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Case Report

Mesenteric Cystic Lymphangioma, an acute presentation in a 9-year-old child ☆

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ABSTRACT

Mesenteric lymphangioma is a rare malformation of the lymphatic system. Misdiagnosis of mesenteric cystic lymphangioma can occur due to its rarity and resemblance to other cystic lesions and ascites. The authors report an acute presentation of a mesenteric lymphangioma in a 9-year-old child. Clinical, radiological, and surgical findings are illustrated accordingly. As authors, we aim to add to the limited data of acute presentations of mesenteric lymphangiomas.

Introduction

Mesenteric lymphangioma and simple mesenteric cysts are both rare presentations. Lymphangioma is a rare malformation of the lymphatic system that is mainly congenital but can also be acquired. Nearly 95% of lymphangiomas are present in the head, neck, and axial regions [1,2], while the remaining 5% occur in the mesenteries, abdominal viscera, lung, retroperitoneum, and mediastinum.

Lymphangiomas in the mesentery are the most reported intra-abdominal lymphangiomas [2,3]; 70% of intra-abdominal cystic lymphangiomas arise from the mesenteries [4]. Mesenteric cystic lymphangiomas (MCLs) have an incidence of 1 per 250,000 [5]. Cystic lymphangioma usually presents in male children under the age of 12 years [6].

The rarity of lymphangiomas and their clinical and radiological presentation makes them challenging to recognize and can lead to adverse outcomes. We report this case with clinical, radiological, surgical, and macroscopical presentation illustrations to add to the scarce data. Thus improving clinical practice with favorable outcomes for the patients involved globally.

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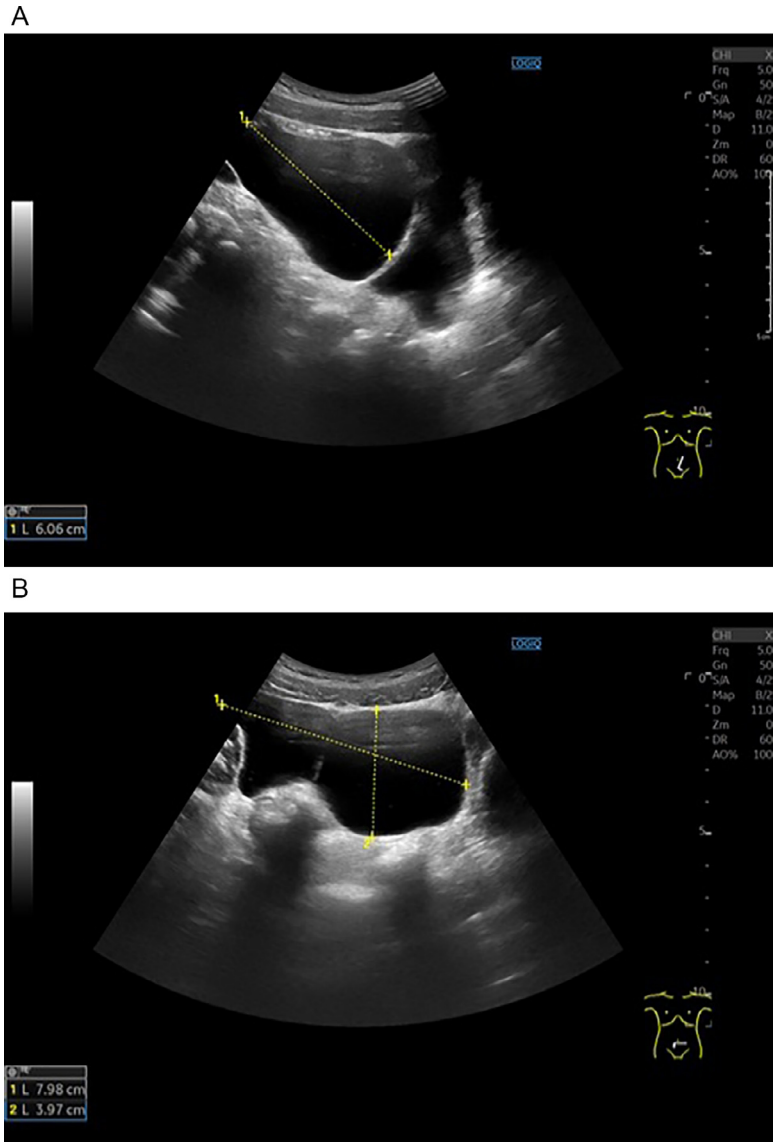

