

Case report

Huge Mesenteric Cystic Lymphangioma Presenting With Small Bowel Volvulus in a 3-Year-Old Child

ABSTRACT

Mesenteric cystic lymphangioma is a rare abdominal mass. Small bowel volvulus caused by mesenteric lymphangioma is exceptional but potentially fatal. We report a case of a 3-year-old boy who complained of recurrent abdominal pain for several months and was admitted to the emergency department for acute abdominal pain with vomiting and bowel obstruction over 24 hours. Clinical examination objectified a peri-umbilical pain. Radiological investigations objectified a small bowel volvulus with individualization of a cystic lesion. Surgical exploration revealed a huge cystic lymphangioma of the mesentery, with a small bowel volvulus over completed common mesentery. The cystic lymphangioma was excised with the 2 supporting bowels, and an ileo-ileal anastomosis was performed. Histological findings confirmed the diagnosis. The child remained asymptomatic at the 6-month follow-up; no recurrence had been noted.

Keywords: small bowel volvulus; mesenteric lymphangioma; abdominal pain; surgical resection

1. INTRODUCTION

Mesenteric cystic lymphangioma is an uncommon benign intraabdominal malformation with unknown origin that mostly affects children, with an incidence of 1/20,000 [1,2]. It's often asymptomatic but can cause acute symptoms due to complications like volvulus, bleeding, or rupture [3,4]. The diagnosis is made radiologically and confirmed on the basis of histological findings.

Here, we present a case of a child admitted for an acute abdominal presentation. Radiological and surgical exploration revealed a huge cystic lymphangioma of the mesentery with a small bowel volvulus.

2. PRESENTATION OF CASE

A 3-year-old child with a history of intermittent abdominal pain 4 months prior to admission was admitted to emergency for acute abdominal pain with vomiting and bowel obstruction over 24 hours. Clinical examination found an afebrile patient with peri-umbilical tenderness without palpation of any obvious mass. Laboratory tests came back normal. The abdominal X-ray showed diffuse greying, with no air-fluid levels or pneumoperitoneum (Figure 1).



Fig. 1. Abdominal X-ray showing a diffuse greying.

Abdominal ultrasound revealed a large, multilocular mesenteric cystic structure containing fine septa and fluid cavities with echogenic content (Figure 2).



Fig. 2. Abdominal ultrasound revealing a large, multilocular mesenteric cystic structure containing fine septa and fluid cavities with echogenic content.

The abdominal CT scan showed a volvulus of the small bowel (whirlpool sign) associated with occlusion of the superior mesenteric artery, and a cystic lesion formation occupying the entire peritoneal cavity, suggesting a cystic mesenteric lymphangioma (Figure 3).

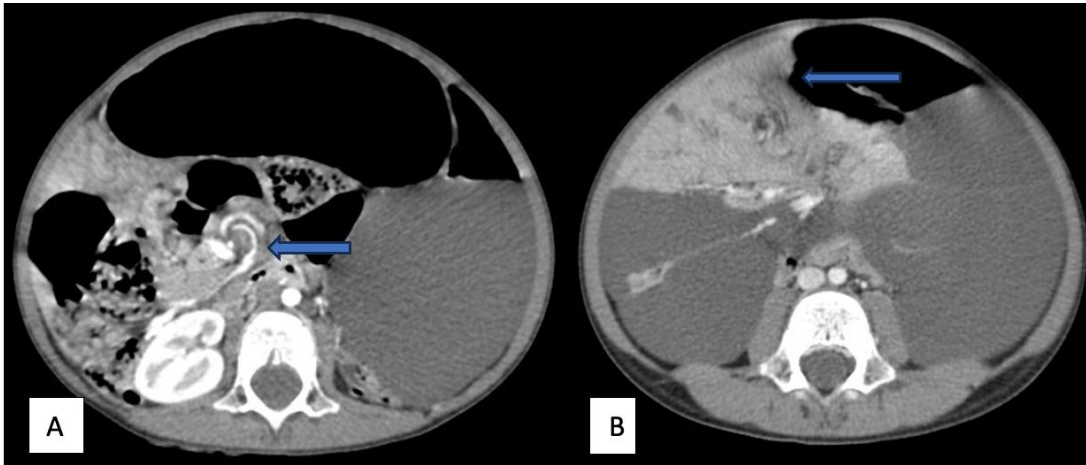


Fig. 3. CT scan showing: (A) whirlpool sign and (B) cystic lesion formation.

Through a supra-umbilical laparotomy, surgical exploration revealed a giant cystic mesenteric lymphangioma measuring 17 x 10 cm attached with the last 2 bowel loops with a volvulus on complete common mesentery. Dilated mesenteric veins were found as a sign of chronic intermittent volvulus (Figure 4).

