

## A study on the significance of abdominal lymphatic malformation

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### Abstract

Lymphatic malformations are congenital vascular malformations with lymphatic differentiation. Although the most common locations for lymphatic malformation are the neck and axilla, they can occur at several locations in the body including the abdomen. The abdominal location is rather rare and accounts for approximately 5% of all lymphatic malformation.

Abdominal lymphatic malformation can arise from mesentery, omentum, gastrointestinal tract, and retroperitoneum. Clinical presentation includes an abdominal lump, vague abdominal discomfort, and secondary complications including intestinal obstruction, volvulus, ischemia, and bleeding. There is a broad spectrum of radiological manifestation. In the present review, we discuss the imaging appearance of abdominal lymphatic malformation. The diagnosis of lymphatic malformation in our series was based on the histopathological examination (in cases who underwent surgery) and fine needle aspiration cytology.

**Keywords:** abdomen, lymphatic malformation, mesentery, retroperitoneum

### Introduction

Lymphatic malformations are a form of congenital vascular malformations. The usual location is head and neck. Abdominal lymphatic malformation are rare, comprising only 5% of all lymphatic malformations. Within the abdominal cavity, they occur in mesentery, retroperitoneum, solid organs (liver, spleen, pancreas) and gastrointestinal tract. Complications including gastrointestinal obstruction, hemorrhage and volvulus mandate treatment in all cases. Surgery remains the treatment of choice. The less invasive form of treatment including per cutaneous sclerotherapy have a greater recurrence rate. In this review we present the imaging spectrum of abdominal lymphatic malformations.

### Embryological Basis

The lymphatic system of the body is derived between 12th and 16th weeks of gestation from endothelial channels located in the neck, root of the mesentery, and the femoral and sciatic vein bifurcation<sup>[1]</sup>. Sequestration of the lymphatic channels leads to the development of lymphatic malformations. Continued growth of these lesions represents both the accumulation of fluid and the proliferation of pre-existing spaces. Histopathological appearance Grossly, lymphatic malformations appear as thin-walled cystic lesions with a smooth external surface.

Cut surface may reveal macroscopic or microscopic cysts<sup>[2]</sup>. The fluid in the cysts may be serous, hemorrhagic, or chylous<sup>[3]</sup>. Microscopically, the cysts are lined by flattened endothelial cells. The cyst fluid is eosinophilic. The stroma shows collagen fibres, lymphocytes, and occasional lymphoid aggregates<sup>[2]</sup>.

### Clinical features

Both male and female predominance has been reported in abdominal lymphatic malformation. The majority of abdominal lymphatic malformations are diagnosed in children. The clinical presentation is highly variable depending on the size and exact location of the lesion<sup>[1]</sup>. They are symptomatic in as many as 88%<sup>[2]</sup>. Presenting features are abdominal pain,

abdominal distension, nausea, vomiting, constipation, diarrhea, and abdominal mass.

### Specific sites

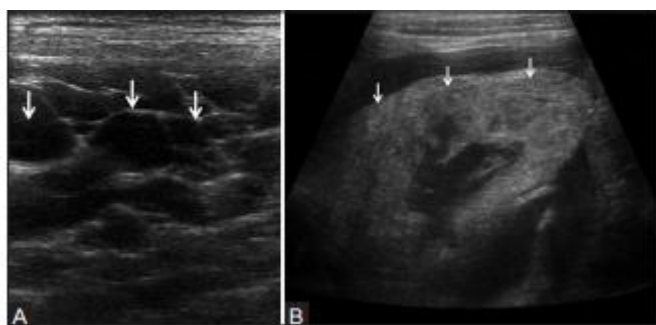
Mesenteric lymphatic malformation the mesentery is the most common site for abdominal lymphatic malformations<sup>[3]</sup>. Mesenteric lymphatic malformations are known to produce serious complications including hemorrhage, intestinal ischemia, obstruction, and volvulus<sup>[4]</sup>.

