

Cystic lymphangioma of the lesser curvature of the stomach - case report

Hyuma A. Leland¹, James T. Lee², Justin H. Tan², Lorene E. Romine², Vishal Bansal^{1*}

1. Department of Surgery, UCSD Medical Center, San Diego, CA, USA

2. Department of Radiology, UCSD Medical Center, San Diego, CA, USA

* **Correspondence:** Vishal Bansal, MD, FACS, Assistant Clinical Professor of Surgery, 200 W. Arbor Dr. #8896, San Diego, CA 92103-8896, USA
(✉ v3bansal@ucsd.edu)

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ABSTRACT

We report a case of a cystic lymphangioma arising from the lesser curvature of the stomach in a 16-year-old female who initially presented with abdominal pain, nausea, and emesis. Contrast enhanced computed tomography and magnetic resonance imaging revealed a large, thin-walled multicystic mass located anteromedial to the stomach, which was predominately supplied by the left gastric artery. Given the imaging appearance and location, a mesenteric cyst, specifically a cystic lymphangioma, was considered. Lymphangioma was the final pathological diagnosis after laparotomy with complete resection of the cystic abdominal mass.

CASE REPORT

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A 16-year-old Hispanic female with no significant medical history presented with two weeks history of left upper quadrant abdominal pain with nausea and non-bilious emesis. On physical exam, her left abdomen was mildly tender to palpation with a palpable mass in the epigastrium and left upper quadrant. No guarding or rebound tenderness was present. She was febrile with leukocytosis on admission.

A contrast enhanced (intravenous iodixanol) abdominal and pelvic computed tomography (CT) examination was performed on a 64-row multidetector CT (Lightspeed VCT, GE Medical Systems, Milwaukee, WI) and demonstrated a lobulated, low attenuation 9.9 x 10.5 x 11.4 cm mass along the lesser curvature of the stomach occupying both the gastrohepatic ligament and lesser sac (Fig. 1a and 1b). No surrounding inflammatory changes were present to suggest infection and there was no proximal gastric distension to suggest obstruction. A contrast-enhanced magnetic resonance (MR) examination (Signa 1.5T, GE Medical Systems, Milwaukee, WI with 10mL of gadobenate dimeglumine) of the abdomen was performed for further characterization. This demonstrated a 12.8 x 8.9 x 11.3 cm multiseptated cystic lesion with thin walls and multiple thin septae, with no enhancing solid components. The fluid within the mass

demonstrated uniform T1 hypointensity and T2 hyperintensity, with no evidence of hemorrhage, proteinaceous material, or focal wall thickening to suggest infection (Fig. 2a, 2b, 3a, and 3b). The left gastric artery was noted to course through the mass on CT and MR, and MR revealed multiple additional smaller vessels traversing through the mass, particularly at the cephalad aspect. No narrowing of the vessels was seen. At the time of interpretation, the differential diagnosis provided was lymphangioma versus mesenteric cyst.

The patient was taken to the operating room for mass excision. Upon examination, the cyst was adherent to the lesser curvature and posterior wall of the stomach and gastrohepatic ligament (Fig. 4). The gastrocolic ligament was divided to allow access to the lesser sac, facilitating dissection of the mass from the posterior wall of the stomach. A combination of electrocautery and silk ties were used to control bleeding from the vasculature of the cyst. The root of the cyst was then determined to include the left gastric artery, which was tied off to facilitate complete removal of the cyst for pathologic analysis. The patient tolerated the procedure well and was discharged without complication on post-operative day 5, tolerating a regular diet. A 7 month post-operative intravenous contrast enhanced CT demonstrated no recurrence of the lymphangioma (Fig. 5).

Upon pathologic examination, the excised mass was a 12.0 x 10.0 x 5.0 cm multilocular cystic mass filled with serosanguinous and hemorrhagic fluid. The size of the loculi within the cyst ranged from 0.5 to 3 cm and wall thickness ranged from 0.1 to 0.5 cm. Histological sections showed variably sized cysts filled with proteinaceous fluid and lined by a flattened layer of endothelial cells, with a patchy background of acute and chronic inflammation (Fig. 6a). Areas of necrosis and xanthrogranulomatous changes with cholesterol clefts were present (Fig. 6b). The final histological diagnosis was lymphangioma.

