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Cystic Abdominal Lymphatic Malformations

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Abstract

Abdominal cystic lymphatic malformations are rare in children. In this article we reviewed our experience of the same for a period of 7 years with respect to their presentation and management. Advanced imaging modalities and regular use of laparoscopy has made diagnosis and management of cystic abdominal lymphatic malformations straightforward and easy.

Keywords: Abdominal Cystic Mass; Lymphangioma; Abdominal lymphatic Malformation.

Introduction

Mesenteric and omental cysts are rare; the incidence is approximately 1 per 105,000 admissions to general hospitals and 1 per 20,000 admissions to pediatric hospitals.[1-3] Modern series show that one fourth occur in patients younger than 10 years.[3-5] These cysts are reportedly more common in females than in males and in white persons than in nonwhite persons.[1,7,8] Mesenteric cysts can occur in the mesentery anywhere along the gastrointestinal tract but cysts arising from stomach are extremely rare.[14,15] They may extend from the base of the mesentery into the retroperitoneum. Omental cysts are located in the lesser or greater omentum. Mesenteric cysts are 4.5 times more common than omental cysts.[6]

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Methods

All cases of intra abdominal cystic lymphatic malformations diagnosed and treated during the period from January 2013 to December 2019 were reviewed. There were totally seventeen children screened for eligibility. Two children with other mesenteric cysts-one in the ileal mesentery (reported as enteric duplication cyst) and one in the root of mesentery (without epithelial lining reported as pseudocyst) were excluded, so the remaining 15 children were included in the study. The clinical features, investigations, surgeries performed and histopathology reports were analyzed.

Results

Out of 15 cases, 7 were female and 8 were male patients. Age of presentation was from infant to 14 years. Two of the patients in this series were infants. Five were less than 3 years, five were from 3 to 5 year group and three were older than 5 years. Five (one third) of the patients presented with acute abdominal symptoms due to intestinal obstruction or hemorrhage inside the cyst and two of them were infants. 7 patients presented with progressive abdominal distension and swelling and the rest (three patients) presented with nonspecific symptoms. Large abdominal cysts were diagnosed during evaluation for these non specific symptoms. Ultrasonogram was the primary investigation for all the cases except the ones who presented with intestinal obstruction. For the patients who

presented with intestinal obstruction abdominal radiograph was the initial imaging followed by CT abdomen with contrast. Almost all patients underwent CT abdomen during evaluation and very few patients underwent MRI. With the combination of USG and CT scan diagnosis was made preoperatively in all the patients. All the children were managed surgically. Laparotomy and cyst excision with or without resection of bowel was the common surgery performed in these children. Eleven children were managed laparoscopically. Complete excision was done for all the patients. Intra operatively the lymphatic malformations were found to have different macroscopic appearances and were found in different locations. All but one had multiloculated cystic appearance. The remaining one was a large single cyst arising from the small bowel mesentery (Fig. 5). Seven of the cysts were arising from small bowel mesentery, four from transverese mesocolon, 3 were in the greater omemtum and one was arising from the gastro hepatic omentum and predominantly retrogastric. The contents were clear or hemorrhagic fluid. Post operative complications were not very significant except for wound infection of the laparotomy wound or port site in about 26.6% of the patients. There were no recurrences.

