Giant mesenteric cystic lymphangioma presenting with abdominal pain and masquerading as a gynecologic malignancy

John Maa,1 Christianne Wa,2 Adnan Jaigirdir,1 Soo-Jin Cho,3 Carlos U. Corvera1

1Department of Surgery, University of California; 2University of Southern California; 3Department of Anatomic Pathology, University of California, San Francisco, CA, USA

Abstract

Lymphangiomas are congenital malformations of the lymphatic system that account for about 5% of all benign tumors in infants and children.1 The most common sites are the neck and axilla, which account for 95% of cases.2 Abdominal cystic lymphangiomas are quite rare, and can arise from either the retroperitoneum, gastrointestinal tract, or the mesentery of the abdominal viscera.3 The presenting symptoms are painless abdominal distension, a palpable mass, or secondary complications in the abdomen such as intestinal obstruction, volvulus, intestinal infarction, or bleeding.4 Typically diagnosed during childhood, these tumors prompt surgical intervention. We describe an atypical case of an abdominal cystic lymphangioma, which did not manifest until adulthood, with atypical symptoms of a rapidly expanding and symptomatic mass.

Case Report

A 36-year-old Gravida 4 Para 2 woman came to the emergency department with vague abdominal discomfort, abdominal distension, and decreased energy. She had noted an abdominal mass while on vacation approximately one month before presentation, which had expanded rapidly over the following 10 days. She was healthy otherwise but had undergone dilatation and curettage for a molar pregnancy three years previously. Though she had immigrated to the United States, she did not report previous exposure to toxic chemical agents. A palpable fullness in the left lower quadrant was noted during her physical examination. A CA-125 blood test for ovarian cancer and serum laboratory studies were normal. A computed tomography (CT) scan revealed a 14.0x5.4x16.2 cm cystic lesion in her pelvis and retroperitoneum, which was suspected to be of ovarian origin. Magnetic resonance imaging (MRI) again demonstrated a multicystic lesion that appeared intense on a T2-weighted image (Figure 1). The mass extended superiorly to the base of the mesentery, abutting the superior mesenteric vessels and occupying the entire pelvis inferiorly. Although the ovaries and uterus appeared to be distinct from this mass, a gynecologic etiology could not be excluded.

Given the concerns of either a gynecologic malignancy, pseudomyxoma peritonei, or a peritoneal mesothelioma, a curative resection was planned. During the operation, a multicystic mass that appeared to engulf the entire sigmoid colon was noted. It was well encapsulated and was contained within the leaflets of the grossly distorted sigmoid mesentery. The transition to normal colon mesentery proximally, and mesorectum distally, was clear, sharp, and abrupt, suggesting a benign etiology. There was no evidence of either carcinomatosis or suspicious metastatic tumor deposits. The fluctuant mass was exteriorized and resected with a wide mesenteric and colon margin. A primary hand-sewn anastomosis was utilized for bowel reconstruction. After an uneventful recovery, the patient was discharged on postoperative day five.

Gross pathologic evaluation revealed an unremarkable segment of colon with an associated large cystic mass (Figure 2). Permanent sections revealed a benign cystic lymphangioma, composed of innumerable thin-walled channels, the walls of which focally contained aggregates of bland lymphocytes. A few clusters of macrophages with moderate amounts of eosinophilic cytoplasm were present in the lumina of the cystic spaces, as is seen commonly in cystic vascular spaces with sluggish flow. Immunohistochemical staining for D2-40 (a marker of lymphatic endothelium) showed positive staining in the attenuated cyst lining cells, supporting the impression of lymphangioma (Figure 3). In addition, immunohistochemical staining for calretinin (a mesothelial marker) was performed and the cyst lining cells were negative, arguing against a cystic mesothelial proliferation. The left ovary, although cystic, was uninvolved by the lymphangioma.

