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CASE REPORT

Pancreatic Cystic Lymphangioma Complicated by Haemorrhagic Infarction: A Case Report

Patricia Jarmin Lopez Pua[1], Florence Los Baños[1], Honey Lee Tan[2], Victor Tatco[2]

[1]Institute of Radiology, St Luke's Medical Center, Quezon City, The Philippines
[2]Institute of Surgery, St Luke's Medical Center, Quezon City, The Philippines

INTRODUCTION

Pancreatic cystic lymphangioma or lymphatic malformation in the pancreas is an extremely rare entity resulting from lymphatic flow obstruction. It comprises <1% of all types of lymphatic malformations and about 0.2% of all cystic lesions of the pancreas.[1] Fewer than 100 cases have been reported in the literature.[2] Currently, there are no guidelines for diagnosis and management. Preoperative evaluation and management depend on clinical presentation. Computed tomography (CT), magnetic resonance imaging (MRI), and ultrasonography have been widely used for initial assessment but histopathology remains the standard for diagnostic confirmation.[3]

We report the case of a patient who presented to the emergency department with severe right-sided abdominal pain. Imaging studies revealed a large complex cystic mass with unclear origin. The patient underwent surgery that revealed pancreatic cystic lymphangioma complicated by haemorrhagic infarction.

CASE PRESENTATION

A 43-year-old female presented to the emergency department of our institution in March 2022 with sudden-onset severe right upper abdominal pain. Bedside ultrasound in the emergency room revealed a large cystic abdominal mass. There was no associated jaundice, vomiting, weight loss or altered bowel movement and no history of abdominal surgeries. Physical examination revealed a slightly globular abdomen with epigastric tenderness. Contrast-enhanced CT scan of the abdomen (Figure 1) showed a lobulated cystic mass with thick and incomplete minimally enhancing septations measuring about 10.6 × 13.6 × 17 cm^3 (anteroposterior × transverse × craniocaudal). Minimal surrounding fat stranding densities were seen. The mass was located in the right subhepatic space, close to the duodenum and pancreatic head, exerting a mass effect on the liver and gallbladder, superiorly, and on the right kidney, renal vessels and inferior vena cava, posteriorly. Marked luminal narrowing of the inferior vena cava was evident. There were no enlarged abdominopelvic lymph nodes.

Correspondence: *Dr PJL Pua, Institute of Radiology, St Luke's Medical Center, Quezon City, The Philippines*
Email: patriciajarmin.pua@gmail.com

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Ethics Approval: The study was approved by the St Luke's Institutional Ethics Review Committee, the Philippines (Ref No.: SL-23172). Informed consent for the study and publication was obtained from the patient.