DISCUSSION

Abdominal cystic lymphangiomas are exceedingly rare benign tumors with incidence reported between 1 in 20,000-250,000 [1]. Most commonly, lymphangiomas occur in the head, neck, and axilla, with less than 5% affecting the intra-abdominal compartment [2, 3]. Age at presentation occurs in a bimodal distribution with greater than 80% presenting before age five [4] and the remainder occurring around age 40 [1]. The etiology of cystic lymphangiomas is not fully understood [5], but anomalous development of the lymphatics [6, 7] or inflammation and obstruction of developed lymphatic channels have both been proposed as possible mechanisms [8, 9]. Cyst contents are variable and may include serous, hemorrhagic, chylous, or mixed fluid. Rarely, calcification may be present [2]. Lymphangiomas can become locally invasive and often require surgical excision, with recurrence rates of 12% and 53% when completely or partially resected, respectively [4]. To our knowledge, only a few case reports describe cystic lymphangiomas occurring intra-abdominally in adolescent patients [10, 11]. Of these reports, only one describes a cystic lymphangioma originating from the lesser curvature of the stomach [12].

Early imaging, detection, and diagnosis of abdominal cystic lymphangiomas is important to definitive treatment of this rare and treatable tumor. Imaging plays an important role in preoperative evaluation as lymphangiomas tend to be insinuating and may make complete surgical resection difficult [2]. We report both CT and MR imaging which clearly detail the morphology and imaging characteristics of a cystic lymphangioma arising from the lesser curvature of the stomach in a 16-year-old female. Histological analysis of the tumor supports the characteristic findings previously reported in the literature [13].

CT of a cystic lymphangioma typically demonstrates a multilocular cystic mass with thin septa that demonstrate enhancement of the septa and cyst wall with intravenous contrast. Cyst contents are of fluid attenuation most commonly, though the density may be less with chylous contents, or increased with hemorrhage or infection [2, 14]. On MR, the same features are seen, typically with cyst content intensity similar to fluid on T1 and T2-weighted images in the absence of hemorrhage or infection [2]. Similarly, the contents may appear anechoic or contain echogenic material on sonographic evaluation [2, 14]. Radiographs may reveal displacement of bowel loops by a soft-tissue mass [15].

Included in the differential diagnosis of an intra-abdominal cystic mass are an enteric duplication cyst, enteric cyst, mesothelial cyst, pancreatic pseudocyst, non-pancreatic pseudocyst, cystic mesothelioma, cystic spindle cell tumor, and cystic teratoma [16], of which the imaging features will be briefly discussed. For a more complete discussion of these entities, readers are referred to an excellent review by Stoupis, et al [16].

Enteric duplication cysts are a rare congenital malformation exhibiting diversity in size, location, and presentation. CT and MRI with contrast characteristically exhibit a thick cyst wall with fluid, debris, or blood contents, while sonographic examination yields a classic "three layer" appearance created by the echogenic mucosa, hypoechoic muscular layer, and echogenic serosa [17].

Enteric cysts are the result of migrated bowel or diverticuli into the mesentery and are lined by enteric epithelium. In contrast to duplication cysts, enteric cysts lack a muscular layer and nervous plexus in the cyst wall. As a result, enteric cysts lack a discernable cyst wall on CT and MR examination. Sonogram reveals a hypoechoic cystic mass with occasional thin septations [16, 18].

Mesothelial cysts arise as a result of a developmental failure in the migration of mesothelial cells and most commonly occur in the mesentery or small bowel of females [18]. By CT and MRI, mesothelial cysts appear as a fluid filled mass without visible septations or cyst walls, and exhibit fluid signal on all pulse sequences. Sonography is notable for an anechoic mass without acoustic enhancement or discernable cyst wall or septations [16, 19].

Pancreatic pseudocysts result as a complication of acute or chronic pancreatitis and consist of a localized, amylase and pancreatic enzyme rich fluid collection that is surrounded by a thick fibrous wall without epithelial lining. CT imaging that reveals a round, thick walled, fluid-filled cyst adjacent to the pancreas in a patient with a history of acute or chronic pancreatitis is diagnostic. MRI provides improved characterization of fluid collections and cyst debris while sonography shows a thick walled cyst [20].

