Case Report
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CT Findings of Ileal Cystic Lymphangioma: A Case Report[1]
회장에 생긴 낭성림프관종의 CT 소견: 증례 보고[1]

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Cystic lymphangioma is a rare benign submucosal tumor which originates from sequestered lymphatic tissue presenting as cystic lesions, but in rare cases originates in the ileum. Herein, we present a case of histologically proven ileal lymphangioma in a 48-year-old woman with an emphasis on the computed tomography (CT) imaging findings and pathological features. We also present a brief review of the relevant literature.

Index terms
Ileum
Lymphangioma
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INTRODUCTION

Cystic lymphangiomas are rare benign tumors originating from a congenital malformation of the lymphatic system and presenting as cystic lesions. The vast majority (~95%) of lymphangiomas are found in the head, neck, and axillary regions (1, 2). It also can present in the gastrointestinal (GI) tract, pancreas, omentum, mesentery, and retroperitoneum in the abdominal cavity (1-4). Among these rare presentations, occurrence in the small intestine is especially rare (2, 5-7). Here, we report the computed tomography (CT) and pathologic findings of a patient diagnosed with cystic lymphangioma of the ileum.

CASE REPORT

A 48-year-old woman presented with diffuse pelvic pain and was referred to the department of obstetrics. She was diagnosed with menorrhagia and dysmenorrhea, and underwent a total abdominal hysterectomy as well as a left ovarian cystectomy upon diagnosis of a uterine myoma and a left ovarian cyst 12 years ago. Following surgery, she recovered without complications.

The patient visited a local clinic where a pelvic cystic mass was found around the right ovary on obstetric ultrasonography two years prior. A review of an outside hospital contrast-enhanced abdominal CT revealed a 6.7 × 6.5 × 6.0 cm sized lobulating-contoured and homogeneous thin-walled cystic mass with 14 Hounsfield units in the mesentery adjacent to the wall of the ileal loop without septa or an enhancing solid portion (Fig. 1A). It was hard to differentiate the origin of the mass; either from the ileum or the mesentery. There was no sign of proximal small bowel obstruction. Moreover, there was no perilesional fatty infiltration or regional lymph node enlargement around the mass. Considering these imaging findings, this mass could arise from either the ileum or mesentery, and the differential diagnosis included a cystic lymphangioma, mesenteric cyst, and other benign cystic tumors. As the patient wanted to avoid surgery and the lesion appeared benign, we decided to first observe the lesion and perform a follow-up study.

Two years later, she revisited our hospital because of lower abdominal pain and tenderness. A physical examination revealed a palpable mass in the lower quadrant of the abdomen with direct tenderness. Her complete blood count, urinalysis, and liver function tests were normal. Her serum level of CA-

125, CA 19-9, and CA 15-3 were within the normal range. The follow-up contrast-enhanced CT revealed an increase in size to 7.2 × 8.8 × 6.5 cm and a change in shape of the mass (Fig. 1B).

A diagnosis could not be made by only these imaging findings and laboratory results. As the mass was enlarged and its shape was changed, we decided to perform a surgical resection of the mass. Intraoperative findings revealed that the mass was attached to the mesenteric side of the ileum, 30 cm proximally from the ileocecal valve, and showing a well-defined exophyting nature. Complete excision of the mass, including segmental small bowel resection, was performed.

Grossly, the mass presented as yellowish white cysts containing a chylous component. The microscopic findings showed intercommunicating cystic spaces in the muscle and serosal layer of the ileum (Fig. 1C). The cystic spaces contained pinkish proteinaceous fluid and the wall is composed of thin fibrous tissue (Fig. 1D). The histopathologic examination showed many fluid-containing cystic lesions lined with endothelial cells on the serosal layer below the normal mucosa. A collection of lymphocytes was also seen and there was no abnormal pathology in the mucosa. The results of pathologic examination were consistent with the diagnosis of cystic lymphangioma.

The patient was discharged one week after surgery without any complications, and the subsequent postoperative period was uneventful.