The details of some of the unique cases are as follows: A Two year old male child was admitted with progressive abdominal distension and intermittent pain for 2 months. On examination,the child was anemic (hemoglobin 7g%)and abdomen was grossly distended. No tenderness or visible intestinal loops were present. The USG abdomen showed large cystic mass occupying the entire abdomen, and MRI confirmed the presence of cyst measuring 20x 17x12 cm and with septations(fig 1). Laparotomy was done under general anesthesia by upper abdominal transverse incision, which revealed a large multi loculated cyst which was twisted and with about 2 liters of hemorrhagic fluid. The cyst was arising from lesser curvature and posterior surface of the body of the stomach with two components, a large component within the lesser sac protruding into the peritoneal cavity (the one which had undergone torsion)and a smaller component along the lesser curvature of stomach.(fig 2). The entire cyst was free except its attachment in the stomach. The larger component was excised with relative ease and the smaller component required careful dissection owing to the proximity to right and left gastric vessels and the vagus and the cyst was excised with seromuscular layers of involved portion of stomach in the posterior wall close to lesser curvature and the same area was repaired with seromuscular inverting stitches. Postoperatively the child had a large volume of gastric aspirate(150 to 200ml) and dilated stomach both lasted for about a week possibly due to operative injury to vagal branches; the same improved with addition of prokinetic agent. The child was discharged home and is on follow up with no complaints. The histopathology of the cyst was inflamed cystic lymphangioma of stomach with hemorrhagic contents.

A 2 year old girl presented with abdominal pain and fever for 1month. Routine USG abdomen showed cyst of size 8x8 cms in lower abdomen. The differential diagnosis was between a mesenteric cyst and ovarian cyst though a subsequent CT showed the ovaries to be normal.(Fig 3a). A diagnostic laparoscopy was done followed by excision of the cyst laparoscopically . Cyst was thick walled with infected fluid.(Fig3b) Histopathology confirmed the cyst to be of lymphatic origin.

A 7 year old boy with progressive abdominal distension and fever on and off for 6 months. USG abdomen was suggestive of a free fluid abdomen with turbidity due to a possible sealed perforation. CT also was suggestive of loculated ascites with calcific material. Laparotomy was done and a 12x18 cm cystic lesion hanging from the transverse colon attached to the greater omentum was seen. The cyst had chylous fluid with macroscopic calcification. The cyst was excised in toto along with greater omentum. Histopathology confirmed it as giant cystic lymphangioma with calcification.(fig 4).

4 year old female child presented with recurrent abdominal pain. CT scan showed cystic structure in the jejunal mesentery occupying the right side of the abdomen and extending to pelvis. Laparoscopy showed a large cystic lesion with a thick wall. (fig 5) The cyst was in the jejunal mesentery near the mesenteric border of the bowel. The cyst was carefully dissected from the blood vessels and bowel and enucleated from the mesentery.

All the children in the present series who underwent emergency surgeries were aged less than three years. 3 (20%) of the five cases who underwent emergency surgeries required resection of the bowel along with cyst. In all other children the cysts were removed successfully either laparoscopically or by laparotomy. The location of the cyst was the important factor determining need for bowel resection. The cysts which are located very close to and inseparable from major arterial branches compromising the blood supply of the bowel or a large cyst causing volvulus of the bowel and gangrene is the reason for resection in most of

Fig 1: MRI picture showing giant cystic lymphatic malformation occupying entire abdomen.

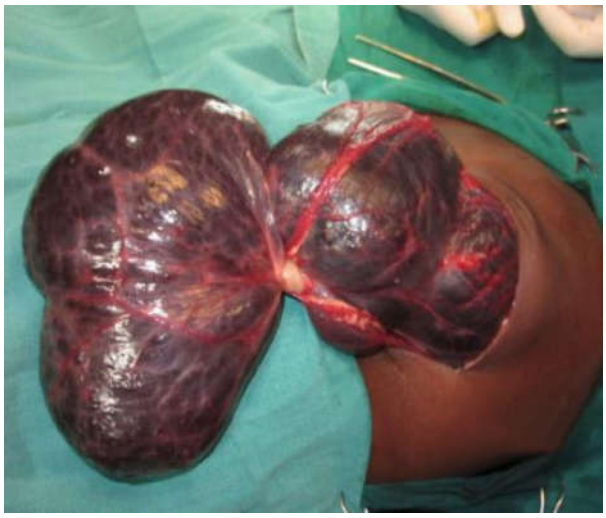

Fig 2: cyst arising from lesser curvature and posterior surface of body of stomach with two components, a large component in the lesser sac partly protruding into the peritoneal cavity along the lesser curvature of stomach.

