J Hepatobiliary Pancreat Surg (2008) 15:436–439
DOI 10.1007/s00534-007-1243-4

Journal of HBP Surgery

Case reports of interest

Epidermoid cyst originating from an intrapancreatic accessory spleen

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Abstract
We report a rare case of an epidermoid cyst originating from an intrapancreatic accessory spleen, in a 40-year-old Japanese man with no clinical symptoms. A cystic tumor in the pancreatic tail was detected incidentally by abdominal ultrasonography. The patient was referred to the KKR Tachikawa Hospital for further examination of the tumor. Preoperative imaging findings suggested that the tumor was an epidermoid cyst originating from an intrapancreatic accessory spleen. On both pre- and post-contrast computed tomography and magnetic resonance images, the solid compartment of the tumor had the same X-ray attenuation and intensity as the spleen. Upon surgical excision, the mass consisted of solid and cystic components that were macroscopically evident on the preoperative images. Microscopic analysis revealed that the solid component was an accessory spleen in the pancreatic tail, whereas the cystic component was lined with stratified epithelium representative of an epidermoid cyst. This is the thirteenth report (in English) of an epidermoid cyst originating from an intrapancreatic accessory spleen, and the first case to be diagnosed prior to surgery.

Key words Epidermoid cyst · Epithelial inclusion cyst · Intrapancreatic accessory spleen · Pancreatic cyst

Introduction

Since Davidson et al.[1] reported the first case of an epidermoid cyst originating from an intrapancreatic accessory spleen, in 1980, there have been 12 additional cases reported in the English-language literature. However, because of the difficulty in differentiating this tumor from cystic neoplasm of the pancreas, none of these cases was diagnosed correctly before surgery. In this article, we report a case of an epidermoid cyst that originated from an intrapancreatic accessory spleen and was diagnosed before surgery. We also highlight key findings required for accurate diagnosis, with a review of similar cases in the literature.

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Received: September 8, 2006 / Accepted: March 5, 2007

Case report

A 40-year-old man with no clinical symptoms underwent a routine health checkup in which a cystic tumor of the pancreatic tail was detected incidentally during abdominal ultrasonography. No history of trauma or pancreatitis was recorded, and he was referred to the KKR Tachikawa Hospital for further examination. Physical examination and blood chemistry findings upon admission showed no abnormalities. Tumor markers such as carcinoembryonic antigen, carbohydrate antigen 19-9 (CA19-9), and Dupan-2 were within normal ranges.

Abdominal ultrasonography showed a 3-cm cystic tumor that started from the tail of the pancreas. When viewed on abdominal computed tomography, a 4.0-cm cystic mass was detected in the tail of the pancreas (Fig. 1A). The tumor was primarily composed of cystic material, but also had a solid component with a small region of calcification in its medial region. On contrast studies, the solid component showed the same homogeneous attenuation as the spleen (Fig. 1B). On magnetic resonance images, the cystic component showed high signal intensities on both T1- and T2-weighted images (Fig. 2A,B), whereas the solid component showed intermediate-low signal intensity on T1-weighted images. The solid component showed the same signal change as normal splenic tissue on contrast studies (Fig. 3). Endoscopic retrograde cholangiopancreatography revealed no change in the pancreatic duct.

