Case report

Mesenteric lymphangiomatosis in children: a distinct clinico-pathological entity

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Abstract: Cystic lymphangiomas occurring within the abdomen are usually classified together with mesenteric cysts. However, they differ by location, histology, and potential for recurrence and should be considered as a separate clinical entity. The present article describes two cases of extensive mesenteric lymphangiomatosis involving variable lengths of small and large bowel and presenting as an acute abdomen. Ultrasonography and computed tomography proved the most reliable imaging modalities. In the first case the lesion could not be completely resected; whereas in the second resection and anastomosis without any residua was possible. Life-threatening complications can arise from these lesions, and excision is facilitated by earlier diagnosis.

Key words: Lymphangiomatosis – Paediatric – Abdominal

Introduction

Intra-abdominal lymphangiomas are uncommon cystic and cavernous lesions that may present clinically in a confusing manner. Reports of isolated cases and collected series have described these cysts as presenting unexpectedly during abdominal surgery and at post-mortem examination [1, 2]. Histological and ultrastructural evidence confirms that these lesions are pathologically distinct [12, 18]. Occasionally, due to their size or position, lymphangiomas may result in symptoms and clinical findings that necessitate surgical intervention. Mesenteric cysts may be relatively symptom-free; lymphangiomas frequently present with acute abdominal findings. In instances of acute presentation, the correct diagnosis is rarely made preoperatively. The aim of this paper is to review our experience with paediatric abdominal lymphangiomatosis and illustrate its bizarre clinical presentation and management options. Two clinical summaries are documented as examples.

Case reports

Case 1. A 6-year-old girl was admitted with complaints of anorexia; abdominal distension, and vomiting. The abdominal distension was evident for 6–8 months prior to admission. Physical examination revealed a malnourished, irritable child with generalised distension of the abdomen, which was firm on palpation. A distinct mass was not felt. The rectal examination was noncontributory. A haemogram, serum electrolytes, liver function tests, amylase, and urinary vanilmandelic acid assays were within normal limits. Ultrasound (US) and computed tomography (CT) of the abdomen demonstrated a diffuse, solid mass with multiple cystic areas of varying sizes occupying the epigastric, umbilical, and right iliac regions. The lesion appeared to be separate from the liver, gall bladder, and spleen but distinction from the jejuno-ileal loops was not possible (Fig. 1).

At laparotomy a diffuse, cystic mesenteric lesion was seen extending from the duodeno-jejunal flexure to the ileo-caecal junction with diffuse oedema of the mesentery. The mass encroached upon the small bowel (Fig. 2), contained chylous fluid, and was firmly adherent to the base of the small-bowel mesentry involving the superior mesenteric artery, superior mesenteric vein, duodenum, and pancreatic surface. Complete excision of the lesion was thus not possible, and internal marsupialisation of the cysts within the peritoneal cavity was done.

Cytological examination of the fluid revealed only mesothelial cells, and histological studies confirmed the diagnosis of lymphangiomatosis with multiloculated areas lined by endothelial cells and variable amounts of lymphatic tissue in the wall of dilated lymphatic channels and cysts. The child had done well with no signs of recurrence after 16 months of follow-up.

Case 2. An 11-year-old girl presented with a 3-year history of intermittent abdominal colics, anorexia, and weight loss. Four months prior to admission she had developed more frequent episodes of abdominal pain, nausea, and vomiting. On physical examination the temperature was 37°C; pulse 110/min; respirations 26/min; and blood pressure 120/80 mmHg. Abdominal examination showed a firm, nontender, mobile mass in the hypogastric, umbilical, and right iliac regions. A haemogram, serum electrolytes, urea, liver function tests, and amylase were within normal limits. US and CT scans revealed a 20×15×10-cm,
Fig. 1. CT of abdomen (case 1) showing diffuse, solid mass with multiple cystic areas indistinguishable from jejuno-ileal loops

Fig. 2. Diffuse cystic mass from mesentery of small bowel encroaching upon intestinal loops (case 1)

Fig. 3. CT of abdomen (case 2) showing multi-loculated, fluid-filled, extensive mass separate from liver, spleen, and kidneys

Fig. 4. Case 2: cystic lesion restricted to mesentery from mid-jejunum to mid-ileum

Mesenteric cysts and cystic lymphangiomas are infrequently encountered intra-abdominal lesions that may be confused on gross appearance but are distinguishable by their histological and ultrastructural features [5, 16, 19, 20]. Mesenteric cysts were first described in 1507 by Benevieni [3] and later by Tillaux [27]. Only 5% to 10% of all intra-abdominal cysts microscopically prove to be cavernous or cystic lymphangiomas [10]. Apart from being larger, cystic lymphangiomas are more frequently multilocular, diffuse, and may require resection of adjacent organs to accomplish complete excision.

A variety of factors have been implicated in the formation of these cystic lesions. Gross considered them to represent malformations secondary to proliferating lymphatic tissue without access to drainage [14]. An early age of presentation, in the 1st decade of life, has been taken as evidence of a congenital etiology [22, 25]. Godart proposed that lymphatic spaces in the embryo fail to join the venous channels [13], while Elliot et al. blamed congenital