Fig.3a

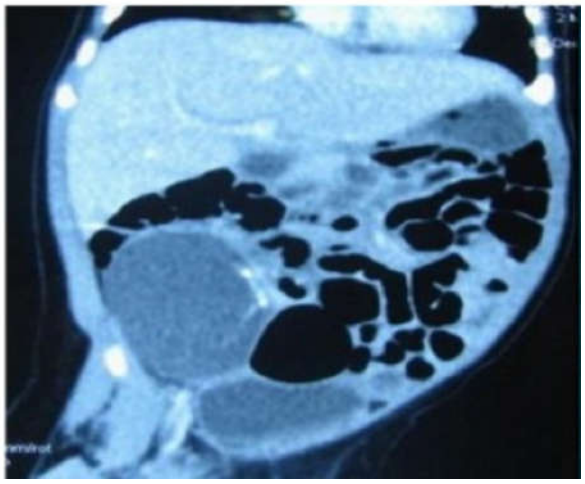

Fig3a: CT picture in three year old girl showing cyst in the right side of abdomen and suspected as an ovarian cyat initially**.**

Fig.3b

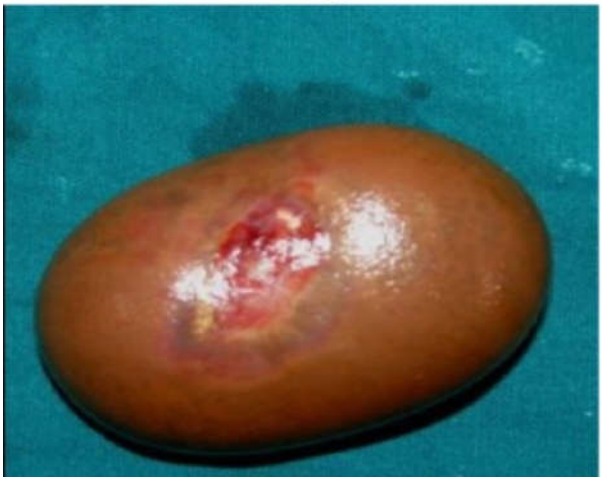

Fig 3b: Specimen of thick walled uni locular cyst with infected fluid.

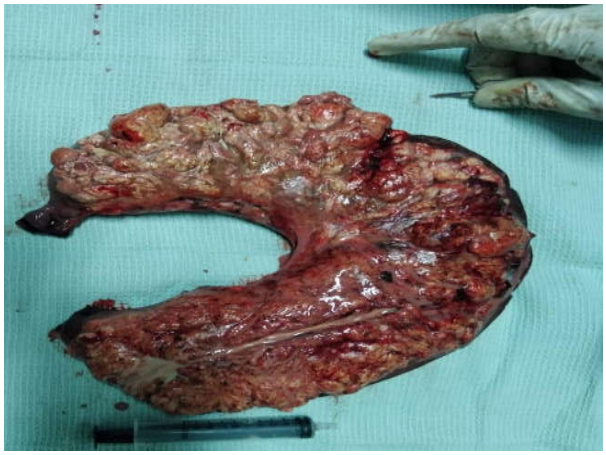

Fig 4: Excised specimen of large mesenteric cyst arising from transverse colon containing chylous fluid with macroscopic calcifications.