Non-pancreatic pseudocysts are believed to be the result of trauma or infection and exhibit a thick fibrous wall without a cellular lining on histology [18]. CT and MRI demonstrate septal enhancement and thick-walled cysts with fluid levels demonstrating hemorrhage or purulence, whereas sonography only demonstrates a hypoechoic mass with variable echogenic debris [16].

Cystic mesotheliomas represent a potentially locally invasive, rare, benign mesothelial neoplasm found most commonly in middle-aged women with history of abdominal or pelvic surgery, trauma, or inflammation. CT, MRI, and sonogram demonstrate a non-calcified multicystic mass with hypointense T1 and hyperintense T2 signal on MRI correlating with serous fluid content [16, 18].

Cystic spindle cell tumor (aka gastrointestinal leiomyoma or leiomyosarcoma) is a potentially locally invasive tumor that may develop a necrotic core. Cystic spindle cell tumors show irregular walls, enhancing solid components, and a high attenuation rim on CT. MRI better characterizes the extent of tumor and cyst contents. Sonogram typically shows a complex mass with echogenic debris and may reveal septations of variable thickness [16].

Cystic teratomas are totipotent germ cell tumors with radiographic findings reflecting cyst contents. CT and MRI demonstrate the mixed content of these tumors, often with foci of fat, fluid, and calcifications. Ultrasound is likewise dependent on the tissue composition, but may demonstrate anechoic cystic areas, hyperechoic fat, or echogenic calcifications with posterior acoustic shadowing [16].

Abdominal cystic lymphangiomas have a variable clinical presentation and should be included in the differential diagnosis when CT or MR imaging reveals a characteristic multi-septated, enhancing, thin-walled cystic mass. Ultrasound can adequately characterize the cystic and septated features of lymphangiomas, but further characterization of local tissue involvement, vasculature, and cyst fluid is necessary by CT or MRI. While CT is an acceptable imaging option, MRI is preferred as it reduces exposure to ionizing radiation in a largely pediatric population. Radiologic characterization of the mass should include the size, location, mass effect on or involvement of neighboring organs, vascular supply, and signs of inflammation in or around the cyst. Histology is diagnostic with characteristic endothelial-lined cysts, myofibroblastic proliferation, and xanthogranulomatous changes [2, 13]. As a rare and treatable disease, radiographic recognition of the cystic lymphangioma is important to early diagnosis and excision of the mass before it becomes locally invasive. Complete surgical excision is the preferred intervention.

TEACHING POINT

Abdominal cystic lymphangiomas have a variable clinical presentation and should be included in the differential diagnosis when CT or MR imaging reveals a characteristic multi-septated, enhancing, thin-walled cystic mass. Complete surgical excision is the preferred intervention.