Fig. 1 – (A and B) Acute Abdominal Ultrasound. It showed a fluid collection with thin mobile incomplete septa in the lower abdominal quadrant. The fluid collection measured about 4 X 6 X 8 cm. The image was considered as free fluid intraperitoneally with debris-mobile septa. However, retrospectively, this is a case of a large cystic lesion /pseudocyst with septa, where some of them were ultra-thin and could not be fully detected Figure 2.

Case report

A 9-year-old healthy Indian boy with a previously operated right-sided inguinal hernia at age 3 presented to the Emergency Department with lower abdominal pain, repeated vomiting, inability to eat nor drink, and no bowel motion in 4 days. He had normal vital signs and a weight loss of 2.7 kg (26 kg from 28.7 kg), sunken eyes, an old 1.5 cm right-sided inguinal hernia incision, and slight abdominal tenderness at the level of the urinary bladder. Oral rehydration therapy had no good effect.

Biochemically: Urin stix: pH 5.5, Blood +1, Ketones +3, protein trace. No leukocytes, glucose, or nitrite. Arterial gas analysis: metabolic alkalosis with Bicarbonate 31.6. Otherwise, normal blood tests.

Abdominal ultrasound showed a large anechoic fluid collection extending from the epigastrium to the pelvis measuring about 5 X 8 cm, with few mobile incomplete thin fibrin septa. There was neither an ileus of the scanned bowels nor a suspicion of an abscess formation. In the clinical context, the near-normal biochemical parameters, abdominal ultrasound imaging, and the symptoms mentioned above led to the tentative diagnosis of a perforated hollow organ with free fluid intraperitoneally, fibrin debris septa, and reactive peritonitis, Figs. 1A and B. The radiologists advised the patient to neither eat nor drink, and an abdominal MRI was scheduled.

An abdominal MRI revealed the presence of a large lesion with internal septa extending from the epigastrium to the lower abdomen, Fig. 2A, B and C, which led to a tentative diagnosis of either internal herniation or a cystic tumor causing