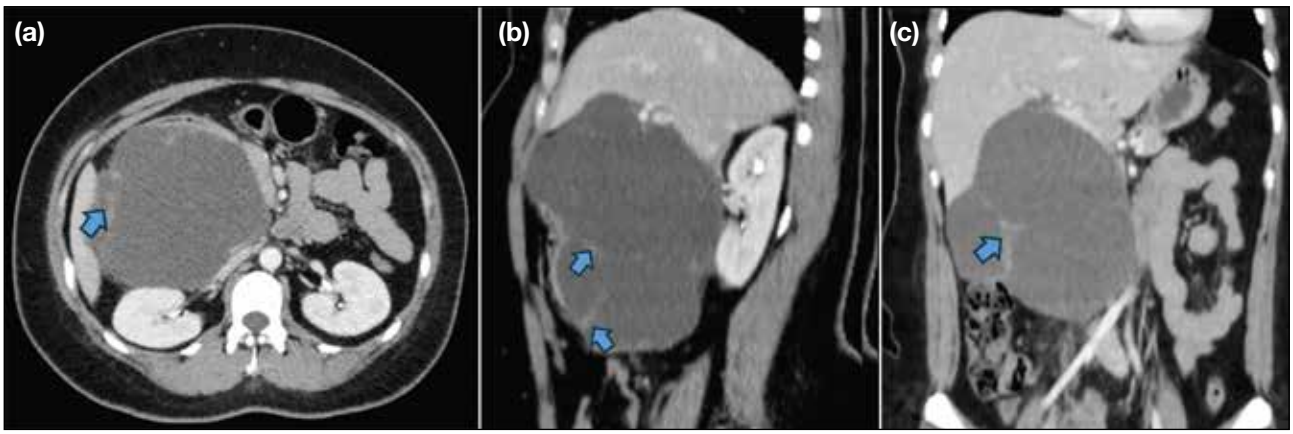

Figure 1. Contrast-enhanced computed tomography scan of the whole abdomen of the patient with a large, lobulated cystic mass lesion with incomplete non-enhancing septations (arrows) seen in the right subhepatic space exerting mass effect on the adjacent structures. (a) Axial view. (b) Sagittal view. (c) Coronal view.

