A lymphoepithelial cyst in the pancreatic accessory spleen: A case report

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Abstract  
We present the first report of a lymphoepithelial cyst. As additional cases will likely be encountered in the future, our study sets the precedent for future research.

KEYWORDS  
a lymphoepithelial cyst, pancreatic accessory spleen

1 INTRODUCTION

There are 57 reports of epidermoid cysts in the accessory spleen in the pancreas, but none of lymphoepithelial cysts (LECs) in this location. The pathological diagnostic criteria for LECs are still unclear and controversial. We describe an LEC in the accessory spleen, including the pathological findings.

Pancreatic cysts are broadly categorized as neoplastic or non-neoplastic. The former category includes intraductal papillary mucinous neoplasms (IPMNs), mucinous cystic neoplasms (MCNs), and serous cystic neoplasms (SCNs), and the latter includes pancreatic pseudocysts and lymphoepithelial, epidermoid, and dermoid cysts.¹ There are also pancreatic neuroendocrine tumors (pNETs), which are tumors that have degenerated into cysts. These classifications are laid out in several documents including the International Association of Pancreatology guidelines,² the European expert consensus statements,³ and the American Gastroenterological Association guidelines.⁴ However, there are no guidelines or clear diagnostic criteria for pancreatic pseudocysts.

Pancreatic lymphoepithelial cysts (LECs) are rare benign lesions initially described by Luchtrath and Schriefers in 1985⁵ and named by Truong et al in 1987.⁶ LECs are typically observed in middle-aged and elderly men; they occur equally in the pancreatic head, body, or tail, and may present...
FIGURE 2  Magnetic resonance imaging (MRI): A, The T1-weighted image shows a cystic lesion measuring 18 × 14 mm with low signal intensity (arrow). B, The same lesion has a high signal intensity on a T2-weighted image (arrow). C, Part of the cyst wall was hyperintense (arrow) on diffusion-weighted imaging. D, Early uptake of the dye by this part of the wall (arrow) on dynamic MRI is shown.

FIGURE 3  Endoscopic imaging: A, Endoscopic ultrasonography (EUS) shows a cyst with calcification in the margin (arrow). B, Endoscopic retrograde cholangiopancreatography shows a normal main pancreatic duct with no communication with the cystic lesion.
as a single or multilocular lesion. This study is the first to document a case of an LEC in the pancreatic accessory spleen, and thus, in this respect, we believe it is valuable. Furthermore, the knowledge acquired in this study will enable the development of international diagnostic criteria for LECs and similar epidermoid cysts and dermoid cysts in the future.

2 | PRESENTATION OF THE CASE

A 43-year-old man was found to have a pancreatic tail cyst on abdominal ultrasonography at a previous hospital. He had no subjective symptoms or medical history. Laboratory examinations showed normal results for the following: complete blood cell counts, hepatic and renal function, and serum levels of carcinoembryonic antigen (CEA), carbohydrate antigen 19-9 (CA 19-9), amylase, lipase, and glucose.

The patient was referred to our hospital, and the presence of the cyst in the pancreatic tail was confirmed via computed tomography (CT) (Figure 1). The cystic lesion was 15 cm in diameter, monocystic, and surrounded by an area of low intensity. Magnetic resonance imaging (MRI) revealed a lesion with depleted and enhanced intensity on T1- and T2-weighted images, respectively (Figure 2A, B). Diffusion-weighted MRI displayed increased signal potency in the peripheral portion of the cystic lesion (the wall and the septa) (Figure 2C); the cystic contents had reduced intensity on enhanced MRI (Figure 2D). Additional observations included a cyst with high-echoic lesions, a pancreatic tail, and calcification on the cyst margin on endoscopic ultrasonography (Figure 3A), and a slight displacement of the pressure superior to the duct with

![Figure 1](image1.jpg)

**FIGURE 1** Computed tomography image of the pancreatic tail cyst.

![Figure 2](image2.jpg)

**FIGURE 2** Magnetic resonance imaging images of the pancreatic tail cyst.

![Figure 3](image3.jpg)

**FIGURE 3** Endoscopic ultrasonography image of the pancreatic tail cyst.

![Figure 4](image4.jpg)

