Case Report

A Rare Case of Cystic Lymphangioma Arising from the Ascending Colon: A Case Report and Literature Review

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Lymphangiomas are rare and benign vascular malformations of the lymphatic system. They may arise in any location and at all ages and have variable presentations, although 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. 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, often requiring resection. Herein, we report a 55-year-old female with cystic lymphangioma of the ascending colon illustrated by imaging modalities and recognized via postoperative pathological examination and review the relevant literature.

[*J Soc Colon Rectal Surgeon* (*Taiwan*) *2024;35:195-200*]

Key Words
Cystic lymphangioma;
Colonic lymphangioma;
Surgical resection;
Case report;
Literature review

Cystic lymphangioma is a non-cancerous vascular malformed tumor consisting of thin-walled cysts. It is a rare benign tumor, with 95% of these cases observed in the neck, head and armpit while 5% occur mainly in the abdomen and mediastinum.[1] Cystic lymphangiomas are common in children and related to 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 as arising from lymphatic obstruction as a result of inflammation, trauma or degeneration.[2]

Received: February 25, 2024. Accepted: April 12, 2024.
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The majority of abdominal lymphangiomas are located in the mesentery and retroperitoneum.[3] These lesions in the colonic wall have been reported even more rarely, accounting for about 0.7% of all abdominal lymphangiomas.[4] Various imaging modalities including ultrasound (US), computed tomography (CT), magnetic resonance imaging (MRI) and endoscopy/endoscopic ultrasound (EUS) are increasingly important in diagnosis and assessment of these lesions before therapeutic intervention. We demonstrate a case of a 55-year-old female presenting with acute abdominal pain, who was found to have a cystic mass in the anti-mesocolic side of the ascending colon on imaging, which was confirmed as a lymphangioma on postoperative histopathology.

Case Report

A 55-year-old female with medical history of hypertension was under well-control. She presented to our outpatient department (OPD) due to intermittent abdominal pain at the right upper quadrant (RUQ) that had lasted for 6 months. She visited a local medical clinic initially, where abdominal echo revealed a cystic mass in the subhepatic area. Under the impression of intra-abdominal cystic mass, she was referred to our OPD, where abdominal physical examination (PE) showed neither peritoneal nor Murphy's indications. Imaging studies including abdominal CT scans and magnetic resonance image (MRI) of the liver were performed where the abdominal CT scans demonstrated a 5.6-cm diameter of cystic mass in the subhepatic area (Fig. 1A) and the MRI revealed a lobular-contour cystic lesion at the right subhepatic region (Fig. 1B). Subsequently, these two reports indicated suspicion of a cystic lymphangioma or a colon duplication cyst.

She received surgical intervention after discussion. During the surgery, a cystic lesion arising from the anti-mesocolonic side of the distal ascending colon was found and the diameter was 6 cm (Fig. 2). A wedge resection with Surgical Staplers Devices was performed (Fig. 3). The macroscopic feature showed a cystic mass lesion with a soft spongy consistency measuring 6.5 × 5.5 × 2.4 cm in size, while microscopic examination disclosed expansion of the colonic submucosa by cystic dilated lymphatics spaces lined by a single simple layer of lymphatic endothelial cells with proteinaceous material (Fig. 4A), and D2-40 staining highlighted the endothelial cell immunohistochemically (Fig. 4B). The overall feature appeared compatible with a diagnosis of lymphangioma of the ascending colon.

Till now, she has regular follow-up at our OPD and the general condition is very stable.

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%.[5] Those aged from the 40 s to 60 s are most commonly diagnosed with a

