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Case Report

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Cystic lymphangiomas of the cecum cause intussusception in adults

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Abstract

Lymphangiomas are rare benign lymphatic malformations. Even though these neoplasms can arise from any location and age, most of them are dominant in the head and neck of children. On the other hand, lymphangiomas are exceedingly rare in adults, especially in the gastrointestinal tract. Gastrointestinal tract lymphangiomas account for only about 1% of lymphatic malformations. Although there have been increasing cases of gastrointestinal lymphangiomas reported in recent years, cecal lymphangiomas causing intussusception in adults are rarely mentioned. In this article, we report a 27-year-old female patient with cecal lymphangiomas detected by imaging modalities and confirmed via postoperative histopathological examination.

Introduction

Lymphangiomas are rare benign lymphatic malformations. According to the 2018 Classification of Vascular Anomalies and Molecular Biology (ISSVA),[1] they are classified as vascular malformations, which do not belong to the vascular tumours group. Although they can occur in any organ of the body, most of these entities are predominant (95%) in the axillary and neck, and the remaining 5% occur in the lungs, mediastinum, and abdomen, while gastrointestinal lymphangiomas account for only 1%.[2-5] Children are the most typical victims and are associated with congenital abnormalities. Lymphangiectasis occurs due to a failure to establish a patent communication of the local lymphatic vessel with the lymphatic system. Conversely, these neoplasms rarely present in adults with the most common location being the mesentery and/or retroperitoneum. The acquired origin is thought to be lymphatic obstruction due to trauma, inflammation, or degeneration.[4,6] The exact pathophysiological mechanisms remain to be fully elucidated. However, Matsuda *et al.* proposed that excessive endothelial proliferation within the walls of lymphatic vessels could be attributed to inflammation, trauma, or radiation, based on their review of 279 cases from Japan.[7] Lymphangiomas often have no clinical symptoms, thus, these entities tend to be incidentally found on routine imaging or other disease surgery.[8] The most common complaints of patients relate to complications which may include abdominal pain, constipation, and vomiting. Lower gastrointestinal bleeding, intussusception, intestinal obstruction, and protein-losing enteropathy are rarely documented complications,[9-13] which require complete surgical excision.[6] Herein, we present a case report of a 27-year-old female who came to the hospital

because of gradually increasing acute abdominal pain in the right hypochondrium and epigastrium region. A cystic mass was discovered inside the cecum causing intussusception of the ileum-cecum-right colon on the image, the patient underwent emergency surgery and postoperative pathologic result confirmed the lesion to be cystic lymphangiomas in the cecum.

Case Report

A 27-year-old female came to the emergency room with acute pain in the right hypochondrium and epigastrium for three days. She described that the pain was intermittent at first, then gradually increased and continued. The patient had no fever, nausea, vomiting, melena, or scant hematochezia. Her medical history was uneventful.

On clinical examination, she had a tenderness in the right hypochondrium region. The clinician revealed a palpable mass measuring 5x3 cm in the right hypochondrium region, with no rigidity, guarding, and rebound tenderness. Blood tests were all within normal limits.

The patient's abdominal ultrasound showed a cystic lesion with a thin septum, vascular proliferation, and no solid components inside. This mass was described as having a target appearance on transverse sonography, and a sandwich appearance on longitudinal sonography, indicating an intussusception due to a mass located in the right hypochondrium region.

Unprepared abdominal radiography in a standing position did not show subdiaphragmatic free gas, air-fluid level, or any other abnormalities.

Subsequently, the patient underwent a contrast-enhanced abdominal Computed Tomography (CT) scan to confirm the diagnosis, evaluate lesion characteristics, and assess associated lesions. On the CT scan, an intussusception mass of the ileum-cecum-right colon was observed in the right hypochondrium region. In the cecum segment of the intussusception mass, there was a large cyst measuring 64x37 mm with a thin capsule, thin septa, and post-contrast enhancement inside. No abnormal enhancing solid component was seen. The intussusception loops were still enhanced uniformly, with no signs of intestinal obstruction or free intraperitoneal fluid (Figure 1).

