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Case Report

Paediatric Mesenteric Chylolymphatic Cyst - Case Discussion and Literature Review

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Abstract

Chylolymphatic cysts are a rare variant of mesenteric cysts. Although very uncommon in both adults and children, they can present with life-threatening complications like intestinal obstruction, volvulus, rupture etc. Hence awareness of existence of these lesions is vital for proper diagnosis and management. There are now well-established histopathological features that differentiate chylolymphatic mesenteric cysts from other cystic abdominal lesions. Complete excision of the cyst ensures excellent prognosis and is curative. We present an 18-month old child with an ileal chylolymphatic cyst who underwent complete surgical resection of the cyst along with the adjacent ileum. Relevant literature is reviewed and discussed which emphasizes the need for awareness, early detection and surgical management of such rare entities to avoid ominous complications.

Keywords: *Chylolymphatic Cyst; Mesenteric Cyst; Pediatric; Infant*

Introduction

Mesenteric and omental cysts are rare; their incidence is about 1 per 140,000 general hospital admissions and about 1 per 20,000 paediatric hospital admissions [1]. Mesenteric cysts can occur anywhere in the mesentery of the gastrointestinal tract from the duodenum to the rectum, and they may extend from the base of the mesentery into the retroperitoneum [2]. A chylolymphatic cyst is a rare variant of a mesenteric cyst and chylolymphatic cysts in the pediatric age group are extremely uncommon [3].

We present an 18-month old child with an ileal chylolymphatic cyst who underwent complete surgical resection of the cyst along with the adjacent ileum. Relevant literature is reviewed and discussed which emphasizes the need for awareness, early detection and surgical management of such rare entities to avoid ominous complications.

Case Report

An 18-month-old male child presented with a vaguely palpable abdominal mass. No other associated symptoms like pain, vomiting, constipation etc were present. Child had no significant antenatal history and was delivered by normal vaginal delivery at 38-weeks-gestation with a birth-weight of 2650 grams. At 12-months-age, the infant was treated by a paediatrician for pneumonia, during the evaluation of which, a screening ultrasonography (USG) of the abdomen revealed an intra-abdominal cystic lesion. Child was sent to our paediatric surgery department for further management.

Clinical examination showed a well-nourished active male child weighing 12 kilograms. Abdominal examination revealed fullness of the right lower abdomen with an irregular soft mobile lesion of approximately 8 cm x 8 cm palpable in the hypogastrium. There were no other positive signs. Hematological and biochemical parameters were normal. Detailed USG abdomen showed an 8.3 cm x 4.1 cm x 2.8 cm multi-septated ill-defined cystic lesion in the hypogastrium. Contrast CT scan abdomen (Figure 1) showed a homogenous fluid-filled peripherally enhancing cystic lesion insinuating between the bowel loops and closely abutting the superior surface of the bladder. Multiple vessels were seen coursing through the lesion. There was no evidence of any calcification or haemorrhagic foci with an overall radiological impression of a mesenteric lymphangioma.

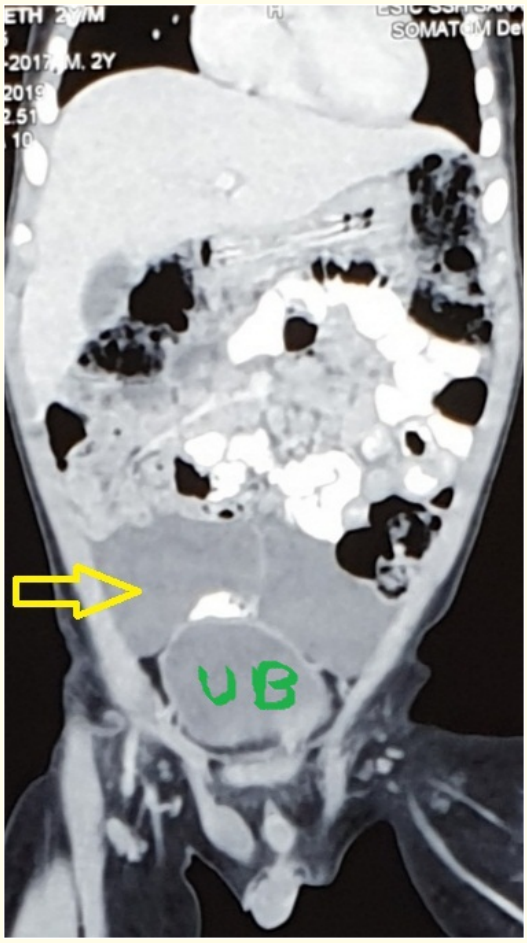

Figure 1: *Contrast CT of abdomen showing a fluid-filled septate cystic lesion (yellow arrow) between the bowel loops and abutting the urinary bladder (UB) with no evidence of calcification or haemorrhagic foci.*

Child was posted for laparotomy under General Anaesthesia. Pfannenstiel incision was given which revealed an 8 cm x 8 cm milky white cystic mass in the small bowel mesentery approximately 40 cm from the duodenojejunal flexure suggestive of a chylolymphatic cyst (Figure 2 and 3). As the lesion was intertwined in the mesentery, we had to perform a resection of about 10 cm of the ileum along with the lesion and bowel continuity was restored by performing an ileo-ileal anastomosis in two layers. Child had an uneventful postoperative recovery and is well at 8 months follow-up.

