Epithelial Cyst Arising in an Intrapancreatic Accessory Spleen: A Diagnostic Dilemma

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Abstract

An epithelial cyst in an intrapancreatic accessory spleen (ECIAS) is a rare disease that is commonly misdiagnosed preoperatively. To identify the clinical and imaging features of ECIAS, we reviewed the relevant medical literature. Twenty-one cases of ECIAS were identified, including our own. The cases were mainly diagnosed as mucinous cystic neoplasm (MCN) preoperatively based on clinical and imaging features, such as, a woman in middle age; elevation of serum CA19-9 levels; location in the tail of the pancreas; and a solid component resembling a mural nodule. ECIAS is another lesion to be considered in the differential diagnosis of MCN.

Key words: epithelial cyst, intrapancreatic accessory spleen, endoscopic ultrasound, mucinous cystic neoplasm, differential diagnosis


Introduction

Although the occurrence of an accessory spleen is not uncommon, an epithelial cyst in an intrapancreatic accessory spleen (ECIAS) is a rare disease and only a few case reports describe its clinical features. To date, 20 case reports of ECIAS have been published in English (1-18). The first case, reported by Davidson et al (1) in 1980 and other subsequent case reports, described the difficulty in discerning these lesions preoperatively from potentially malignant cystic neoplasms of the pancreas such as mucinous cystic neoplasm (MCN), intraductal papillary mucinous neoplasm, serous cystic neoplasm, or solid pseudopapillary neoplasm. There is no consensus on the clinical and imaging features of ECIAS. However, it is crucial to differentiate a case of ECIAS from these cystic neoplasms using various imaging modalities, as surgical management should be avoided if the ECIAS is small and asymptomatic.

Herein we report a case of a preoperative MCN diagnosis originating from the tail of the pancreas and characterized by a thick cystic wall and solid component resembling a mural nodule on endoscopic ultrasonography (EUS). However, the postoperative diagnosis was ECIAS, indicating the level of difficulty in a preoperative diagnosis of this disease. The clinical and imaging features of ECIAS are thus reviewed and discussed.

Methods

We conducted a MEDLINE review of the relevant medical literature published from January 1960 to December 2009 and identified all reported cases of ECIAS using the following key words: epidermoid cyst and pancreas, epithelial cyst and pancreas, and intrapancreatic accessory spleen. Relevant publications were evaluated and demographic data

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Received for publication February 17, 2011; Accepted for publication May 22, 2011
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as well as findings on imaging and preoperative diagnoses were collected. This collective information, which also includes the present patient, is summarized in Table 1.

#### Case Report

A 55-year-old asymptomatic Japanese woman with no history of alcohol consumption, trauma, or pancreatitis was admitted for further investigation of a cystic lesion in the tail of the pancreas that had been detected upon abdominal ultrasonography during a medical checkup. The physical examination was not significant. Hematology, bilirubin levels, transaminases, and pancreatic enzymes were normal. Among tumor markers, carbohydrate antigen 19-9 (CA19-9) in the sera was slightly elevated at 90 U/mL (normal, <37 U/mL), whereas carcinoembryonic antigen, Duke pancreatic monoclonal antigen type 2, and s-pancreas-1 antigen levels were within normal limits. Abdominal ultrasound as well as plain computed tomography (CT) revealed an approximately 3.0×1.5 cm, well-demarcated cystic lesion in the tail of the pancreas that was not adjacent to the spleen (Fig. 1a). A contrast-enhanced CT identified no solid component in the cystic lesion, and the wall of the cyst was relatively thick but not enhanced (Fig. 1b). On magnetic resonance images (MRI), the cystic component showed slightly high intensity and strongly high intensity on T1- and T2-weighted images, respectively (Fig. 2a, b). A 5.8-mm solid component suggesting a mural nodule in the cyst was detected on EUS (Fig. 3). A dilation of the main pancreatic duct and communication with the cyst were not observed on endoscopic retrograde pancreatoscopy (ERP).

