

CASE NOTE

Giant mesenteric cystic lymphangioma in an infant presenting with acute bowel obstruction

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Abdominal cystic lymphangiomas (ACLs) are rare congenital benign malformations of the lymphatic system that are usually located in the small-bowel mesentery, followed by the omentum, mesocolon and retroperitoneum.¹ Most lymphangiomas are found in the head and neck; intra-abdominal locations are very unusual. Abdominal cystic lymphangiomas are more frequent in boys (5:2) with a mean age at presentation of 2 years.² Clinical presentation varies from asymptomatic masses to acute abdominal pain. We report the unusual case of a 14-month-old boy with a giant, cystic, small bowel mesenteric lymphangioma who presented with intestinal occlusion.

CASE REPORT

A 14-month-old boy presented with diffuse abdominal distention, nausea, vomiting and severe pain after a 3-week history of abdominal discomfort. Physical examination revealed severe abdominal distention with a palpable, soft and fluid-filled mass. Abdominal palpation was painful and tympanic. Bowel sounds were hyperactive.

Plain abdominal radiographs showed air–fluid levels and distended bowel loops, suggesting intestinal occlusion. Ultrasonography demonstrated a hypo-echoic cystic mass measuring 15 × 16 × 12 cm in size located in the inferior abdominal cavity. Computed tomography (CT) revealed a huge cystic lesion compressing the bowel loops (Fig. 1). The tumour was located in both iliac spaces. We suspected a mesenteric cyst and planned a surgical intervention.

At laparotomy, we found a giant, soft, cystic, milky fluid-filled mass in the mesentery of the ileum (Fig. 2). Intestinal resection of the involved loops was necessary. Pathological examination confirmed a diagnosis of mesenteric uniloculated cystic lymphangioma containing chylous milky fluid. The mass weighed 1.12 g. At 5 years old, the patient had experienced no recurrence of disease noticed on long-term follow-up.

DISCUSSION

Lymphangiomas account for about 5%–6% of all benign tumours in infants and children.^{1,3} Fifty percent of cases involve the head and neck, with only 10% occurring in internal organs.² Sixty percent of these masses are present at birth. Abdominal cystic lymphangiomas are very uncommon. Almost 90% are detected by the mean age of 2 years, and most occur in the mesentery of the small bowel.⁴ They result from an embryological failure of the lymphatic system: lack of communication between small bowel lymphatic tissue and the main lymphatic vessels during fetal

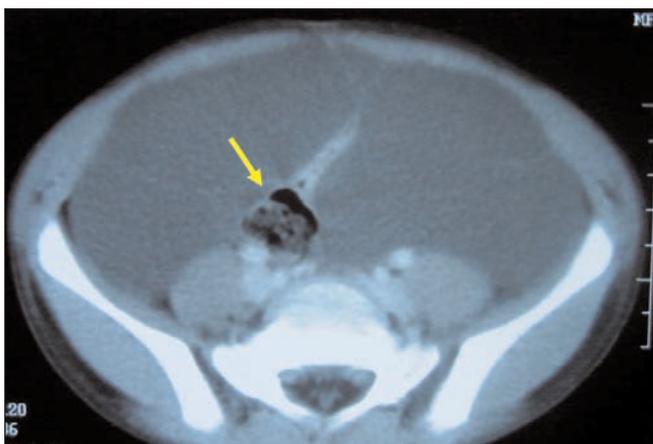


Fig. 1. Computed tomography scan illustrating a large cystic mass pushing against a loop of small bowel (arrow).