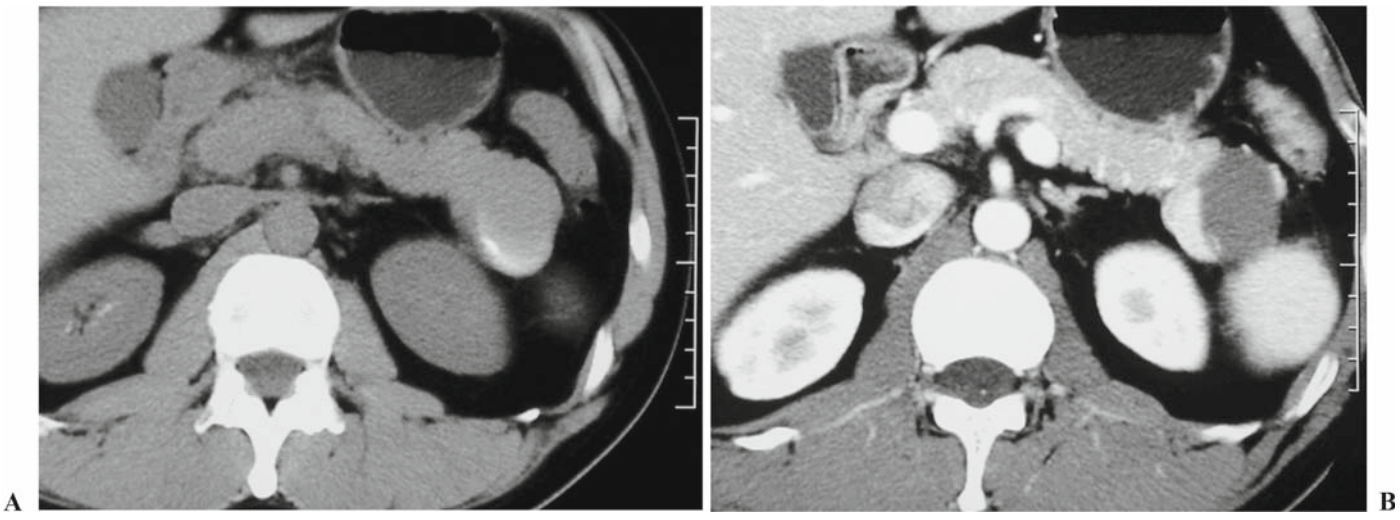

Fig. 1. A Abdominal computed tomography reveals a 4.0-cm cystic mass with a region of calcification in the tail of the pancreas. **B** On contrast studies, the solid component shows the same homogeneous attenuation as the spleen

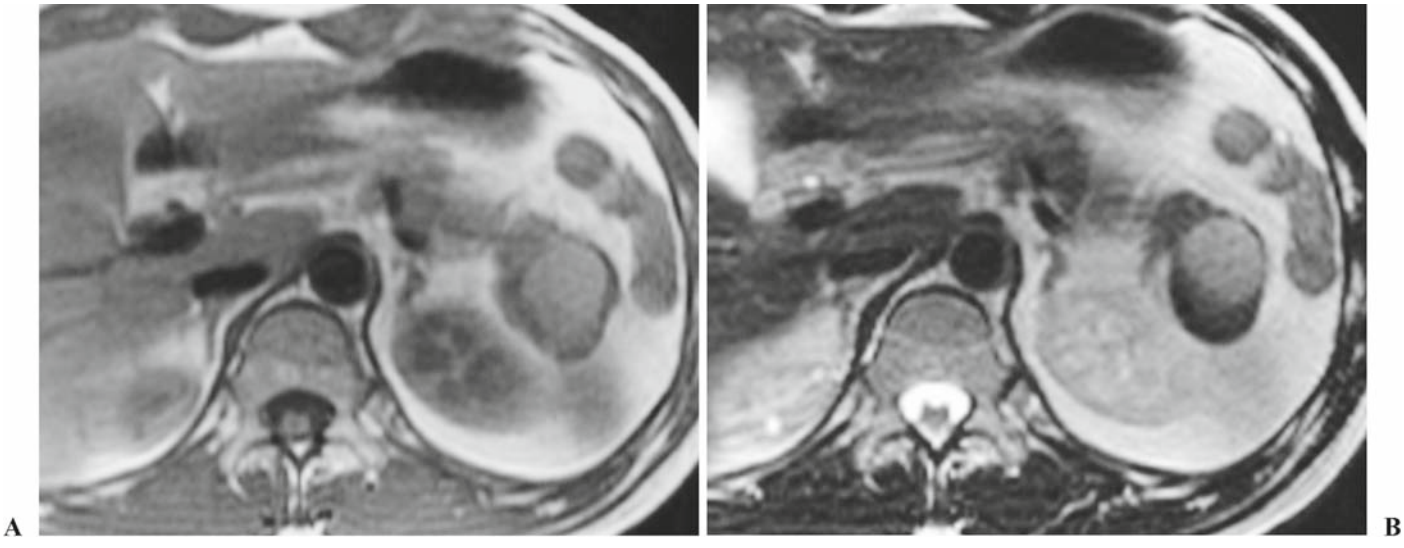

Fig. 2A,B. Magnetic resonance images. Axial T1-weighted (**A**) and T2-weighted (**B**) magnetic resonance images reveal that the cystic component shows high signal intensity on both T1- and T2-weighted images

These findings suggested the highly probable preoperative diagnosis of an epidermoid cyst originating from an intrapancreatic accessory spleen. Because the patient was eager to have the tumor resected and malignancy could not be completely ruled out, distal pancreatectomy with splenectomy was performed. Macroscopically, the surgical specimen was a well-demarcated, solitary, encapsulated mass, measuring 4.0 × 3.2 × 3.0 cm, in the pancreatic tail. Bisection across the tumor revealed a unilocular cyst with a brown solid component that resembled normal spleen (Fig. 4). Histopathological examination demonstrated that the cyst was lined with stratified squamous epithelium (Fig. 5) and was surrounded by normal splenic tissue. The final pathological diagnosis was in agreement with the preoperative diagnosis of an epidermoid cyst originating from an intrapancreatic accessory spleen.

