Giant Chylolymphatic Mesenteric Cysts in Children: A Preoperative Enigma

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Abstract

Although mesenteric cysts in general have been reported in the literature fairly frequently, chylolymphatic cysts in the pediatric age group are extremely rare in the modern medical literature. Ultrasonography and computed tomography suggest the diagnosis but the picture is not clear until demonstrated intraoperatively. Intra-operatively, similar findings can be seen in cystic lymphangioma, retroperitoneal cystic teratoma, caseating tubercular lymph nodes, and hydatid cysts. Even lymphoma and duplication cysts may also give similar appearances. Excision biopsy is then recommended to differentiate these cases. Histopathology is confirmatory and differentiates chylolymphatic cysts from all these lesions. We present our experience with two such cases that presented to our center.

Keywords: Chylolymphatic; Mesenteric cyst; Ileum; Abdominal pain

Introduction

A chylolymphatic cyst is a rare variant of a mesenteric cyst [1,2]. These cysts present within the mesentery, lined with a thin endothelium or mesothelium and filled with chylous and lymphatic fluid [3]. Although mesenteric cysts in general have been reported in the literature fairly frequently, chylolymphatic cysts in the pediatric age group are extremely rare in the modern medical literature [2], therefore very little information is available regarding their presentation which can vary from asymptomatic to an acute abdomen with intestinal obstruction. Ultrasonography and computed tomography can provide an idea regarding the diagnosis but the intraoperative findings are often quite different. Histopathological examination of the specimen is required for confirmation. Complete excision of the cyst yields excellent result. In an attempt to reinforce the diagnostic and treatment strategy, we present our experience with two such cases that presented to our center within a short span of two weeks.

Case Presentation

Case 1

A 4 years old female child presented with abdominal pain of 6 months duration. The pain was dull, aching and continuous in nature. It was associated with mild abdominal distension noted for the last 1 month. The general physical examination was unremarkable. The abdomen was mildly distended but soft and suggestive of diffuse ascites rather than a discrete palpable mass. An ultrasonography (USG) and computed tomography scan (CT scan) of the abdomen revealed a large, septated, cystic mass with hypodense fluid attenuation. The biochemical parameters including serum amylase and lipase were within normal limits. With a preoperative suspicion of mesenteric cyst or pelvic cyst, the patient was posted electively for exploratory laparotomy but had to be taken up in the emergency because of a 10 days history of increasing abdominal pain and decreased oral intake. There was no history of vomiting or constipation. Upon surgical exploration, there was a huge cystic mass about 15cm by 18cm in dimension, filled with white milky fluid suggestive of a chylolymphatic cyst, occupying the mesentery of proximal ileum (Figure 1). The cyst was enucleated from the mesentery and excised completely along with 5cm of ileum closely related to it (Figure 2). An ileoileal end to end anastomosis was done. Feeds were started from 5 th postoperative day and the patient was discharged on 7 th postoperative day without any postoperative complications. The histopathological examination of resected specimen confirmed the diagnosis of chylolymphatic mesenteric cyst. At 6 months of follow up the patient is feeding adequately and doing well.

Case 2

A 5 years old male child presented with history of abdominal pain for the last 3 years. The pain was dull, aching and continuous in nature. It was associated with mild abdominal distension noted for the last 1 month. The general physical examination was unremarkable. The abdomen was mildly distended but soft and suggestive of diffuse ascites rather than a discrete palpable mass. An ultrasonography (USG) and computed tomography scan (CT scan) of the abdomen revealed a large, septated, cystic mass with hypodense fluid attenuation. The biochemical parameters including serum amylase and lipase were within normal limits. With a preoperative suspicion of mesenteric cyst or pelvic cyst, the patient was posted electively for exploratory laparotomy but had to be taken up in the emergency because of a 10 days history of increasing abdominal pain and decreased oral intake. There was no history of vomiting or constipation. Upon surgical exploration, there was a huge cystic mass about 15cm by 18cm in dimension, filled with white milky fluid suggestive of a chylolymphatic cyst, occupying the mesentery of proximal ileum (Figure 1). The cyst was enucleated from the mesentery and excised completely along with 5cm of ileum closely related to it (Figure 2). An ileoileal end to end anastomosis was done. Feeds were started from 5 th postoperative day and the patient was discharged on 7 th postoperative day without any postoperative complications. The histopathological examination of resected specimen confirmed the diagnosis of chylolymphatic mesenteric cyst. At 6 months of follow up the patient is feeding adequately and doing well.
was continuous, dull, aching but intermittently increased in intensity and the child needed analgesics for the relief. The pain increased in intensity and in frequency over the last 2 months. The patient was investigated with USG and CT scan of abdomen suggesting a huge intraperitoneal cystic mass and posted electively for exploration (Figure 3). This patient also underwent emergency exploratory laparotomy due to features of acute intestinal obstruction. Upon surgical exploration, there was a huge 10cm by 15cm cystic mass filled with white milky fluid in the mesentery of jejunum 35 cms from duodenojejunal flexure (Figure 4). Resection of about 4cm of jejunum with end to end anastomosis was needed in this case also with uneventful postoperative course. The patient was discharged by 7th postoperative day and is doing well after 6 months of follow up. The histopathology confirmed a chylolymphatic mesenteric cyst.