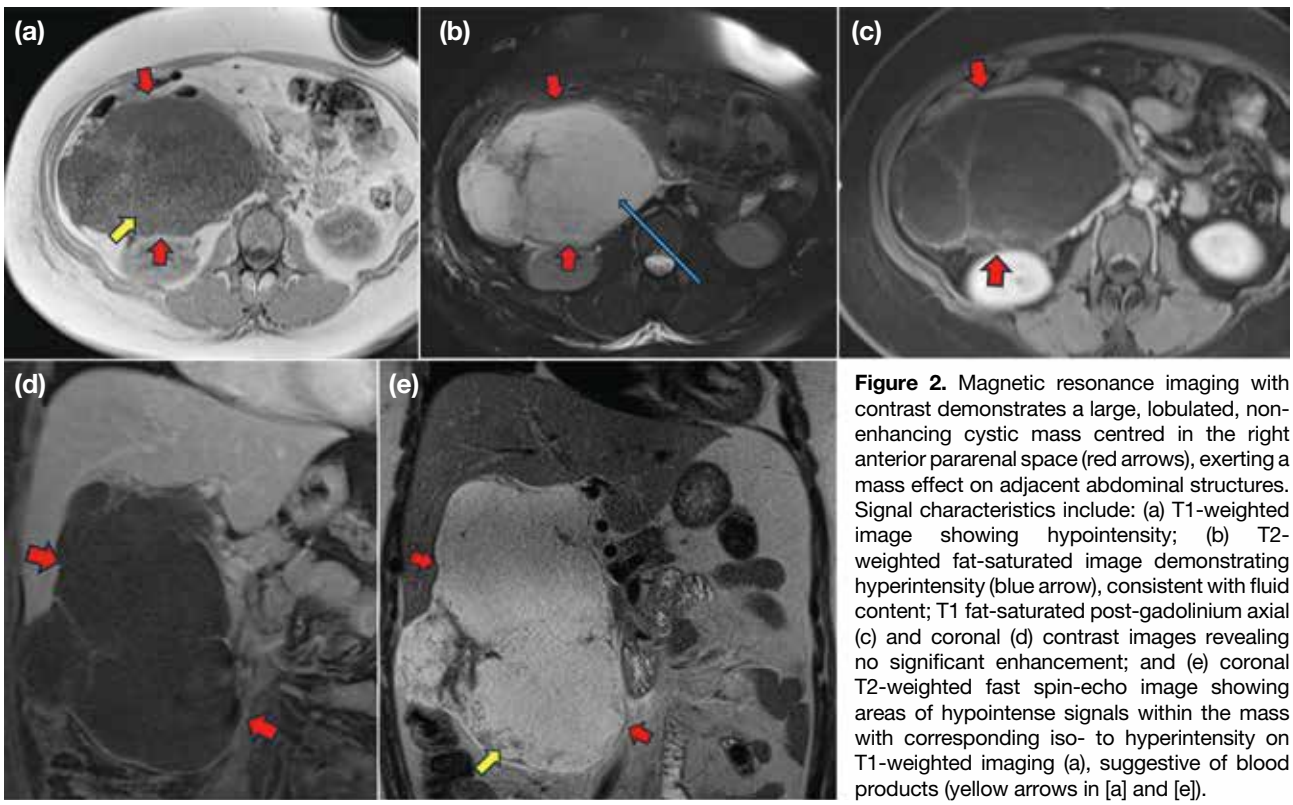

Figure 2. Magnetic resonance imaging with contrast demonstrates a large, lobulated, non-enhancing cystic mass centred in the right anterior pararenal space (red arrows), exerting a mass effect on adjacent abdominal structures. Signal characteristics include: (a) T1-weighted image showing hypointensity; (b) T2-weighted fat-saturated image demonstrating hyperintensity (blue arrow), consistent with fluid content; T1 fat-saturated post-gadolinium axial (c) and coronal (d) contrast images revealing no significant enhancement; and (e) coronal T2-weighted fast spin-echo image showing areas of hypointense signals within the mass with corresponding iso- to hyperintensity on T1-weighted imaging (a), suggestive of blood products (yellow arrows in [a] and [e]).

Differential diagnoses were mesenteric cyst, duplication cyst and cystic neoplasm of the pancreas.

Subsequent magnetic resonance cholangiopancreatography with contrast (Figure 2) of the patient showed a predominantly T2 hyperintense lobulated mass lesion measuring 10.5 × 14 × 17 cm^3 (anteroposterior × transverse × craniocaudal) in the right anterior pararenal space, extending superiorly to the subhepatic region. It impinged on the inferior surface of the liver and displaced the right mesocolic region inferiorly. Some T1-weighted iso- to hyperintense signals with corresponding T2-weighted hypointense signals were also seen, suggestive of blood products. There was

no diffusion restriction abnormality nor significant enhancement. Compression of the adjacent duodenum, pancreatic head/uncinate process, inferior vena cava, right kidney and renal vessels was evident. The pancreatic duct was not dilated. The main portal vein, common hepatic artery and common bile ducts were partially encased but did not appear infiltrated by the mass. There were no enlarged lymph nodes. Mild abdominopelvic ascites was present.

Laboratory examinations showed elevated level of carbohydrate antigen 19-9 at 269.40 U/mL (normal value = 0.00-37.00). Carcinoembryonic antigen level was normal at 1.12 ng/mL (normal value < 2.5). Initial considerations were retroperitoneal lymphatic malformation or mucinous cystic neoplasm of the pancreas.

The patient underwent exploratory laparotomy (Figure 3) that revealed a cystic mass adherent to the first to the third duodenal segments and posterior aspect of the pancreatic head. It measured about 10 × 15 cm^2 (width × length) and displaced the portal vein and superior mesenteric vein anteromedially, the duodenum, pancreatic head, vena

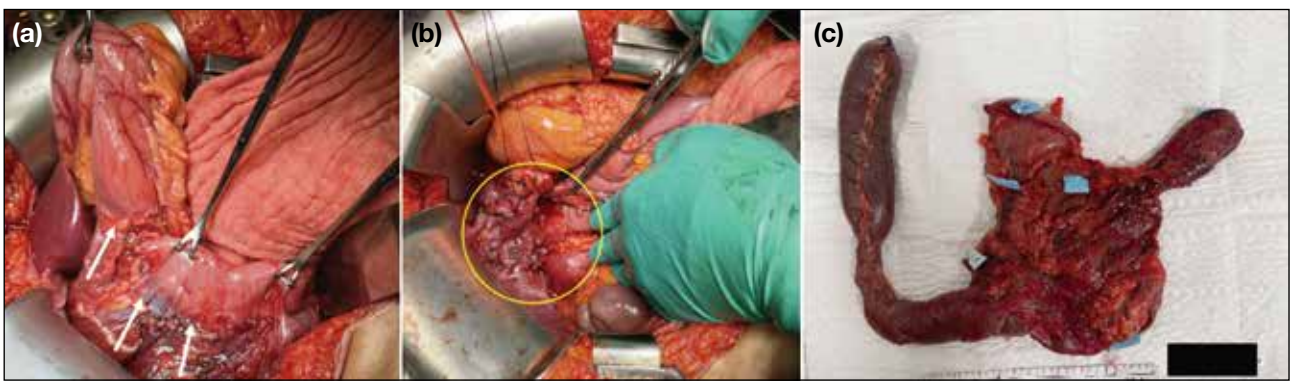

Figure 3. Intraoperative findings showing a large retroduodenal mass. (a) Cystic mass measuring approximately 10 × 15 cm^2 adherent to the first to the third duodenal segments (arrows) and posterior aspect of the pancreatic head (not shown). (b) Cyst wall rupture of the mass (circle). (c) Gross surgical specimen.

