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Cystic lymphangioma of the mesentery causing intestinal obstruction

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ABSTRACT

Mesenteric cystic lymphangioma is a rare lesion that is not often described in the literature. A four-year-old boy, who presented with abdominal distension and pain, is reported. At surgery, a huge mesenteric cyst was found to be the cause of the intestinal obstruction and was completely excised. Histology was consistent with a cystic lymphangioma. Abdominal lymphangioma is a rare cause of bowel obstruction. Clinical presentation varies and may be misleading due to a lack of awareness of the clinical condition. Occasionally, the diagnosis is made during surgery. General awareness of this entity with a high index of suspicion is needed to avoid complications.

Keywords: abdominal lymphangioma, acute intestinal obstruction, cystic lymphangioma, lymphangioma, mesenteric lymphangioma

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INTRODUCTION

Cystic lymphangiomas occur most frequently in the head and neck or axilla of young children. Intraperitoneal and retroperitoneal cystic lymphangiomas are uncommon benign tumours of congenital origin. Cystic lymphangioma presents either with chronic abdominal distension (detected by palpation of a cystic mass) or acutely with bowel obstruction or signs of peritonitis. We report one such case that presented with acute intestinal obstruction, along with a brief review of the literature.

CASE REPORT

A four-year-old boy presented with abdominal distension and pain of a two-week duration. On examination, the abdomen was uniformly distended, with no tenderness and no palpable mass. Per rectal examination revealed an empty non-dilated rectum. Abdominal radiographs showed evidence of dilated large bowel loops with no gas within the rectum.

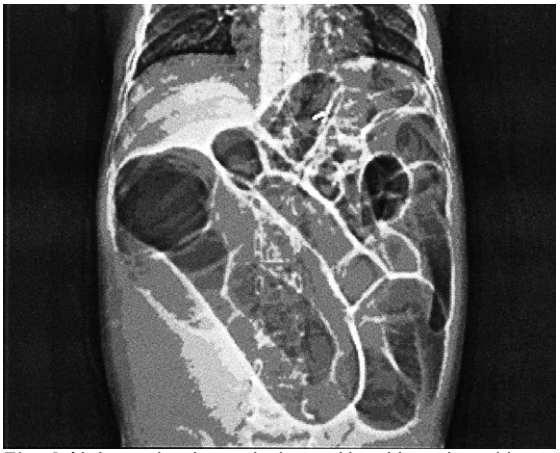

Fig. 1 Abdominal radiograph shows dilated large bowel loops with no gas within the rectum.

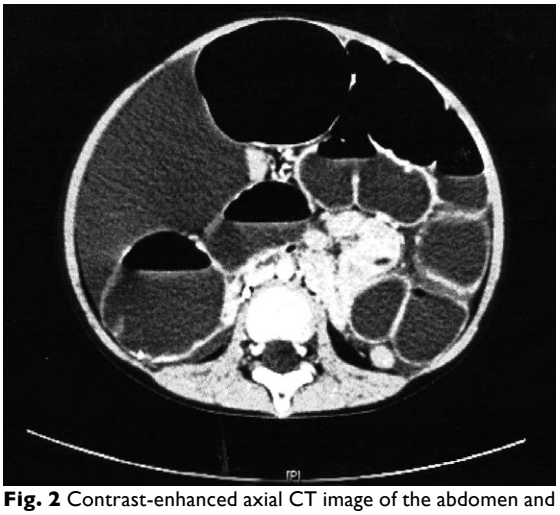

Fig. 2 Contrast-enhanced axial CT image of the abdomen and pelvis shows markedly-dilated small and large bowel loops with air-fluid levels. A uniform density of fluid collection was seen on the right side of the abdomen as well as inferiorly in the lower abdomen and upper pelvis.

Air-fluid levels were noted, mainly within the large bowel (Fig. 1). With conservative measures, the distension of abdomen settled, to reveal a non-uniform central abdominal mass. Contrast-enhanced axial computed tomography (CT) of the abdomen and

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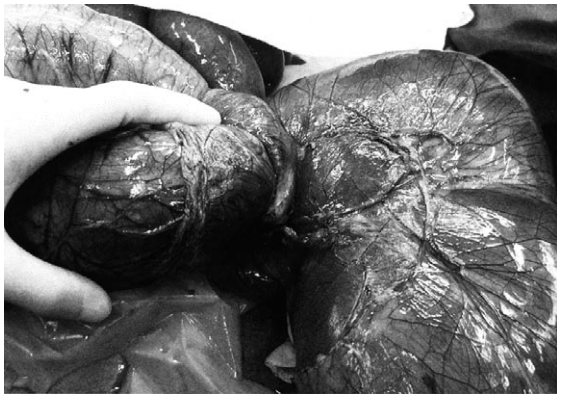

Fig. 3 Operative photograph shows a 15 cm × 20 cm × 7 cm mesenteric cyst arising from the mid-transverse colon. There was a 270° clockwise torsion of the cyst, leading to a constrictive narrowing of the adjacent large bowel loop.

pelvis showed markedly dilated small and large bowel loops with air-fluid levels. The transition point appeared to be in the region of the splenic flexure. A fluid collection of uniform density was seen on the right side of the abdomen, as well as inferiorly in the lower abdomen and upper pelvis (Fig. 2). Exploratory laparotomy revealed a 15 cm × 20 cm × 7 cm mesenteric cyst arising from the mid-transverse colon. There was a 270° clockwise torsion of the cyst, leading to a constrictive narrowing of the adjacent large bowel loop (Fig. 3). Fortunately, the bowel was healthy and the cyst was excised completely. Postoperative recovery was uneventful. Histology was consistent with cystic lymphangioma.

