CASE REPORT

Omental Lymphangioma with Torsion: Case Report

FCY Lam,1 TW Fan,1 KF Chan,1 WY Leung,2 NSY Chao2

1Department of Radiology and Imaging, and 2Department of Surgery, Queen Elizabeth Hospital, Hong Kong

ABSTRACT

Lymphangioma is an uncommon benign lesion that usually occurs during childhood. Its presentation in the abdomen is rare. Its occurrence at the omentum, with complication of torsion, is even rarer. This report describes a 4-year-old boy who presented with abdominal pain and fever. Subsequent imaging, operative, and histological findings revealed omental lymphangioma with torsion.

Key Words: Diagnosis, differential; Lymphangioma; Omentum; Tomography, X-ray computed; Torsion

INTRODUCTION

Lymphangioma is an uncommon benign lesion that has a vascular origin and shows lymphatic differentiation.1 Lymphangiomas can occur in many anatomical locations. Most cases (95%) are found in the neck and axillary regions, whereas other sites, such as the mesentery, retroperitoneum, abdominal viscera, bone, lung, and mediastinum, are unusual.2-6 Although rare, abdominal lymphangiomas are more common in boys than in girls, and they usually occur during childhood.7 They are reported to occur most commonly in the mesentery, followed by the omentum, mesocolon, and retroperitoneum.1 We report a case of childhood lymphangioma presenting as abdominal pain, which was finally diagnosed as omental lymphangioma with torsion.

CASE REPORT

A 4-year-old boy presented to the Accident and Emergency Department of the Queen Elizabeth Hospital in August 2004 with a 2-day history of fever and abdominal pain. There was no vomiting or diarrhoea. The patient had unremarkable medical and family histories. Physical examination revealed right lower abdominal tenderness. Laboratory tests showed a normal white cell count. The abdominal X-ray showed a soft-tissue mass in the right abdomen and paucity of bowel gas at the pelvic and left lower abdominal regions. The intestinal loops were displaced to the upper part of the abdomen. No free gas was visible, and there were no obvious calcified areas (Figure 1).

Because the patient presented with an acute abdomen and fever, the presumptive diagnosis was abscess related to a ruptured acute appendicitis. Thus, no ultrasound

Figure 1. Supine abdominal radiograph showing displacement of bowel gas at the upper abdomen and paucity of bowel gas in the lower abdomen.
examination was performed at that time. Instead, computed tomography (CT) was performed (Figures 2 and 3), which revealed a large homogeneous mass with fluid attenuation mainly at the right and lower abdomen and pelvis. The mass extended from the subhepatic region to the pelvic and left lower abdominal regions. There was rim enhancement; no mural mass or calcification was detected. The loops of the small intestine were displaced to the left side of the abdomen. The appendix could not be located. In view of the patient’s history of fever, abscess probably due to a ruptured acute appendicitis was considered. Other differential diagnoses included duplication cyst, mesenteric cyst, and lymphangioma.

During emergency laparotomy, a large omental cyst with a stalk arising from the greater omentum was found. The cyst adhered to the serosa of the sigmoid colon and was twisted around itself by 360°. Omental cystectomy was performed; bowel loop resection was not necessary. The appendix appeared mildly erythematous, so appendectomy was also done, but pathological examination revealed a normal appendix.

The gross specimen of the omental cyst measured 14 x 8 x 2 cm. It contained multilocular cystic spaces and a central portion of firm whitish tissue measuring 3.0 x 2.0 x 1.5 cm that was surrounded by haemorrhagic tissue. This site corresponded to the site of torsion in the coronal reformatted CT image (Figure 4). Under the microscope, the cyst was lined by a single layer of endothelial cells. The wall was markedly congested and had very prominent dilated lymphatic vessels in a fibrous stroma (Figures 5 and 6). Furthermore, there were areas of interstitial haemorrhage and foci of neutrophil infiltration (Figure 7). After consideration of

Figure 2. Plain axial computed tomograms showing the lymphangioma as a large cystic lesion of fluid attenuation that occupied the mid and lower abdomen (a and b); no calcification or internal soft-tissue mass was detected.

Figure 3. Postcontrast computed tomograms showing enhancement at the rim and internal septa (a and b).
these and the operative findings, the final diagnosis was omental lymphangioma with torsion.

**DISCUSSION**

The aetiology of lymphangioma remains unclear. Because lymphangiomas occur mainly in children, the majority of cases are thought to derive from a congenital abnormality of the lymphatic system. Clinical presentation can be variable and non-specific. Acute symptoms include acute abdominal pain, distension, vomiting, fever, and peritonitis. Chronic symptoms include progressive abdominal distension and pain.

Plain radiographs may show a non-calcified soft-tissue mass, displacement of intestinal loops, as in our case, or features of small bowel obstruction. Ultrasonography and CT are highly sensitive tests that can be used in the diagnosis. Sonographically, lymphangiomas are anechoic cystic masses that have posterior acoustic enhancement. They can be multilocular with internal septa. Sometimes, internal debris or even solid echogenicity with a honeycomb pattern can be demonstrated. Their variable internal echogenicity is accounted for by the various contents that are possible.

CT can provide information regarding anatomical location, adjacent organ involvement, size, and complications. On CT scans, lymphangiomas are thin-walled multiseptated cystic masses. The attenuation of the fluid
ranges from that of clear or complicated fluid to that of fat, depending on the various contents. The cyst wall and septa can show enhancement after intravenous injection of contrast agent. Calcification is uncommon.

For the patient in our case, the omental cyst was large and it was difficult to trace its origin. On magnetic resonance imaging, lymphangiomas have a low signal intensity on T1-weighted images and a high signal intensity on T2-weighted images. If haemorrhage or infection occurs, the CT attenuation and magnetic resonance imaging signal pattern may alter.

Occasionally, it is difficult to differentiate lymphangioma from duplication cyst, mesenteric cyst, or abscess by imaging modalities. As in our case, in which the patient presented with acute abdomen, surgery was necessary no matter which of the above differential diagnoses applied. Imaging tests, such as CT, can help define the extent and site of the origin of the lesion. However, if the lesion is too large (as in our case), it may be difficult to delineate the origin.

Complications of lymphangioma include haemorrhage, infection, torsion (as in our case), or small-bowel obstruction. Konen et al. suggested that progressive enlargement, multiplication, thickening of the septa, and increased echogenicity of cystic fluid are signs that indicate complications that require urgent treatment. Ultrasonography and CT are helpful to detect such complications. However, the patient in our case had not undergone any previous imaging studies, so no comparison was available to suggest a complication. In addition, the stalk of the cyst was short and was difficult to detect during CT.

Differential diagnoses include other fluid-containing masses, such as duplication cysts, enteric cysts, pseudo-cysts, cystic teratoma, cystic leiomyoma, and leiomyosarcoma, or ovarian cystic masses in female patients. However, there can be no specific radiological features to differentiate between these options, and histological evaluation may be necessary. Ascites and lymphangioma can also be difficult to differentiate. The presence of septa, compression on adjacent intestinal loops, and lack of fluid in a dependent recess of the peritoneum and between leaves of the small-bowel mesentery suggest lymphangioma.

Malignant degeneration to low-grade sarcoma has been reported, but is rare. Complete surgical excision is the treatment of choice. Prognosis is excellent if the resection is complete. The recurrence rate is lower for complete resection compared with incomplete resection, aspiration, or injection of sclerosing agent, such as bleomycin or OK-432. The patient in our case underwent complete resection of the lymphangioma. Long-term follow-up is still necessary to detect recurrence or malignant change.

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REFERENCES