Figure 4. Cystic lymphangioma with haemorrhagic infarction. (a) Sectioning of the pancreas and duodenum along the plane of the ampulla of Vater reveals gross adherence of the cystic lesion to the superior and posterior surfaces of the pancreas. (b) Cross-section of the cystic lesion with haematoxylin and eosin stain (×40) shows a cystic lymphangioma (asterisk) with dilated lymphovascular channels, haemorrhage, and necrosis, adjacent to the posterosuperior surface of the pancreas (arrowhead) and peripancreatic soft tissues.

cava, and the right kidney posteriorly. The cystic mass insinuated in between the portal vein and common bile duct.

Surgical rupture of the cyst drained approximately 1 L of dark serosanguineous fluid. Pancreaticoduodenectomy was then performed. Histopathology revealed a pancreatic cystic lymphangioma with haemorrhagic infarction (Figure 4). There was no evidence of dysplasia or malignancy. The patient's postoperative course was unremarkable.

DISCUSSION

Lymphangiomas are benign lymphatic malformations in the pancreas as a result of blockage of lymphatic outflow. Most cases are asymptomatic and an incidental finding on imaging. Nonetheless, the patient may present with symptoms secondary to mass effect such as abdominal pain, discomfort, and a palpable abdominal mass.[3,4] There is a female preponderance with mean age at presentation of 28.9 years.[5] Laboratory tests including tumour markers are usually normal.[5] In our case, there were signs and symptoms of acute abdomen and an elevated level of carbohydrate antigen 19-9, characteristics that may be atypical for pancreatic lymphangioma.

Radiological imaging has a role in preoperative assessment of pancreatic lymphangiomas. These lesions may mimic other pancreatic cystic masses including mucinous cystic neoplasm, pseudocysts and cystic neuroendocrine tumours, making preoperative diagnosis difficult.[6,7] On CT and MRI, pancreatic lymphangiomas appear as encapsulated homogeneous cystic masses, as in our patient. These lesions may show enhancing septations and occasionally contain phleboliths.[7] The presence of T1 hyperintense signals within cystic masses indicates lipid, highly proteinaceous, or haemorrhagic content.[8] In our case, it is interesting to note that the suggestive blood products present within the mass may have been congruent with the haemorrhagic infarction seen intraoperatively. To the best of our knowledge, there have been no prior reported cases of haemorrhagic lymphangiomas arising in the pancreas.

On histopathology, lymphangiomas appear as multilocular cysts representing dilated lymphatic channels containing serous, serosanguineous, or chylous fluid. The cyst walls are lined with endothelial cells and are composed of varying degrees of collagenous connective tissue and smooth muscle.[7]

Endoscopic ultrasound-guided fine needle aspiration may help establish a definitive preoperative diagnosis and enable conservative management of asymptomatic patients.[9] Future studies are encouraged to establish guidelines for management of pancreatic lymphatic malformations.[9]

The treatment of choice of pancreatic lymphangiomas is complete surgical resection accompanied by a low chance of recurrence. Overall prognosis is excellent. Surgery is often required for symptom control or diagnosis.[10] An imaging study on follow-up is an alternative for asymptomatic patients.

CONCLUSION

Although rare, pancreatic lymphangiomas should be included in the differential diagnoses of complex pancreatic cystic lesions, especially in asymptomatic women. Preoperative diagnosis with diagnostic imaging is usually inconclusive but may determine the extent of the mass and its relationship to adjacent structures. A combination of imaging studies and analysis on endoscopic ultrasound-guided fine needle aspiration can provide a definitive preoperative diagnosis.

Since lymphangiomas are considered benign, a conservative approach may be reasonable in asymptomatic patients and stable lesions once tissue diagnosis is confirmed. Nonetheless these lesions may be locally invasive. Radiological imaging, particularly CT and MRI, may play a role in determining complications such as obstruction, rupture, or infection that warrant surgical intervention. Guidelines for appropriate selection of conservative approach versus surgical intervention should be established.

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