On ultrasound, lymphatic malformations appear as multilocular cystic masses [Figure 1A]. They are often anechoic but may also contain echogenic debris. Less commonly, the lesions may appear as predominantly solid [Figure 1B]<sup>[5]</sup>. On computed tomography, the fluid component of the lymphatic malformation is homogeneous and shows low attenuation values [Figure 2A]. Following administration of intravenous contrast, enhancement of the cyst wall and septa is seen [Figure 2B]<sup>[5]</sup>. Less common computed tomography (CT) manifestation includes negative attenuation values due to the presence of predominantly chylous fluid<sup>[4]</sup>.

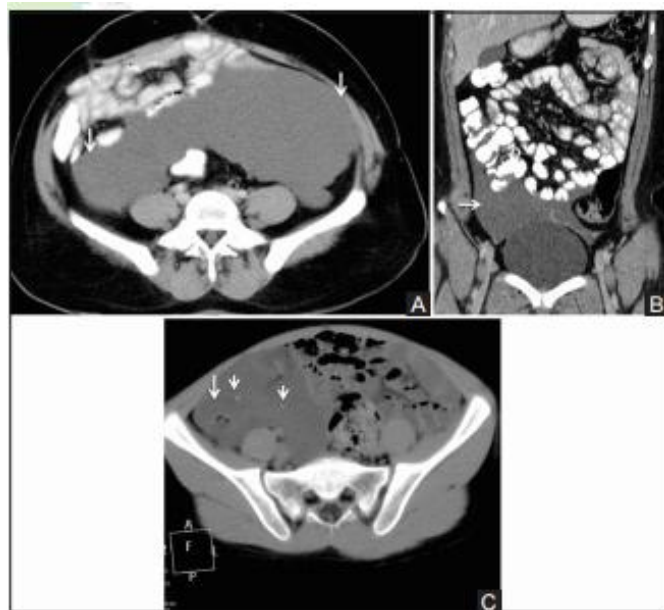
Calcification and hemorrhage are other uncommon imaging findings on CT [Figure 2C]<sup>[6-8]</sup>. Magnetic resonance imaging (MRI) signal intensity parallels that of the fluid appearing hypointense on T1-weighted images and markedly hyperintense on T2-weighted images [Figure 3A]. Heterogeneous signal intensity is seen in the presence of hemorrhage and infection [Figure 3B].

In addition to suggesting a diagnosis of lymphatic malformation, imaging provides information regarding the extent of lesions, aiding in accurate surgical planning<sup>[9]</sup>. Percutaneous biopsy is not recommended in typical cases as there is low cellularity and negative yield in most cases. In addition, in rare cases of cystic malignancy, there is a theoretical risk of needle tract seeding. Fine needle aspiration cytology (FNAC) is a less invasive technique compared to biopsy. It allows confident diagnosis of lymphatic malformation, especially in patients with atypical presentation, age, and location. It is a safe alternative to more cumbersome

and time-consuming surgical modalities of diagnosis [10]. The differential diagnoses of mesenteric lymphatic malformations include uncomplicated ascites (free of septations) and fluid-containing masses in the abdomen [Figure 4] [4, 11]. Ascites is characterized by the lack of septations or displacement of bowel loops. Moreover, ascitic fluid tends to gravitate to the dependent sites including paracolic gutters, Morison's pouch, and pelvis. Mesothelial cysts, enteric duplication cysts, and pseudocysts mimic mesenteric lymphatic malformations closely, and preoperative imaging diagnosis may be impossible. Several other conditions simulating mesenteric lymphatic malformations have been described. These include malignant gynecological lesions (ovarian malignancies) and abdominal tuberculosis. Overall, the features that support lymphatic malformation on imaging include the absence of solid areas, fine septa, low-level septal vascularity, and absence of significant mass effect on adjacent structures.