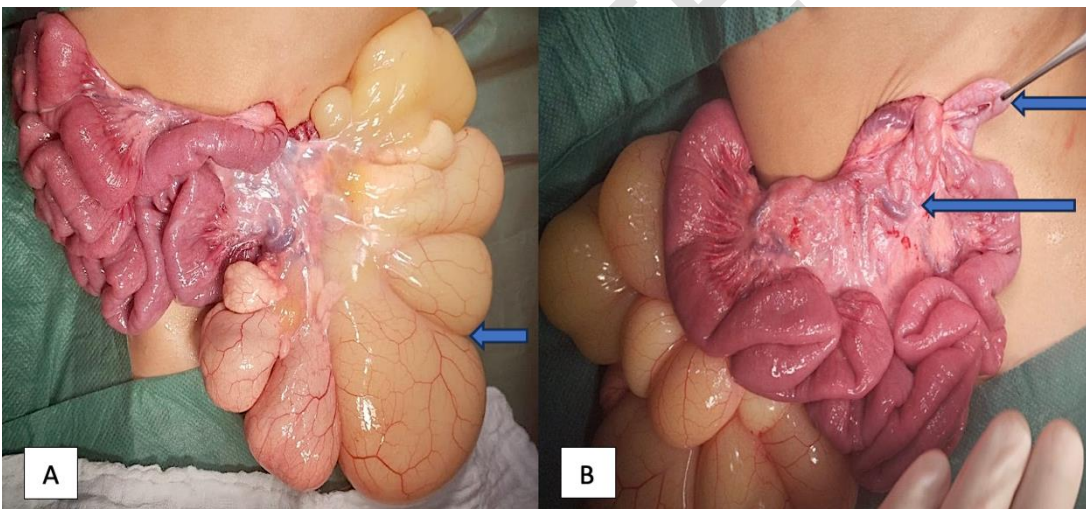


Fig. 4. Operative images revealing: (A) giant cystic mesenteric lymphangioma, (B) an appendix on the left and dilated mesenteric veins.

There were no signs of bowel suffering. The cystic lymphangioma was excised with the 2 holding bowels, and an ileo-ileal anastomosis was performed. Then we returned the bowels as an incomplete common mesentery; appendicectomy was not performed. Immediate postoperative follow-up was straightforward; the patient was discharged after 3 days.

Histological findings confirmed the diagnosis of mesenteric cystic lymphangioma. Lymphatic endothelium was positive for D2-40 and CD31 expression (Figure 5).

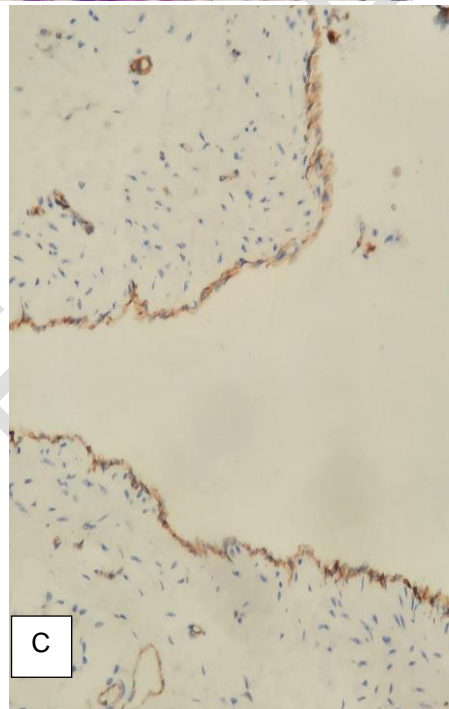
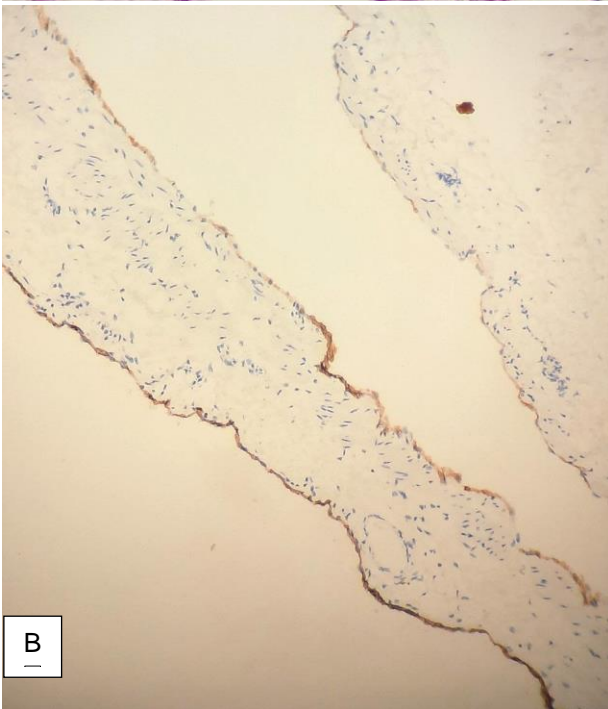
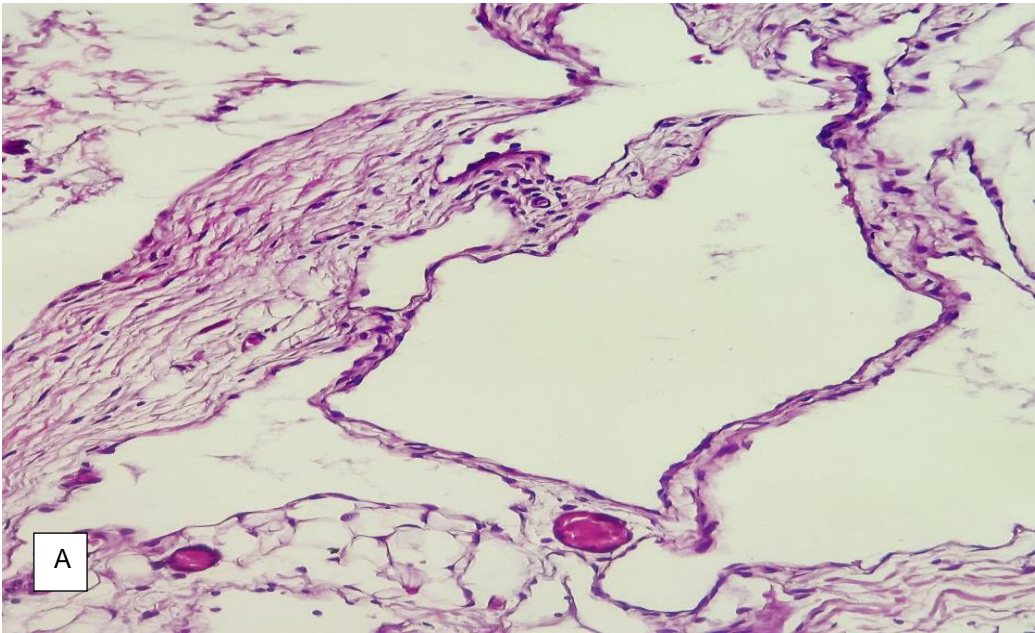