The patient's postoperative course was uneventful. During 3 years of follow-up, he has been doing well.

Discussion

A total of 12 epidermoid cysts originating from intrapancreatic accessory spleens have been reported to date in the English-language literature.[1–12] The characteris-

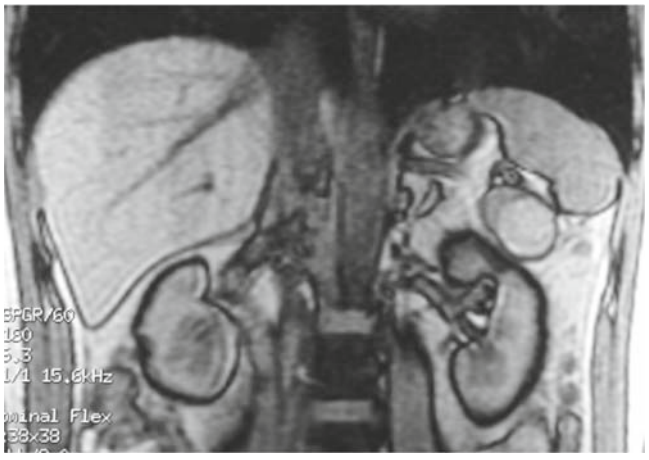

Fig. 3. Coronal T1-weighted magnetic resonance image. The solid component of the mass shows the same signal change as normal splenic tissue, although it is completely separate from the spleen

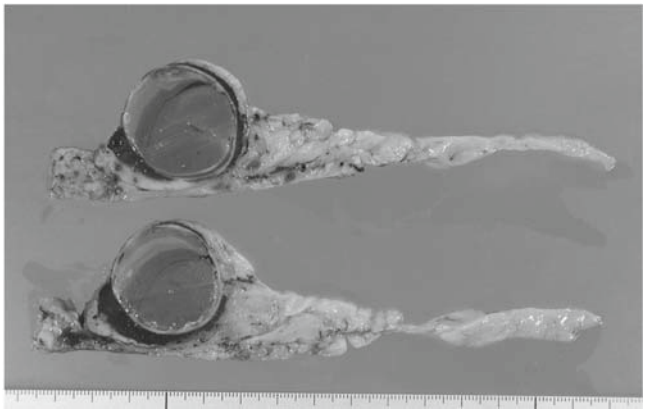

Fig. 4. Gross findings of the cystic mass in a cut section. A portion of the outer surface of the cyst was covered with the accessory spleen in the pancreatic tail

tics of our case and the previously reported cases are summarized in Tables 1 and 2. The 13 patients had a mean onset age of 45 years and the sex ratio was six males to seven females, suggesting that this type of tumor tends to develop in middle age without preference for a particular sex.

Seven patients had no symptoms and their tumors were found during regular checkups. Six patients had abdominal pain or discomfort or weight loss and nausea. Although blood chemistry findings showed no abnormalities, four patients had slight increases in the serum CA19-9 level. We presumed that this increase in serum CA19-9 was a natural response of the surrounding pancreatic tissue to the tumor. Another possibility, suggested in two previous reports,[7,10] is that the epithelium of epidermal cysts of an accessory spleen may originate

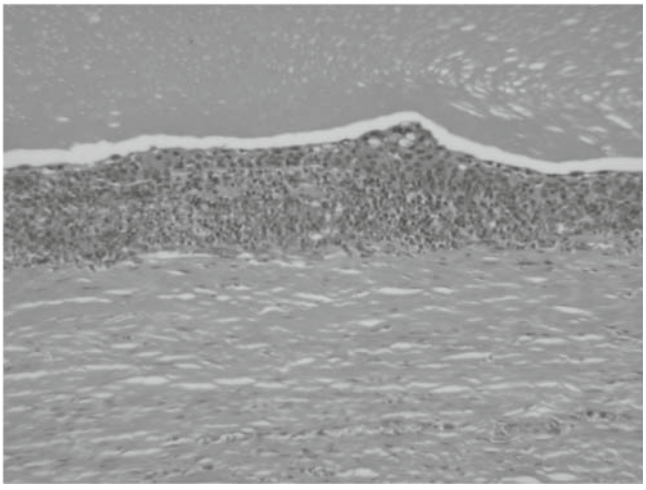

Fig. 5. Histopathological examination shows that the cyst is lined with stratified squamous epithelium. H&E, High-Power magnification