**Figure 1 (A and B):** Gray scale ultrasound (US) image (A) showing mesenteric lymphatic malformation as multiple anechoic cystic spaces (arrows). US image (B) showing the mesenteric lymphatic malformation as a solid echogenic lesion (arrows). The diagnosis in later case (B) was confirmed at surgery



**Figure 2 (A-C):** Axial contrast-enhanced computed tomography (CECT) image (A) showing mesenteric lymphatic malformation as a low attenuation lesion (arrows). Coronal reformatted CECT image (B) showing a mesenteric lymphatic malformation with peripheral thin contrast enhancement (arrow). Axial non-contrast CT image (C) showing punctate calcific focus (short arrow) within the lymphatic malformation (arrow)

MRI and ultrasound are better at discriminating the cystic nature of the lesions. The role of chemical shift MRI to differentiate mesenteric lymphatic malformations from other cystic masses has also been described [12]. As described above, in certain cases with atypical clinical features, FNAC may aid in diagnosis.

**Retroperitoneal lymphatic malformations**

Retroperitoneal lymphatic malformations are extremely rare and comprise less than 1% of the abdominal lymphatic malformations [13, 14]. They usually present in older children and adults. Clinically, they present as palpable abdominal masses, abdominal pain, intestinal or ureteric obstruction, and hematuria.

Asymptomatic lesions may be detected incidentally during imaging evaluation for unrelated indications. Histologically, the retroperitoneal lesions are almost always of cystic variety [1]. The imaging features are similar to those of cystic mesenteric lymphatic malformations [Figure 5A and B]. An important feature on imaging that differentiates these lesions from other pathologies is the insinuating nature crossing multiple compartments. Retroperitoneal cystic teratoma is an important differential diagnosis [1].

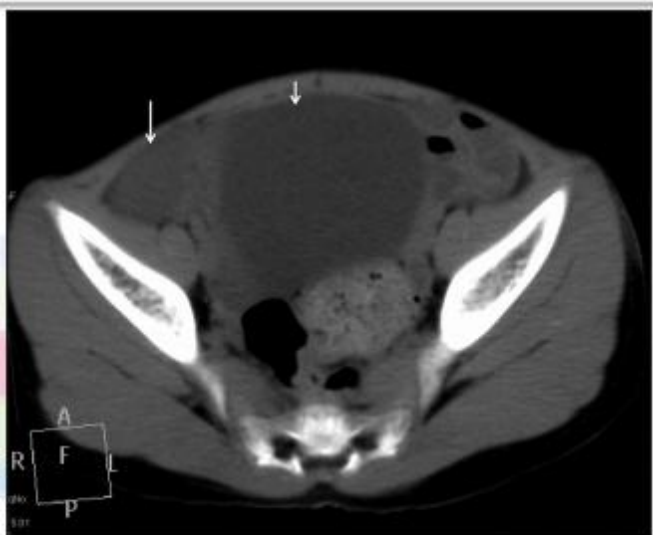
**Gastrointestinal tract lymphatic malformations**

Gastrointestinal tract (GIT) is an infrequent location for abdominal lymphatic malformations [15]. These intramural lesions are frequently asymptomatic and detected incidentally on radiologic studies or at endoscopy [15]. The lesions appear as smoothly margined intramural masses on barium studies and deform with compression. Homogeneous low-attenuation is seen at CT [Figure 6A and B]. Endoscopic ultrasound confirms the cystic nature of these lesions.

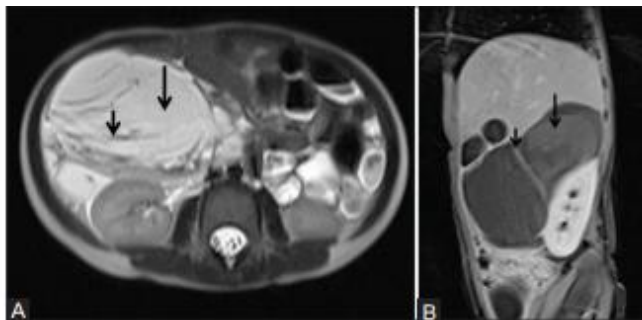
**Hepatic lymphatic malformations**

Primary hepatic lymphatic malformations are extremely uncommon. Hepatic lymphatic malformations may be seen in isolation or as a part of systemic lymphatic malformation or hepatosplenic lymphatic malformation [16]. Isolated hepatic lesions are seen in relatively older children and adults compared to systemic or hepatosplenic lymphatic malformation that usually present during infancy, or early childhood. Hepatic lymphatic malformations present as single or multiple cysts [Figure 7].