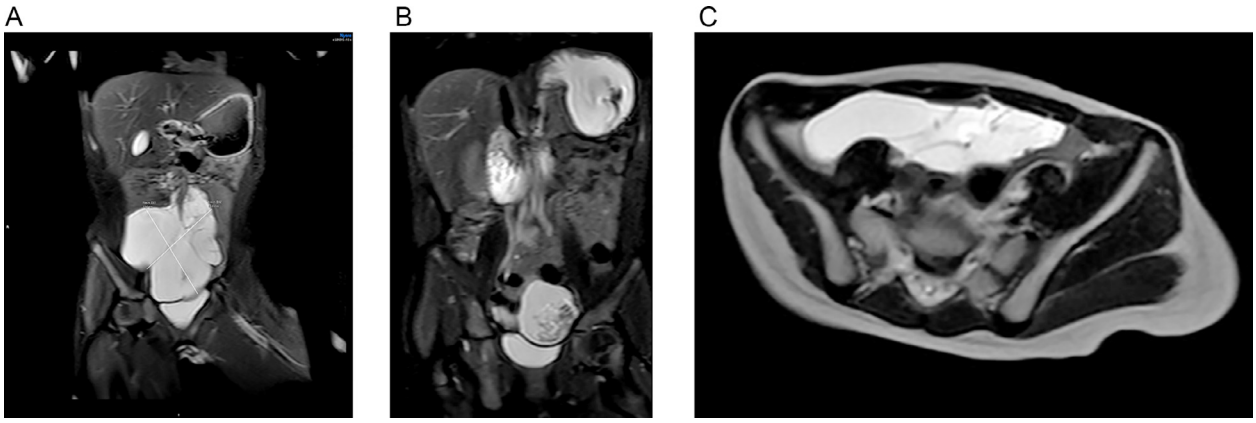

Fig. 2 – (A-C) Acute Abdominal MRI without Contrast. The patient could not cooperate in executing the entire investigation—Coronal 2B - SPAIR sequence, and Coronal (A) and Axial (C) T2-weighted images showed the presence of a large fluid-filled lesion with internal septa with a possibility of hemorrhage/calcification and no signals in the diffusion restriction sequences. The lesion extended from the epigastrium to the lower abdomen measuring 9 cm X 11 cm. Fluid retention and dilatation of the stomach and the 1. st and 2nd parts of the duodenum with a probability of being followed by a whirl sign- raising the suspicion of internal herniation. Also, a possibility of the lesion being a benign cystic tumor.

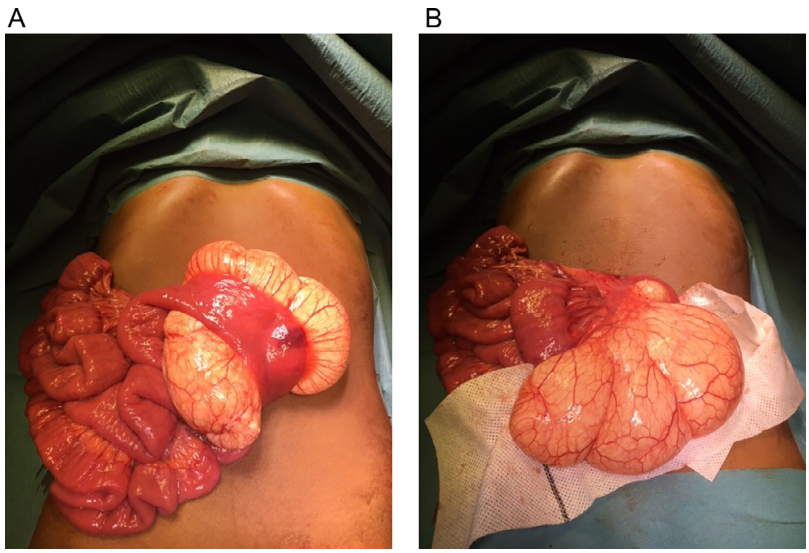

Fig. 3 – (A and B) Open laparotomy. A well-defined 15 cm large cyst was seen in the mesenteryextending from the proximal jejunum, and insinuating the blood vessels. Normal bowel with no signs of ischemia. Total resection of the cyst and about an adjacent 10 cm long bowel from the jejunum with primary enteroenteric anastomosis located about 5 cm distally from the ligament of Treitz. The cyst burst unintentionally outside the abdomen, revealing white fluid. Therefore, the pathology was initially diagnosed as a chylolymphatic mesenteric cyst.

proximal bowel obstruction. Later, within hours, the images were reviewed by another set of consultants at another larger university hospital, which is also a center for pediatric radiology. The colleagues also raised the suspicion of internal herniation or a nonmalignant cystic tumor. As a result, the patient was operated acutely, Fig. 3.

A laparoscopic exploration revealed a large pathology in the mesenteries; therefore, an open laparotomy was favored. A well-defined 15 cm large cyst in the mesentery extending from the proximal jejunum and insinuating the blood vessels (Fig. 3) was identified. The cyst was resected entirely along with about an adjacent 10 cm from the jejunum- diagnosed as a chylolymphatic mesenteric cyst. However, the histopathology examination diagnosed the cyst as a mesenteric cystic lymphangioma, Fig. 4.