Discussion

Cystic lymphangiomas are rare congenital benign malformations of the lymphatic system and account for about 5-6% of all benign tumors in infants and children.1 Abdominal cystic lymphangiomas are more frequent in boys than girls (5:2), and approximately 80-90% are diagnosed within the first few years of life. Approximately 50% of cases involve the retroperitoneum, which was suspected to be of ovarian origin.

Correspondence: John Maa, 521 Parnassus Avenue, San Francisco, CA 94131-0790, USA. E-mail: maaj@surgery.ucsf.edu

Key words: cystic lymphangioma, mesenteric lymphangioma, retroperitoneal lymphangioma.

Acknowledgments: the authors thank Pamela Derish, UCSF Department of Surgery, for editorial assistance and Dr. Annemieke van Zante, UCSF Department of Anatomic Pathology, for review of the pathologic findings.

Contributions: JM, AJ, and CC were involved in the design of the study, perioperative care of the patient, and obtaining operative photographs of the tumor. CW, JM, and CC drafted and revised the manuscript, and approved the final version. SC was involved in the pathologic examination of the resected specimen and critical revision of the manuscript.

Conflict of interest: the authors report no conflicts of interest.

Received for publication: 15 October 2009. Accepted for publication: 21 October 2009.

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Rare Tumors 2009; 1:e48
doi:10.4081/rt.2009.e48

Figure 1. Magnetic resonance image demonstrating the mesenteric cystic lymphangioma. (A) Axial T2-weighted image. White arrow points to the sigmoid colon engulfed by the tumor mass. (B) Sagittal view. White arrow points to the engulfed sigmoid colon.
head and neck, with only 10% occurring in internal organs. These tumors result from an abnormal embryonic development of the lymphatic system causing sequestration of lymphatic tissue and fluid, as in our patient.

Clinical manifestations are variable, without characteristic signs and symptoms that can be attributed solely to the mesenteric lymphangioma. They may present as nonspecific but painful abdominal symptoms. When a cystic lymphangioma is suspected, either an abdominal ultrasound examination, CT scan, or MRI can be used for diagnosis. Percutaneous biopsy is not recommended because the diagnostic value is likely to be low owing to the low cellularity index of these tumors. Moreover if the tumor is demonstrated to be malignant, the risk of needle tract metastasis or peritoneal dissemination of the fluid is unacceptably high. In our patient, the preoperative imaging studies suggested the mass to be technically resectable even if malignancy would have been encountered, thus obviating the need for preoperative tissue biopsy. Although laparoscopic approaches utilizing partial aspiration intraoperatively have been described, we selected the open approach via laparotomy given the large size of the lesion and the concerns of a possible malignancy.

Our patient is unusual because she was diagnosed as an adult and experienced the rapid onset of abdominal swelling and distension as her presenting symptoms. Given her previous history of a molar pregnancy three years before, a possible gynecologic origin of trophoblastic tissue accounting for the mass was strongly considered. No abnormalities had been visualized on her previous abdominal sonograms for a subsequent pregnancy. The cause of the rapid expansion of the tumor remains unknown. No free fluid was found at surgery to suggest perforation.

Surgeons should be aware of this rare entity. In our case, the low Hounsfield units on CT scan raised suspicion of lymphangioma, a diagnosis supported by the MRI characteristics, particularly on the T2-weighted images. However although imaging studies favored a benign cystic condition, it was important to consider that the patient might have a malignant mucinous tumor. Accordingly she was counseled preoperatively that if malignancy was found, an aggressive surgical approach would be warranted, potentially involving a multiorgan resection to achieve gross complete resection or a diverting colostomy.

In conclusion, cystic abdominal mesenteric lymphangiomas are rare tumors that can present in adult life occasionally, and masquerade as a malignant gynecologic lesion. Continued advances in cross-sectional imaging undoubtedly will help further distinguish benign cystic lesions from malignant ones. In the meantime, we think these cystic tumors should be managed as malignant until proven otherwise.

References