**FIGURE 4** Macroscopic findings and histopathology: A, The pancreatic parenchyma at the tail of the pancreas (white arrowhead) contains spleen tissue (white arrow) and a multilocular cyst with a maximum diameter of 17 mm (yellow arrow). B, The cystic wall is lined by mature keratinized squamous epithelium and underlying lymphoid tissue (×20 magnification, hematoxylin and eosin staining). C, The epithelial lining is surrounded by splenic pulp and pancreatic tissue (×40 magnification, hematoxylin and eosin staining). D, Lymphoid follicles and lymphoid tissue are observed (×100 magnification, hematoxylin and eosin staining).
| Case | Authors    | Sex/age | Symptom    | Location | Size(cm) | Cyst       | Serum markers | CEA CA19-9 | CT | MRI | Preoperative diagnosis | Surgery | Pathology | Final diagnosis                                      |
|------|------------|---------|------------|----------|----------|------------|---------------|-------------|-----|-----|------------------------|---------|------------|-----------------------------------------------------|
| 1    | Davidson   | M/40    | Nausea     | Tail     | 5.5      | Multilocular|               |             |     |     | NI                     |         |            | DP Cystic lesion surrounded by thin rim of tissue   |
| 2    | Hanada     | M/51    | Abdominal pain | Tail     | 6        | NI          |               |             |     |     | NI                     |         |            | NI Pseudocyst, cystadenoma, and cystadenocarcinoma   |
| 3    | Morohoshi  | F/32    | Abdominal pain | Tail     | 6        | Unilocular |               |             |     |     | Normal               |         |            | DP The wall of the cyst was composed of dense, hypocellular, collagenous tissue. The inner surface of the cyst was lined by flattened stratified squamous type of epithelium. The lining epithelium was mature stratified squamous epithelium. The cystic wall consisted of three tissue elements: The inside was lined by stratified epithelium, the middle layer was composed of some splenic pulp, and the peripheral layer consisted of dense fibrous connective tissue containing some involutional pancreatic ducts and islets. |
| 4    | Nakae      | F/37    | Epigastric pain | Tail     | 6.5      | Unilocular |               |             |     |     | NI                     |         |            | SPDP The cyst is surrounded with fibrous tissue and a thin layer of splenic tissue containing a germinal center, adjacent to normal pancreatic tissue. |
| 5    | Tang       | M/38    | Asymptomatic | Tail     | 1.4      | Multilocular|               |             |     |     | NI                     |         |            | DP The cyst is lined by stratified squamous epithelium. Cyst is present within normal splenic tissue, which is surrounded by pancreatic parenchyma. |
| 6    | Furukawa   | M/45    | Asymptomatic | Tail     | 2        | Multilocular|               |             |     |     | NI                     |         |            | DP The cyst is surrounded by accessory splenic tissue. The cyst wall was lined with stratified squamous epithelium. |
| 7    | Higaki     | F/46    | Left back pain | Tail     | 3        | Multilocular| CA19-9:high, 201 U/mL |             |     |     |                        |         |            | Epidermoid cyst                                      |
| CT                                      | MRI  | Preoperative diagnosis                  | Surgery | Pathology                                                                                                                                                                                                 | Final diagnosis |
|-----------------------------------------|------|-----------------------------------------|---------|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-----------------|
| Cystic lesion surrounded by thin rim of tissue | NI   | Pseudocyst, cystadenoma, and cystadenocarcinoma | DP      | Cyst wall focally shows a transition from low cuboidal to low stratified squamous epithelium. A giant-cell granulomatous reaction in the underlying red pulp is surrounded by a band of fibrous tissue like that beneath the epithelium of the cyst wall | Epidermoid cyst |
| Cystic mass with a rim of dense density | NI   | Pseudocyst                              | DP      | The wall of the cyst was composed of dense, hypocellular, collagenous tissue. The inner surface of the cyst was lined by a flattened stratified squamous type of epithelium                                                                 | Epidermoid cyst |
| Well-demarcated cystic lesion           | NI   | Pancreatic cyst                          | Cyst removal | The cystic wall consisted of three tissue elements: The inside was lined by stratified epithelium, the middle layer was composed of some splenic pulp, and the peripheral layer consisted of dense fibrous connective tissue containing some involutional pancreatic ducts and islets. The lining epithelium was mature stratified squamous epithelium | Epidermoid cyst |
| Cystic lesion with a thin wall of high density | T1 low, T2 high | Pancreatic cyst                         | SPDP    | The cyst is surrounded with fibrous tissue and a thin layer of splenic tissue containing a germinal center, adjacent to normal pancreatic tissue                                                                 | Epidermoid cyst |
| Well-demarcated hypodense lesion        | NI   | NI                                      | DP      | The cysts are lined by nonkeratinizing, stratified squamous epithelium. Mucic-containing cells are scattered among the epithelium                                                                                     | Epidermoid cyst |
| Peripherally enhanced area, its density is equal to the spleen | NI   | Primary cystic neoplasm                 | DP      | The cyst is lined by stratified squamous epithelium. Cyst is present within normal splenic tissue, which is surrounded by pancreatic parenchyma.                                                                   | Epidermoid cyst |
| Oval nodule with a distinct margin      | NI   | Malignant tumor                          | DP      | Cyst surrounded by accessory splenic tissue The cyst wall was lined with stratified squamous epithelium                                                                                                      | Epidermoid cyst |

