Cystic Lymphangioma of the Small Intestine in a Young Girl

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Abstract

We describe a case of a 12-year old girl who presented with abdominal pain. A large abdominal mass was discovered on physical examination. Preoperative studies including abdominal ultrasonography and CT scan were performed, but they could not accurately determine the nature of the tumor. At laparotomy, a large cystic tumor of the small bowel was found. Histopathologic examination revealed a tumor of cystic lymphangioma. Although lymphangiomas are rare, especially in the abdomen, they may sometimes present as abdominal pain and acute abdomen, causing complications that require emergent surgery.

Keywords: Lymphangioma; Small bowel; Abdominal mass

Introduction

Cystic lymphangioma is a cystic lymphatic lesion that can affect any anatomic subsite in the human body. Approximately 20% of cystic lymphangiomas occur in the axilla, more infrequent subsites including the mediastinum, groin, and retroperitoneum.¹ Ninety percent of cases are symptomatic by the age of two years. Cystic lymphangioma was first described by Wernher in 1843. Lymphangiomas are uncommon and occur mainly in children; approximately 80-90% of them are diagnosed within the first few years of life and adult cases are rare.² They are commonly found in the head, neck, and axillary regions. However, isolated occurrence in the small intestine is encountered even less frequently, comprising less than 1% of all lymphangiomas.³ The most common location of such intraperitoneal lymphatic tumors appears to be the mesentery of the small bowel, accounting for 70% of all such tumors.⁴,⁵ The etiology of lymphangiomas is probably a congenital abnormality of the lymphatic system, causing sequestrations of lymphatic tissue during embryologic development.³ This theory would explain why lymphangiomas occur primarily in children. However, it is suggested that abdominal trauma, lymphatic obstruction, inflammatory process, surgery, or radiation therapy may lead to the secondary formation of such a tumor.⁵,⁶ Traditionally, lymphangiomas are classified as simple, cavernous, or cystic. The simplest type is usually situated superficially in the skin and composed of small thin-walled lymphatic vessels. The cavernous type is composed of dilated lymphatic vessels and lymphoid stroma, and has a connection with spaces of various normal adjacent lymphatics. Finally, the cystic type consists of lymphatic spaces of various sizes and contains fascicles of smooth muscle and collagen bundles, but has no connection with the bowel. The tumor is not usually adhered to the small intestine and adjacent normal lymphatics. However, cystic lymphangioma is not always clearly differentiated from cavernous type because the cystic type may also contain cavernous areas.⁷

Case Report

A 14-year old girl was referred to Department of Surgery on March, 2008 due to abdominal mass found on physical examination and ultrasonography. Recently, she had begun to experience hypogastralgia and ab-
dominal fullness, especially after having meals. Her family and medical history were unremarkable. She had no history of previous abdominal surgery. On physical examination, a soft, non-tender mass was palpated in the lower abdomen. Auscultation of the abdomen revealed normal bowel sounds. Laboratory data including tumor markers were within normal limits (Hb:13.2, WBC:4.7, Plt:229). Abdominal ultrasonography revealed a large cystic mass measuring 30×28×9 cm in size with a well differentiated margin with right ovarian origin. CT scan of the abdomen showed a large homogeneous mass measuring 33×30×10 cm in size. Based on such findings, the probable diagnosis was ovarian cyst. This lesion was located at the anterior aspect of the right ovary and the descending colon, causing compression and stretching of the small bowel (Figure 1).

At laparotomy, a yellowish cystic tumor with soft consistency was found in the mesenteric aspect of the ileum. The tumor was adhered to the wall of the intestine. No ascites was seen in the peritoneal cavity, nor was there any dilatation or inflammatory change of the intestines or mesentery. At first, the cyst was punctured and opened. It was filled with a milky liquid. The tumor was resected completely with 15 cm resection of the intestine. The specimen consisted of a multilocular cyst on the mesenteric aspect of the intestine, measuring 14×9 cm. Macroscopically, it was yellowish in color and soft in consistency. The cyst wall was smooth, measuring up to 0.2 cm in thickness. Microscopically, the small intestine had a normal mucosa. In the mesenteric area, ill-defined cystic neoplasm composed of multiloculated cysts lined by a flat layer of endothelium on the loose stroma was seen. Some foamy macrophages scattered in the luminal side of the cyst were present. Also, infiltration of the lymphocytes in the stroma was seen. These findings were consistent with a cystic lymphangioma (Figures 2A-C). The patient had an uneventful postoperative course with no evidence of recurrence, 3 months after the operation.

Discussion

Abdominal lymphangiomas are usually asymptomatic until they get enlarged or become complicated. Abdominal pain and distention seem to be the most common symptoms, but the clinical presentation of the abdominal lymphangiomas varies. Although abdominal lymphangiomas are benign in nature, they may cause significant morbidity or mortality due to their large size and critical location, when compressing the adjacent structures. In addition, complications such as secondary infection, rupture with hemorrhage, volvulus, or intestinal obstruction have all been reported.

The ultrasonographic presentation of an abdominal lymphangioma is described as a cystic lesion with multiple thin septa. In our case, abdominal ultrasonography revealed a large cystic mass measuring 30×28×9 cm in size with a well differentiated margin. On CT imaging, abdominal lymphangiomas appear as a uni- or multilocular mass with enhancement of the wall and septum by contrast medium. In our case, CT scan of the abdomen showed a large homogeneous mass measuring 33×30×10 cm in size. Such studies will help to determine whether the tumor is cystic, how large it is, and where its location is,
but these findings are insufficient to establish an accurate diagnosis preoperatively. One report suggested that fine needle aspiration of the milky fluid containing lymphoid cells could confirm a preoperative diagnosis of lymphangioma.\textsuperscript{14} Abdominal lymphangiomas may behave in an aggressively invasive manner and grow to an enormous size. The optimal treatment is radical excision, even when asymptomatic. However, abdominal lymphangiomas may cause complications such as infiltration of the intestine, or involvement of the main branch of mesenteric arteries or adjacent organs that necessitate segmental resection of the intestine. Sometimes, radical resection might be technically impossible.\textsuperscript{15} Abdominal lymphangiomas are very rare, but they can cause acute abdomen that requires an emergent surgery. Therefore, they should be included in the differential diagnosis of the acute abdomen.

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References