An emergency surgery was performed to remove the cystic lesion in the cecum causing intussusception of the ileum-cecum-right colon and restore intestinal circulation. Intraoperatively, the surgeon described a large cystic mass located in the cecum, causing intussusception of the ileum-cecum-right colon (Figure 2).

Microscopically, lymphatic structures are covered by a layer of flattened endothelium and thick septum. There are scattered areas of thrombosis with no malignant cells. Pathology results confirmed benign cecal submucosal lymphangiomas (Figure 3).

Coronal images a and b show an intussusception mass located at the level of the right hypochondrium, terminal ileum (arrow), and a cystic lesion within the cecum in intussusception mass (*).

Axial image c focuses on the lesion and shows a cystic lesion (*) located in the lumen of the cecum with the thin septum (arrows) pushing the air in the cecum forward. Axial images d and e show a cystic lesion (*) with a thin septum, a thin capsule that enhances after contrast injection.

Discussion

Lymphangiomas are rare and benign malformations of the lymphatic vessels. They are classified as vascular malformations, which do not belong to the vascular tumours group, according to the 2018 ISSVA classification of vascular abnormalities.[1] They are reported mainly in children.[14] Children in the first few years of life account for approximately 80-90% of all diagnosed cases, and adult cases are exceedingly atypical.[15] Men have a higher incidence of disease than women with a male/female ratio of 6/4.[16,17] Gastrointestinal lymphangiomas tend to be located in the right colon, often in the submucosa covered with normal mucosa.[18,19]

Colon lymphangiomas are classified into simple (or capillary), cavernous, and cystic types, with the cystic types being the most commonly reported.[20-22] Cystic lymphangiomas can be further divided into 3 types: small cysts, large cysts, and mixed cysts depending on the size of the mass. Macroscopically, cystic lymphangiomas often appear as multiple cysts or spongy masses with watery or milky fluids and can also be yellow-pink, yellow, or greyish when complications such as infection and bleeding.[9-13,23] Microscopically, the appearance is similar to those of normal lymphatic tissue, they consist of cysts lined by flat endothelial cells. The surrounding stroma aggregates of lymphatic tissue and is usually irregular enlarged lymphatic spaces. Diagnosis may require immunohistochemistry in cases of atypical histology. Endothelial cells stain positive with lymphatic endothelial markers such as marker D2-40 and vascular markers including CD31-, CD34-, VEGFR3-related antigen, and factor VIII.[24,25]

Ultrasound is considered the first diagnostic imaging tool because it is a low-cost, non-invasive method that does not entail ionizing radiation. On ultrasound, gastrointestinal lymphangiomas can appear cystic with a septum and thin capsule located in the lumen of the digestive tract. Some lesions may contain debris show increased echogenicity, and develop a fluid-fluid level due to infection or intracystic hemorrhage.[26] Lymphangiomas are mainly avascular masses, but on Doppler ultrasound, small flow signals of arteries and veins in the septum and/or capsule of the lesion can be seen.[26] However, this technique needs to be combined with CT or Magnetic Resonance Imaging (MRI) to evaluate the overall disease and also to obtain additional information such as the degree of enhancement, structural characteristics, as well as the spread of the lesion.

An abdominal CT scan plays an important role in diagnosis in an emergency. In addition, CT with multiple slices allows the assessment of information about the composition of the lesion, size, and relationship with adjacent structures with multi-planar reconstruction using different methods (MPR, MIP, SSD, VR). In typical cases, the cystic mass has a fluid density, and well-defined capsule enhancement after contrast injection.[21,27,28] Sometimes it is also observed that the fibrous septum inside the cyst has a relatively uniform thickness.[21] Lymphangiomas have high density when bleeding within the cyst, and lower density when containing chylous fluid. Intracystic calcification is uncommon for abdominal lymphangiomas, and the presence of sediment at the bottom of the cyst is highly suggestive of the diagnosis of cystic lymphangiomas.[6]

MRI is often valuable in differential diagnosis and exclusion of other gastrointestinal lesions before surgery. Furthermore, MRI is useful in surgical planning due to its multiplanarity and high contrast

resolution, which can accurately show the loco-regional lesion spread.[29] On MRI, lymphangiomas have fluid signal with low signal on T1-weighted images and high signal on T2-weighted images. When chylous fluid is present, hyperintensity can be seen on T1-weighted images. Since lymphangiomas are surrounded by flattened endothelium, cyst capsules are often thin.[26] Fluid-fluid levels may be seen when there are complications of bleeding and intracystic hemorrhage.