(Continues)
| Case | Authors | Sex/age | Symptom          | Location | Size(cm) | Cyst          | Serum markers | CEA CA19-9 |
|------|---------|---------|-----------------|----------|----------|---------------|---------------|-------------|
| 8    | Tateyama| F/67    | Abdominal pain  | Tail     | 3        | Multilocular  |               |             |
| 9    | Sasou   | F/49    | Asymptomatic    | Tail     | 4.3      | Multilocular  | NI            |             |
| 10   | Choi    | F/54    | Epigastric pain | Tail     | 15       | Multilocular  | NI            |             |
| 11   | Tsutumi | M/51    | Asymptomatic    | Tail     | 2.5      | Multilocular  | Normal        |             |
| 12   | Horibe  | M/48    | Asymptomatic    | Tail     | 2        | Unilocular    | CA19-9:high 53 U/mL |             |
| 13   | Sonomura| F/45    | Epigastric pain | Tail     | 3.5      | Multilocular  | NI            |             |
| 14   | Fink    | F/12    | Fever           | Tail     | 10       | Multilocular  | NI            |             |
| Case | Authors | Sex/Age | Symptom | Location | Size (cm) | MRI | CT | Preoperative diagnosis | Surgery | Pathology | Final diagnosis |
|------|---------|---------|---------|----------|-----------|-----|----|------------------------|---------|-----------|-----------------|
| 8    | Tateyama | F/67    | Abdominal pain | Tail     | 3         | NI  | NI | Multilocular Cystic mass of low density | DP      | The cyst is lined by stratified squamous epithelium. The epithelial lining is surrounded by hyalinized fibrous tissue with scattered lymphoid tissue, splenic pulp, and pancreatic tissue | Epidermoid cyst |
| 9    | Sasou   | F/49    | Asymptomatic | Tail     | 4.3       | NI  | NI | Multilocular Cystic mass of low density | DP      | The cyst is surrounded by splenic tissue in the pancreas. The inside of the cyst in the accessory spleen was lined with stratified squamous epithelium and a layer of flat cells | Epithelial cyst |
| 10   | Choi    | F/54    | Epigastric pain | Tail     | 15        | NI  | Major cystic component, small solid component with the same homogeneous Cyst: T1 low, T2 high: solid lesion: T1 low, T2 intermediate-high | DP      | The cyst is lined by stratified squamous epithelium with keratinization. A thin fibrous capsule separates the intrapancreatic accessory spleen from the pancreas | Epidermoid cyst |
| 11   | Tsutumi | M/51    | Asymptomatic | Tail     | 2.5       | NI  | Well-demarcated cystic lesion containing a solid portion | DP      | Cystic lesions are seen in the splenic tissue surrounded by pancreatic tissue. Cystic walls are lined by stratified squamous epithelium | Epidermoid cyst |
| 12   | Horibe  | M/48    | Asymptomatic | Tail     | 2         | NI  | No substance in the cyst by enhanced image | NI      | The cyst is surrounded with fibrous tissue and a thin layer of splenic tissue containing a germinal center, adjacent to normal pancreatic tissue. The cyst wall is lined with stratified and a few layers of squamous epithelium | Epidermoid cyst |
| 13   | Sonomura | F/45    | Epigastric pain | Tail     | 3.5       | NI  | Parenchymal medial lesion with calcification and cystic lateral lesion | NP      | The parenchymal region of the mass showed historic features of the spleen, and the multilocular cyst was lined with several layers of stratified squamous epithelium | Epidermoid cyst |
| 14   | Fink    | F/12    | Fever | Tail     | 10        | NI  | Rim enhancing cystic lesion, with a medial mural nodule | NP      | The cyst was surrounded by nonkeratinizing squamous epithelium. The wall was composed of fibrous tissue with prominent hemosiderosis surrounded by splenic tissue. The pancreas was compressed by the splenic capsule | Epidermoid cyst |