Discussion

Mesenteric cysts and mesenteric lymphangiomas are challenging to tell apart clinically, radiologically, and intraopera-

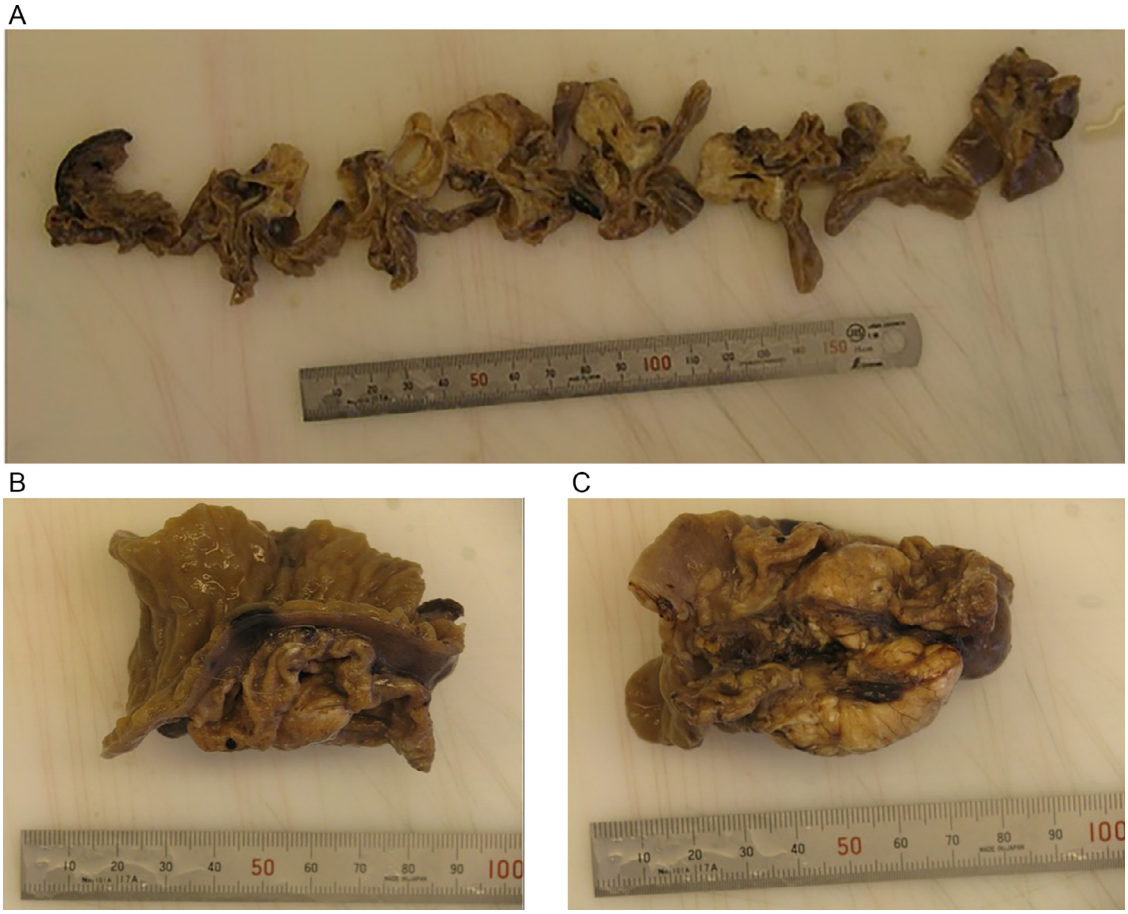

Fig. 4 – (A-C) Histopathological investigation. A lobulated cystic mass with a whitish-yellowish tan appearance with vascular streaks protruded from both sides of the mesentery and was surrounded by the mesenteric peritoneum. The cyst contained a thin white fluid. Microscopically, the resected bowel showed normal mucosa, submucosa, and muscular layers. At the side of the serosal layer of the bowel, there was cystic dilation lined with a single layer of endothelial-like cells with neither dysplasia nor malignancy—positive immunohistochemical staining to transcription factor ETS-related gene (ERG) and D2-40, and negative to Calretinin. ERG; is a highly specific endothelial marker. D2-40; a selective marker of lymphatic endothelium in normal tissues and vascular lesions. Final diagnosis- Benign Mesenteric Cystic Lymphangioma. Microscopic illustrations are not available.

tively [7]. Therefore, misdiagnosis can occur. A chylolymphatic mesenteric cyst is a rare variant of a mesenteric cyst [7]. Cystic lymphangioma and chylolymphatic mesenteric cysts have a striking resemblance both macroscopically and microscopically [7]. All types of mesenteric cysts have no smooth muscles and lymphatic spaces in their wall. These microscopical features help to distinguish mesenteric cysts from cystic lymphangioma [7,8].