DISCUSSION

Cystic lymphangiomas are uncommon benign tumors that are usually found in the head, neck, and axillary regions (1, 2, 8). They also involve GI tract, though the incidence is low (9). Lymphangiomas in the GI tract have a predilection for the colon,

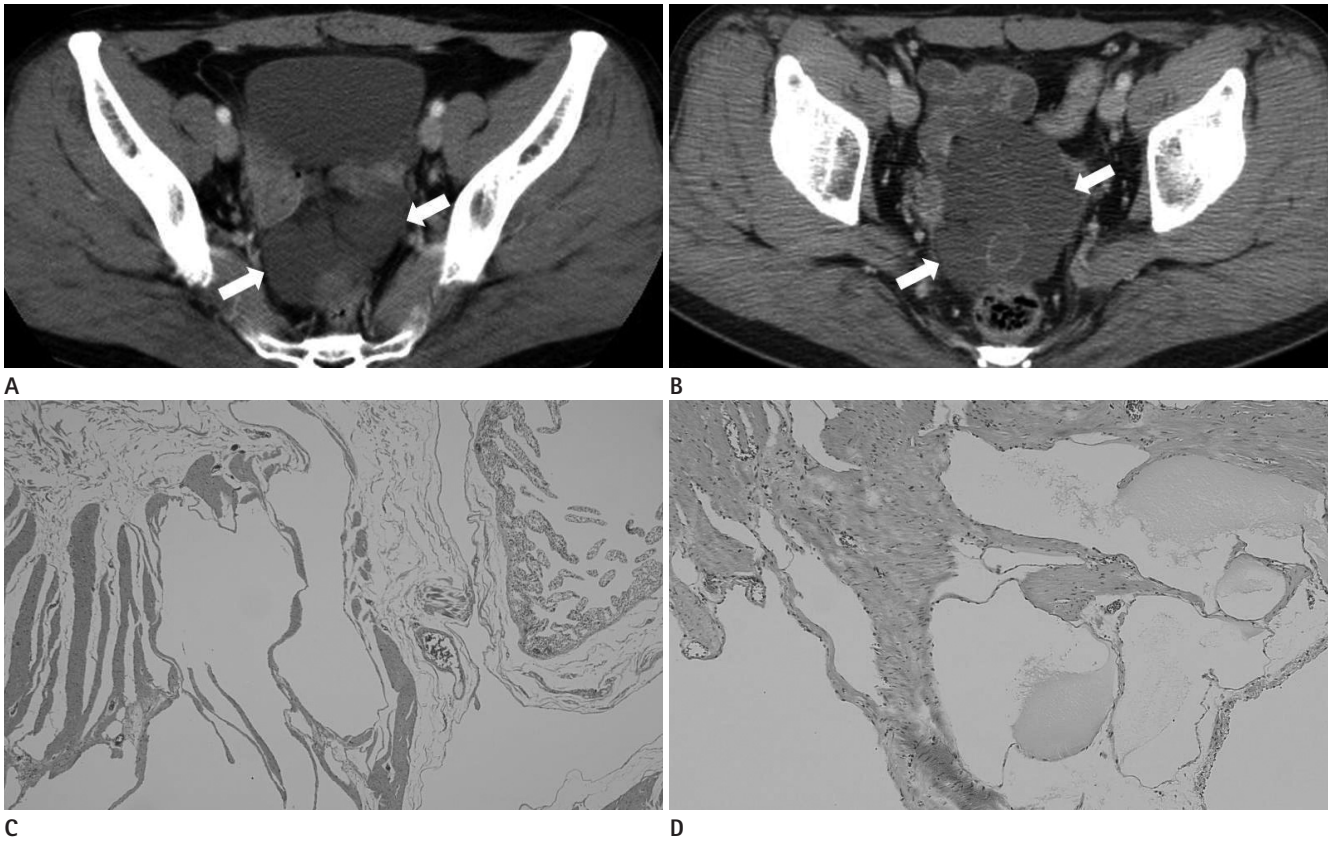

Fig. 1. Cystic lymphangioma of the ileum in a 48-year-old woman.
A. Contrast-enhanced axial CT image shows a lobulating-contoured and homogeneous thin-walled cystic mass (arrows) without internal septation or calcification in the pelvic cavity.
B. On 2-year follow-up contrast-enhanced axial CT imaging, the mass (arrows) shows an increase in size and change in shape without significant enhancement in the pelvic cavity.
C. Microscopic findings showed intercommunicating cystic spaces in the muscle and serosal layer of the ileum (Hematoxylin & Eosin stain, × 40).
D. The cystic spaces contain pinkish proteinaceous fluid with a wall composed of thin fibrous tissue (Hematoxylin & Eosin stain, × 200).

followed by the duodenum and stomach (2), but are relatively rare in the ileum. They generally have a single cavity and rarely present as a multilocular mass (3). In the literature, only a few reported cases of ileal lymphangiomas exist (2, 5-7).

The tumors occur as a result of developmental failure in communication between lymphatic vessels during fetal life, which resulted in dilated cystic tumors (4). On histologic examination, the tumors were lined with a single layer of flat endothelium and also contained some lymphoid tissue, small lymphatic spaces, smooth muscle, and foam cells with walls composed of thin fibrous connective tissue (4, 5, 10). Based on the pathologic findings which are described above, our case was diagnosed as cystic lymphangioma arising from the ileum.

Depending on their location and size, cystic lymphangiomas in the GI tract can cause symptoms including abdominal pain, a palpable mass, and even cause obstructive symptoms or intussusceptions. However, they are usually detected as incidental findings without symptoms (4, 5, 10). A high risk of ileus due to the torsion of the small intestine has been reported (10). In our case, the patient complained of chronic lower abdominal pain.