(Continues)
| Case | Authors | Sex/age | Symptom | Location | Size(cm) | Cyst | Serum markers | CEA | CA19-9 |
|------|---------|---------|---------|----------|----------|-----|---------------|-----|---------|
| 15   | Yokimizo M/38 | Asymptomatic | Tail | 3 | Multilocular | CA19-9:high | 410 U/mL |
| 16   | Kanazawa F/58 | Asymptomatic | Tail | 2.5 | Multilocular | CA19-9:high | 62 U/mL |
| 17   | Watanabe F/55 | Posyprandialepogastralgia | Tail | 2.5 | Multilocular | CA19-9:high | 176 U/mL |
| 18   | Won M/32 | Asymptomatic | Tail | 7.5 | Unilocular | CA19-9:high | 53 U/mL |
| 19   | Won F/49 | Abdominal pain | Tail | 2 | Multilocular | Normal |
| 20   | Ru M/41 | Asymptomatic | Tail | 2.5 | Unilocular | Normal |
| 21   | Itano M/40 | Asymptomatic | Tail | 4 | Unilocular | Normal |
| 22   | Servais F/52 | Asymptomatic | Tail | 11.5 | Multilocular | CA19-9/CEA:high |
| CT                          | MRI                               | Preoperative diagnosis                        | Surgery | Pathology                                                                 | Final diagnosis |
|-----------------------------|-----------------------------------|-----------------------------------------------|----------|---------------------------------------------------------------------------|------------------|
| NI                          | Cyst:T2 super-high,               | MCN, adenocarcinoma and ECIPS                 | DP       | The cyst wall was lined by nonkeratinizing stratified squamous epithelium and containing splenic tissue | Epidermoid cyst  |
|                             | Cyst wall: delineated enhancement |                                 |          |                                                                           |                  |
| Septated low-density area   | Cystic component: T1 hypo, T2 hyper | MCN                                           | SPDP     | The cyst was lined with stratified squamous epithelium and was surrounded by normal splenic tissue | Epidermoid cyst  |
| Multilocular cystic tumor. | No protruded lesion in the inner lumen | T1 low, T2 high | DP       | The cystic lesions were surrounded by fibrous tissue and a thin layer of splenic tissue with a germinal center and separated from the adjacent pancreatic parenchyma | Epidermoid cyst  |
| Well-circumscribed cystic  | NI                                | Pancreatic pseudocyst                        | SPDP     | The cyst was surrounded by splenic parenchyma with was lined by nonkeratinizing squamous epithelium and flattened or cuboidal epithelial cells that continued to the stratified squamous epithelium | Epithelial cyst  |
| mass with inner fluid debris |                                  |                                               |          |                                                                           |                  |
| or hemorrhagic fluid        |                                  |                                               |          |                                                                           |                  |
| Well-circumscribed cystic   | NI                                | Serous or mucinous cystadenoma                | Laparoscopic DP | The epithelial lining showed a mixture of flattened mesothelial-like cells, ciliated cuboidal cells, and stratified squamous epithelial cells. Red pulps in the cystic walls were identifiable | Epithelial cyst  |
| tumor septation             |                                  |                                               |          |                                                                           |                  |
| Well-circumscribed tumor    | NI                                | Cystic lesion of the pancreas                 | DP       | The epithelial lining appeared focally stratified without atypia. Scattered mucinous cells were identified occasionally. A fibrotic band with sclerosis lay underneath the epithelial lining, and overlying spleen tissue | Epidermoid cyst  |
| which partially compressed  |                                  |                                               |          |                                                                           |                  |
| the spleen                  |                                  |                                               |          |                                                                           |                  |
| Solid component with        | Cyst:T1/ T2high;solid component:T1 intermediate-low | ECIPAS                                      | DP       | The cyst was lined with stratified squamous epithelium and was surrounded by normal splenic tissue | Epidermoid cyst  |
| the same homogeneous        |                                  |                                               |          |                                                                           |                  |
| attenuation as the spleen   |                                  |                                               |          |                                                                           |                  |
| Cystic mass which was thin  | NI                                | Malignant pancreatic neoplasm                 | DP       | Epithelium-lined cyst with a dense hyalinized fibrous wall surrounded by a normal rim of native pancreatic tissue. The cyst wall demonstrated splenic pulp tissue | Epidermoid cyst  |
| walled and contained single |                                  |                                               |          |                                                                           |                  |

