

Cystic Lymphangioma Involving the Mesentery and the Retroperitoneum: A Case Report¹

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Cystic lymphangioma is uncommon angiomatous tumor that mainly occurs in the neck. Less than 1% of these tumors affect the mesentery, retroperitoneum and greater omentum. In particular, the cystic lymphangioma involving the mesentery and the retroperitoneum is a rare lesion. We report here on an uncommon case of cystic lymphangioma that presented as a multilocular mass involving the mesentery and the retroperitoneum, and we also present a brief review of the relevant literature.

Index words : Mesentery, neoplasms
Retroperitoneum, neoplasms
Cysts, CT
Lymphangioma

Cystic lymphangiomas involving the mesentery and the retroperitoneum are rare tumors. Most of the cystic lymphangiomas occur in the neck, and they generally have a single cavity with only a small proportion of these tumor being multilocular. The cystic spaces are lined with a single layer of endothelium and there are small lymphoid aggregates in the cyst's wall that help distinguish lymphangiomas from the simple cysts of the mesentery. We describe here an uncommon case of cystic lymphangioma presenting as a multilocular mass involving the mesentery and the retroperitoneum.

Case

A 20-year-old male patient was referred to the depart-

ment of general surgery for an operation on an intraabdominal mass that was incidentally detected. He had back pain after minor trauma 4 weeks earlier, and magnetic resonance imaging (MRI) demonstrated a huge cystic mass extending from the upper intraabdominal cavity to the pelvic cavity, and there was herniation of the nucleus pulposus at the L4 - 5 level (Fig. 1A). The patient's past medical history was unremarkable and he was asymptomatic. The physical examination revealed a blood pressure of 100/80 mm Hg, a pulse of 80 beats/min, respiration of 24 breaths/min, and a body temperature of 36.5 °C. There was no palpable mass in the abdomen and the bowel sounds were hypoactive. The laboratory data on admission, including the blood chemistry profile, coagulation studies and complete blood count revealed unremarkable findings. An admission abdominal X-ray showed diffuse increased opacity that suggested a soft tissue mass in the abdomen. The transabdominal ultrasonography (US) revealed a multi-septated cystic mass without any definite wall (Fig. 1B). The computed tomography (CT) scan showed a large cyst involving the mesentery and the retroperitoneum

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(Figs. 1C, D). The possible clinical and radiological diagnoses at that time included mesenteric cystic lymphangioma, multicystic mesothelioma, other mesenteric cysts and lymphoma. Exploratory laparotomy revealed

a broad based cyst in the mesentery measuring $19 \times 15 \times 6$ cm; there were dense adhesions to the adjacent bowel loop, and the mass involved the retroperitoneum through the duodenal recess. The cyst's wall was yel-