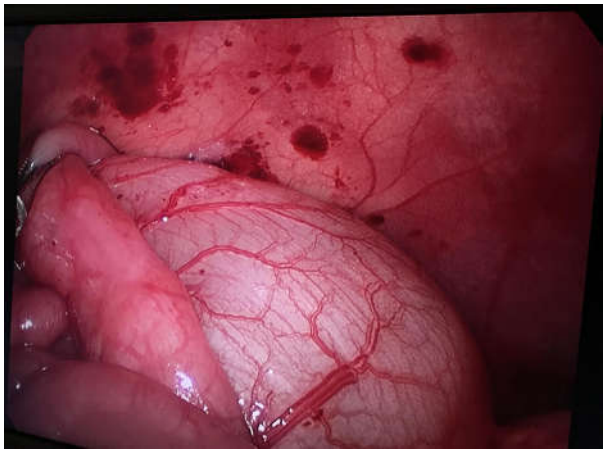

Fig 5:Laparoscopic view of a large mesenteric cyst arising from small bowel mesentery.

the cases. Volvulus occurs when the cyst is located near the intestinal border of the ileal or jejunal mesentery allowing free movement of the bowel along with the cyst during peristalsis.

Eleven out of fifteen patients in this series were managed laparoscopically. The usual technique followed in these cases was a diagnostic laparoscopy by three 5mm ports, one umbilical and other depending on the location of the cyst. In most of the cases after initial inspection the cyst fluid will be aspirated, then the bowel segment with partly or fully collapsed cyst will be delivered by extending the umbilical incision to about three centimeters and either the cyst alone or along with small segment of bowel will be resected. Anastomosis was done externally and the bowel placed back and the incision sutured.

Histopathology in all our cases were carefully reviewed in order to categorize them as the lymphatic malformation and not in other differential diagnoses. All of them had unequivocal features of lymphatic origin, like endothelial lining either in part or full, endothelial lined microscopic spaces called lymphatic spaces, areas of smooth muscles and foam cells.

Discussion

In our series the sex distribution was almost equal (7 female and 8 male). Five of the cases (one third) presented with acute symptoms. Four with features of acute intestinal obstruction like bilious vomiting and abdominal distension and one with severe abdominal pain and anemia suggestive of bleeding within the cyst. The rate of emergency operation in our series is 33.3% which is similar to most other studies in the literature.[20] Bowel resection was required in our series in 20% of the cases. Some series have a high intestinal resection rate upto 50% (20,24). 73.3% of the patients were operated laparoscopically.

Beahrs and colleagues(1950) classified cystic disease of the mesentery into four categories on the basis of possible etiology: embryonic and developmental cysts, traumatic cysts, neoplastic cysts, and infective cysts. The classification is quite confusing. Recent classification of mesenteric cysts, proposed by de Perrot,[12] is based on the origin of the cyst: 1. lymphatic origin (lymphangioma); 2. mesothelial origin (mesothelial cyst, cystic mesothelioma); 3. enteric origin (duplication cysts); 4. urogenital cysts, 5. cystic teratomas (dermoid), and 6. nonpancreatic pseudocysts (traumatic and infectious). However, most of the mesenteric cysts in children are benign proliferation of lymphatic tissue and they are of embryonic origin.

Multiple theories of origin of mesenteric lymphatic cysts have been proposed. The most commonly accepted theory, proposed by Gross,[10] is benign proliferation of ectopic lymphatics in the mesentery that lack communication with the remainder of the lymphatic system. These cysts are thought to arise from lymphatic spaces associated with the embryonic retroperitoneal lymph sac, analogous to cystic hygromas of the neck arising in association with the jugular lymph sac.[10] The role of lymphatic obstruction is questionable. Because mesenteric and omental cysts are all from anterior extensions of what were retroperitoneal structures during the embryonic stage, they share common characteristics along with retroperitoneal cysts. Since mesenteric, omental, and retroperitoneal Cysts of lymphatic origin share common characteristics, the term "cystic lymphatic malformations" may be more appropriate.[11]