(Continues)
| Case | Authors         | Sex/age | Symptom           | Location | Size(cm) | Cyst       | Serum markers CEA CA19-9 |
|------|----------------|---------|-------------------|----------|----------|------------|--------------------------|
| 23   | Gleeson        | F/32    | Abdominal pain    | Tail     | 1.5      | Unilocular | NI                       |
| 24   | Zhang and Wang | F/26    | Asymptomatic      | Tail     | 2.5      | Unilocular | Normal                   |
| 25   | Reiss          | M/49    | Asymptomatic      | Tail     | 3.6      | Multilocular| NI                       |
| 26   | Kodota         | F/57    | Asymptomatic      | Tail     | 6        | Multilocular| Normal                   |
| 27   | Kodota         | F/70    | Asymptomatic      | Tail     | 1.7      | NI         | CA19-9: high 48 U/mL, CEA: normal |
| 28   | Kodota         | M/73    | Asymptomatic      | Tail     | 10       | NI         | CA-19-9: high 647 U/mL, CEA: normal |
| 29   | Itano          | M/67    | Epigastric pain   | Tail     | 1.5      | Unilocular | CA19-9: high 182 U/mL    |
| CT | MRI | Preoperative diagnosis | Surgery | Pathology | Final diagnosis |
|----|-----|------------------------|---------|-----------|----------------|
| Demarcated cyst without septation calcification and satellite lesion | NI | Pancreatic cystic neoplasm | DP | The epithelial cyst lining at the top, with surrounding splenic tissue inferior to it, and adjacent pancreas demonstrating chronic pancreatitis inferior to the splenic tissue | Epidermoid cyst |
| Cystic wall revealed a density similar to that of the pancreas | NI | Primary MCN | SPDP | The cyst contained homogenous eosinophilic fluid and was lined with stratified squamous epithelium. Accessory spleen tissue was found under the epithelium and surrounded by a complete fibrous capsule | Epidermoid cyst |
| Heterogeneously enhancing mass | NI | MCN | DP | The lesion revealed an intrapancreatic accessory spleen lined with stratified squamous epithelium with occasional goblet cells | Epidermoid cyst |
| Cystic wall : a partial enhancement | NI | Pancreatic cystic tumor | DP | The cyst is surrounded by normal splenic tissue and hyalinized fibrous tissue consisting of spleen tissue. The cystic walls were lined with nonkeratinizing stratified squamous epithelium and focally cuboidal epithelium | Epidermoid cyst |
| Cystic mass lesion | NI | MCN | DP | The cysts are surrounded by normal splenic tissue and hyalinized fibrous tissue. The lining epithelium shows a nonkeratinizing stratified squamous epithelium | Epidermoid cyst |
| Cystic mass lesion with a partial enhancement of the cystic wall | NI | Serous cystic tumor or lymphoepithelial cyst | DP | The cysts are lined with the keratinizing stratified squamous epithelium and focally several layers of the cubical epithelium. The cuboidal epithelium revealed transitional findings to the stratified squamous epithelium | Epidermoid cyst |
| Cystic tissue and smooth solid component | Cyst:T1 intermediate, T2 high. Solid lesion:T1 intermediate | ECIPAS | Laparoscopic DP | The cyst was surrounded by ectopic splenic tissue with a normal appearance and atrophic pancreatic tissue. The cyst was lined with a stratified squamous epithelium | Epidermoid cyst |

(Continues)
| Case | Authors            | Sex/age | Symptom              | Location | Size(cm) | Cyst         | Serum markers CEA CA19-9 |
|------|--------------------|---------|----------------------|----------|----------|--------------|--------------------------|
| 30   | N.Panagiotopoulos | M/51    | No                   | Tail     | 2.3      | NI           | Normal                   |
| 31   | Horn and Lele      | M/62    | Abdominal pain       | Tail     | 4.8      | Multilocular | NI                       |
| 32   | Iwasaki            | F/36    | Asymptomatic         | Tail     | 3.4      | Unilocular   | CA19-9: high 79 U/mL     |
| 33   | Yamanishi          | F/55    | Asymptomatic         | Tail     | 2.5      | Unilocular   | CA19-9: high 90 U/mL     |
| 34   | Urakami            | F/50    | Asymptomatic         | Tail     | 3        | Unilocular   | NI                       |
| 35   | Khashab            | F/49    | Abdominal pain       | Tail     | 2.3      | Unilocular   | NI                       |
| 36   | Harris             | F/39    | Asymptomatic         | Tail     | 2.5      | NI           | NI                       |
| 37   | Hong               | F/54    | Abdominal discomfort | Tail     | 2        | Multilocular | NI                       |
| 38   | Hamidian           | F/36    | Asymptomatic         | Tail     | 5        | Multilocular | NI                       |
| 39   | Zavras             | F/63    | Nausea and vomiting  | Tail     | 12.6     | NI           | CA19-9: high 5000 U/mL, CEA: high 180.4 ng/mL |
| CT                                                                 | MRI  | Preoperative diagnosis                        | Surgery       | Pathology                                                                 | Final diagnosis                        |
|-------------------------------------------------------------------|------|-----------------------------------------------|---------------|---------------------------------------------------------------------------|----------------------------------------|
| Well-defined low attenuation lesion arising exophytically from the tail of the pancreas | NI   | potential pancreatic malignancy               | DP            | The cyst was lined by nonkeratinising stratified squamous epithelium. The cyst was revealed within accessory splenic tissue | Epithelial cyst                         |
| Left-sided retroperitoneal mass with a possible cystic component  | NI   | NI                                            | DP            | The cyst was revealed within accessory splenic tissue. The cysts were lined by stratified squamous epithelium | Epidermoid cyst                         |
| Septate low-density lesion, with an area showing higher degree of enhancement than the pancreas | NI   | MCN                                           | Laparoscopic DP| Cystic lesion lined with stratified squamous epithelium and surrounded by an intrapancreatic accessory spleen | Epidermoid cyst                         |
| Cyst wall was relatively thick, but not enhanced                  | Cyst: T1 slightly high, thick, but not enhanced | MCN            | DP            | The cyst was surrounded by a dense, hyalinized fibrous and a thin layer of splenic tissue | Epidermoid cyst                         |
| Single cyst with a contrasted mass beside it                       | Cyst: T1 low, T2 high | ECIPAS | Laparoscopic DP | The cyst was surrounded by fibrous tissue and a thin layer of splenic with a general center, adjacent to normal pancreatic parenchyma | Epidermoid cyst                         |
| Solid                                                             | NI   | PNET                                          | Laparoscopic DP| The mass had a well-defined capsule within which was splenic parenchyma and a small cyst lined by a layer of benign squamous epithelium | Epidermoid cyst                         |
| Stable hypodense lesion                                           | Pancreatic cyst neoplasm | Malignant cystic tumor | Laparoscopic SPDP | The cyst was surrounded by accessory splenic tissue in the pancreas parenchyma. The cyst was lined by multilayered cuboidal epithelium | Epidermoid cyst                         |
| Cystic mass                                                       | NI   | NI                                            | SPDP          | A cyst is surrounded by accessory splenic tissue in the pancreas parenchyma. A cyst is lined by multilayered cuboidal epithelium | Epidermoid cyst                         |
| Cystic lesion                                                     | NI   | NI                                            | DP            | pancreas tissue, spleen tissue, fibrous capsule                           | Epidermoid cyst                         |
| Mass lesion with solid and cystic components                      | NI   | Malignant tumor of the pancreas               | DP            | The cyst was lined with multilayered flattened epithelium, reminiscent of squamous epithelium above a red pulp splenic parenchyma | Epidermoid cyst                         |