With the development of colonoscopy, many lymphangiomas can be discovered incidentally when they are small and have not yet caused symptoms. The lesions are submucosal cystic masses characterized by a steep rising margin and a narrow base, covered with normal colon mucosa but tense, lustrous, translucent surface, and pinkish color.[3] When we apply force with the forceps against the lesion, results in depression of the mass, it is called the "cushion sign", which is the typical colonoscopic feature.[30-33] Endoscopic ultrasound is a reliable diagnostic method. Endoscopic ultrasound, with the advantage of directly accessing the colon mucosal surface, can provide accurate information about the boundary of the submucosal tumor and the layer of origin.[19] However, endoscopic ultrasound makes it difficult to assess the spread and size of the lesion when the lesion is large and its correlation with adjacent organs.

The appropriate diagnostic regimen for cystic lymphangiomas should include ultrasound and MRI with contrast injection to fully assess the lesion characteristics, and correlation with adjacent structures as well as rule out other cystic lesions.[29] CT scan should be used to evaluate emergency cases such as intussusception, intestinal obstruction, bleeding, *etc*.

There are a few differential diagnoses that need to be made with a cystic lesion in the gastrointestinal tract such as enteric duplication cysts, lymphatic metastasis, gastrointestinal tumours (colorectal adenocarcinoma, lymphoma, mesothelial cyst, and myogenic or neurogenic submucosal tumour…) and colorectal cavernous hemangiomas. Enteric duplication cysts are uncommon congenital abnormalities that may occur anywhere in the gastrointestinal tract, are rare in the large intestine, and are usually located in the rectum. Like gastrointestinal stromal tumours, duplication cysts generally have a common exoenteric location. Unlike cystic lymphangiomas, which are only surrounded by flat endothelium and therefore appear thin-walled at imaging, enteric duplication cysts contain all enteric layers and appear thick-walled. Ultrasound plays an important role in these patients, and many authors have demonstrated that the combination of a hyperechoic inner mucosal layer and a hypoechoic outer muscular layer on the cyst wall is highly suggestive of enteric duplication cysts.[26,34,35] The next differential diagnosis is gastrointestinal tumours, such as colorectal adenocarcinoma, lymphoma, mesothelial cyst, and myogenic or neurogenic submucosal tumour.... Gastrointestinal tumour lesions manifest as solid masses, enhance after injection, and may have an accompanying degenerative/cystic necrosis component. Additionally, malignant tumours also exhibit invasive features, invading adjacent organs destroying the layered structures of the intestinal tract, and causing narrowing of the intestinal lumen. Colorectal cavernous hemangiomas are a differential diagnosis to consider, which are very rare vascular malformations. The rectosigmoid is the most common site of this disease in the gastrointestinal tract. They often cause clinical symptoms of gastrointestinal bleeding, and the lesions can also invade adjacent

structures. Most gastrointestinal hemangiomas present as pedunculated polyps in the gastrointestinal tract, but occasionally the lesions may present as submucosal infiltrates. The presence of phleboliths is common in colorectal hemangiomas and is a useful diagnostic sign in younger patients who have gastrointestinal bleeding. Phleboliths appear in clusters and have an atypical distribution. Lesions may appear as soft, serpentine masses, polypoid lesions, and circumferential lesions, and some patients may present with narrowing of the digestive lumen. CT may show transmural thickening of the involved colon, vascular engorgement within the adjacent mesentery, and post-contrast enhancement and phleboliths. MRI characteristics of colorectal hemangiomas are focal thickening of the colonic wall with high signal intensity on T2-weighted images in both the lesion and perilesional fat.[36,37]

Lymphangiomas are benign lesions of the lymphatic system. Therefore, collaboration among the interprofessional team can optimize the treatment plan for the patient. With expertise in lymphangioma, radiologists can provide an accurate diagnosis prior to surgery, differentiate it from other similar lesions, and identify potential complications. This enables the treating physicians to anticipate and devise the most effective treatment strategy for the patient. If the patient has no symptoms, they can be monitored and do not need treatment. If there are symptoms, large size, and complications, complete surgical excision is the best option to treat and reduce the risk of future complications of large masses.[6] Endoscopic surgery is the main treatment modality for this disease, but endoscopic treatment of the digestive tract has been increasingly prescribed recently because this is a benign disease, so only the lesion needs to be removed, to avoid overly invasive treatment.