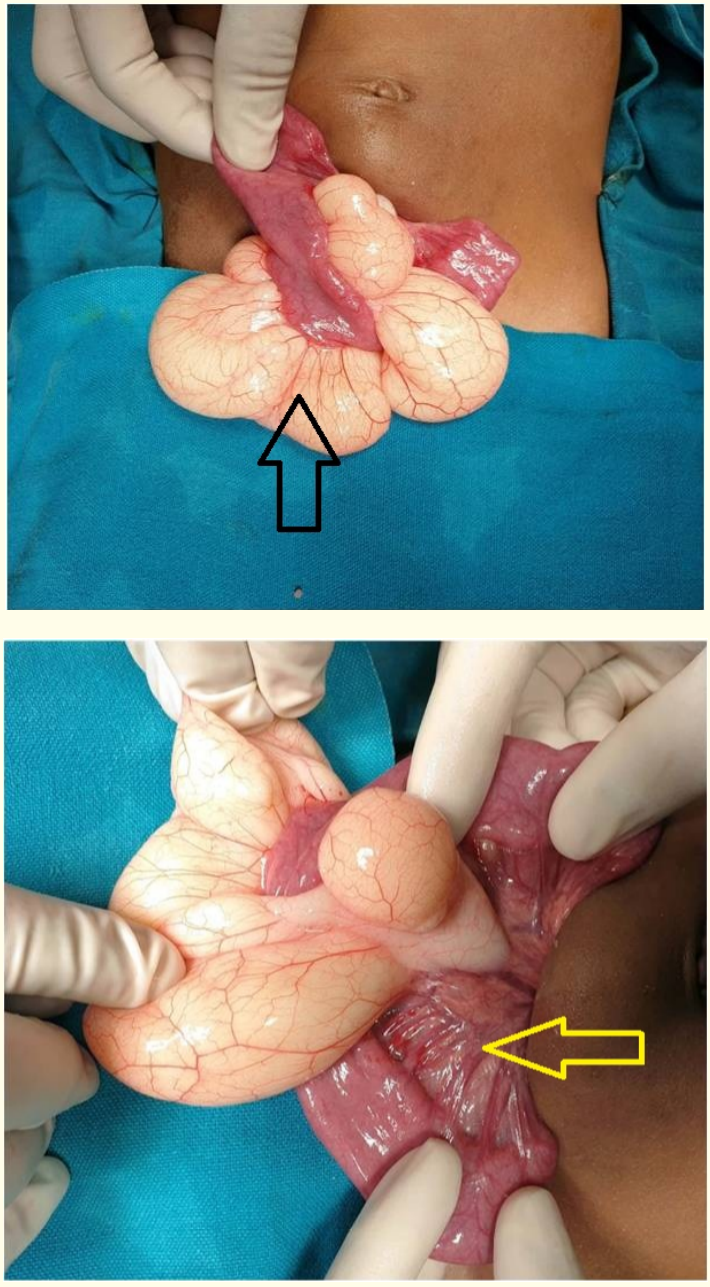

Figure 2: *Approximately 8 cm x 8 cm milky white cystic mass (black arrow) in the ileal mesentery approximately 40 cm from the duodenojejunal flexure and intertwined in the mesenteric blood vessels (yellow arrow).*

Grossing of the specimen showed a milky white chylous fluid in the multiloculated cystic lesion. Histopathological examination showed normal ileal mucosa with congested blood vessels in the sub-mucosa. There was a cyst arising from its serosal surface composed of compressed connective tissue with a lining of single layer of flattened epithelial cells (Figure 4) suggestive of a chylolymphatic cyst.

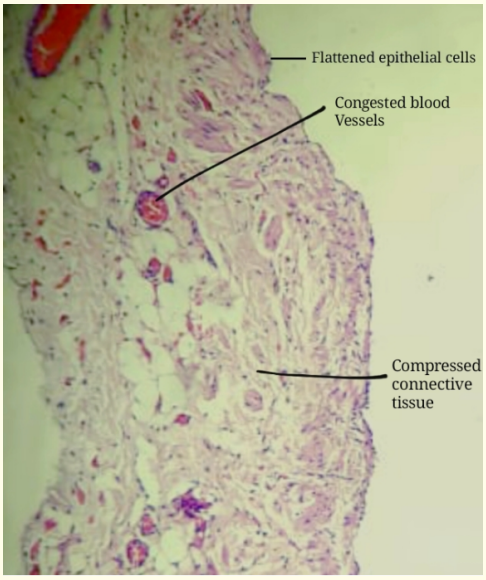

Figure 4: *Microphotograph showing cyst wall composed of compressed connective tissue with a lining of single layer of flattened epithelial cells. (Hematoxylin-eosin, 40X).*

Discussion

Mesenteric cyst was first described in 1507 by Florentine anatomist Benevieni, while performing an autopsy on an 8-year-old boy. In 1842 Rokitansky published the first accurate description of a chylous cyst [1]. Mesenteric cysts most commonly occur in the ileal mesentery of the small bowel or the sigmoid mesentery of the colon [4].