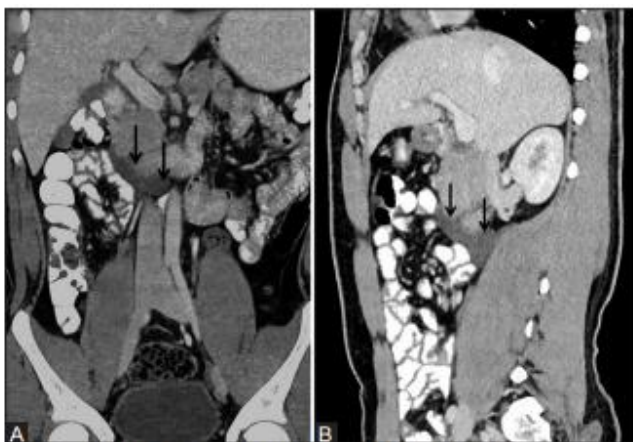
The imaging features are similar to lymphatic malformations elsewhere in the body. Hyperechoic lesions resembling hemangiomas are also seen infrequently [5]. This appearance is related to small size lymphatic spaces separated by abundant fibrous tissue. The differential diagnosis includes other hepatic cysts including polycystic liver disease, hydatid cysts, and cystic tumors including cystic metastasis and mesenchymal hamartoma [5].



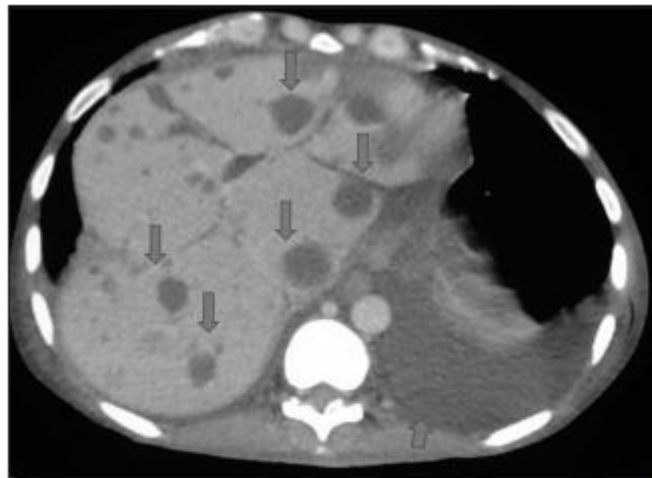
**Figure 4:** Axial non-contrast CT image showing lymphatic malformation as a hypodense lesion (arrow) mimicking ascites. Shorter arrow points toward urinary bladder. Aspiration cytology of the fluid supported the diagnosis of lymphatic malformation



**Figure 5 (A and B):** Axial T2-weighted magnetic resonance (MR) image (A) showing lymphatic malformation in the anterior pararenal space along with fluid component (arrows) and septations (short arrows). Parasagittal T1-weighted postcontrast image (B) shows the lesion to be hypointense (arrows) with enhancing septum (short arrow)



**Figure 6 (A and B):** Coronal (A) and sagittal (B) reformatted images showing duodenal lymphatic malformation as an ill-defined hypodense lesion along the second and third parts of duodenum (arrows). The diagnosis was confirmed following surgery