Endoscopic treatment indications should be applied to lesions with a diameter of 2 cm or less.[7] If the lymphangioma is complicated by torsion, total resection of the involved intestine is required.[38]

Conclusions

Report of a rare case of lymphangiomas in the cecum causing ileocecal intussusception in an adult. When there is a cystic lesion in the gastrointestinal tract with a thin wall, there may be a fluid-fluid level that requires consideration of the diagnosis of lymphangiomas, and this may be one of the lesions that cause intussusception in adults, even though it is a rare lesion to occur in the gastrointestinal tract in adults. Large lesions need to be surgically removed to avoid future complications.

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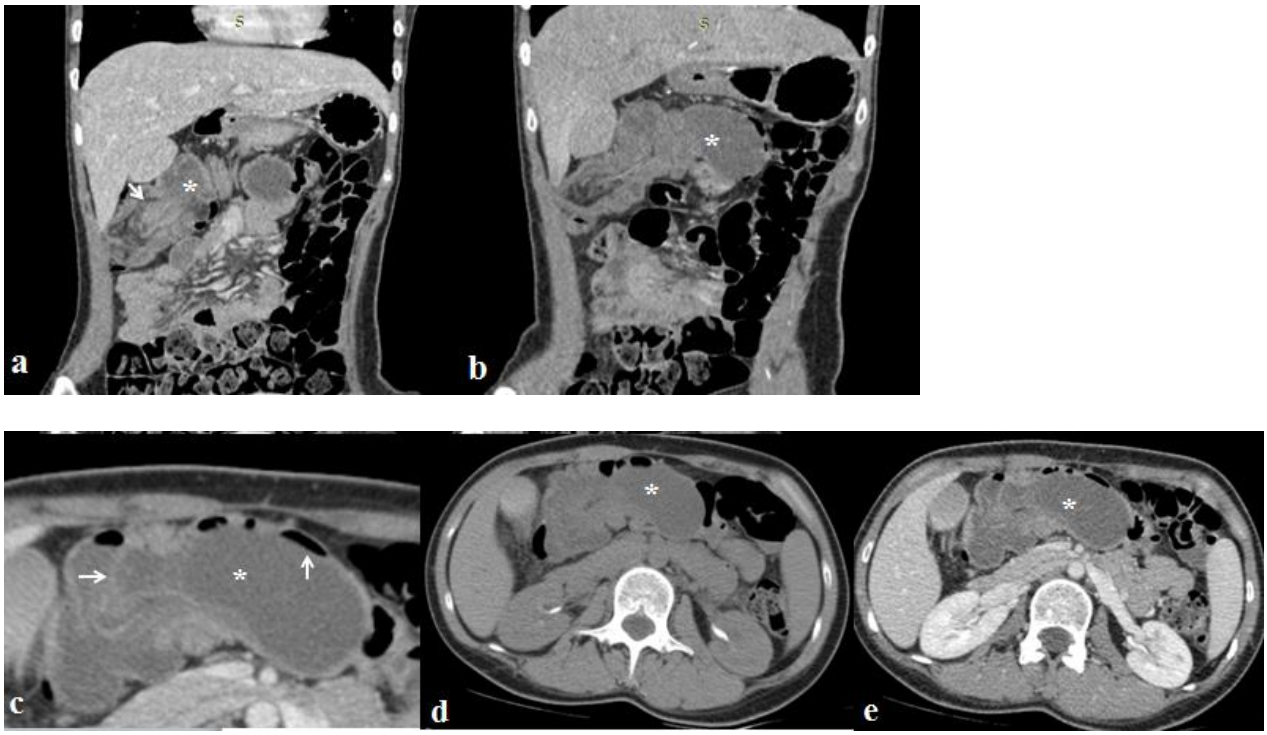

Figure 1. Computed Tomography (CT) scan of the abdomen with contrast injection.