DISCUSSION

Cystic lymphangiomas are benign congenital masses that occur most frequently in the head and neck or axilla of young children, although rare cases have been detected in adulthood at various anatomical sites.[1-4] Intra-abdominal cystic hygromas are rare. The incidence is approximately one per 20,000 admissions to paediatric hospitals.[5,6] Nearly one-third of mesenteric cysts occur in children younger than 15 years of age, and one-fourth occur in patients younger than ten years of age.[7,8] The age range is from birth to 18 years (mean 4.9 years) and the cysts occur more frequently in males (55%).[4,7,8-10] Clinical presentation is diverse and can range from an incidentally-discovered abdominal mass to symptoms of an acute abdomen.[9,11,12] It may be diagnosed antenatally on prenatal ultrasonography.[13] Symptoms depend on the size, location, and complications, such as torsion, haemorrhage, infection, or rupture. The "classic presentation" is that of a low-grade, partial-intestinal obstruction combined with a palpable, freely-mobile abdominal mass.[4,7-9] The most common mode of acute presentation in children is a small bowel obstruction, sometimes associated with volvulus and intestinal infarction.[8,10] On physical examination, the majority of children with mesenteric and omental cysts have abdominal distension, with or without a palpable mass.[4,7,9,10] A definite mass may be difficult to palpate because of its large size, soft and fluid consistency, and great mobility. The mass can be huge, filling the abdominal cavity and simulating ascites.

Modern imaging studies can usually establish the diagnosis of a mesenteric-omental cyst or at least raise a strong suspicion. Abdominal radiograph shows a gasless, homogeneous, water-dense mass that displaces bowel loops around it.[14] Ultrasonography (US) and CT are very sensitive and relatively specific for evaluation of a cystic abdominal mass. On US, cystic lymphangioma appears as a sharply-defined cystic or multicystic mass, often with internal septations.[14] The fluid may be anechoic with enhanced through-transmission or have scattered internal echoes.[14] US is also useful in the follow-up of those patients. CT may add important preoperative information regarding size, anatomical location, adjacent organ involvement, and complications.[14] The typical CT appearance is of a large, thin-walled multiseptated cystic mass. The fluid contents are usually homogeneous, and attenuation values may range between those of fat and those of fluid.[14] Complete surgical excision is the treatment of choice.[11] The prognosis is excellent if the resection is complete.[12] US is the modality of choice for follow-up, especially in children.

In summary, abdominal cystic lymphangiomas have a varied clinical spectrum. In children, they usually present with a wide range of non-specific symptoms and clinical signs. General awareness of this entity with a high index of suspicion is needed to avert complications. CT and US are very sensitive in making the diagnosis. Not surprisingly, the radiologist may be the first to suggest the correct diagnosis.

REFERENCES

1. Bill AH Jr, Sumner DS. A unified concept of lymphangioma and cystic hygroma. Surg Gynecol Obstet 1965; 120:79-86.
2. Alqahtani A, Nguyen LT, Flageole H, Shaw K, Laberge JM. 25 years' experience with lymphangiomas in children. J Pediatr Surg 1999; 34:1164-8.
3. Weiss SW, Goldblum JR. Tumors of lymph vessels. In: Weiss SW, Goldblum JR. Enzinger and Weiss's Soft Tissue Tumors. 4th ed. St Louis: Mosby, 2001: 955-81.
4. Galonje E, Fletcher CDM. Tumors of blood vessels and lymphatics. In: Fletcher CDM, ed. Diagnostic Histopathology of Tumors. 2nd ed. London: Churchill Livingstone, 2000: 45-86.
5. Chung MA, Brandt ML, St-Vil D, Yazbeck S. Mesenteric cysts in children. J Pediatr Surg 1991; 26:1306-8.

6. Vanek VW, Phillips AK. Retroperitoneal, mesenteric, and omental cysts. Arch Surg 1984;119:838-42.
7. Bliss DP Jr, Coffin CM, Bower RJ, Stockmann PT, Ternberg JL. Mesenteric cysts in children. Surgery 1994; 115:571-7.
8. Mollitt DL, Ballantine TVN, Grosfeld JL. Mesenteric cysts in infancy and childhood. Surg Gynecol Obstet 1978; 147:182-4.
9. Hebra A, Brown MF, McGeehin KM, Ross AJ 3rd. Mesenteric, omental, and retroperitoneal cysts in children: a clinical study of 22 cases. South Med J 1993; 86:173-6.
10. Kosir MA, Sonnino RE, Gauderer MWL. Pediatric abdominal lymphangiomas: a plea for early recognition. J Pediatr Surg 1991; 26:1309-13.
11. Steyaert H, Guitard J, Moscovici J, et al. Abdominal cystic lymphangioma in children: benign lesions that can have a proliferative course. J Pediatr Surg 1996; 31:677-80.
12. Freud E, Farkash U, Cassella R, Freire C, Zer M. Childhood retroperitoneal lymphangioma presenting following minor trauma. Injury 1999 Jun; 30:380-3.
13. Mentzel HJ, Schramm D, Vogt S, et al. Intra-abdominal lymphangioma in a newborn. J Clin Ultrasound 1998; 26:320-2.
14. Cutillo DP, Swayne LC, Cucco J, Dougan H. CT and MR imaging in cystic abdominal lymphangiomatosis. J Comput Assist Tomogr 1989; 13:534-6.