Pediatric abdominal chylolymphatic cysts: Sneaky abdominal masqueraders

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ABSTRACT

Chylolymphatic mesenteric cysts of the abdomen are a rare differential for cystic tumors, more so in the pediatric age group, and they have various clinical presentations. There have been very few cases reported worldwide. To the best of our knowledge, a multicystic chylolymphatic cyst of the abdomen as demonstrated below is the first reported occurrence worldwide in the pediatric age group. We herein report the case of a 1-year-old who presented to us with gradual distension of the abdomen for 4 months with occasional episodes of vomiting and a lobulated cystic abdominal mass on palpation. He was mistakenly treated with anti-tuberculous treatment for suspected loculated ascites. The imaging revealed a multiloculated mass in the abdomen in close relation to the small bowel. He was also worked up for tuberculosis, and the findings were negative. Laparotomy revealed multiple cysts closely applied to the jejunum, starting at 4 cm from the duodenojejunal flexure, which were resected and anastomosed. To the best of our knowledge, this is the first reported case of an abdominal chylolymphatic cyst occurring as a chain of multiple cysts in the pediatric age group, and we want to emphasize the importance of further imaging in a case of suspected loculated tuberculous ascites as this is a differential diagnosis with a treatable surgical option. Surgical excision of these cysts should be done in toto as an individual excision of each of these cysts in an attempt to preserve the bowel length may end up jeopardizing the blood supply of the affected bowel.

Keywords: Abdominal cysts, atypical presentation, chylous cyst, mesenteric cyst

INTRODUCTION

Mesenteric cysts of the abdomen are exceedingly rare, more so in the pediatric population, with an incidence of 1:100,000 in adults and 1:20,000 in children (1). They are most commonly found in the small bowel as compared to the large bowel, the ileum being the relatively more common site (2). Chylolymphatic cysts of the mesentery are chyle-filled cysts characterized by the absence of smooth muscle and lymphatic spaces in the cyst wall (1, 3). The exact theory of origin of these cysts is not known, but a variety of possible explanations have been proposed, the most widely accepted being Gross’ theory of benign proliferation of ectopic mesenteric lymphatics that have been excommunicated from the main lymphatic channels (4). They commonly present as solitary cysts or multiloculated cysts. Multiple lymphatic cysts of the small bowel arranged in a chain tightly attached to a segment of the small bowel is rare as described in the case below.

CASE PRESENTATION

Informed consent has been obtained from the patient’s guardian before the submission of this article.

A 1-year-old male child presented to us with gradually progressing abdominal distension for the past 4–5 months with occasional episodes of vomiting that was non-bilious. There was a history of occasional constipation. On evaluation at his hometown, he was treated with anti-tuberculous treatment for a mistaken diagnosis of abdominal tuberculosis as his ultrasound revealed loculated ascites. After the medication, his mother did report some decrease in size. A clinical examination revealed a large mass 10X12cm in size, which was lobulated, variegated in consistency (cystic in some areas, firm in others), and partly mobile (Figure 1).

Ultrasound imaging of the abdomen revealed a well-defined large, heterogenous, predominantly multilocystic lesion 9.4 cm x 12.2 cm, extending from the epigastrium up to the urinary bladder with no significant interval vascularity. On computed tomography (CT) (Figure 2), the same lesion showed multiple enhancing septae within the lesion with the bowel loops enmeshed inside. The patient’s blood findings were within the normal limits.

Intraoperatively, the patient was found to have multiple cysts of varying sizes and colors, some hemorrhagic, arranged in a chain along the mesentery of the duodenum commencing at 4 cm from the duodenojejunal flexure and extending up to 20 cm (Figure 3). An attempt was made to individually excise the cysts; however, that led to rupture of the cysts leading to leakage of a milky chylous fluid,
and proceeding would have rendered the mesenteric vascular supply precariously compromised. A resection was hence performed on the affected segment with the multiple cysts and an end-to-end double layer bowel closure, with closure of the mesenteric defect, was performed after confirming a good vascular supply to the bowel. The entire specimen was sent in toto for a histopathological examination, which revealed that the wall of the excised cyst was composed of a single layer of flattened cells with compressed connective tissue, consistent with the chylolymphatic cyst.

His postoperative period was uneventful.

DISCUSSION

Mesenteric cysts were first reported in the 16th century (2). They are rare with an incidence of 1:100,000 in adults and 1:20,000 in children (3). Chylolymphatic cysts comprise 7.3% of all mesenteric cysts and are characterized by the absence of the smooth muscle and lymphatic spaces in the cyst wall (3, 4, 5). They are most commonly found in the small bowel as compared to the large bowel, the ileum being the relatively more common site (6). The exact theory of origin of these cysts is not known, but a variety of possible explanations has been proposed; the most widely accepted being Gross’ theory of benign proliferation of ectopic mesenteric lymphatics that have been excommunicated from main lymphatic channels (2). The usual occurrence is that of a solitary or multicystic lesion closely applied to the bowel wall that to the best of our knowledge, has never been reported before. A similar case has been described in an adult aged 35 years, published in 1992 (7).

Clinically, the patient may be asymptomatic or present with an abdominal mass, acute abdomen due to rupture, infection, hemorrhage into a cyst, or volvulus or intestinal obstruction, or other atypical presentations. Especially in endemic areas like ours, the presence of large chylolymphatic cysts can be mistaken for the more common loculated tuberculous ascites (8). Although tuberculosis would be a more common differential diagnosis to consider, emphasis has to be made that the diagnosis of these multiple chylolymphatic cysts, albeit rare, has to be kept in mind while dealing with patients who present with multiple cystic lesions in the abdomen. A CT is a useful adjunct in the evaluation of patients with abdominal distension.

Differential diagnoses considered in this case include mesenteric lymphangioma and multilocular peritoneal inclusion cysts (MPIC). These are distinct histologically; the lack of the smooth muscle and lymphatic spaces in the cyst wall seen in chylolymphatic cysts will be demonstrated in a lymphangioma (3). The histopathology of MPIC shows mesothelial cells (4).

Current management guidelines recommend complete resection of the cyst either by enucleation, or if the blood supply to the bowel has to be sacrificed, resection and anastomosis of the closely applied bowel loop should be performed as in our case.

CONCLUSION

Chylolymphatic cysts should be borne in mind as one of the differential diagnoses of the cystic masses in the abdomen in the pediatric age group. Multiple chylolymphatic cysts are rarely found, but their clustered appearance with a negative work up for tuberculosis should warrant further imaging and a reconsideration of management options. In an endemic coun-
try for tuberculosis such as India, this differential should also be borne in mind. An adequate surgical treatment includes a complete excision with resection and anastomosis of the affected bowel.

Take-home messages:
1- Chylolymphatic cysts are a rare cause of abdominal distension in children.
2- CT is a useful adjunct in the evaluation of abdominal distension of unknown etiology (especially if the diagnosis is questionable).
3- En bloc bowel resection with the cyst may be required.

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REFERENCES