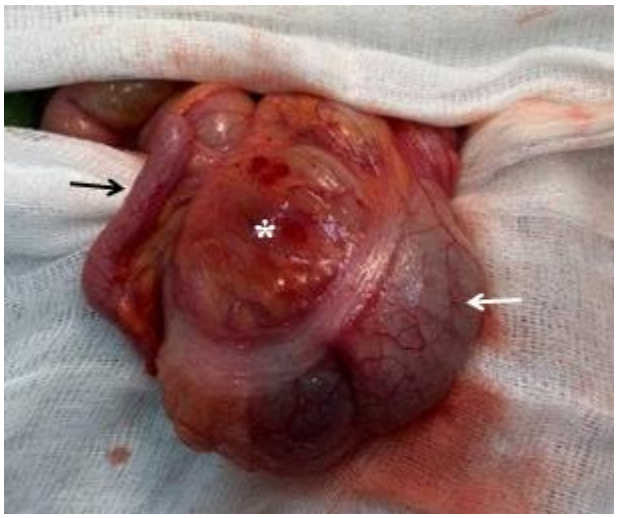

Figure 2. Macroscopic image in surgery shows intussusception at the ileocecal location. Looking outside the cecum, there are areas of congestion and bruising (white arrows), inside the cecum there is a cyst-like structure pushing the cecum wall (*), a non-inflamed appendix (black arrow).

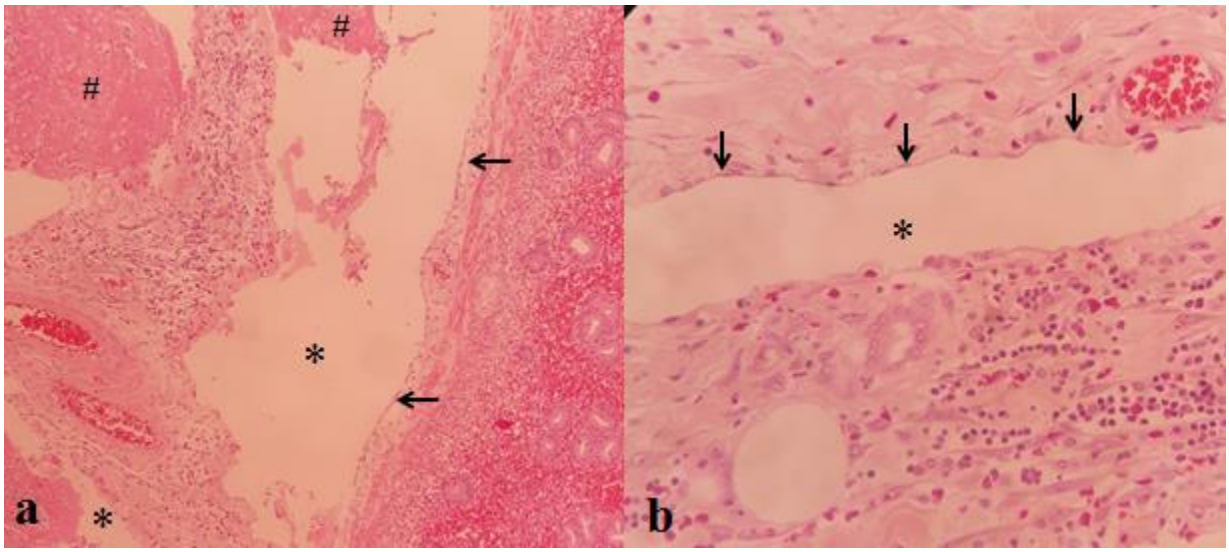

Figure 3. Photomicrograph of histopathologic specimen (HE stain) with H&E, 20X **(a)** and 40X **(b)**. **a)** Lymphatic cysts (*) covered by a layer of flattened endothelium (arrow) with areas of intracystic haemorrhage (#). **b)** Lymphatic cyst (*) covered by a layer of flattened endothelium (arrow).