Fig. 5. Histological sections: (A) showing cavities lined by flattened endothelial cells H&Sx200, these cells express D2-40 (B) and CD31 (C).

At 6-month follow-up, the child remained asymptomatic; no recurrence had been noted.

3. DISCUSSION

Lymphangioma is a rare benign condition characterized by a proliferation of lymphatic spaces. Mesenteric cystic lymphangioma is rare, accounting for less than 1% of all lymphangiomas; its incidence is estimated at 1/20,000 in children [2,5]. According to Morris-Stiff et al., only 19 cases of small bowel lymphangioma were noted in the literature from 1960 to 2009 [6]. It can cause complications such as volvulus, requiring surgical intervention. Almost 90% are discovered at the age of two years [7]. The clinical presentation is variable, ranging from an asymptomatic presentation to an acute abdominal condition following a volvulus, hemorrhage, or rupture [3,8]. According to the studies [9], the majority of children complained of intermittent abdominal pain. In our case, the patient suffered from episodic abdominal pain and vomiting 4 months prior to admission, then he presented with acute abdominal pain. Radiological diagnosis remains

challenging. It helps to identify the characteristics of the mass, its size, and its location without confirming the diagnosis [4,10]. MRI is the most reliable imaging modality for differentiating between mesenteric cysts and lymphangiomas [8,10]. There are two opinions that explain the causal association between small bowel volvulus and lymphangioma. The mobile nature of the mesenteric lymphangioma may cause rotation, leading to small bowel volvulus. Another hypothesis suggests that long-term or intermittent volvulus might produce lymphatic congestion, leading to lymphatic cysts [8,11].

This pathology is treated surgically, although some authors suggest surveillance for asymptomatic patients [8,12]. Complete surgical removal must be done to prevent the risk of recurrence through laparotomy or laparoscopy; segmental bowel resection is generally performed when the cyst adheres intimately to the bowel [8,12]. In the case of an unresectable mass due to vessel invasion, some authors recommend intra-cystic sclerotherapy using doxycycline and bleomycin [2,3,13]. In our patient, a complete surgical resection of the mass was carried out with the 2 supporting bowel loops.

The diagnosis of lymphangioma is established by histopathological study [10]. The presence of an endothelial covering, connective tissue, and smooth muscle fibers allows lymphangiomas to be differentiated from mesenteric cysts, which are characterized by the presence of a cuboid or columnar covering with no smooth muscle [8]. The immunohistochemical study confirms the diagnosis by the presence of CD31, CD43, CD45, and D2-40 markers [2,4,8].

Regular follow-up is necessary due to the risk of recurrence [8,12]. The principal limitation of this case was the absence of a long-term follow-up.

4. CONCLUSION

Mesenteric cystic lymphangioma is an uncommon surgical condition mostly asymptomatic. It may sometimes produce life-threatening conditions such as volvulus, which may need urgent surgery. Complete mass resection is the best choice to reduce the risk of recurrence. However, additional studies are needed to clarify the pathogenesis of this type of tumor. Laparoscopy is becoming increasingly useful, enabling rapid recovery and avoiding complications.

CONSENT

All authors declare that 'written informed consent was obtained from the patient family for publication of this case report and accompanying images.

ETHICAL APPROVAL

All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

Disclaimer (Artificial intelligence)

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc.) and text-to-image generators have been used during the writing or editing of this manuscript.

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