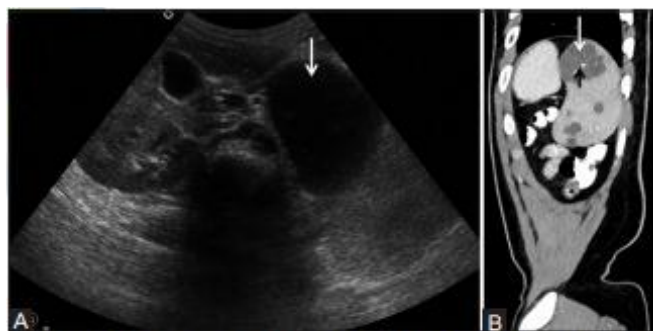
**Figure 7:** Axial computed tomography image shows multiple well-defined hypodense lesions (arrows) in the liver as a part of hepatic lymphatic malformation. Also note left moderate pleural effusion. The diagnosis of lymphatic malformation was supported by aspiration cytology

### Splenic lymphatic malformations

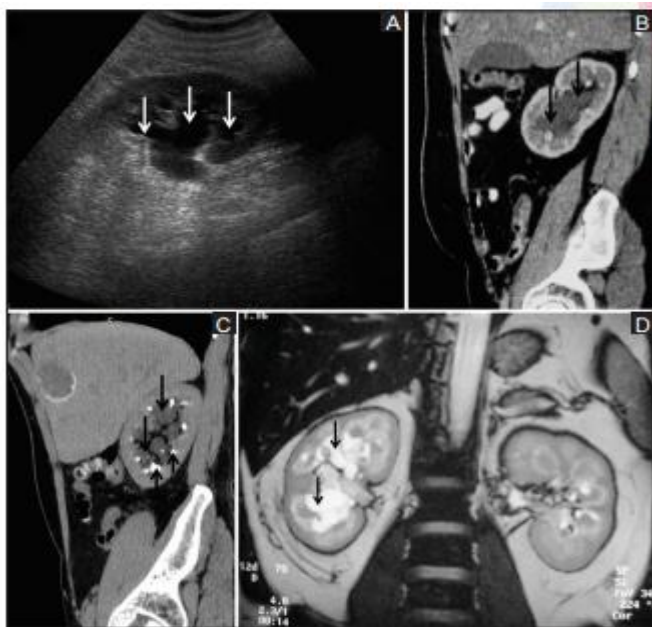
Splenic lymphatic malformations usually occur in subcapsular locations [Figure 8A]. This location reflects the anatomic distribution of lymphatics draining splenic parenchyma. Less common sites for splenic lymphatic malformations include intraparenchymal location presenting as well defined lesions [Figure 8B] or global splenic enlargement [17]. There are no distinct imaging features of splenic lymphatic malformations. Lesions closely resembling splenic lymphatic malformations include true splenic cysts, pseudocysts, infective lesions including pyogenic abscesses and hydatidcysts, infarction, peliosis, and neoplastic lesions including hemangioma, lymphoma, and cystic metastasis [17].

### Lymphatic malformations of extrahepatic biliary tree

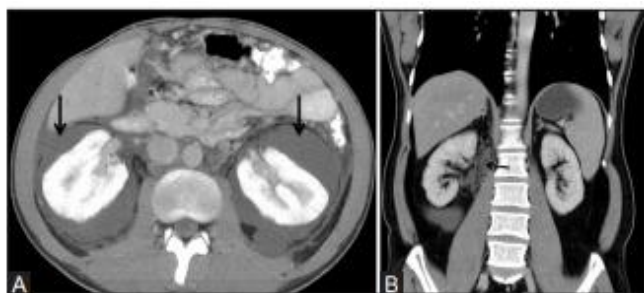
This is a rare site for abdominal lymphatic malformations. These lesions usually arise from the gallbladder and are characterized by multilocular cystic appearance compressing the gallbladder lumen [18].



