Differential Diagnosis of Cystic Lymphangioma of the Pancreas Based on Imaging Features

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Lymphangioma is a benign tumor, which is a consequence of lymphatic malformation with blockage of lymphatic flow. Most lymphangiomas occur in the neck and axillary region, and < 1% occur in the mesentery or retroperitoneum. Lymphangiomas arising from the pancreas are extremely rare. We report the case of a 34-year-old woman with cystic lymphangioma of the pancreas without major symptoms or signs. A 6 × 6 cm intra-abdominal cystic mass was incidentally revealed by sonography during a health examination. It is always a challenge to differentiate the lesion from other possible cystic-like pancreatic neoplasms. Differential diagnosis of cystic lymphangioma from other cystic-like tumors of the pancreas can be performed based on their imaging characteristics, including presence of septa, cystic or wall calcification, soft tissue, wall thickness, single or multiple loculation, and dilatation of the pancreatic duct. Post-gadolinium magnetic resonance imaging is excellent in defining the origin of intra-abdominal cystic mass and intracystic septa. [J Formos Med Assoc 2006;105(6):512–517]

Key Words: cystic-like neoplasm, cystic lymphangioma, pancreas

The availability of medical imaging modalities such as sonography, computed tomography (CT) and magnetic resonance imaging (MRI) has led to the increasing discovery of cystic lesions of the pancreas in asymptomatic patients. Although only a minority of pancreatic cystic tumors are true neoplasms, proper diagnosis and accurate staging is important for early treatment and to increase the length of survival in patients.1 In the majority of cases of cystic pancreatic tumor, the differential diagnosis includes non-neoplastic cystic lesions such as pseudocysts,3 and neoplastic cystic lesions such as serous cystadenomas, mucinous cystic neoplasms, nonfunctioning islet cell tumors with cystic degeneration, and cystic solid and papillary epithelial neoplasms. Malignant cystic tumors are rare and comprise only about 1% of all pancreatic malignancies.3 They are sometimes potentially curable by surgery. Many benign pancreatic cystic neoplasms are considered premalignant and should be resected. Pseudocysts do not always require resection, and many may be treated with observation, internal drainage or other methods. Correct diagnosis of these lesions is essential to determining appropriate treatment. We report a case of cystic pancreatic lymphangioma, including its preoperative diagnosis and treatment.

Case Report

A 34-year-old woman with no history of systemic disease was referred to our hospital because of the incidental sonographic finding of an intra-abdominal cyst during a health check-up. According to the admission note, there was no abdomi-
Cystic lymphangioma of the pancreas

with characteristic appearance of low signal intensity on T1WI and high signal intensity on T2WI without post-gadolinium enhancement. Unlike the CT study, MRI in the coronal view of T2WI could distinguish a clear border from the stomach. MRI also revealed some thin curvilinear septations separating cystic areas on T2WI and post-gadolinium T1WI (Figure 1B,C). Since both the imaging and laboratory studies did not favor a pseudocyst or advanced malignancy, surgical exploration was arranged. Intraoperatively, the lesion was found to originate from the pancreatic tail and to be composed of multiloculated cystic spaces and thin transparent walls. Some clear fluid content was found within the cystic spaces (Figure 1D). Microscopically, the resected cyst showed dilated lymphatics with flattened lining cells associated with lymph-like fluid content, focal lymphocytic infiltration and supporting fibrous stroma (Figure 2). Immunohistochemical studies of the lining cells

![Figure 1](image-url)

**Figure 1.** (A) Enhanced computed tomography reveals a thin encapsulated intraperitoneal cyst continuous with pancreatic tail. (B) T1-weighted (TR = 120 ms, TE = 2.2 ms) axial magnetic resonance imaging (MRI) reinforces our imaging diagnosis of a pancreatic tail cystic mass (homogeneous hypointense appearance). (C) Gadolinium-enhanced axial MRI shows a hyperintense thin septum (arrows) within the cystic mass. (D) Grossly, the major cyst is composed of multiloculated cystic spaces with transparent thin wall and thin septum (arrows), which corresponds to the findings in (C).
atic cystic mass but are unable to classify the type of cystic lesions. Our review of the literature suggested that the imaging characteristics of cystic lymphangioma may be useful in the differential diagnosis from other cystic tumors of the pancreas (Table 2). Resembling cystic lymphangioma, serous adenoma has multiple cysts and very thin septa, some of them contain a central stellate scar and sunburst calcification. Scott et al reported that about 75% of mucinous cystadenomas are unilocular or multilocular with thin septa. In 66% of mucinous cystadenocarcinomas, the lesions are multilocular with thick walls and solid excrescences. Overall, 62% of mucinous cystic neoplasms of the pancreas exhibit cyst wall calcification, focal thickening of the cyst wall and papillary projections. The imaging characteristics of cystic-like neoplasms of the pancreas are summarized in Table 2.

Differential diagnosis using serum tumor markers may also be useful, because mucinous cystic neoplasms of the pancreas are immunoreactive for CEA and CA19-9. Most cases of nonfunctioning islet cell tumors with cystic degeneration have a thick wall and an irregular inner surface. Over 90% of cystic solid and papillary epithelial neoplasms were associated with dilatation of the pancreatic duct. Patients with pseudocysts often have a history of acute or chronic pancreatitis, while the majority of other cystic tumors may lack such an antecedent factor. The radiographic characteristics of pseudocysts are usually unilocular and there is associated pancreatic duct dilatation. In addition, pseudocysts have a different appearance from cystic lymphangiomas in terms of CT findings, with absence of septa, loculation, solid component, and cyst wall calcification. Initially, medical imaging modalities, sonography or spiral CT may have the advantages of lower cost and shorter acquisition time, in addition to higher sensitivity to cystic calcification. However, this case suggests the advantage of post-gadolinium MRI studies to define the thin septa, which contrast-enhanced CT could not demonstrate. In addition, the coronal and sagittal views of MRI could further distinguish the structural interface with other

Figure 2. Histologic examination shows dilated lymphatics and lymph-like fluid content (hematoxylin & eosin, x 200).

using a Factor VIII-R antigen test were positive. The pathologic diagnosis was cystic lymphangioma.