The patient underwent a distal pancreatectomy with a preoperative diagnosis of MCN based on the characteristic features: a woman in middle age; a mural nodule detected on EUS; location of the cyst in the tail of the pancreas; and elevation of serum CA19-9 levels. The surgical specimen showed a well-demarcated cystic lesion located in the tail of pancreas, measuring up to 3.3 cm in diameter. The cyst was

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### Table 1. Reported Cases of an Epithelial Cyst Arising in the Intrapancreatic Accessory Spleen (n=21)

<table>
<thead>
<tr>
<th>Study Author</th>
<th>Age / sex</th>
<th>Symptom</th>
<th>Serum CA 19-9 (U/mL)</th>
<th>Country</th>
<th>Location</th>
<th>Size</th>
<th>Locular type</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Davidson et al.</td>
<td>40/M</td>
<td>Weight loss, nausea</td>
<td>Nil</td>
<td>United States</td>
<td>Tail</td>
<td>7 × 3 × 2.5 cm</td>
<td>multilocular</td>
</tr>
<tr>
<td>2. Morokoshi et al.</td>
<td>32/F</td>
<td>Left abdominal pain</td>
<td>WNL</td>
<td>Japan</td>
<td>Tail</td>
<td>6 × 5 cm</td>
<td>unilocular</td>
</tr>
<tr>
<td>3. Nakae et al.</td>
<td>37/F</td>
<td>Epigastric pain</td>
<td>Nil</td>
<td>Japan</td>
<td>Tail</td>
<td>6.5 × 6.5 × 4.5 cm</td>
<td>unilocular</td>
</tr>
<tr>
<td>4. Tang et al.</td>
<td>38/M</td>
<td>Asymptomatic</td>
<td>WNL</td>
<td>Japan</td>
<td>Tail</td>
<td>2.3 × 2.1 cm</td>
<td>multilocular</td>
</tr>
<tr>
<td>5. Furukawa et al.</td>
<td>45/M</td>
<td>Asymptomatic</td>
<td>WNL</td>
<td>Japan</td>
<td>Tail</td>
<td>2 cm</td>
<td>multilocular</td>
</tr>
<tr>
<td>6. Higaki et al.</td>
<td>46/F</td>
<td>Left back pain</td>
<td>201</td>
<td>Japan</td>
<td>Tail</td>
<td>3 × 3 cm</td>
<td>multilocular</td>
</tr>
<tr>
<td>7. Sato et al.</td>
<td>49/F</td>
<td>Asymptomatic</td>
<td>WNL</td>
<td>Japan</td>
<td>Tail</td>
<td>4.3 × 2.6 cm</td>
<td>multilocular</td>
</tr>
<tr>
<td>8. Choi et al.</td>
<td>54/F</td>
<td>Epigastric discomfort</td>
<td>Nil</td>
<td>Korea</td>
<td>Tail</td>
<td>15 × 11 cm</td>
<td>multilocular</td>
</tr>
<tr>
<td>9. Tsuchimoto et al.</td>
<td>51/M</td>
<td>Asymptomatic</td>
<td>WNL</td>
<td>Japan</td>
<td>Tail</td>
<td>2.5 × 2.0 cm</td>
<td>multilocular</td>
</tr>
<tr>
<td>10. Hori et al.</td>
<td>48/M</td>
<td>Asymptomatic</td>
<td>WNL</td>
<td>Japan</td>
<td>Tail</td>
<td>2 × 1 cm</td>
<td>multilocular</td>
</tr>
<tr>
<td>11. Sonomura et al.</td>
<td>45/F</td>
<td>Epigastric discomfort</td>
<td>159</td>
<td>Japan</td>
<td>Tail</td>
<td>3.5 cm³</td>
<td>multilocular</td>
</tr>
<tr>
<td>12. Kanazawa et al.</td>
<td>58/F</td>
<td>Asymptomatic</td>
<td>WNL</td>
<td>Japan</td>
<td>Tail</td>
<td>2.5 cm</td>
<td>multilocular</td>
</tr>
<tr>
<td>13. Ru et al.</td>
<td>41/M</td>
<td>Asymptomatic</td>
<td>Nil</td>
<td>United States</td>
<td>Tail</td>
<td>2.5 cm</td>
<td>Nil</td>
</tr>
<tr>
<td>14. Servais et al.</td>
<td>52/F</td>
<td>Asymptomatic</td>
<td>Nil</td>
<td>United States</td>
<td>Tail</td>
<td>11.5 × 10.5 × 6.5 cm</td>
<td>Nil</td>
</tr>
<tr>
<td>15. Itano et al.</td>
<td>40/M</td>
<td>Asymptomatic</td>
<td>WNL</td>
<td>Japan</td>
<td>Tail</td>
<td>4.0 × 3.2 × 3.0 cm</td>
<td>unilocular</td>
</tr>
<tr>
<td>16. Zhang et al.</td>
<td>28/F</td>
<td>Asymptomatic</td>
<td>WNL</td>
<td>China</td>
<td>Tail</td>
<td>2.5 × 2.5 cm</td>
<td>unilocular</td>
</tr>
<tr>
<td>17. Kadota</td>
<td>57/F</td>
<td>Asymptomatic</td>
<td>WNL</td>
<td>Japan</td>
<td>Tail</td>
<td>6.0 × 5.0 × 4.0 cm</td>
<td>unilocular</td>
</tr>
<tr>
<td>18. Kadota</td>
<td>70/F</td>
<td>Asymptomatic</td>
<td>48</td>
<td>Japan</td>
<td>Tail</td>
<td>1.7 × 1.0 × 0.8 cm</td>
<td>unilocular</td>
</tr>
<tr>
<td>19. Kadota</td>
<td>37/M</td>
<td>Asymptomatic</td>
<td>647</td>
<td>Japan</td>
<td>Tail</td>
<td>10.0 × 7.0 × 7.0 cm</td>
<td>unilocular</td>
</tr>
<tr>
<td>20. Itano</td>
<td>67/M</td>
<td>Epigastric pain, weight loss</td>
<td>182</td>
<td>Japan</td>
<td>Tail</td>
<td>1.5 × 1.5 × 1.2 cm</td>
<td>unilocular</td>
</tr>
<tr>
<td>21. Our patient</td>
<td>55/F</td>
<td>Asymptomatic</td>
<td>90</td>
<td>Japan</td>
<td>Tail</td>
<td>2.5 × 1.5 cm</td>
<td>unilocular</td>
</tr>
</tbody>
</table>

