominal X-ray displayed no specific findings. The abdominal US showed no abnormalities of the visceral abdomen as well as no peritoneal fluid, however, in the supra umbilicus, there was a focal lesion involving the transverse colon wall. This lesion manifested as a multilocular anechoic cystic mass with internal septa of varying thickness, and no hypervascularity on echocardiography (**Figure 1**). An abdominal CT scan revealed a low-density multilocular mass, with a low contrast-enhanced wall and internal septa located in the transverse colon; the proximal transverse colon showed thickened submucosal edema (**Figure 2**). The rest of the abdomen revealed no remarkable abnormalities. Although CT was suggestive of cystic lymphangioma with colonic segmental inflammation upstream of this cyst, it was not sufficient to rule out other more frequent diagnoses. Therefore, the patient was taken for endoscopy to better define the characteristics of the lesion.

Flexible colonoscopy documented a broad-based smooth polypoid mass with glossy pinkish mucosal coating in the middle part of the transverse colon (**Figure 3**). Because this lesion was covered by near-normal mucosa and was suggestive of a submucosal tumor, a biopsy was not performed. Based on the diagnosis of submucosal tumor of the

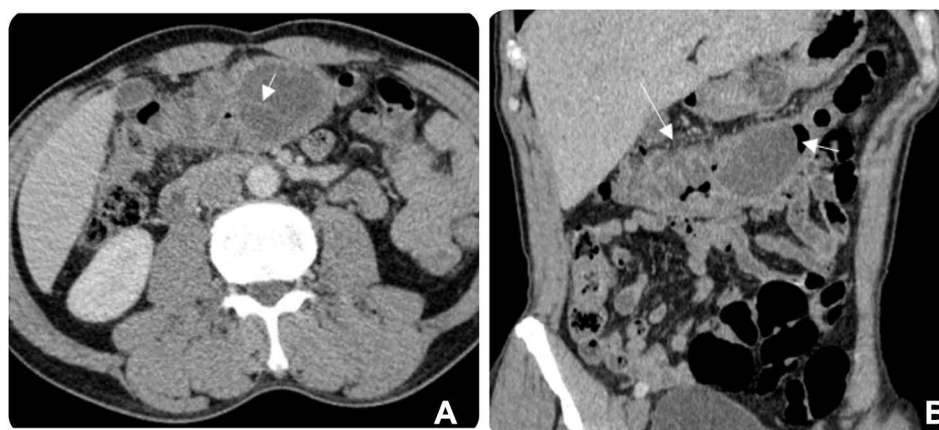


Figure 2 Abdominal CT with late venous phase, axial (**A**) and coronal (**B**) views of the transverse colon found fluid-attenuation density mass (10–12HU) with low enhanced septa ((**A**), arrow) measuring 42×47 × 63 mm. The mass is partially surrounded by air in the colon and this site revealed a very thin unenhanced wall ((**B**), short arrow). The colonic segment upstream to the mass is thick-walled with enhanced thin mucosa and without clear pericolic fat stranding ((**B**), long arrow).

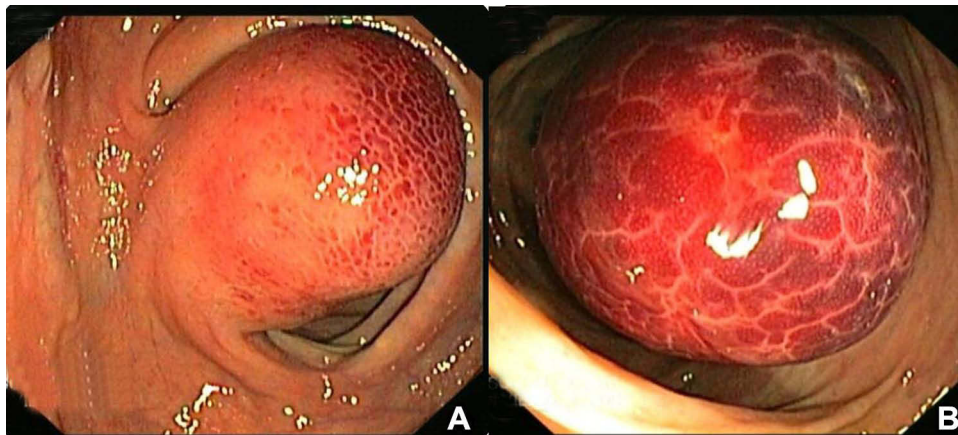


Figure 3 Colonoscopic view (A and B) demonstrated a large smooth-surfaced sub-pedunculated polyp with glossy, pinkish mucosal coating located in the transverse colon presenting as a submucosal tumor.

transverse colon, laparoscopic surgery with right hemicolectomy was performed, which showed a 4 x 6 cm, soft and spongy submucosal polypoid lesion on palpation, confined to intramural colon and non-invasion out of the serosa (Figure 4). Macroscopically, the cystic mass had multiple septa fluid-filled compartments. Histopathological examination revealed thin-walled dilated spaces consisting of only a single layer of endothelial cells (Figure 5). These findings were consistent with the diagnosis of cystic lymphangioma. The patient recovered well and was discharged on the 6th post-operative day. The 6-months subsequent follow-up was unremarkable.