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FIGURES

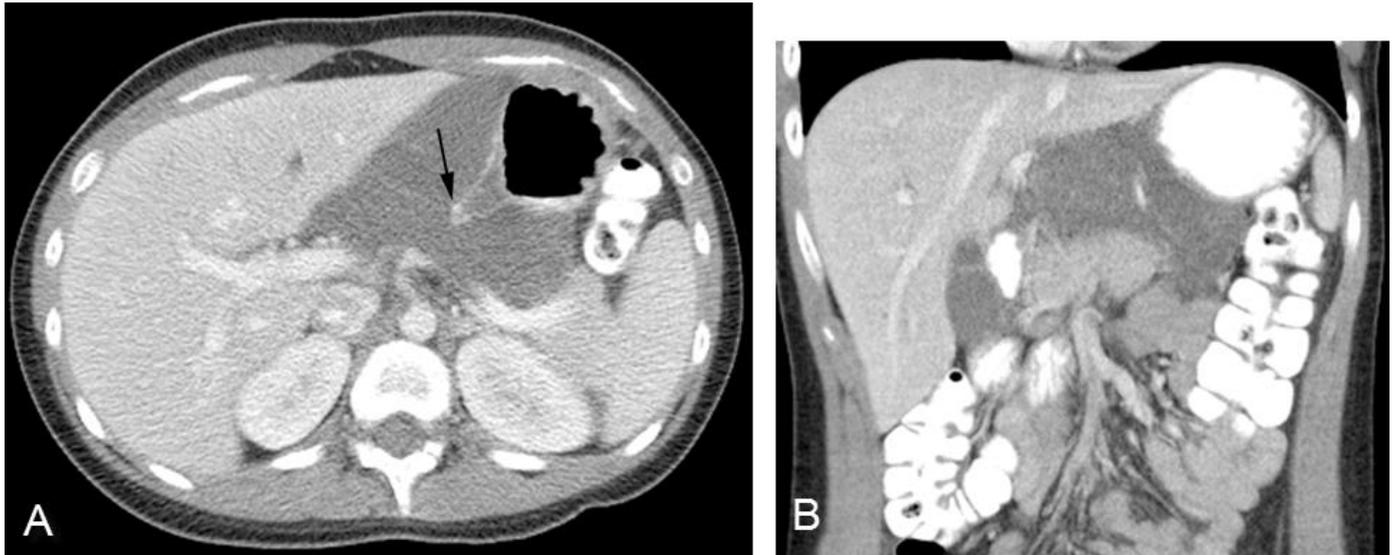


Figure 1: 16-year-old female with an intra-abdominal cystic lymphangioma. Contrast-enhanced axial (a) and reformatted coronal (b) CT scan of the abdomen shows a lobulated 9.9 x 10.5 x 11.4 cm multi-septated, low attenuation mass compressing the stomach, in the region of the gastrohepatic ligament. Cyst contents are of homogeneously low attenuation and septae are thin. The left gastric artery is shown coursing through the central portion of the mass (solid black arrow). (Protocol: 64-slice, 120 kV, 101 mA, 5 mm slice thickness, intravenous iodixanol)

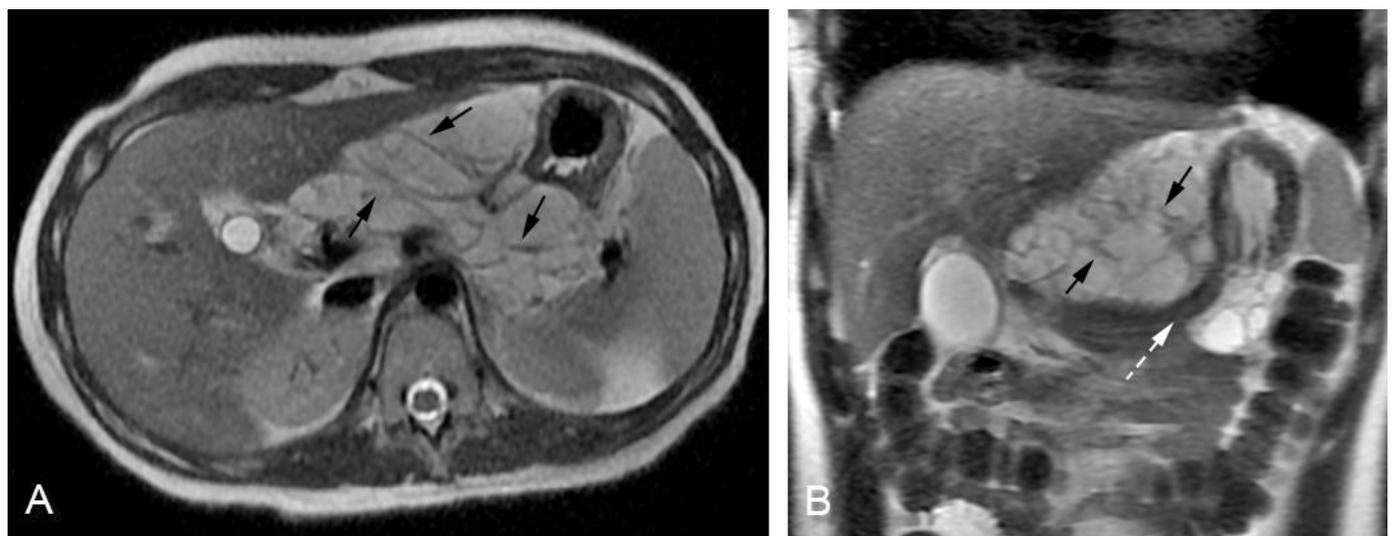


Figure 2: 16-year-old female with an intra-abdominal cystic lymphangioma. T2 weighted single-shot fast spin echo axial (a) and coronal (b) abdominal MRI demonstrates the cystic nature of the 12.8 x 8.9 x 11.3 cm mass with multiple thin septa (solid black arrows) along the lesser curvature of the stomach (broken white arrow). Cyst contents are of uniform hyperintensity. (Protocol: 1.5T, TE: 71.52, TR: 667.5)