(Continues)
| Case | Authors | Sex/age | Symptom | Location | Size(cm) | Cyst | Serum markers | CEA CA19-9 |
|------|---------|---------|---------|----------|----------|-----|---------------|-------------|
| 40   | Kumamoto| M/39    | Diarrhea| Tail     | 3.8      | NI  |               | CA19-9:high 286 U/mL |
| 41   | Kwak    | F/21    | Abdominal pain and fever | Tail | 2.5 | Multilocular | Normal |
| 42   | Kato    | F/33    | Asymptomatic | Tail | 3 | Multilocular | Normal |
| 43   | Modi    | F/62    | Abdominal pain | Tail | 2.4 | Unilocular | NI |
| 44   | Fujii   | F/50    | Asymptomatic | Tail | 4 | Unilocular | CA19-9: high 43.1 U/mL |
| 45   | Fujii   | F/60    | Back discomfort | Tail | 3.5 | Multilocular | Ca:19-9: high 52.9 U/mL |
| 46   | Guo-Dong Shan | M/39 | Epigastric pain | Tail | 3.5 | NI | Normal |
| 47   | Anghela Paredes | F/17 | Abdominal pain, nausea, vomiting | Tail | 4.3 | NI | Normal |
| 48   | Hirabayashi | M/38  | Asymptomatic | Tail | 3 | Multilocular | Normal |
| CT | MRI | Preoperative diagnosis | Surgery | Pathology | Final diagnosis |
|----|-----|------------------------|---------|-----------|----------------|
| A cyst lesion, surrounded by a crescent-like solid component with the same | Typical findings of an intrapancreatic accessory spleen | ECIPAS | Laparoscopic SPDP | The cyst was lined by nonkeratinizing stratified squamous epithelium and a few layers of atrophic squamous epithelium. The outside cystic wall was composed of relatively thick fibrous connective tissues. The brown solid component was composed of both red and white pulp, locating in the pancreatic parenchyma | Epidermoid cyst |
| | | | | | |
| The wall of the cyst was relatively regular, thick and enhanced | Cyst: T1 iso, T2 hyper. Rim showed hyperintensity in DWI | SPT | Laparoscopic DP | The cyst was lined by stratified squamous epithelium within splenic parenchyma | Epidermoid cyst |
| | | | | | |
| The densities of the solid component and spleen on enhanced CT were similar | The intensity of the solid component T1 and T2 was similar to that of the spleen | SPT and NET | Laparoscopic SPDP | The solid component included splenic tissue with typical red and white pulp. The cyst was lined with a multilayered of nonkeratinized stratified squamous epithelium without any skin appendage, and the squamous epithelium was covered with a hobnail-like growth epithelium | Epidermoid cyst |
| | | | | | |
| Cystic lesion | NI | NI | Laparoscopic DP | The cyst within an intrapancreatic accessory spleen showing a thin epithelial layer with ectopic splenic tissue | Epidermoid cyst |
| | | | | | |
| A unilocular cystic lesion with same enhancement as the adjacent spleen | T1 low/T2 high | MCN | Laparoscopic SPDP | The cyst wall showed a thin multilayered squamous epithelium, with small patches of splenic epithelium | Epidermoid cyst |
| | | | | | |
| A multilocular cystic lesion, solid component with enhancement similar to the spleen | Low T1 and high T2 | IPMN | Laparoscopic DP | The cyst wall showed a thin multilayered squamous epithelium, with small patches of splenic epithelium | Epidermoid cyst |
| | | | | | |
| A cystic lesion in tail of pancreas | Low T1 and high T2 | Pancreatic cystadenoma | DP | Old hemorrhage in spleen tissue and formation of capsule wall, surrounded by pancreatic tissue | Hematoma |
| | | | | | |
| A cystic lesion in tail of pancreas | NI | MCN,IPMN | Laparoscopic SPDP | A cyst was lined by squamous epithelium arising in an accessory spleen | Epidermoid cyst |
| | | | | | |
| NI | NI | NI | DP | cyst in intrapancreatic accessory spleens are lined by stratified squamous or urothelial epithelium | Epidermoid cyst |

