Case Report

Diagnosis of cystic lymphangioma of the colon by endoscopic ultrasound: Biopsy is not needed!

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ABSTRACT

Cystic lymphangioma of the colon (CLC) is a rare benign lesion that is usually asymptomatic and found incidentally during colonoscopy. Limitations in the conventional noninvasive diagnostic techniques have led to surgical resection of these lesions for diagnostic confirmation. Classic endoscopic ultrasound (EUS) findings of colonic cystic lymphangioma are submucosal anechoic cystic spaces with septations, intact *muscularis propria*, and no solid component. Patients who are asymptomatic with lesions having classic appearance as cystic lymphangioma with EUS can be observed without any intervention. We herein report a case of cystic lymphangioma of distal transverse colon in an asymptomatic patient diagnosed noninvasively using 20-MHz miniprobe EUS and managed conservatively without any surgical intervention.

Key words: Colon, cystic lymphangioma, endoscopic ultrasound (EUS)

INTRODUCTION

Lymphangiomas are developmental malformations of the lymphatic system with 5% of these affecting the abdomen.[1] Cystic lymphangioma of the colon (CLC) is very rare and have increasingly been reported during the recent years with the usage of colonoscopy. Given the benign nature, some authors have suggested that asymptomatic lesions <2 cm in size can be left alone without any intervention.[2-4] But the lack of effective noninvasive diagnostic techniques has led to the resection or endoscopic biopsy in most instances to make a histologic diagnosis. An accurate noninvasive diagnostic method is essential for such lesions to prevent unwanted surgical intervention. Recent advancements in endoscopic techniques, especially endoscopic ultrasound (EUS) have enabled us to accurately diagnose these lesions preoperatively without any invasive procedures. It is suggested that EUS appearance of colonic lesions confined to the submucosal layer with well-defined margins as anechoic cystic spaces with septations and intact *muscularis propria* is highly diagnostic of cystic lymphangioma of the colon.[5-9]

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In this report, we present a case of CLC diagnosed using a miniprobe EUS and relevant literature review emphasizing the role of EUS in making preoperative noninvasive diagnosis of CLC and thereby preventing unwanted surgical resection in asymptomatic patients.

CASE REPORT

A 61-year-old patient with a history of colonic polyps who underwent colonoscopy for follow-up was found to have a large subepithelial cystic lesion in the distal part of the transverse colon [Figure 1a and b]. The patient denied any abdominal pain, fatigue, weight loss, hematochezia, or change in bowel habits. Physical examination was unremarkable. Computed tomography (CT) scan of the abdomen performed to evaluate the lesion; the CT revealed no such lesions or any enlarged lymph nodes that could be attributed to the cystic lesion. The patient was referred to us for a colonoscopy with EUS for further characterization of the lesion. Colonoscopy/EUS on the endoscopic views revealed a cystic appearing smooth subepithelial lesion of 3-4 cm size in the distal transverse colon. The lesion was soft and compressible [Figure 1a and b]. The lesion partially flattened with air insufflation and the overlying mucosa appeared smooth and thinned out without any ulceration or erosion. A 20-MHz miniprobe was passed through the biopsy channel of the colonoscope and imaging was performed. High frequency ultrasound probe imaging demonstrated the lesion to be located in the submucosal layer with well-defined margins and intact *muscularis propria* underneath it. It appeared to have multiple anechoic spaces with septations and without any solid components [Figures 2a and b]. No lymph nodes seen near the lesion. Given the EUS findings being classical for a benign cystic lymphangioma of the colon and the patient being asymptomatic, no further intervention was made.

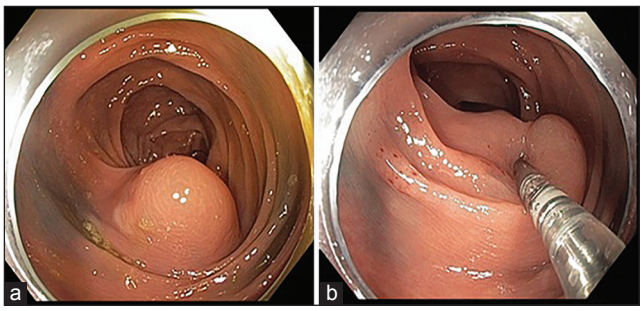

Figure 1. (a) Colonoscopic view of the cystic-appearing colon lesion with smooth thinned out mucosa (b) The lesion in figure 1a was soft and compressible

DISCUSSION

Lymphangiomas are developmental malformations of the lymphatic system composed of multiple lymphatic channels lined by endothelial cells. Possible pathogenesis includes failed communication of sequestered lymphatics with the normal lymphatic system, lack of fusion of lymphatic sacs with the venous system, abnormal budding of the lymphatic anlage, or the obstruction of efferent lymph vessels. Lymphangiomas are primarily classified into simple, cystic, and cavernous subtypes based on histologic appearance. Lymphangiomas can affect any part of the body with 50%-75% of them affecting the head, neck, and axilla with 90% of these becoming evident by 2 years of age. Intraabdominal lymphangiomas are very rare and most commonly affect the mesentery followed by the omentum, mesocolon, and retroperitoneum.[2,10]