Presentation, whether a cyst or a lymphangioma, varies and depends on the anatomic site and size. It can range from being asymptomatic and detected incidentally to being life-threatening [9]. Unspecific abdominal symptoms such as nausea, vomiting, digestive disorders, melena, palpable abdominal mass, increasing abdominal girth, or pseudo-ascites have been reported [4,5]. Patients with intra-abdominal cystic lymphangioma (ICL) have shown unspecific intestinal disorders in 59%, while 17% presented with acute abdominal symptoms [4]. Especially mesenteric lymphangiomas seem to become apparent more frequently by acute abdominal symptoms [4].

Cystic lymphangioma is one of these benign cystic diseases/lesions that may grow large enough to 'mimic loculated ascites'. Differentiating lymphangiomas from other fluid-containing masses and ascites is radiologically challenging [2]. Ultrasound can detect septa in ascites/pseudo ascites in 85% of cases and defined chambers in 15%, but using ultrasound for presurgical diagnosis remains challenging [4]. Large cysts, including cystic lymphangiomas, renal cysts, mesenteric cysts, ovarian cysts, pseudocysts, etc., can be misdiagnosed as ascites [6,10].

Accurate diagnosis through immunohistochemical and histopathological exams is necessary because cystic lymphangioma can become invasive and proliferate into its surroundings, leading to life-threatening complications such as bowel obstruction, volvulus, and invasion of the bowel walls and vessels [1,8]. It is also of prognostic relevance due to re-

currence risk in incomplete resection of an intra-abdominal cystic lymphangioma (ICL), for example [4]. Complete surgical resection is the only curative therapy for cysts and lymphangiomas [4,5].

Lymphangiomas of the small bowel mesentery represent less than 1% [11,12]. Acute intestinal obstruction in the form of volvulus is the most common manifestation of mesenteric lymphangioma [11]. Suthiwartnarueput et al. [11] reported a mesenteric lymphangioma presenting as a volvulus in a 2-year-and-9-month-old boy who was operated acutely with bowel resection [11]. [12] Kumar et al. reported a similar presentation of acute intestinal obstruction in 8-year-old and 10-year-old boys.

Generally, when such fluid collections are seen radiologically, other differential diagnoses should also be considered, such as an abscess, cysts of foregut/urothelial origin, and malignant tumors or metastasis with cystic degeneration. In the retrospective analysis, our patient had compression symptoms from the mass effect exerted by the lymphangioma, leading to obstruction of the duodenum and the stomach and thus manifesting itself as abdominal pain, recurrent vomiting, and subsequent dehydration with metabolic alkalosis. The acute operation executed in the specialized center was in the light of the given presentation. The patient was discharged, followed up, and did not manifest postoperative complications with the histopathological analysis/examination consistent with a benign mesenteric cystic lymphangioma. The patient was doing well at the time of writing this manuscript.

Conclusion

1- Including large-size cysts/ pseudoascites in the differential diagnosis of ascites when it demonstrates loculations and septations is necessary when doing ultrasound exams especially in pediatric patients.
2- MRI is of diagnostic value in children presenting with fluid collections. Therefore, it can be used for a second step after ultrasound.

Patient consent

Written consent to access the electronic journals, tests, and images, and to publish the clinical case and all figures anonymously were obtained from both parents. This consent was archived and if requested by the journal, the consent can be provided.

Written consent to access the patient's electronic journals and tests was obtained from both the Department of Radiology/Kolding hospital and the hospital. This consent can also be provided upon request.

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