Discussion

Chylous cysts are rare variants of mesenteric lesions and constitute 7.3% to 9.5% of all abdominal cysts [1]. There are very few cases of pediatric chylolymphatic cysts reported in the literature. The clinical presentation is varied ranging from asymptomatic to palpable abdominal mass, diffuse abdominal fullness, abdominal pain and in extreme cases an acute abdomen with features of intestinal obstruction. These two cases presented here stand testimony to the fact that a mesenteric cyst with torsion, hemorrhage or intestinal obstruction can present as an acute abdomen and should be kept in mind as one of the differential diagnoses. Beahrs et al. [4] classified mesenteric cysts into four groups based on etiology: embryonic or developmental; traumatic or acquired; neoplastic; and infective or degenerative. Recently, a pathologic classification system has been proposed [5]. Type1 (pedicled) and Type2 (sessile) are limited to the mesentry and can be excised completely with or without resection of the involved gut. Type3 and Type4 (multicentric) extend into the retroperitoneum and require complex operations and often sclerotherapy as well. Based on the contents of the cyst, the mesenteric cyst can be divided into serous, chylous, hemorrhagic and chylolymphatic cyst. The chylolymphatic cyst, as indicated by its name, contains both chyle and lymph. The accumulation of chyle and lymph is considered to be the result of an imbalance between the inflow and outflow of fluid [1]. Radiological investigations form an integral part of the management of these lesions. A plain abdominal radiograph may show a gasless, homogenous mass defect displacing the bowel loops around it. In a child with an obstructed intestine, multiple air-fluid levels will be seen on an erect abdominal radiograph. Barium studies are now only of historical interest; abdominal ultrasonography is currently the imaging procedure of choice. This delineates the nature of the mass, organ or site of the origin, and the extent and associated mass effects on the kidney or liver, if any. Computed tomography adds little additional information; however, contrast-enhanced film can show the relationship of the bowel and other vital structures to the lesion. Some authors have described the characteristic appearance of a chylolymphatic cyst on computed tomography and

Figure 1: Intraoperative picture – Case 1- Huge chylolymphatic mesenteric cyst attached to mesentery of ileum.

Figure 2: Resected specimen – Case 1.

Figure 3: CT scan-Arrows pointing at huge hypodense fluid attenuation of chylolymphatic mesenteric cyst– Case 2.

Figure 4: Resected specimen – Case 2.
ultrasonography in the form of the presence of fluid levels of differing echodensities, that is to say, an upper fatty echodensity of chyle on top of the water echodensity of lymph in a well-defined cystic lesion [6,7]. The different surgical approaches used are marsupialization, sclerotherapy, drainage, enucleation, percutaneous aspiration, and excision of the cyst with or without resection of the involved gut [8,9]. Due to high recurrence rates associated with marsupialization and drainage, complete excision of the cyst should be attempted whenever possible [4]. In adults, the cyst can often be enucleated or 'shelled out' from between the leaves of the mesentery, however, in children, bowel resection is frequently required [9]. Literature mentions instances where laparoscopic removal of mesenteric cysts has been tried successfully, but this might have been difficult in our patients, especially with large sized chylolymphatic mesenteric cysts [10].

Intra-operatively, similar findings can be seen in cystic lymphangioma, retroperitoneal cystic teratoma, caseating tubercular lymph nodes, and hydatid cysts. Even lymphoma and duplication cysts may also give similar appearances. Excision biopsy is recommended to differentiate these cases. Histopathology is confirmatory and differentiates chylolymphatic cysts from all these lesions. 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, but the medical literature also shows some authors describing chylolymphatic cysts as a variant of mesenteric cysts [3,5]. The absence of smooth muscle and lymphatic spaces in the wall of the cyst differentiates mesenteric cysts from cystic lymphangioma [3].

**Conclusion**

Although very rare, chylolymphatic mesenteric cysts should be kept in mind as one of the differential diagnosis of cystic masses in children and can have a varied presentation ranging from asymptomatic to abdominal pain, mass or an acute abdomen with features of intestinal obstruction.

**References**