CLC most commonly affects the transverse colon followed by the ascending colon, cecum, and descending colon.[11,12] Colonic lesions are solitary in 95% of the cases with few reported cases of lymphangiomatosis. Patients are mostly asymptomatic except for a few reported cases with complications such as abdominal pain, protein-losing enteropathy, intusseption, and bleeding particularly corresponding to larger tumor size.[13-17] CLC may range from 0.1 cm to 15 cm in diameter in size and are multicystic in 80% of the cases. Treatment options suggested in previous cases for pedunculated or semi-pedunculated lesions that are 2 cm or smaller in size include endoscopic polypectomy and for lesions of size >2 cm include segmental resection.[6,18]

Since colonic cystic lymphangioma is a benign lesion, a few authors have suggested that asymptomatic patients with lesions <2 cm in size can be managed

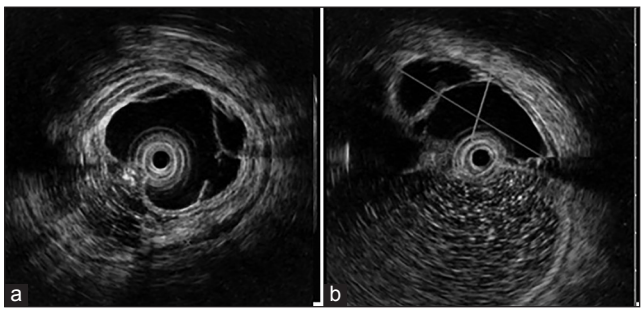

Figure 2. (a and b) EUS revealed 22 mm × 8 mm lesion originating/located in the submucosa with intact *muscularis propria,* with multiple anechoic cystic spaces with septations consistent with a colonic cystic lymphangioma

conservatively by observation.[2-5,19,20] however, many lesions were surgically resected to confirm the diagnosis histologically due to ineffective noninvasive diagnostic techniques. Although radiological classification for colonic submucosal tumors with double contrast barium enema was performed, lymphangiomas, along with lipomas and hemangiomas, are classified as type C (wide-based sessile with lobulated surface) and type D (pedunculated lesion with smooth or granular surface) as they share common features, making it difficult to differentiate from other lesions.[21] Endoscopic features suggestive of cystic lymphangioma are smooth-appearing broad-based subepithelial compressible fluid filled cysts covered with normal mucosa, but the differentials include lipoma, leiomyoma, submucosal cysts, submucosal tumors, and others.[4,18,22,23] CT imaging cannot generally identify such lesions if they are <2 cm and even with modified air enema technique, it can only detect lesions larger than 1 cm.[8]

A case was previously reported by our group[24] of a 4-cm cecal, subepithelial mass seen on colonoscopy with through-the-scope, miniprobe EUS demonstrating an anechoic, multiseptated, submucosal cecal cystic lesion with intact *muscularis propria*. These images were consistent with CCL. However, to document the benign nature of the overlying mucosa standard endoscopic pinch biopsy was performed, resulting in the drainage of whitish fluid. Unfortunately, the following day the patient presented with abdominal pain, chills, and fever. Emergency CT scan showed an increase in the size of the cecal cyst with air pockets and pericecal stranding consistent with an abscess. The patient fortunately recovered with 2 weeks of antibiotics without the need for surgery or abscess drainage. The pathologic findings of the endoscopic pinch biopsy (that caused the complication above) revealed colonic mucosa with markedly dilated lymphatic channels consistent with a CCL. In retrospect, the mucosal wall of the CCL was so thin that the endoscopic biopsy unroofed it and introduced infection into the multicystic lesion. However, the EUS images of this case of CCL (definitely proven by the endoscopic biopsy) that became infected were identical to the current case. Given our prior experience and learning from this prior case, we are now avoiding even endoscopic biopsies for such lesions and relying solely on the classic EUS appearance of anechoic, multiple septated cystic spaces in the submucosa to make the diagnosis of CCL.

Current advancements in the imaging modalities using EUS have enabled us to make non- invasive diagnosis of CLC and thereby avoiding the need for resection. The EUS imaging can clearly identify the layer of origin, echogenicity, internal echo, and borders, which not only helps to accurately diagnose CLC but also allows us to differentiate from other submucosal lesions.[7,9] Based on our experience and a few prior reports, the classic anechoic cystic appearance of a submucosal colonic lesion with septations and intact *muscularis propria*, is sufficient to diagnose cystic lymphangioma of the colon using EUS.[3-6,22]

CONCLUSION

In conclusion, asymptomatic patients with classic findings of cystic lymphangima on EUS imaging independent of the size can be managed conservatively.

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Conflicts of interest
There are no conflicts of interest.

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