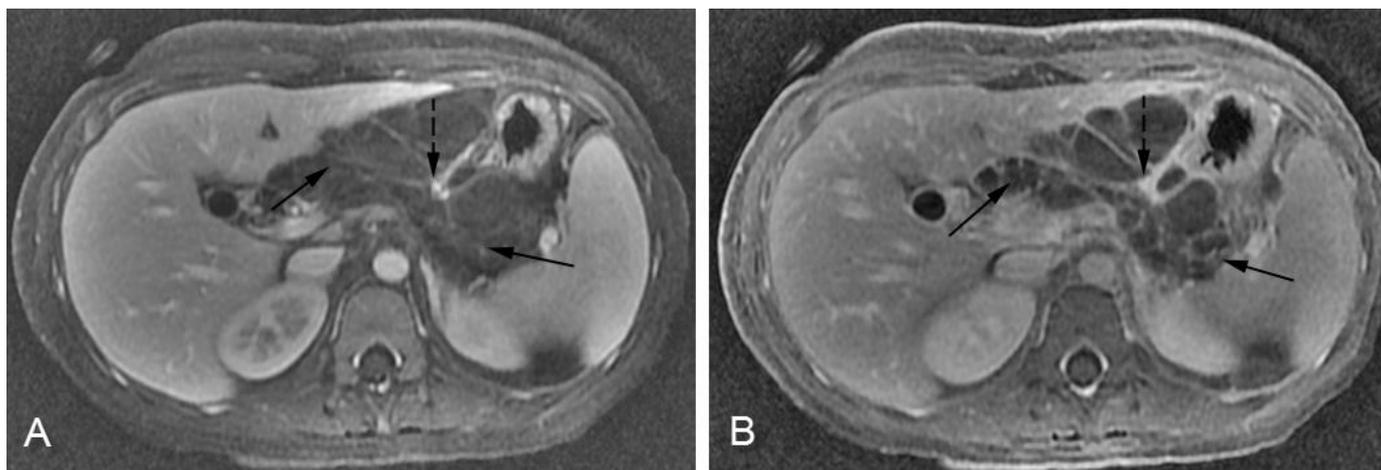


Figure 3: 16-year-old female with an intra-abdominal cystic lymphangioma. Axial contrast enhanced, T1 weighted, fat-suppressed, 3-D spoiled gradient recall echo during the arterial (a) and delayed (b) phase of imaging demonstrates thin enhancing septa (solid black arrows) and the left gastric artery (broken black arrows) in the central portion of the 12.8 x 8.9 x 11.3 cm mass. (Protocol: 1.5T, TE: 1.452, TR: 3.057, 10mL of intravenous gadobenate dimeglumine)

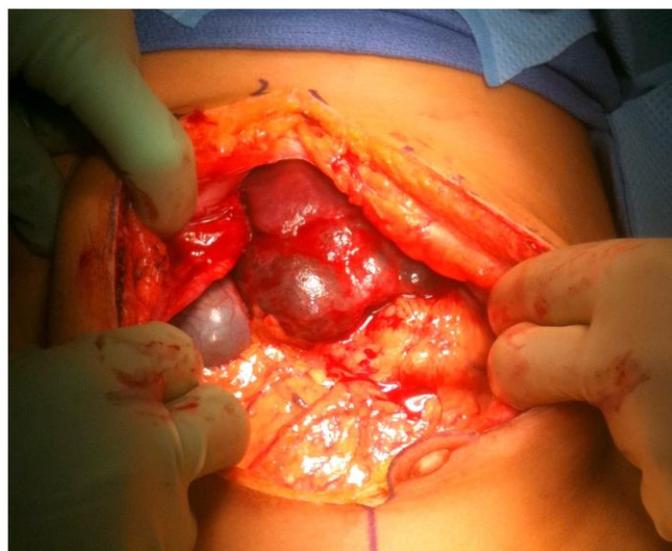


Figure 4: Intra-operative photograph of an intra-abdominal cystic lymphangioma adherent to the gastrohepatic ligament in a 16-year-old female.