NI, no information; WNL, within normal limits.

* measured on the CT image, † measured on the AUS image.
Figure 2. Magnetic resonance images revealed that the cystic component was slightly hyperintense on T1-weighted MRI (a) and hyperintense on T2-weighted MRI (b).

Figure 3. Endoscopic ultrasound demonstrated a cystic lesion (C) with a solid component suggesting a mural nodule (arrow) in the tail of the pancreas.

unilocular without a mural nodule (Fig. 4a). Histologically, a fibrotic band with sclerosis lay under the epithelial lining and included splenic tissue with typical red and white pulp. The cystic wall was covered with stratified squamous cells without hair and skin appendages (Fig. 4b). Ovarian-type stroma was not identified. The histologic diagnosis was ECIAS due to the absence of differentiation into dermoid in the cystic epithelium and infiltration of lymphocytes into the cystic wall. The solid component that was considered a mural nodule by EUS, of which the size and shape was compatible with postoperative pathological findings, was a thick fibrotic band with hyaline degeneration and splenic tissue (Fig. 4a, c, d). Immunohistochemically, CA19-9 was positive in the monolayer and surface layer of the squamous epithelium (Fig. 4e). After the operation the serum CA19-9 levels were decreased to normal. The patient’s postoperative course was uneventful and she has been doing well after two years of follow-up.

Literature Review

Clinical features of patients with ECIAS

Twenty-one cases of ECIAS including the present case were identified in the literature: 18 from Asia and 3 from the United States. The mean age at onset of all of the patients was 47, and the sex ratio was 8 males to 13 females. Although 14 (67%) patients were asymptomatic, the pancreatic tumors in all of the patients were noted during a medical checkup. Seven patients had various symptoms including abdominal pain or discomfort (n=5); weight loss (n=2); nausea (n=1); and left back pain (n=1). Serum CA19-9 levels were elevated in 8 of 16 cases. Of the 19 cases that described preoperative diagnoses, only 2 (15, 18) gave a precise diagnosis of ECIAS. The remaining diagnoses were as follows: MCN, including a mucin-producing pancreatic tumor and cystadenoma/cystadenocarcinoma (n=7); benign/malignant pancreatic tumor (n=5); pancreatic cyst (n=3); serous cystic tumor (n=1); or pancreatic pseudocyst (n=1).