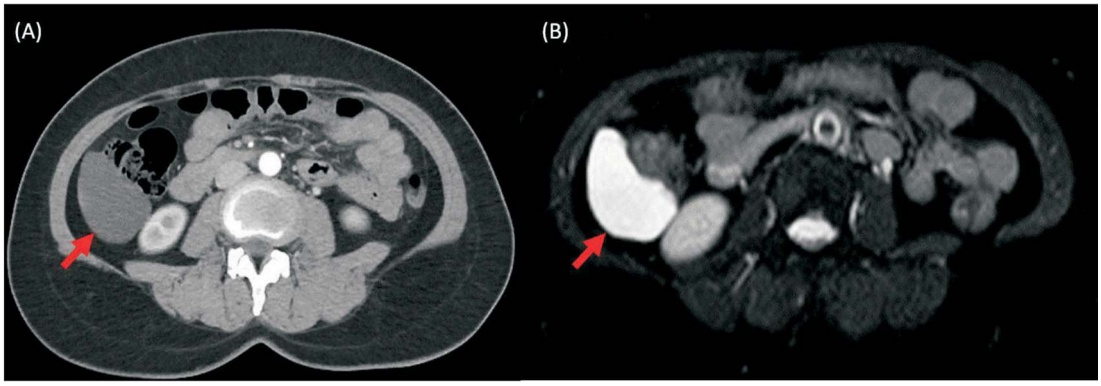

Fig. 1. The image studies. (A) Axial view of CT showed a poor enhanced mass in the right subhepatic region (red arrow). (B) T2 fat suppression MRI revealed lobular-contour cystic lesion (red arrow).

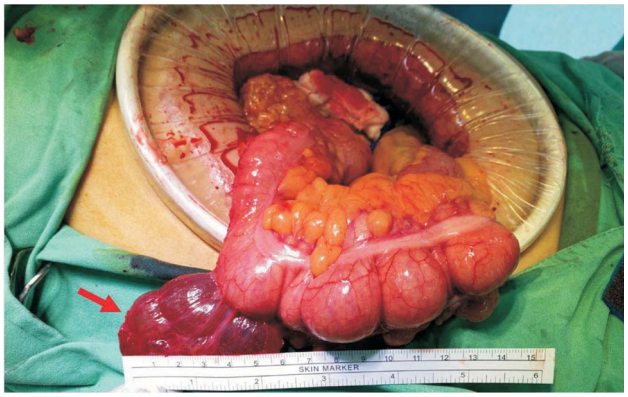

Fig. 2. Intra-operative image. A cystic lesion arising from ascending colon (red arrow).

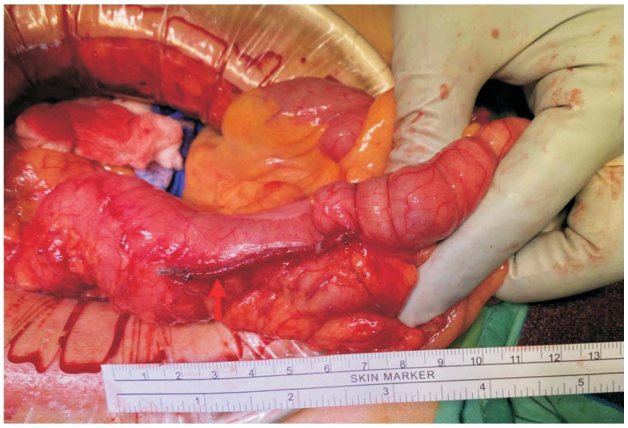

Fig. 3. Intra-operative image. After wedge resection of cystic lesion of ascending colon with surgical stapler device (red arrow).

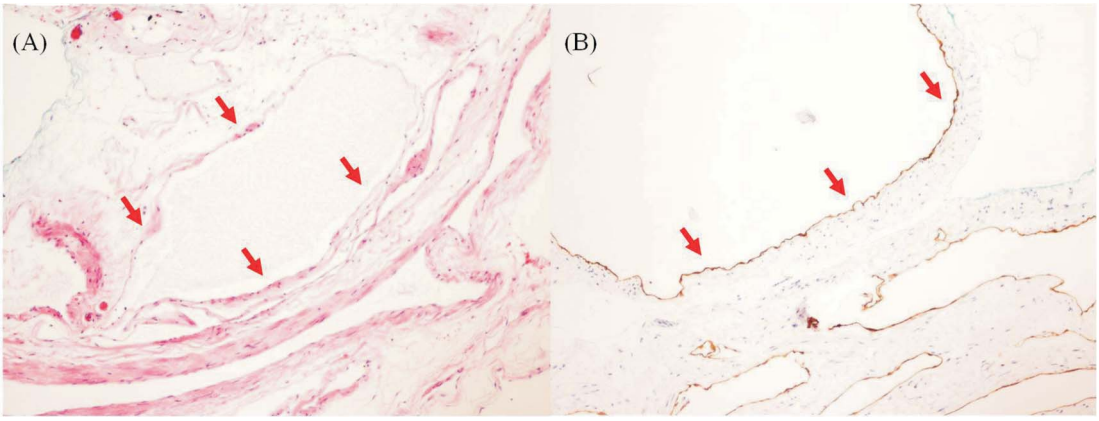

Fig. 4. Pathological section. (A) cystic dilated lymphatics spaces lined by a single simple layer of lymphatic endothelial cells filling with proteinaceous material (hematoxylin-eosin; original magnification ×100) (red arrows). (B) Immunohistochemically, the D2-40 stain highlighted the endothelial cells (hematoxylin-eosin; original magnification ×100) (red arrows).