The pathologic features of these cysts can vary considerably. They can be single or multiple, unilocular or multilocular; they can have serous, chylous, hemorrhagic, or mixed fluid contents. Cystic lymphangiomas are simple cysts with an endothelial cell lining, foam cells, and thin walls that contain small lymphatic spaces, lymphoid tissue, and smooth muscle. The wall of a non-lymphatic mesenteric cyst lacks smooth muscle and lymphatic spaces, and the lining cells are cuboidal or columnar with patchy fibrosis.[9,13,19] Rarely, the cyst wall contains calcium. Unlike duplication cysts, mesenteric and omental cysts contain no mucus-producing cells.[8]

The clinical presentation of mesenteric and omental cysts can vary from an incidental finding during laparotomy performed for another reason to an acute, life-threatening intra-abdominal catastrophe. In adults, these cysts are found incidentally in approximately 40% of patients[1,13] and present as acute abdominal emergencies in up to 60% of patients. The classic presentation is that of a low-grade, partial intestinal obstruction combined with a palpable, freely movable abdominal mass.

The most common mode of acute presentation in children is a small bowel obstruction, sometimes associated with volvulus and intestinal infarction[7,9] The differential diagnoses include intestinal duplication cyst, ovarian cyst,choledochal cyst, hydronephrosis, ascites, pancreatic or splenic cyst or hydatid cyst. Complications associated with mesenteric and omental cysts include intestinal

obstruction (most common), volvulus, hemorrhage into the cyst, infection, rupture, torsion of the cyst,[13] obstruction of the urinary or biliary tract,and malignancy.[14] The reported incidence of malignant conditions (sarcoma, lymphangioendothelioma, or, rarely, adenocarcinoma) is 3%.

The goal of surgery is complete excision of the mass. Omental cysts are easily removed and almost never require bowel resection.[9] Lymphatic cysts can be easily excised using laparoscopic techniques.[6] Partial drainage of the cyst may be necessary to confirm the site of origin of the cyst and allow its removal through the umbilical port. Most of our patients(73.3%) were managed by minimally invasive techniques which significantly reduced cost, number of days in the hospital, complication rate and size of the post operative scar.

The preferred treatment of mesenteric cysts is enucleation. In adults, the cyst can often be "shelled out" from between the leaves of the mesentery in children, however, bowel resection is required to totally eradicate the mass and ensure that the blood supply to the bowel is not compromised. A bowel resection is necessary in only about 33% of adults, but 50% to 60%

of children with mesenteric cysts require resection.[4,9,14,20,24] Bowel resection was required in our series in 20% of the cases only. Recurrence is a known complication with a range from 1 to 13%.Most recurrences occur in patients with retroperitoneal cysts or those who had a partial excision.[13] None of our cases recurred in the follow up period.

Very few cysts arising from the stomach or mesentery of stomach have been reported so far in literature and mostly in adult patients.[14,16-18] The cysts so huge as with one of our cases arising from the posterior surface of stomach and extending to peritoneal cavity through lesser omentum are difficult to excise as it may involve resection of stomach and can injure the motor supply of the pylorus of the stomach. A complete denervation may require a drainage procedure in the form of pyloroplasty.

Conclusion

Mesenteric and omental cysts are rare, but they are more commonly encountered in children as congenital abnormalities of the lymphatic system. The cysts can be located in the mesentery or omentum. Very few cysts arising from the stomach or mesentery of stomach have been reported so far in literature. The age, sex clinical features and histological characteristics in our study corresponds to the similar studies in the past. CT scan and MRI made the diagnosis preoperatively in all the cases. Most of our cases were operated electively and one third of our patients(33.3%) required emergency operations. 73.3% of the patients were operated laparoscopically. 20% of our cases required bowel resection as against the literature reported rate of 50 to 60%. The present study is aimed at emphasising the importance of making the diagnosis of cystic abdominal lymphatic malformation and its anatomical location preoperatively through good imaging techniques and thus facilitating management of the children with minimally invasive modality. Most of these children can be managed by laparoscopy who would have otherwise required a laparotomy and associated longer postoperative stay in the hospital and higher complication rate.

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