Discussion

Colorectal lymphangioma used to be considered an extremely rare disease. Geographically, they have been reported mainly in Eastern countries (95%), with Japan alone accounting for 85%.² The most common age at diagnosis is in the 40s to 60s; and slight predominance in men with 1.4 to 1.6 times more than women.^{2,7} In cases of lymphangioma presenting in late adulthood and old age, the occurrence of the lesion may have developed secondary to local disturbance



Figure 4 Gross morphology during surgery revealed a 4×6 cm, soft and spongy submucosal polypoid mass (arrow) in the transverse colon.

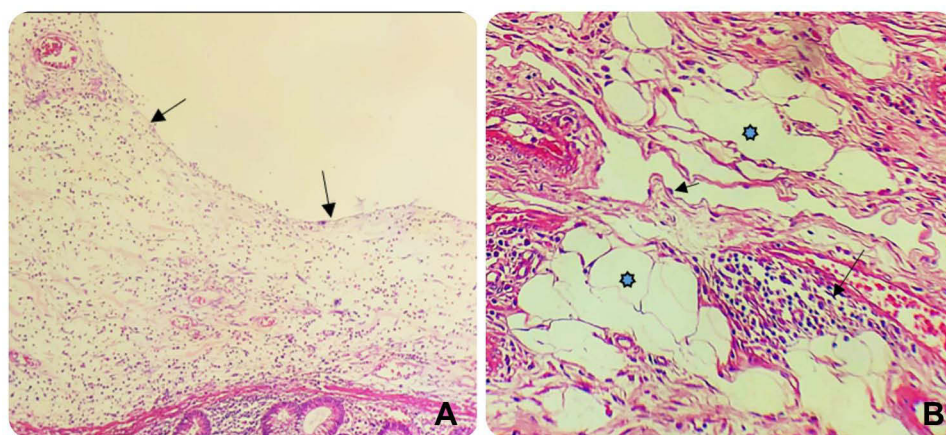


Figure 5 Photomicrograph of histopathologic specimen (HE stain). **(A)** (x20) showing the submucosal cystic lymphangioma with the interior wall lined by flattened endothelium (arrows). **(B)** (x40) showing a wall that has multiple dilated spaces (asterisks), lined by a thin layer of endothelial cells (short arrow); between these lymphatic spaces are smooth muscles and foci of lymphocyte clusters (long arrow).

of lymphatic circulation.⁷ Factors such as certain intestinal inflammations, surgical procedures, and radiation may be responsible for the appearance of these tumors.¹

Common sites of colorectal lymphangiomas are the transverse colon, ascending colon, and cecum; and most of them are single lesions.^{2,7} The size of the mass is variable, up to 23 cm, and not related to the patient's age nor to colonic location.^{2,7} Lymphangioma of colon is usually located in the submucosa with the overlying mucosa remaining intact. Depending on the size of the lymphatic spaces, this tumor has been classified into simple (or capillary), cavernous, and cystic types.³ Of these, cystic types are the most common - 70%,⁷ with 80% being multilocular.²

Cystic lymphangiomas of colon (CLCs) are often asymptomatic with small tumors. Symptoms, when present, are usually nonspecific and, depending on their size and location, may be acute abdominal pain with or without bloody stools, diarrhea or constipation.⁷ There have also been several cases reported of complicated acute abdomen such as infection,⁸ intussusception,^{9,10} anemia^{1,3,11} and protein-losing enteropathy.^{12,13} To date, to our knowledge, there have been no reported cases in which these tumors transformed into malignancy.

Practically, US is often the initial examination used for the symptomatic abdomen. CLCs typically show a multilocular cystic mass with septa, thin wall, containing anechoic fluid which are highly suggestive of the diagnosis¹⁰ (Figure 1A). Some lesions may be complicated by an intracystic infection or hemorrhage, causing internal echoes or sedimentation, with fluid–fluid levels caused by debris.¹⁴ CLCs are mostly avascular masses, but Doppler imaging may depict seemingly normal tiny flow signals of veins and arteries in the margin and/or septa without solid components associated with vascularity¹⁴ (Figure 1B). Although US is considered the first level of imaging investigation for a suspicious mass suggestive of a cystic mass, confirmation of a cystic lesion within or outside the bowel wall needs to be supplemented by CT or MRI because of the need for panoramic views and also to obtain additional information such as structural features, contrast enhancement, as well as to exclude locoregional extent of a malignant lesion.¹²