**Figure 8 (A and B):** Axial gray scale ultrasound image (A) shows a splenic subcapsular lymphatic malformation (arrows). Short arrows in (A) outline the spleen. Sagittal contrast-enhanced computed tomography image (B) shows splenic lymphatic malformation as multiple focal lesions (arrows). Also note punctate calcification (short arrow). The diagnosis in this case was confirmed by fine needle aspiration cytology



**Figure 9 (A-D):** Gray scale ultrasound image shows anechoic structures within the renal sinus (arrows) mimicking hydronephrosis. Sagittal contrast-enhanced computed tomography image (B) of the same patient shows the lesion as hypodense tubular structures (arrows) that were seen distinct from the pelvicalyceal system in the excretory phase (short arrows, C). Also note a well-defined liver lesion with peripheral hyperdense rim suggestive of a liver hydatid. Coronal T2-weighted magnetic resonance image (D) shows the abnormality as hyperintense channels in the right renal sinus (arrows)



**Figure 10 (A and B):** Axial contrast-enhanced computed tomography (CECT) (A) image shows bilateral diffuse perinephric lymphatic malformations (arrows). Coronal CT image (B) shows right perinephric lymphatic malformation (arrows)

### Renal lymphatic malformations

Renal lymphatic malformations are rare lesions and usually present as focal lesions [Figure 9A-D] [19]. Less commonly, these present as diffuse lymphatic malformations or renal lymphangectasia [Figure 10]. They appear as cystic lesions (uni or multilocular) on imaging. Rare cases of solid appearing lymphatic malformations have been reported [7]. The proposed mechanism for such appearance is the numerous interfaces between microcysts of the lymphatic malformation.

Differential diagnosis on imaging in adults includes hydronephrosis, benign renal cysts, renal abscess, and cystic renal tumors including multilocular cystic nephroma and renal cell carcinoma. Multilocular cystic nephroma is the chief differential diagnosis in children.

### Pancreatic lymphatic malformations

Lymphatic malformations are said to be of primary pancreatic

origin only when they are located within the pancreatic parenchyma, are attached to the pancreas by a pedicle, or have a broad area of contact with the pancreas [Figure 11] [20].

The most common cystic lesion in relation to the pancreas is a pseudocyst. Other differential considerations are cystic neoplasms including cystic neuroendocrine neoplasms, mucinous or serous neoplasms, solid pseudopapillary epithelial neoplasms, and intraductal papillary mucinous neoplasms. Definite imaging diagnosis is difficult as these lesions closely resemble lymphatic malformations.

### Treatment

Treatment is recommended in all (even asymptomatic) cases as the incidence of several complications such as bleeding, infection, bowel obstruction, and volvulus tends to increase with time as the lesion size increases [21, 22]. Complete resection has conventionally been advocated as the treatment of choice for all types of abdominal lymphatic malformations. For all sites combined, a recurrence rate of 40% after incomplete resection and 17% after macroscopically complete resection was shown. Moreover, recurrence is more common in larger lesions [23].

Sclerotherapy is the less invasive form of treatment. A variety of sclerosing agents, including ethanol, sodium tetradecylsulfate, bleomycin, doxycycline, and OK-432 (picibanil), has been shown to be effective in the treatment of macrocystic lesions [21]. Variable recurrence has been reported following sclerotherapy, with up to 100% in some series [24, 25]. The rate of recurrence depends on the lesion location, size, and complexity [26, 27].

### Conclusion

Abdominal lymphatic malformations, although rare, should be kept in the differential diagnosis of cystic lesions occurring in the various abdominal compartment and abdominal viscera, both solid and GIT. Clinical presentation is extremely variable. Preoperative diagnosis may be facilitated by FNAC. Definite treatment is required in most cases due to the fear of complications. Surgery is the mainstay of treatment. Other forms of treatment may have a higher recurrence rate.



**Figure 11:** Axial contrast-enhanced computed tomography image shows a well-defined hyperdense lesion (arrow) having a broad area of contact with the distal pancreatic body and tail. Fine needle aspiration cytology of the lesion led to a diagnosis of lymphatic malformation

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