Figure 5: 16-year-old female with no evidence of intra-abdominal lymphangioma recurrence. Contrast-enhanced coronal CT at 7 months post-operative reveals normal findings with no evidence of recurrent disease. (Protocol: 64-slice CT, 200 mA, 100 kV, 2.5 mm slice thickness, 2 mL Omnipaque 350 intravenous contrast)

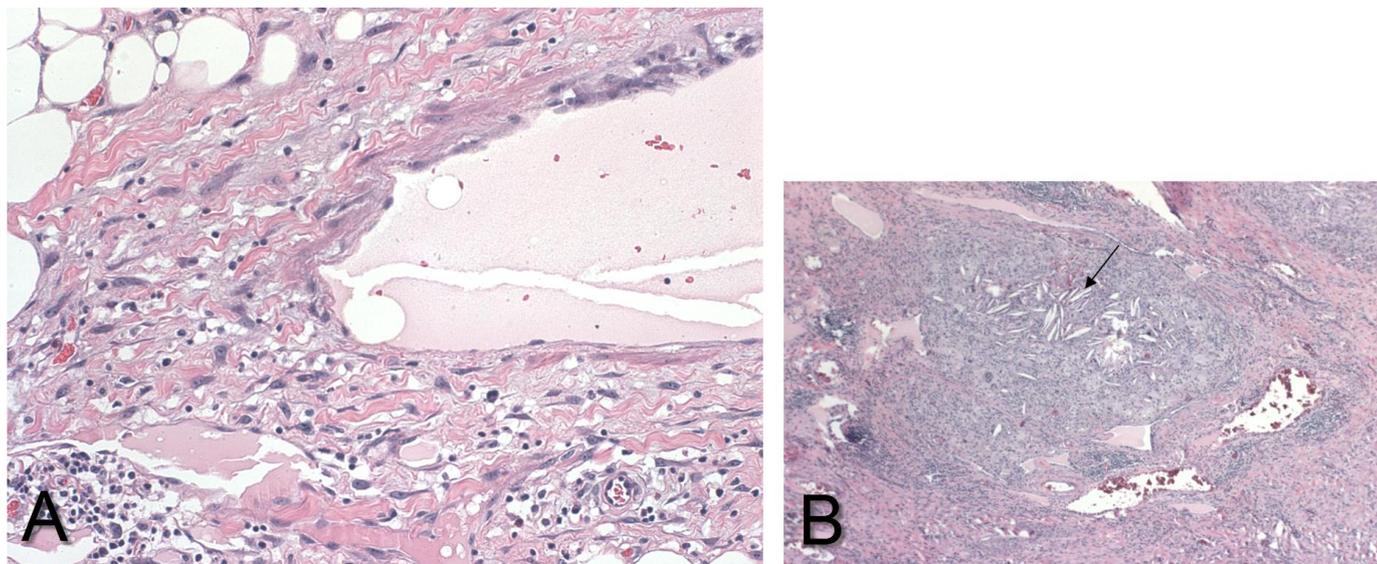


Figure 6: Hematoxylin and Eosin (H&E) stained preparations of an excised intra-abdominal cystic lymphangioma in a 16-year-old female. (a) The mass consisted of variably sized cystic spaces lined by bland flattened endothelial cells and filled with thin, proteinaceous fluid. The cyst walls were notable for myofibroblastic proliferation admixed with adipose and inflammatory cells (40x). (b) Areas of xanthogranulomatous inflammation with cholesterol clefts (solid black arrow) were noted (10x).

Etiology	Proposed mechanisms are anomalous development of lymphatic channels [7] or obstruction of developed lymphatic channels [9]
Incidence	1 in 20,000-250,000 [1]
Gender	Male > Female [16]
Age predilection	> 80% present before age five [4] <20% occur around age 40 [1]
Risk Factors	Chromosomal anomalies [6, 7]
Treatment	Complete surgical excision [4]
Prognosis	Excellent with complete surgical excision
Findings on Imaging	<u>CT</u> - multilocular cystic mass with thin enhancing septa with IV contrast. Cyst contents most commonly show fluid attenuation, hypodensity with chylous contents, or hyperdensity with hemorrhage or infection [2, 14]. <u>MR</u> - similar to CT, typically with cyst content intensity similar to fluid on T1 and T2-weighted images in the absence of hemorrhage or infection [2]. <u>Ultrasound</u> - contents appear anechoic with or without echogenic material [2, 14].

