Pancreatic Lymphangioma Mimicking Mucinous Cystadenoma: A Report of a Rare Case

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Case report

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Abstract

Background: Pancreatic lymphangioma (PL) is a rare benign neoplasm arising from the lymphatic system. The imaging features of PLs are valuable but not pathognomonic, distinguishing PLs from other pancreatic cystic lesions is still a great challenge.

Case presentation: In our study, we present a 62-year-old woman with PL showing tumor in the distal pancreas, which was misdiagnosed as mucinous cystadenoma preoperatively. Laparoscopic cystectomy was performed after onsite assessments intraoperatively and the recovery was uneventful.

Conclusions: PLs should be included in the differential diagnosis of pancreatic cystic neoplasms and a minimally invasive way for PL treatments is also of great importance. This case will be a good complement for the whole PL cohort.

Background

Lymphangiomas are benign slow-growing neoplasms mostly occurring in the neck and axilla (95%) of the pediatric population[1]. Other onset sites reported in literatures are lung, mediastinum, liver, spleen, colon, omentum, genital organs, and so on[2]. Pancreatic lymphangioma (PL) is extremely rare in clinical practice, accounting for less than 1% of all lymphangiomas and only 0.2% of all pancreatic lesions[3]. It was usually encountered in pancreatic parenchyma or peripancreatic soft tissues with a female predominance[4] and presented as large, solitary, multi- or unilocular lesions. Most PL patients were asymptomatic, their masses were usually detected occasionally in medical checkups or treatments for other unrelated diseases[5]. The diagnosis for PL remains a great challenge without histopathological supports due to the confusion of other cystic neoplasms in pancreas, although radiological studies can offer some implications preoperatively[6].

In our study, we present a female patient with PL which was considered as pancreatic mucinous cystadenoma preoperatively. This will be a good complement for the whole PL cohort.

Case Presentation

A 62-year-old woman was referred to our hospital due to the finding of pancreatic cyst for two years. At the admission, she presented no abdominal symptoms such as abdominal pain, bloating, fever, or jaundice. The physical examination was negative without tenderness, rebound tenderness, or palpable mass. The patient had a history of cholecystectomy, but denied smoking, alcohol abuse, abdominal trauma or pancreatitis. Laboratory investigations (blood routine, biochemical analysis, serum amylase, and tumor markers) were unremarkable. B-ultrasonography revealed a hypoechoic lesion (7.5cm × 6.2 cm) located in the body/tail of the pancreas (Fig. 1A). Enhanced computed tomography (CT) scan was performed and confirmed a homogeneous cystic mass (7.5cm × 7.0 cm) in the same site of the pancreas. The mass was unilocular and well-circumscribed, without mural nodules and calcifications in
the cystic wall (Fig. 1B). The Wirsung duct was normal. No enlarged lymph node and no vessel invasion were observed peripherally. In terms of these imaging features, a diagnosis of pancreatic mucinous cystadenoma was given preoperatively.

Laparoscopic distal pancreatectomy associated with splenectomy was planned preoperatively. During the operation, a cystic lesion (8 cm in diameter) arising from the distal pancreas was detected. The mass was oppressing adjacent organs (stomach, omentum, and retroperitoneal soft tissues) with mild adhesion, and could be detached with a harmonic scalpel easily. The stub of the mass was implanting into the pancreatic parenchyma superficially without invasion of the main pancreatic duct and splenic vessels, which met the preoperative assessments on the CT scan (Fig. 1C). Given these intraoperative situations, radical local resection of the mass was performed preserving the distal pancreas and spleen (Fig. 1D). Grossly, the tumor presented a cystic unilocular appearance (7.8cm × 7.5 cm) with white clear fluids in it. Histopathological examination revealed a dilated cystic structure, within which thin and flat endothelial cells were lining along the lumen, no atypical cells were detected (Fig. 2A). The Immunohistochemical staining displayed CD31(+), D2-40(+), and CK(-) for these endothelial cells (Fig. 2B,2C,2D).

The patient recovered uneventfully, and no pancreatic fistula or other severe complications were detected postoperatively. She discharged home on day 8 after surgery and did not develop recurrence during the follow-up time (41 months).

Discussion

Pancreatic lymphangioma (PL), a rare benign neoplasm of pancreas was first described by Koch in 1931[7]. PL can affect all age groups especially female adults, with an incidence of less than 1% of all lymphangiomas and only accounting for 0.2% of all pancreatic lesions[3,4]. The etiology of PL is still not well defined. Most studies consider that it may originate from congenital malformations of lymphatic channels, which can block the lymphatic flow and induce cystic dilation[8]. While others favor the lymphatic obstruction secondary to inflammations[9]. Patients with PL usually showed a favorable prognosis provided that the mass was radically excised. However, Surlin et al reported that the recurrence can still occur even after complete resection, and the rate is approximately 7%[10]. It is still a great challenge to carry out prospective studies as it shows low morbidity.

