Case Report

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Pediatric Chylo Lymphatic Mesentric Cyst : A Rare Entity

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ABSTRACT

Chylolymphatic cysts are rare variants of mesenteric lesions constituting 7.3% to 9.5% of all abdominal cysts displaying variable presentation and having surgical implications in the pediatric age group. The preoperative imaging studies are suggestive however histopathological examination remains the gold standard for diagnosis. A two months old male child was admitted with complaints of bilious vomiting for three days which was initially nonbilious in nature.

There were no other significant medical or surgical complaints. X ray abdomen showed evidence of intestinal obstruction.Ultrasound revealed a large cystic lesion in abdominal cavity with internal echos arising from the ileal mesentery on ultrasonography. Subsequently exploratory laparotomy with excision of ileal segment involved by cyst and ileoileal anastomosis was performed and cyst was sent for histopathological examination. On syringing milky white fluid was aspirated which on biochemical analysis confirmed the chylous nature. The histopathological examination revealed an unilocular cyst having a fibrous wall infiltrated by lymphocytes and macrophages and lined by single layer of flattened epithelium with few dilated lymphatic channels consistent with chylolymphatic cyst. Although vary rare, chylomesenteric cyst should be kept as one of the differential diagnosis of cystic masses of the abdomen.

Keywords: *Neonate, chylous cyst, mesentery, histopathology.*

Introduction

A mesenteric cyst is defined as a cyst that is located in the mesentery of the gastrointestinal tract and may extend from the base of the mesentery into the retroperitoneum [1,2]. Mesenteric cyst are among surgical rareities and chylolymphatic cyst is a rare variant of a mesenteric cyst [3]. It was first described by Von Rokitansky in the year 1942 [4]. It is reported both in adult and children. Median age of presentation is 46 years [4]. In children incidence is 1 in 35000 of hospital admission with mean age of presentation of 4-5 years [3]. In most of the cases, the cysts are located in the mesenterium of small intestine, but they can also be found in the descending colon and rectum [5]. These cysts 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 thought to result from an imbalance between the inflow and outflow of fluid across these channels [1]. They may be asymptomatic or may present as abdominal distension, abdominal lump or may present as a case of acute abdomen secondary to a volvulus, intestinal obstruction, hemorrhage, infection, rupture of the cyst. Radiological investigations form an integral part of the diagnosis 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. Abdominal USG is currently the imaging procedure of choice. A "air fluid level" can be seen on USG due to formation of an upper fluid level by lighter chyle over a lower fluid level of heavier lymph [6]. CT with contrast enhanced film can show the relationship of the bowel and other vital structures to the lesion. A fat fluid interface on CT is indicative of a chylous cyst [7] . The treatment of choice is complete surgical excision of the cyst. This can be done either by laparotomy or laparoscopy. The diagnosis is confirmed on histopathology.

Case Report

A two months old male child was admitted with complaints of bilious vomiting for three days. X ray abdomen showed evidence of intestinal obstruction.Subsequently exploratory laparotomy with excision of ileal segment involved by cyst and ileoileal anastomosis was performed and cyst was sent for histopathological examination. Post operative period was uneventfull and patient was dischrged with oral medication. Grossly cystic structure measuring 8x7x6 cm, externally multilobulated with dilated blood vessels on surface was received(Fig1) which on syringing reveal milky white aspirate of aproximately 70 ml and subsequently cyst collapsed(Fig2). Biochemical analysis confirmed the chylous nature of aspirate and cytological examination reveal proteineceous background along with few inflammatory cells (Fig3). The histopathological examination revealed an unilocular cyst having a fibrous wall infiltrated by lymphocytes and macrophages and lined by single layer of flattened epithelium with few dilated lymphatic channels and reported as consistent with chylolymphatic cyst (Fig4).

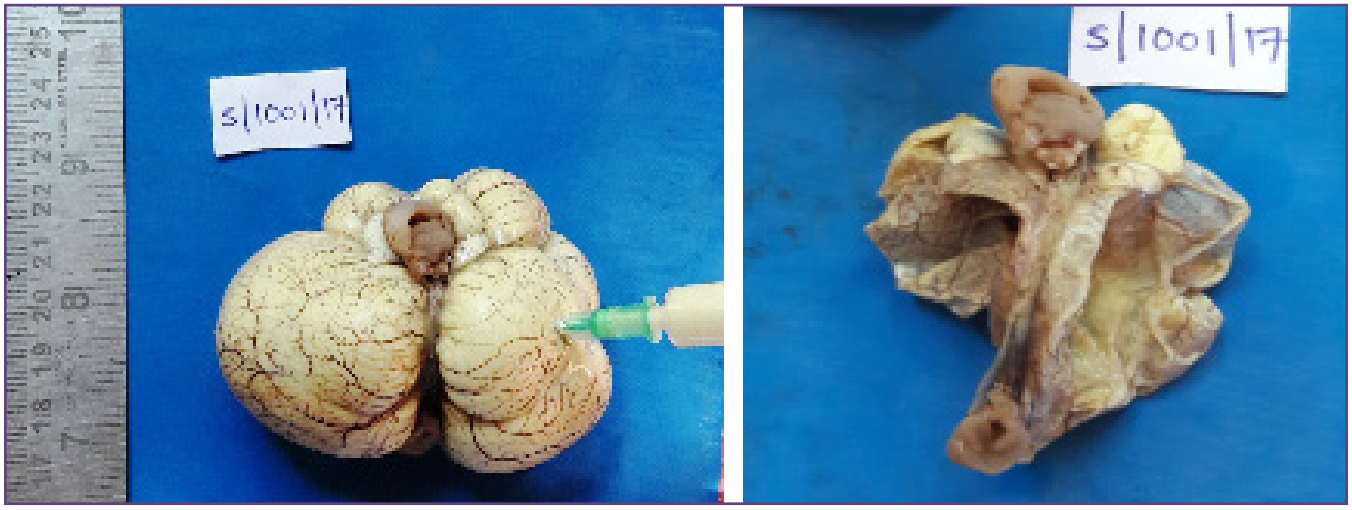

Fig. 1 &2: Grossly cystic lesion with dilated blood vessels with milky white aspirate [1] and collapsed cyst.