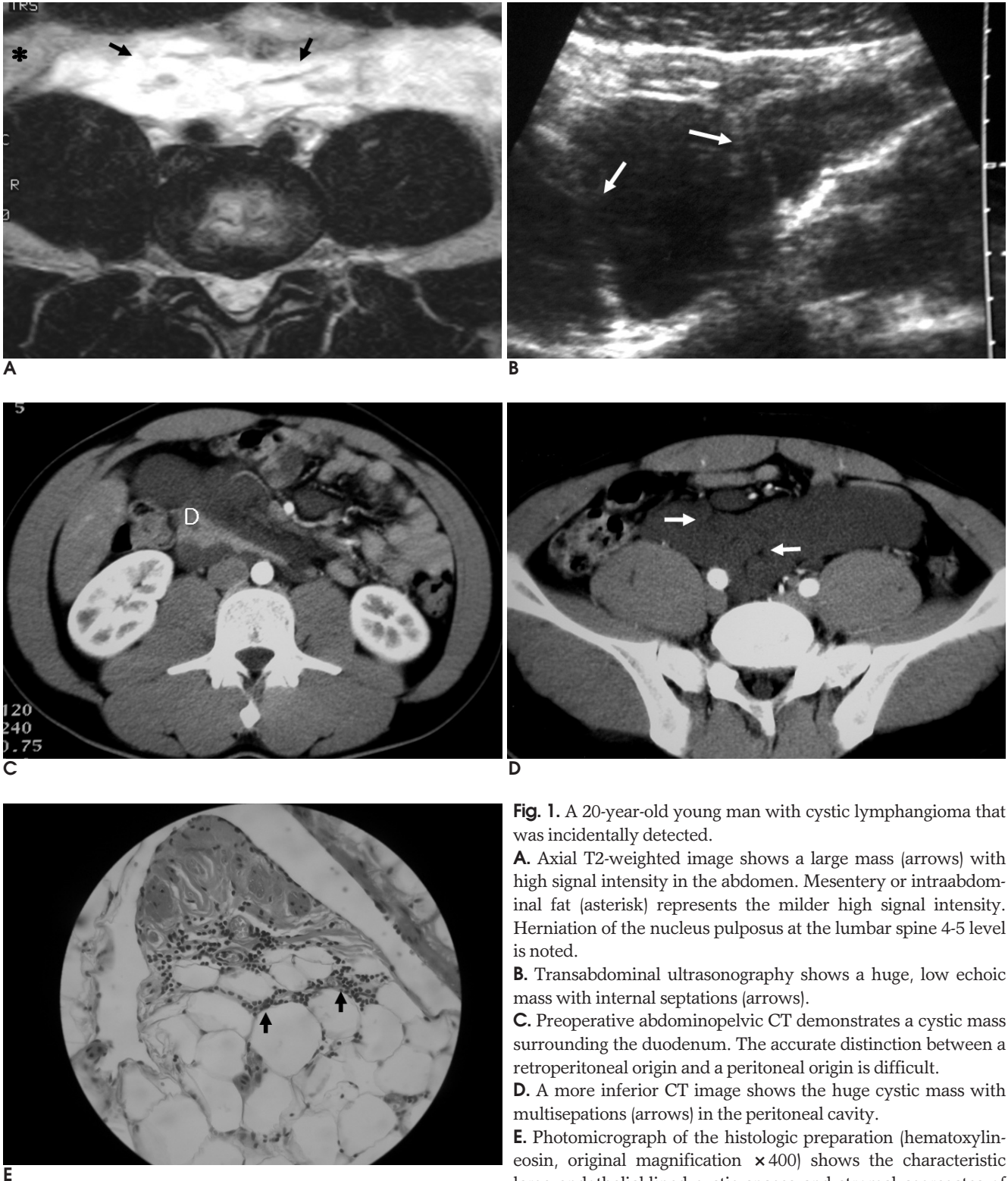


Fig. 1. A 20-year-old young man with cystic lymphangioma that was incidentally detected.

A. Axial T2-weighted image shows a large mass (arrows) with high signal intensity in the abdomen. Mesentery or intraabdominal fat (asterisk) represents the milder high signal intensity. Herniation of the nucleus pulposus at the lumbar spine 4-5 level is noted.

B. Transabdominal ultrasonography shows a huge, low echoic mass with internal septations (arrows).

C. Preoperative abdominopelvic CT demonstrates a cystic mass surrounding the duodenum. The accurate distinction between a retroperitoneal origin and a peritoneal origin is difficult.

D. A more inferior CT image shows the huge cystic mass with multiseptations (arrows) in the peritoneal cavity.

E. Photomicrograph of the histologic preparation (hematoxylin-eosin, original magnification $\times 400$) shows the characteristic large endothelial-lined cystic spaces and stromal aggregates of lymphocytes (arrows).

lowish-gray and glistening; no enlarged lymph nodes were found. The rest of the abdomen was normal and any free intraabdominal fluid was not found. The mass was resected and upon sectioning, multicystic lobules with chylous whitish fluid were noted. Microscopically, the cyst's wall showed an endothelial lining, smooth muscle fibers and fibrovascular adipose tissue. Lymphocytic aggregates were seen throughout the cyst's wall along with diffuse chronic inflammatory infiltration being seen below the endothelial lining (Fig. 1E).