In an emergency setting, CT scans play a central role in the diagnosis. On CT lymphangiomas are cystic masses, typically attenuation values of water if the content is serious, limited and continuous with the colonic wall. This is further confirmed after administration of intravenous contrast and multiplane reconstructions showed slow or non-enhancement of the capsule and fibrous septations of the lesion as well as of the colonic wall with which it was involved^{1,14,15} (Figure 2). This cystic lesion may contain a low- to water-density content of the chylous fluid.¹⁶ High-density CT manifestations of intracystic bleeding and cystic wall calcification are uncommon in intra-abdominal lymphangiomas;⁵ and there seem to be no reports of these features in CLCs. Furthermore and for non-urgent settings, MRI better clarifies the cystic nature of masses and provides a better differentiation from other cyst-like masses,¹⁷ which we will discuss in the endoscopic ultrasound paragraph. On MRI, lymphangiomas are similar to signal intensity of fluid with low-signal

intensity on T1-weighted images and high signal intensity on T2-weighted images. In the presence of chylous fluid, a signal drop may be seen in the opposed-phase chemical shift MR images.¹⁴

Most CLCs are preoperatively diagnosed as submucosal tumors using colonoscopy, which is the most commonly used diagnostic modality. Colonoscopic features of CLCs are characterized by a steep rising margin and a somewhat narrow base, covered with normal colon mucosa but pinkish color, translucent, tense and lustrous surface⁷ (Figure 3). The shape is altered by postural change and compression with forceps, and the fluctuation is called “cushion sign”.^{15,16,18,19}

The role of endoscopic ultrasound (EUS) in the definitive and differential diagnosis of CLCs has been widely reported.^{11,15,18–20} EUS with the advantage of direct access to the surface of the colonic mucosa, can provide precise information about the layer of origin and contour of the submucosal tumor, especially with high accuracy in the diagnosis. On EUS, CLCs are confined to the submucosal layer with well-defined margins containing anechoic cystic spaces with septa and intact muscularis propria. These features are important for differential diagnosis of CLCs from other cystic-like masses located in submucosa of the colon such as leiomyomas, schwannomas, GISTs, lymphoma, and carcinoid tumor; these lesions are usually hypoechoic structures.^{20,21} Also based on these characteristic features of CLCs on EUS, Bhutani et al consider that endoscopic biopsies are unnecessary for diagnosis; moreover, they pose a risk of infection for these multicystic lesions.¹⁹

Other cystic lesions, although rare, that should be noted for differential diagnosis are duplication cysts and colitis cystica profunda. Cross-sectional modalities such as MRI and transrectal US are capable of demonstrating the cystic nature of these lesions. Both are more common anorectal site. Duplication cysts of colon generally have a significant exoenteric location, and the demonstration of mural layers in the cyst wall on US.²²

Treatment of CLCs is nonconsensual and depends on their various presentations, sizes and locations. Traditionally, CLCs presenting asymptomatic or <2 cm in size can be managed conservatively by observation.¹⁹ However, many lesions need to be confirmed via histological diagnosis by removal of tumor due to ineffective noninvasive diagnostic techniques. Endoscopic polypectomy described in the literature so far has been done for lesions 2–3.5 cm in size.^{8,10} Moreover, there are also cases where the mass size up to 5cm has been successfully removed by this method.²³ For cases that are difficult to manage endoscopically or require a differential diagnosis from other malignancies or are symptomatic, laparoscopic surgery should be considered as one choice to avoid complications such as superinfection, progressive growth, rupture or bleeding.^{15,16}

The final diagnosis requires histological examination. Macroscopically, CLCs present as yellow, greyish, or yellow-pink, multilocular cysts or spongy masses, containing watery/milky fluids at sectioning¹⁸ (Figure 4). Microscopically, they consist of cysts lined by flat endothelial cells devoid of atypia, similar to those of normal lymphatic tissue. The surrounding stroma is usually irregular enlarged lymphatic spaces and aggregates of lymphoid tissue. The presence of smooth muscle bundles and adipocytes are additional features^{4,6} (Figure 5). In the case of atypical histology, the diagnosis might require immunohistochemistry. Endothelial cells stain positive with lymphatic endothelial markers such as D2-40 marker and also for vascular markers including CD34-, CD31-, factor VIII-related antigen and VEGFR3.^{11,13}

Conclusion

Cystic lymphangiomas are very uncommon and benign lesions of the colon. The clinical presentation varies from incidental findings on imaging to presentation with acute abdomen. Typical features are usually multilocular cyst located in intramural colon on cross-sectional imaging and submucosal mass on endoscopy. The low frequency and acute abdominal condition can make it difficult for physicians to diagnose colonic lymphangiomas even with typical imaging features. Resection is essential to prevent complications, confirm the diagnosis histologically, and definitively exclude malignancy.

Ethics Approval and Consent to Participate

The case report received approval for publication from the Ethics Committee of Saint Paul Hospital of Ha Noi.

Consent for Publication

Written informed consent for publication of the clinical details and clinical images was obtained from the patient.

Disclosure

The authors declare no conflicts of interest in this work.

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