Discussion

Lymphangiomas are benign lymphatic malformations that lead to blockage of lymphatic flow and lymphangiectasia. Gui et al described these sequestered lymphatic channels as a developmental abnormality rather than a true neoplasm. Most lymphangiomas occur in the neck and axillary region. Abdominal lymphangiomas are rare, accounting for < 1% of all lymphangiomas, and mainly occur in the mesentery and retroperitoneum. Our review of the literature from 1995 to 2005 revealed only a few cases of pancreatic lymphangioma (Table 1). Most reported cases, including the present case, were cystic type lymphangioma. One case of cavernous lymphangioma was reported. Pancreatic lymphangioma occurs predominantly in females, with a mean age of 28.9 years at initial presentation.

The initial clinical symptoms are variable and may include abdominal pain, nausea, vomiting, and a palpable abdominal mass. In some cases, however, the cysts are symptomless and discovered as an incidental finding, as in the present case. No specific or significant laboratory abnormalities have been reported. Previous reports of pancreatic lymphangioma have suggested that imaging studies including CT and MRI could confirm a pancreatic cystic mass but are unable to classify the type of cystic lesions. Our review of the literature suggested that the imaging characteristics of cystic lymphangioma may be useful in the differential diagnosis from other cystic tumors of the pancreas (Table 2). Resembling cystic lymphangioma, serous adenoma has multiple cysts and very thin septa, some of them contain a central stellate scar and sunburst calcification. Scott et al reported that about 75% of mucinous cystadenomas are unilocular or multilocular with thin septa. In 66% of mucinous cystadenocarcinomas, the lesions are multilocular with thick walls and solid excrescences. Overall, 62% of mucinous cystic neoplasms of the pancreas exhibit cyst wall calcification, focal thickening of the cyst wall and papillary projections. The imaging characteristics of cystic-like neoplasms of the pancreas are summarized in Table 2.

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adjacent organs in the cystic lesion, which was too large to define on axial spiral CT. MRI is superior to CT in ruling out communication between cyst and pancreatic duct \(^{14,15}\) (Table 2). The imaging features of pancreatic lymphangioma can be used systematically to eliminate the other possible cystic-like tumors of the pancreas.

**References**

<p>| Table 2. Imaging characteristics of other cystic-like neoplasms of the pancreas 14-16 |
|---------------------------------------------|---------------------------------|---------------------------------|---------------------------------|---------------------------------|---------------------------------|</p>
<table>
<thead>
<tr>
<th>Common imaging findings</th>
<th>Cystic wall calcification</th>
<th>Calcification or central scar</th>
<th>Focal thickening of cyst wall (&gt; 3 mm)</th>
<th>Dilatation of pancreatic duct</th>
<th>Communication between cyst and pancreatic duct</th>
<th>Soft tissue component or papillary projection</th>
<th>Presence of septa</th>
<th>Loculation</th>
<th>Higher incidence of pancreatic location</th>
</tr>
</thead>
<tbody>
<tr>
<td>Other cystic pancreatic tumor</td>
<td></td>
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<td></td>
<td></td>
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<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Serous adenoma</td>
<td>Less frequent</td>
<td>More frequent</td>
<td>Less frequent</td>
<td>Less frequent</td>
<td>Rare</td>
<td>Less frequent</td>
<td>Non prominent or appear as delicate fibrous septum</td>
<td>Multiloculate (more frequent)</td>
<td>More in body and tail</td>
</tr>
<tr>
<td>Mucinous cystic neoplasm</td>
<td>More frequent</td>
<td>Less frequent</td>
<td>More frequent</td>
<td>Less frequent</td>
<td>More frequent</td>
<td>Less frequent</td>
<td>More (thick septum)</td>
<td>Uniloculate (more frequent)</td>
<td>Predominantly in head</td>
</tr>
<tr>
<td>Nonfunctioning islet cell tumor</td>
<td>Frequent</td>
<td>More frequent</td>
<td>Variable</td>
<td>Absent</td>
<td>More frequent</td>
<td>Variable</td>
<td>Variable</td>
<td>Variable</td>
<td>Head:body and tail (1:1)</td>
</tr>
<tr>
<td>Cystic solid and papillary epithelial neoplasm with cystic degeneration</td>
<td>Not frequent</td>
<td>Not frequent</td>
<td>More frequent</td>
<td>May or usually present</td>
<td>More frequent</td>
<td>Less frequent</td>
<td>Variable</td>
<td>Variable</td>
<td>Predominantly in tail</td>
</tr>
<tr>
<td>Pseudozyst</td>
<td>Less frequent</td>
<td>Less frequent</td>
<td>More frequent</td>
<td>More frequent</td>
<td>Usually present</td>
<td>May mimic by presence of necrotic pancreatic or peripancreatic debris or blood</td>
<td>Less frequent</td>
<td>Uniloculate</td>
<td>Predominantly in body and tail (extra-pancreatic cysts)</td>
</tr>
<tr>
<td>Cystic lympathan-gioma (according to our 14 review cases)</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>3</td>
<td>12</td>
<td>Multiloculate (13)</td>
<td>Uniloculate (1)</td>
<td>Head (3) Body-tail (9) Extra-lobular connective tissue of pancreas (1)</td>
</tr>
</tbody>
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