(Continues)
| Case | Authors | Sex/age | Symptom       | Location | Size(cm) | Cyst      | Serum markers | CEA CA19-9 |
|------|---------|---------|---------------|----------|----------|-----------|---------------|------------|
| 49   | Hirabayashi | F/40   | Abdominal pain | Tail     | 3.5      | Multilocular | CA19-9: high, 198.7 U/mL |
| 50   | Hirabayashi | F/39   | Asymptomatic   | Tail     | 2        | Multilocular | CA19-9: high, 31.9 U/mL |
| 51   | Hirabayashi | M/54   | Asymptomatic   | Tail     | 2.7      | Multilocular | Normal         |
| 52   | Hirabayashi | M/55   | Asymptomatic   | Tail     | 3.5      | Multilocular | CA19-9: high, 50.6 U/mL |
| 53   | Hirabayashi | M/36   | Asymptomatic   | Tail     | 13.4     | Multilocular | CA19-9: high, 47.2 U/mL |
| 54   | Matsumoto   | F/40   | Asymptomatic   | Tail     | 1.5      | Multilocular | Normal         |
| 55   | Bo Zhou     | M/32   | Asymptomatic   | Tail     | 3.5      | Multilocular | Normal         |
| 56   | Takagi      | M/73   | Asymptomatic   | Tail     | 2.4      | Multilocular | CA19-9: high, 901 U/mL |
| 57   | Current     | M/43   | Asymptomatic   | Tail     | 1.5      | Multilocular | Normal         |
| Case | Authors | Sex/age | Symptom | Location | Size (cm) | Preoperative diagnosis | Surgery | Pathology | Final diagnosis |
|------|---------|---------|---------|----------|-----------|------------------------|---------|------------|----------------|
| 49   | Hirabayashi | F/40    | Abdominal pain | Tail     | 3.5       | Multilocular           | NI       | NI         | Enucleation Cyst in intrapancreatic accessory spleens are lined by stratified squamous or urothelial epithelium | Epidermoid cyst |
| 50   | Hirabayashi | F/39    | Asymptomatic | Tail     | 2         | Multilocular           | NI       | NI         | DP cyst in intrapancreatic accessory spleens are lined by stratified squamous or urothelial epithelium | Epidermoid cyst |
| 51   | Hirabayashi | M/54    | Asymptomatic | Tail     | 2.7       | Multilocular           | NI       | NI         | Enucleation Cyst in intrapancreatic accessory spleens are lined by stratified squamous or urothelial epithelium | Epidermoid cyst |
| 52   | Hirabayashi | M/55    | Asymptomatic | Tail     | 3.5       | Multilocular           | NI       | NI         | Enucleation Cyst in intrapancreatic accessory spleens are lined by stratified squamous or urothelial epithelium | Epidermoid cyst |
| 53   | Hirabayashi | M/36    | Asymptomatic | Tail     | 13.4      | Multilocular           | NI       | NI         | DP Cyst in intrapancreatic accessory spleens are lined by stratified squamous or urothelial epithelium | Epidermoid cyst |
| 54   | Matsumoto | F/40    | Asymptomatic | Tail     | 1.5       | Multilocular           | NI       | NI         | A multilocular cystic lesion, solid periphery, with the same enhancement as the spleen | Epidermoid cyst |
| 55   | Bo Zhou  | M/32    | Asymptomatic | Tail     | 3.5       | Multilocular           | NI       | NI         | A well-defined cystic neoplasm without enhancing mural nodes | Epidermoid cyst |
| 56   | Takagi    | M/73    | Asymptomatic | Tail     | 2.4       | Multilocular           | NI       | NI         | A cystic mass | Epidermoid cyst |
| 57   | Current   | M/43    | Asymptomatic | Tail     | 1.5       | Multilocular           | NI       | NI         | A cystic lesion in tail of pancreas | Epidermoid cyst |