On ultrasonography, the tumors commonly appeared as a well-defined cystic or multicystic mass, and often with internal septations (1). The appearance of cystic lymphangiomas on CT is variable but is most often described as a well-demarcated, thin-walled unilocular or multicystic mass with a submucosal location, containing septa with possible multiple calcifications (4). In addition, CT and MRI can differentiate between chylous fluid, blood, and pus, as well as provide a specific anatomical location with adjacent organ involvement and even accompanied complications. In our study, CT showed a large, lobulated, thin-walled, low attenuation mass in the ileal loops. In addition, there were no calcifications, septations, or proximal intestinal canal dilatation. Previously reported cases of ileal lymphangimas showed similar CT findings (4). These imaging studies may also be valuable in the preoperative diagnosis; however, the preoperative diagnosis of cystic lymphangiomas is difficult due to non-specific imaging findings. Differential diagnosis of cystic lymphangiomas of the small intestine should be including many other cystic tumors such as duplication cysts, teratomas, gastrointestinal submucosal tumors, cystic mesotheliomas, lymphangiosarcomas, and hemangiomas (1, 3, 6). Several authors have reported that fine needle aspiration is useful for confirming a preoperative diagnosis of a cystic lymphangioma (10).

Surgery is the treatment of choice for cystic lymphangiomas of the small intestine. Although lymphangiomas are benign, they often grow in an aggressively invasive manner and have the potential for recurrence. Partial or incomplete tumor removal may also be associated with complications such as infection, fistula, and hemorrhage. Hence, surgeons usually aim for complete removal of the tumor (7, 9, 10).

In conclusion, although ileal cystic lymphangiomas are relatively rare and their clinical manifestations and their radiologic appearance are non-specific, they can cause acute symptoms. Therefore, they should be considered in the differential diagnosis of abdominal cystic masses with an uncertain origin of the surgical abdomen.

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회장에 생긴 낭성림프관종의 CT 소견: 증례 보고[1]

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낭성림프관종은 드문 양성점막하종양으로 분리된 림프조직에서 기원하며 낭성병변으로 나타나는데, 회장에서는 매우 드물게 발생한다. 이에 저자들은 병리학적으로 확진된 48세 여자 환자에서 회장에 발생한 낭성림프관종의 CT 영상 소견과 병리학적 소견을 기술하고자 한다. 또한 관련 문헌을 간단히 정리하고자 한다.

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