After surgery, the patient complained of diffuse abdominal discomfort and repetitive vomiting for 2 weeks. Postoperative US and CT demonstrated a hematoma surrounding the duodenum and residual cystic mass. Close observation was done and the patient made a recovery without residual sequelae.

Discussion

Mesenteric cystic lymphangioma is an uncommon mesenteric mass. The gross and histological findings (Fig. 1E) in our case are similar to those findings reported in other series (1 - 5). However, when we clinically compare our case with the previous studies, our case demonstrated no symptoms, nor did it radiologically demonstrate any of the unusually involved sites like the mesentery and retroperitoneum.

Cystic lymphangiomas are usually located in the neck and axilla, and they rarely occur in the mediastinum, lungs, esophagus, diaphragm, duodenum, stomach, small and large bowels, spleen and liver. Less than 1% of these tumors affect the mesentery, greater omentum and retroperitoneum. Lymphangioma involving the mesentery and retroperitoneum, such as our case, is rare. Meyer et al. have reported on a retroperitoneal lymphangioma in which the cystic tumor originated from the retroperitoneum with a broad attachment to the pancreas, and it had extended anteriorly into the root of the mesentery, thereby mimicking the radiological features of a mesenteric cyst (6). In our case, the tumor was clearly separable from the pancreas and the tumor was intraoperatively defined as a mesenteric mass growing posteriorly and taking up a retroperitoneal portion through the duodenal recess. Many other cystic tumors can involve the mesentery or retroperitoneum, and cystic mesothelioma, lymphangiosarcoma, myxoid degeneration of lymphangioma and also hemangioma must be included in the differential diagnosis.

Lymphangiomas are usually restricted to the mesentery, omentum, mesocolon and retroperitoneum. In contrast, benign cystic mesothelioma tends to have a pelvic location and there is involvement of the upper abdomen and retroperitoneum in some cases (7, 8).

Histologically, lymphangioma displays dilated lymphatic vessels that are lined by flattened endothelium in between lobules of adipose, fibrous and lymphoid tissue. In contrast, the majority of mesenteric cysts often display a cuboidal or columnar epithelial lining that lacks smooth muscle cells or lymphatic elements.

The most common finding of mesenteric cystic lymphangioma on plain abdominal radiographs is a soft-tissue mass with displacement of the bowel loops. Transabdominal ultrasonography is a very sensitive imaging modality, and the mass generally appears on US as a sharply defined cystic or multicystic mass, and there are often internal septations such as was seen in our case (Fig. 1B). The fluid can be anechoic, or there are scattered internal echoes that represent infection or hemorrhage. CT and MRI can give important preoperative information regarding the anatomical location, the cyst size, organ involvement and the possible complications. In addition, these two modalities can differentiate between chylous fluid, blood and pus.

The standard therapy for patients with mesenteric cystic lymphangioma is surgery. Total removal of the mass that invades the potentially respectable intraabdominal structures such as the bowel, spleen or pancreas is possible. However, adhesions to vital structures can sometimes make resection dangerous or even impossible. Other treatment methods are palliative surgical treatment or sclerosing agents. There are still high recurrence rates for the totally excised and incompletely resected mesenteric cystic lymphangiomas, and this indicates a continuing need to develop new and effective treatment options to supplement surgery (9).

In conclusion, cystic lymphangioma involving the mesentery and the retroperitoneum should be included in the differential diagnosis of cystic intraabdominal lesions. Even when the patient is asymptomatic and this tumor is discovered incidentally, mesenteric cystic lymphangioma must be treated surgically because of its potential to grow and invade vital structures, and this tumor can also develop life-threatening complications.

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