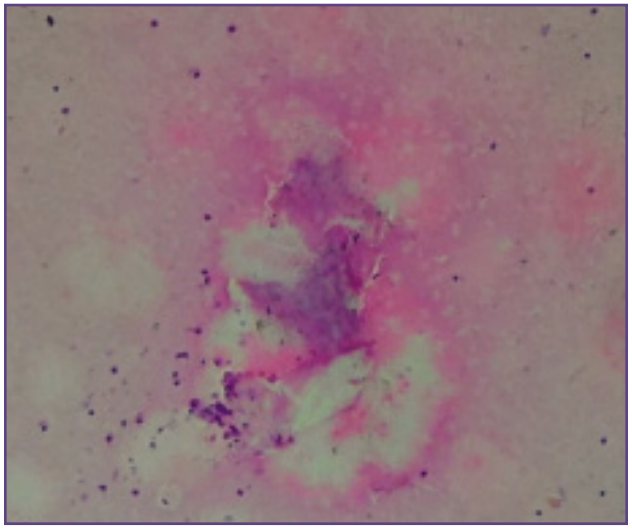

Fig. 3: Cytological smear shows few inflammatory cells against a proteineceous background.[H&E ,40X].

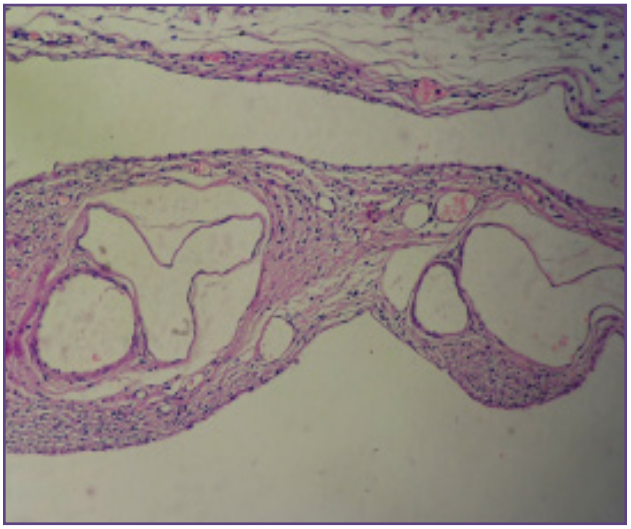

Fig. 4: Histopathological sections [H&E,10X] shows fibrous wall in_iltrated by lymphocytes and macrophages and lined by single layer of flattened epithelium with few dilated lymphatic channels.

Discussion

Chylolymphatic cysts constituting 7.3% to 9.5% of all abdominal cysts [1] displaying variable presentation and having surgical implications in the pediatric age group. There are very few cases of chylolymphatic cysts reported in the literature in neonatal age group [8] . Clinical presentation is not characteristic so the diagnosis of mesenteric cysts pose a challenge as it need to be differentiated from other fluid filled abdominal cystic masses such as mesenteric, omental and dermoid cysts. This is because lymph fluid is normally found in the mesentery of the small bowel and the thoracic duct. True chylous cyst originates from the mesentery of the small bowel whereas false chylous cyst occurs from nonlymphatic sources or regional lymphatics. Chylous cysts are often congenital but may be related to previous abdominal surgery, pelvic diseases, and trauma. Mostly it located in small intestine mesentery and as many as 50–60% occur in association with the ileal mesentery [4]. In our case cyst involved ileal segment and causing intestinal obstruction with perforation. The preoperative imaging studies are imporatnt part of management. A plain radiograph can detect intestinal obstruction, USG and CT scan can suggest mesenteric cyst of chylous nature. In our case only plain radiograph was available suggesting intestinal obstruction. So in most cases diagnosis is confirmed after surgical exploration and removal of cyst.

Management of these cysts involves their removal which may or may not involve resection of the adjacent bowel. Most cysts can be enucleate; however, in some this is not possible without sacrifice of the blood supply to the adjacent bowel and hence necessitates resection. Procedures like marsupialization and drainage are associated with high recurrence rates and are best avoided [9] . In our case excision of ileal segment involved by cyst was done with ileoileal anastomosis. 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 appearance [10]. Histopathology is confirmatory and reveals either unilocular or multilocular cysts.The cysts are usually lined with single layer of flattened endothelium with a fibrous wall in which dilated lymphatics may be observed and may contain lymphoid tissue and foam cells. The presence of cholesterol clefts further supports the pathological diagnosis of a chylous cyst. Microscopy can 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. Malignant transformation is rare but not unknown. Surgical removal is curative.

Conclusion

Although vary rare, chylomesenteric cyst should be kept as one of the differential diagnosis of cystic masses of the abdomen in neonatal period. Complete excision of the cyst ensures excellent prognosis and is curative. Histopathological examination remains the gold standard for diagnosis.

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