slight predominance in men being 1.4-1.6-fold more than women.[5]

Common sites of colorectal lymphangiomas are the transverse colon, ascending colon and cecum, with most being single lesions.[5] Lymphangioma of the colon is usually located in the submucosa with the overlying mucosa remaining intact, and depending on the size of the lymphatic spaces, this tumor has been classified into simple (or capillary), cavernous and cystic types.[2] Among these types, the cystic type is the most common (70%) with 80% being multilocular.[5] Cystic lymphangiomas of the colon (CLCs) are often asymptomatic with small tumors; symptoms, when present, are usually nonspecific and depending on their size and location, can present with acute abdominal pain with or without bloody stools, diarrhea or constipation.[6] To date, to our knowledge, there have been no reported cases in which these tumors have transformed into malignancy. Although ultrasound (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 scans 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.[7]

Under colonoscopy, CLCs usually appear as smooth sessile, or pedunculated solitary lesions. Biopsy may not yield results due to the submucosal location. En-

doscopic ultrasound plays a role in diagnosis, where these lesions are anechoic, septate, and submucosal.[8]

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.[9] Endoscopic removal of lesions has been reported for lesions ranging in size up to a measurement of 2-3.5 cm. The use of a ligating device for removal and unroofing are described for the successful removal of lesions.[8] For cases that are difficult to manage endoscopically or require a differential diagnosis from other malignancies, laparoscopic surgery should be considered as one choice to avoid complications such as superinfection, progressive growth, rupture or bleeding.[10]

The final diagnosis requires histological examination where macroscopically, CLCs present as multilocular cysts or spongy masses and microscopically, they presentas cysts lined by flat endothelial cells devoid of atypia, similar to those of normal lymphatic tissue. In the case of atypical histology, the diagnosis might require immunohistochemistry, where endothelial cells are stained as positive with lymphatic endothelial markers such as the D2-40 marker.[11]

Conclusion

Cystic lymphangiomas are very uncommon and benign lesions of the colon, with clinical presentation varying from incidental findings on imaging to presentation with acute abdomen. The low frequency and acute abdominal condition can make it difficult for physicians to diagnose colonic lymphangiomas even with typical imaging features. CLCs with small size may be observed conservatively. However, for cases with larger size or with symptoms, endoscopic or surgical resection is essential to prevent complications, confirm the diagnosis histologically, and definitively exclude malignancy.

Ethical Approval

The study was approved by the Institutional Review Board of Kaohsiung Medical University Hospital (KMUHIRB-E(I)-20200036).

Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms where the patient has given her consent for the images and other clinical information to be reported in the journal while understanding that her name and initials will not be published and all due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Data Availability Statement

Data sharing is not applicable to this article because no data sets were generated or analyzed during the current study.

Financial Support and Sponsorship

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Conflicts of Interest Statement

The authors declare that they have no conflict of interest with regard to the content of this article.

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病例報告

升結腸淋巴管瘤之罕見案例報告及文獻回顧

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淋巴管瘤是淋巴系統少見且良性的血管，它們可能出現在人體各個部位，並於各年齡階段都可能發生，而在腸道中的淋巴管瘤更為罕見。結腸的淋巴管瘤常見於中年和老年，普遍被認為與局部淋巴循環有關，繼發於炎症、退化、手術、創傷或輻射。由於可能具有非典型的影像並因疾病罕見，會使得術前的診斷變得困難。另一方面，儘管這些腫瘤不會癌化，它們可能具有局部侵襲性或產生併發症而需要進行切除手術。因此，我們報告了一例 55 歲女性，患有升結腸囊性淋巴管瘤，並通過影像學檢查呈現，經由手術後的病理檢查確診，及其相關文獻回顧。

關鍵詞 囊性淋巴管瘤、結腸淋巴管瘤、手術切除、病例報告、文獻回顧。