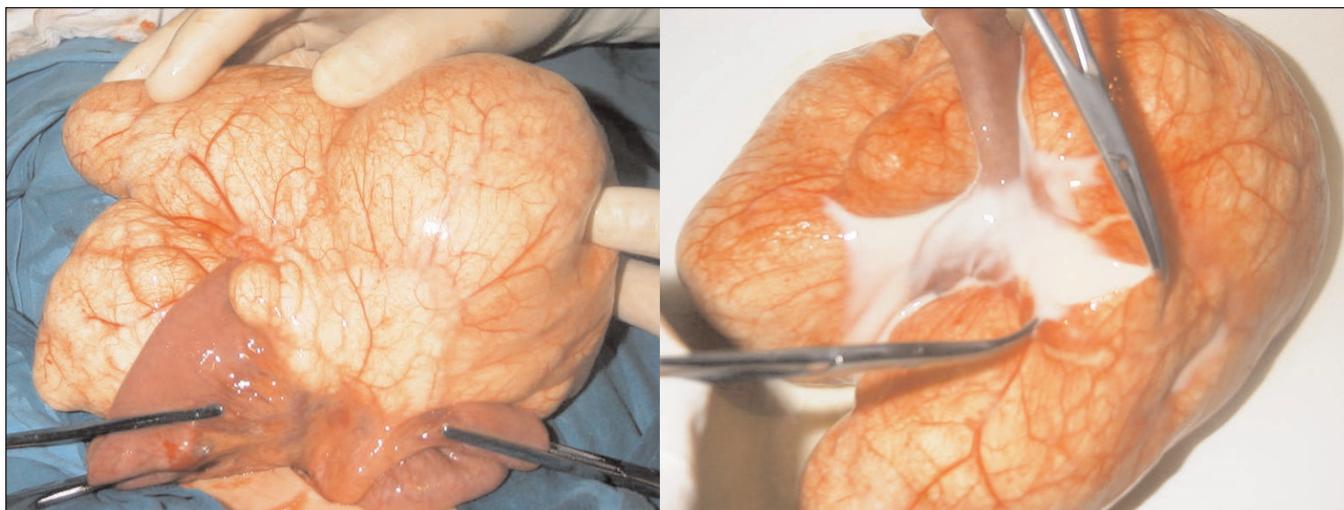


Fig. 2. Photographs demonstrating (left) a giant single-cavity mesenteric cystic lymphangioma compressing the overlying small bowel causing obstruction and (right) the mesenteric cystic lymphangioma after surgical removal showing the chylous milky fluid.

development result in blind cystic lymphatic spaces lined by endothelial layers.

Histopathologically, ACLs are characterized by a thin irregular wall covered by endothelium and containing smooth muscle, foam cells and lymphatic tissue.^{1,4} Clinical presentation is variable and depends on mass size and location. Most ACLs present with a large, slow-growing and mobile mass along with abdominal distention.² Abdominal discomfort is common, with acute peritoneal symptoms due to rupture, volvulus, hemorrhage or infection occurring infrequently.

The differential diagnosis of ACLs must include other fluid-filled lesions such as pseudocysts, dermoid cysts, enteric duplications, lymphoceles or neoplasms like mesotheliomas, pancreatic tumours, lipomas, teratomas, leiomyosarcomas, neurofibromas or liposarcomas.⁵

Abdominal ultrasound is the diagnostic procedure of choice when ACL is suspected. On ultrasound, ACLs appear as a well-circumscribed cystic anechoic structures with irregular thin walls often containing septa. On CT scans, they usually appear as a smooth-margined unilocular or multilocular cystic mass with homogeneous fluid attenuation and density ranging from -4 to -34 Hounsfield units.² The cystic walls show strong enhancement after contrast injection. Magnetic resonance imaging (MRI) may be useful for diagnosis but cannot differentiate between dermoid cysts, cystic teratomas, cystic lymphangiomas or lymphoceles. It is, however, the most useful modality to assess mass extension, as it can demonstrate low signal intensity (similar to muscle) on T_1 -weighted images and high signal intensity on T_2 -weighted images. Occasionally, complicated ACLs may show high signal intensities on both T_1 - and T_2 -weighted images owing to infection, volvulus or hemorrhage.¹ The combined use of ultrasonography, CT and MRI is very helpful when there is a high preoperative index of suspicion for ACL.

The definitive treatment for ACL is complete surgical excision. During surgery, as was the case with our patient,

it is often necessary to perform a bowel resection because of the close relation between the cyst and the intestinal wall. Some authors recommend conservative management in asymptomatic patients, based on a 10% spontaneous regression rate reported in these patients.^{1,5} Aspiration and injection of sclerosant agents may be recommended for emergency decompression, but as definitive therapy they have a high recurrence rate. Other treatments such as OK-432, bleomycin, steroids, fibrin glue and ethibloc have not demonstrated superior results to surgery in ACL.^{3,5} Some recent reports emphasize the role of laparoscopy for surgical resection. However, it should be noted that the 10% postoperative recurrence rate is due to incomplete resection as evinced by positive microscopic resection margins. Surgeons are right to worry about the invasive nature of these lesions.

In summary, ACLs frequently affect young children and are usually symptomatic. The diagnosis is well established by ultrasound, CT and MRI. To prevent recurrence, complete excision of the ACL with or without intestinal resection is mandatory.

Competing interests: None declared.

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