Table 1. Summary table for mesenteric cystic lymphangioma

	CT	MRI	Ultrasound (US)
Lymphangioma	<ul style="list-style-type: none"> Cystic masses with water or fat attenuation (altered if hemorrhagic or infected). Wall and septal enhancement. 	<ul style="list-style-type: none"> Useful to identify mesenteric origin. Typically T1- hypointense T2- hyperintense Altered if hemorrhagic, proteinaceous, or chylous contents. 	<ul style="list-style-type: none"> Cystic, multi-septated, anechoic with or without echoic debris in lobules.
Enteric duplication cyst	<ul style="list-style-type: none"> Cystic mass with enhancing thick wall after contrast injection. Can contain fluid, debris, or blood levels. 	<ul style="list-style-type: none"> Cystic mass with enhancing thick wall after contrast injection. Can contain fluid, debris, or blood levels. 	<ul style="list-style-type: none"> Classic 3 layer appearance with echogenic mucosa, hypoechoic muscular layer and echogenic serosa.
Enteric cyst	<ul style="list-style-type: none"> Fluid-filled mass, no discernable cyst wall. Serous contents give low attenuation. 	<ul style="list-style-type: none"> Follows fluid signal on all pulse sequences. Fluid-filled mass, no discernable cyst wall. Serous contents are hypointense on T1. 	<ul style="list-style-type: none"> Hypoechoic cystic mass with occasional septations.
Mesothelial cyst	<ul style="list-style-type: none"> Fluid-filled mass, no discernable cyst wall or septations. 	<ul style="list-style-type: none"> Follows fluid signal on all pulse sequences. Fluid-filled mass, no discernable cyst wall or septations. Hypointense on T1. 	<ul style="list-style-type: none"> Anechoic mass with acoustic enhancement. No discernable cyst wall or septations.
Pancreatic pseudocyst	<ul style="list-style-type: none"> Thick walled, fluid-filled cyst adjacent to the pancreas in a patient with history of pancreatitis. 	<ul style="list-style-type: none"> Thick walled, fluid-filled cyst with internal debris. T2- hyperintense 	<ul style="list-style-type: none"> Thick walled peripancreatic cyst.
Non-pancreatic pseudocyst	<ul style="list-style-type: none"> Thick walled cyst may have fluid-fluid level from hemorrhage or purulence. Wall and septal enhancement. 	<ul style="list-style-type: none"> Thick walled cyst may have fluid-fluid level from hemorrhage or purulence. Wall and septal enhancement. 	<ul style="list-style-type: none"> Hypoechoic mass with echogenic debris.
Cystic mesothelioma (peritoneal inclusion cyst)	<ul style="list-style-type: none"> Non-calcified multiloculated mass. 	<ul style="list-style-type: none"> Multiloculated mass. T1- hypointense. T2- hyperintense. 	<ul style="list-style-type: none"> Large multicystic mass.
Cystic spindle cell tumor (gastrointestinal leiomyoma or leiomyosarcoma)	<ul style="list-style-type: none"> Mass with irregular walls and high attenuation rim. Enhancing solid components. 	<ul style="list-style-type: none"> Tumor extension and characteristic fluid contents. Enhancing solid components. 	<ul style="list-style-type: none"> Complex mass with echoic debris. May have septa of variable thickness.
Cystic teratoma	<ul style="list-style-type: none"> Variable and mixed composition, often with fluid and fat attenuation and calcifications. 	<ul style="list-style-type: none"> Variable and mixed composition, often with hyperintense fat and proteinaceous material and hypointense serous fluid or calcification on T1-weighted images. 	<ul style="list-style-type: none"> Variable and mixed; anechoic cystic areas, hyperechoic fat, or echogenic calcifications with posterior acoustic shadowing may be seen.

Table 2. Differential diagnosis table for mesenteric cystic lymphangioma

ABBREVIATIONS

CT = Computed Tomography
 MR = Magnetic Resonance
 H&E = Hematoxylin and Eosin
 US = Ultrasound

KEYWORDS

lymphangioma; lesser curvature; cyst; abdominal mass; benign tumor; adolescent

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