Findings on imaging in patients with ECIAS

The findings on imaging in patients with ECIAS (n=21) are summarized in Table 2: ultrasound (n=12); EUS (n=4); CT (n=18); MRI (n=7); and ERP or MRP (n=9). In all of the cases the cysts occurred in the pancreatic tail and were detected as cystic lesions on abdominal imaging. Of the 19 cystic lesions, 9 unilocular and 10 multilocular cysts ranged from 1.5 to 15 cm in diameter. On MRI, the cystic component showed low-signal intensity in 5 cases (71%) and high intensity in all cases (n=7) on T1- and T2-weighted images, respectively. The solid component, confirmed on surgical specimen, showed low-intermediate signal and high-intermediate signal intensity on the T1- and T2-weighted images, respectively. In two of the cases, including the present case, the cystic component showed high-signal intensity on both T1- and T2-weighted images. Out of nine cases in which an ERP or MRP was performed, seven, including the present case, revealed normal pancreatic ducts; however, two cases had a smooth tapering stenosis or displacement in the tail of the pancreatic duct. A solid component was con-
confirmed based on the surgical specimen in nine cases, but it was not always detected on preoperative imaging. The component was confirmed by the following imaging: abdominal ultrasound (0/7, 0%), EUS (3/3, 100%), CT (7/9, 78%) and MRI (4/6, 67%). Of these solid components detected on imaging, correlation of splenic enhancement was detected on CT (3/7, 43%) and MRI (2/4, 50%). Of the 13 cases reported since the World Health Organization (WHO) classified mucinous cystic tumors in 1996 (19), 3 cases were preoperatively diagnosed as MCN. In two of the three cases, mural nodules were suspected on EUS but histologically, they were comprised of a thick fibrotic band with hyaline degeneration and splenic tissue.

Discussion

Herein we presented a case of ECIAS that was diagnosed as MCN preoperatively based on several characteristic clinical features: a woman in middle age; located in the tail of the pancreas; no communication with the pancreatic duct; bearing thick-walled cysts; detection of a solid component suggesting a mural nodule; a small volume of accessory splenic tissue; and elevation of CA19-9 in the sera. However, a final diagnosis of ECIAS was given.

The literature review indicated the significance with which to assess the correlation of enhancement between the solid component of the cystic mass and the spleen in the differential diagnosis (11, 15). In this regard, contrast-enhanced CT and MRI were useful tools; the same homogenous attenuation between the solid component and the spleen was observed in three of seven cases and two of four cases, respectively. However, an accurate diagnosis of ECIAS depends on the amount of enhanced splenic tissue in the pancreas. On the other hand, EUS was useful to detect the solid component itself but unsuitable in assessing the correlation of enhancement between the solid component of the cystic mass and the spleen due to the difficulty in observing them concurrently. In the present case, the amount of solid component, which had been detected only through EUS, was of insufficient size to assess on contrast-enhanced CT and/or MRI. In fact, only two patients were initially given a precise diagnosis of ECIAS (both reported by the
same author): one (15) showing a correlation of enhancement and the other (18) showing similar imaging findings to the former case. From our literature review, we offer the following reasons.

First, the information was insufficient: not all patients had undergone EUS, contrast-enhanced CT, and/or MRI. Second, the imaging findings of ECIAS varies, and both the clinical and imaging features resemble that of MCN, such as the location in the tail of pancreas, no communication with pancreatic duct, bearing thick-walled cysts, and a solid component. Given the fact that MCN with a mural nodule harbors a malignant potential (20), it is difficult to completely distinguish ECIAS from MCN, especially when CA19-9 in the sera is elevated, as in the present case. The authors state that they have no Conflict of Interest (COI).

References