In line with our study, most PL patients were asymptomatic, their lesions were usually detected incidentally by medical checkups or in treatments for other unrelated diseases[5]. While others exhibited featureless symptoms (abdominal pain, nausea, vomiting, and palpable mass, etc) depending on the location, size, and mass effects of the lesion[11,12]. PLs are usually manifested as a hypoechoic septate lesion in ultrasound images[13], and on CT scans, they are solitary well-circumscribed cystic masses within which multiple fine septations can be seen. The walls and septations can be mildly enhanced during the arterial phase, however, mural nodules and calcifications are rarely detected in cystic walls[6,13].
Magnetic resonance imaging (MRI) can offer similar imaging traits, but may not have an advantage over CT scans\cite{12}. In our study, the PL lesion was unilocular and not lobulated, presenting an entirely round homogeneous cyst, which was a rarer type of PLs and was easily misdiagnosed as mucinous cystadenoma. Some studies advocated the usefulness of endoscopic ultrasonography and fine-needle aspiration (EUS-FNA) preoperatively\cite{14}. However, in our case, EUS-FNA was not performed since the preconceived misdiagnosis of mucinous cystadenoma via the imaging features. And the operation was planned directly. The definite diagnosis of PL is still a clinical dilemma preoperatively, owing to the rarity of PLs and the confusion of other pancreatic cystic lesions\cite{11}.

Generally, radical resection is considered the first-line treatment for PL patients\cite{15}. The surgical mode including local resection, distal pancreatectomy (associated with splenectomy if necessary), and even Whipple procedure can be selected depending on the tumor site, size, and the relationship with adjacent organs\cite{1}. In our case, given the clear anatomic structure and the low risk of malignancy, we performed local cystectomy under the laparoscopic way, preserving the main pancreatic duct, distal pancreatic parenchyma, and the spleen. Though incomplete excision was supposed to be the major reason for recurrence\cite{16}, we suggest that it is equally of great importance to employ a minimally invasive way after onsite assessments intraoperatively. For patients unsuitable for surgery, EUS-guided drainage might relieve symptoms, but recurrence was inevitable\cite{17}.

Macroscopically, most PLs are multilocular cystic lesions containing turbid yellow or milky white fluids\cite{18}. The size of mass can vary from 3 to 20cm\cite{13}. In our case, the aspirated fluid was white and clear which was close to previous studies. Microscopically, PLs usually consist of dilated septate lymphatic channels lined by flat endothelial cells and smooth muscles in the wall. Lymphocytes can be detected in cyst fluid aspirated from the PL lumen\cite{7,19}. The immunophenotype of PLs can be characterized by VIII-R antigen(+), CD31(+), D2-40(+), and CD34(-), which are the specific and reliable markers for lymphatic endothelium\cite{7,13}. In our study, all the pathological results were in line with the above features and supported the PL diagnosis.

**Conclusions**

PLs are rare benign entities originating from the lymphatic system. Basing on imaging features, definite diagnosis is still difficult to give preoperatively. Though PLs have a low morbidity in clinical practice, they should be taken into consideration for differential diagnosis of pancreatic cystic neoplasms, especially those presenting as unilocular lesions. Radical resection is the first-line treatment for PLs, and in our opinion selecting a minimally invasive way is also of great importance after intraoperative onsite assessments.

**Abbreviations**

PL Pancreatic lymphangioma
CT Computed tomography
MRI Magnetic resonance imaging
EUS Endoscopic ultrasonography
FNA Fine-needle aspiration

**Declarations**

**Availability of data and materials**

All data generated or analyzed during this study are included in this manuscript.

**Ethics approval and consent to participate**

The report was approved by the Ethics Committee of Shaoxing people's Hospital, reference number (2020) Ethics clearance NO (75).

**Competing interests**

The authors declare that they have no competing interests.

**Consent for publication**

Written informed consent for publication of the clinical details and clinical images was obtained from the patient.

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**Authors' contributions**

HQ collected the clinical data and wrote the manuscript; BL did the operation and was the whole treatment plan maker; FL diagnosed the case histologically and helped with the pathological section of this manuscript; HS consulted the relevant literatures to analyze the case and helped in preparation of draft manuscript.
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Figures
Figure 1

Imaging findings of the PL patient. (A) BUS showed a hypoechoic lesion located in the distal pancreas. (B) CT scan confirmed a homogeneous cystic mass in the distal pancreas. (C) The stub of the mass was implanting into the pancreatic parenchyma superficially without invasion of the main pancreatic duct and splenic vessels. (D) The cystic mass was excised completely, preserving the distal pancreas and spleen on the CT scan postoperatively.
Figure 2

Pathological results of the PL patient. (A) Histopathological examination revealed thin and flat endothelial cells were lining along the lumen of the dilated cyst, no atypical cells were detected (under a magnification of 100×). (B) Immunohistochemical staining for CK was negative (under a magnification of 200×). (C) Immunohistochemical staining for CD31 was positive (under a magnification of 200×). (D) Immunohistochemical staining for D2-40 was positive (under a magnification of 200×).