Beahrs., *et al.* classified mesenteric cysts into four groups based on etiology: embryonic or developmental; traumatic or acquired; neoplastic; and infective or degenerative while Losanoff., *et al.* proposed a classification system wherein types 1 (pedicled) and 2 (sessile) are limited to the mesentery and can be excised completely with or without resection of the involved gut. Types 3 and 4 (multicentric) extend into the retroperitoneum and require complex operations and often sclerotherapy as well [5,6].

Based on the contents of the cyst, the mesenteric cyst can be divided into serous, chylous, hemorrhagic and chylolymphatic cyst [1]. Among these, chylolymphatic cysts are extremely rare. Engel et al reported four cases of cystic lymphangioma in the pediatric age group and only two of them were of the chylous variety [7]. Singh., *et al.* reported five cases of cystic lymphangioma in the pediatric age group though only one of them was diagnosed to be a chylolymphatic cyst on histopathology [8].

The chylolymphatic cyst, as indicated by its name, contains both chyle and lymph. These cysts present within the mesentery, lined with a thin endothelium or mesothelium and filled with chylous and lymphatic fluid. These cysts are thought to arise in sequestered lymphatic channels or ectopic lymphatic tissue in the small bowel mesentery and enlarge by accumulating both lymph and chyle. The accumulation of chyle and lymph is considered to be the result of an imbalance between the inflow and outflow of fluid [7].

Cystic lymphangioma has a striking resemblance to chylolymphatic mesenteric cysts both grossly and microscopically. Some authors consider chylolymphatic mesenteric cysts to be a type of cystic lymphangioma, while some authors have described chylolymphatic cysts as a variant of mesenteric cysts [3,6,9]. There are now well-established histopathological features that differentiate chylolymphatic mesenteric cysts from other cystic lesions including cystic lymphangioma. The absence of smooth muscle and lymphatic spaces in the wall of the cyst differentiates mesenteric cysts from cystic lymphangioma [9].

The cyst may be asymptomatic or may cause abdominal distension with an abdominal lump or may present with ominous complications such as intestinal obstruction, hemorrhage, infection, cyst rupture, volvulus or obstruction of the urinary or biliary tract [3,7]. In a series of 8 patients, Rattan., *et al.* reported that 4 presented with an abdominal lump and 2 each with abdominal pain and acute intestinal obstruction [3].

The definite diagnosis of these lesions is difficult prior to surgical exploration as there are no pathognomonic symptoms or characteristic imaging findings. Abdominal radiographs are usually non-contributory, however may reveal dilated bowel loops with air-fluid levels in the very rare patients with intestinal obstruction which may result from volvulus or compression of the adjacent bowel by the cyst [10]. The diagnosis may be suggested by an ultrasound of the abdomen, which may reveal a cystic lesion in relation to the bowel loops. A fluid-fluid level has been reported as a characteristic finding of these cysts which results from an upper fluid level due to the chyle, and a lower fluid level due to the heavier lymph [11]. CT scan demonstrates the fluid attenuation of the lesion and its relationship with the adjacent viscera. A characteristic chyle-lymph fluid level has also been described [12]. However, in our patient, the USG and CT scans were able to detect a cystic lesion in the lower abdomen but a definitive preoperative diagnosis of chylolymphatic cyst could not be made.

There is no role of medical management in mesenteric cysts. In children with mesenteric or omental cysts, the most common indication for surgical intervention is the presence of an abdominal mass with or without signs of intestinal obstruction. Management of these cysts involves their removal which may or may not involve resection of the adjacent bowel. While some cysts can be enucleated, in some this is not possible without sacrifice of the blood supply to the adjacent bowel and hence necessitates bowel resection as was the case in our patient. Procedures like marsupialization and drainage are associated with high recurrence rates and are best avoided [4].

Conclusion

Cysts of the mesentery are among surgical rarities of which chylolymphatic cysts are rarer. The clinical presentation is not characteristic and preoperative imaging although suggestive is not diagnostic. In most cases, the diagnosis is confirmed after surgical exploration and removal of the cyst. Resection of the involved gut is frequently required in children and one should not unduly prolong the surgery to avoid gut resection. Complete excision of the cyst ensures excellent prognosis and is curative. Awareness, early detection and surgical management of such rare entities helps avoid grave complications like intestinal obstruction, volvulus, haemorrhage and rupture.

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