Table 1. Summary of case reports: clinical findings and demographics

Patient number	Author	Year	Age (years)/ sex	Site	Size (cm)	Symptoms	Serum CA19-9 (IU/ml)
1	Davidson et al.[1]	1980	40/M	Tail	7 × 3 × 2.5	Weight loss, nausea	NI
2	Morohoshi et al.[2]	1991	32/F	Tail	6 × 5	Left abdominal pain	WNL
3	Nakae et al.[3]	1991	37/F	Tail	6.5 × 5.5 × 4.6	Epigastric pain	NI
4	Tang et al.[4]	1994	38/M	Tail	2.3 × 2.1	None	WNL
5	Furukawa et al.[5]	1998	45/M	Tail	2[a]	None	WNL
6	Higaki et al.[6]	1998	46/F	Tail	3 × 3	Left back pain	201
7	Sasou et al.[7]	1999	49/F	Tail	4.3 × 2.6	None	WNL
8	Choi et al.[8]	2000	54/F	Tail	15 × 11	Epigastric discomfort	NI
9	Tsutsumi et al.[9]	2000	51/M	Tail	2.5 × 2.5 × 2.0	None	WNL
10	Horibe et al.[10]	2001	48/M	Tail	2 × 1	None	53
11	Sonomura et al.[11]	2002	45/F	Tail	3.5[a]	Epigastric discomfort	159
12	Kanazawa et al.[12]	2004	58/F	Tail	2.5[b]	None	62
13	Our patient		40/M	Tail	4.0 × 3.2 × 3.0	None	WNL

NI, no information; WNL, within normal limits
[a] Measured on the CT image
[b] Measured on the echo image

Table 2. Summary of case reports: imaging findings

Case	Form of cyst	Detectable spleen part on CT or MRI	Cystic lesion on MRI T1	Cystic lesion on MRI T2	Preoperative diagnosis
1	Multilocular	(–)	NI	NI	Pancreatic pseudocyst or cystadenoma or cystadenocarcinoma
2	Unilocular	(–)	NI	NI	Pancreatic tail cyst
3	Unilocular	(–)	Hypointense	Hyperintense	Cystic mass in tail of pancreas
4	Multilocular	(+)	NI	NI	NI
5	Multilocular	(–)	NI	NI	Malignant pancreatic tumor
6	Multilocular	(+)	NI	NI	Cystadenoma or cystadenocarcinoma
7	Multilocular	NI	NI	NI	Cystic tumor of the pancreas
8	Multilocular	(+)	NI	NI	Benign cystic tumor of the pancreas
9	Unilocular	(+)	Hypointense	Intermediate high	Pancreatic cystic tumor or accessory spleen
10	Unilocular	(–)	NI	NI	Mucin-producing pancreatic tumor
11	Multilocular	(+)	NI	NI	Cystadenocarcinoma or solid cystic tumor
12	Multilocular	(+)	Hypointense	Hyperintense	Mucinous cystic tumor
13	Unilocular	(+)	Hyperintense	Hyperintense	Epidermoid cyst in intrapancreatic accessory spleen

NI, no information; CT, computed tomography; MRI, magnetic resonance image; T1, T1-weighted image; T2, T2-weighted image

from the pancreatic duct, because the cyst epithelium is immunohistochemically positive for CA19-9, and negative for antibodies against mesothelial cells.[7,10]

Macroscopically, all 13 tumors developed in the tail of the pancreas, with sizes ranging from 2 to 15 cm (mean, 4.7 cm) in diameter. There were five unilocular and eight multilocular cysts. Microscopically, the tumor reported in our patient demonstrated features similar to all of the previously reported tumors, in that the cyst was lined with stratified squamous epithelium and was surrounded by normal splenic tissue.

For our patient, we used information from magnetic resonance images to diagnose the cystic portion of the tumor as an epidermoid cyst. We assumed that the tumor had originated from the spleen because the density of the tumor's solid component matched the density of the spleen on post-contrast computed tomography and magnetic resonance images. In the three patients previously reported in whom the cysts were viewed by magnetic resonance imaging, the cystic component showed low signal intensity on T1-weighted images and high signal intensity on T2-weighted images; however, in our patient, the cystic component showed high signal intensity on both T1- and T2-weighted images. Epidermoid cysts, which commonly show low signal intensity on T1-weighted images, do occasionally show high signal intensity.

In 7 of the 13 patients, a solid tumor component was shown on computed tomography or magnetic resonance images, and several reports mentioned that, retrospectively, the images of the solid component were similar to those of the spleen. These findings suggest that, in the presence of a relatively large amount of splenic tissue, a correct diagnosis should be possible with careful examination of images prior to surgery. However, if the amount of splenic tissue is relatively small, accurate diagnosis before surgery may not be possible.

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