**Notes:**
- **CT** and **MRI** columns indicate imaging results.
- **Preoperative diagnosis** includes specific characteristics of the cystic lesion.
- **Surgery** column specifies the type of surgical procedure.
- **Pathology** describes the histological findings.
- **Final diagnosis** confirms the confirmed diagnosis post-surgery.
no narrowing or disruption on endoscopic retrograde cholan-

giopancreatography (Figure 3B).

The CT and T1- and T2-weighted MRI data suggested
cyst degeneration, which is a characteristic of pNETs. Hence,
the preoperative diagnosis was a pNET. Distal pancreatec-
tomy was performed with concomitant splenectomy; the
guidelines mentioned above recommend surgery as the first-
line treatment for pNETs.

The postoperative course was uneventful. The excised
surface of a resected specimen indicated a multilocular cyst
with solid nodules (Figure 4A). Histopathological examina-
tion revealed the presence of spleen tissue, both red and white
pulp, in the parenchyma of the pancreatic tail. Hence, we
diagnosed the tumor as an intrapancreatic accessory spleen
cyst, with the tumor originating in the accessory spleen rather
than in the tissue between the tail of the pancreas and the
spleen. The major and minor multilocular cysts had a maxi-
mum diameter of 17 mm. The luminal epithelium consisted
of mature squamous epithelium and subepithelial lymphoid
tissue (Figure 4B), and the cyst lumen contained keratin and
cholesterol deposits in the clefts. The lymphatic tissue occu-
pied the majority of the cyst (Figure 4D), which is a diagno-
sic criterion for LECs. Ultimately, the patient was diagnosed
with an LEC of the pancreatic accessory spleen.

3 | DISCUSSION

An accessory spleen is not uncommon. Halpert et al reported
291 (10.8%) accessory spleens in 2700 autopsied cases; 215
(62.1%) were in the vicinity of the splenic hilum, followed
by 78 (22.5%) in the pancreatic tail. However, a cyst occurr-
ing in a pancreatic accessory spleen is extremely rare, with
only 57 reported cases (Table 1). The lesion site was the
pancreatic tail in all 57 cases. The primary complaints were
abdominal pain and vomiting, although 33 of the 57 cases
were asymptomatic. Tumors with smaller diameters are often
asymptomatic.

The cyst in the accessory spleen in our study was 1.5 cm
in diameter, making it the smallest of those previously re-
ported (range, 15-134 mm), and apparently had no symptoms.
Levels of CEA or CA 19-9 were elevated in some previous
cases, but within normal limits in others. In many cases, the
lesion was asymptomatic and detected by imaging. Cyst mor-
phology was multilocular in 33 cases and unilocular in 15.
Most cysts had low intensity on T1-weighted MRI and high
intensity on T2-weighted MRI. Preoperative diagnoses, such
as an IPMN, pNET, epidermoid cyst, and malignant tumor,
suggest case complexity. The preoperative diagnosis in our
case was a pNET, whereas the histopathologically confirmed
diagnosis was an LEC in the accessory spleen. This is the
first report of an LEC in this location.

Pancreatic LECs are extremely rare, accounting for only
0.5% of pancreatic cysts. Mege et al examined pancreatic
LECs in 91 middle-aged to elderly men (mean age, 55 years;
range, 20-82 years); the lesion was occasionally accompa-
nied by abdominal pain (43%) and an elevated serum CA
19-9 level (55%). Pancreatic cysts are classified as true
cysts, pseudocysts, or cystic neoplasms. LEC is considered
to be a type of true cyst, with a lining of squamous epithe-
ilum and dense subepithelial lymphoid tissue. The cystic
contents are typically white in color and may include kerati-
tized material or cholesterol crystals. Adsay et al classified
cystic lesions covered by the squamous epithelium of the
pancreas as LECs as epidermoid (those occurring in the
subpancreatic epithelium) or dermoid (those with cutaneous
appendages).

The pathological diagnostic criteria for LECs are ambigu-
ous. Presently, the predominant diagnostic criterion of an LEC
is a lumen surface with a stratified squamous epithelium and
abundant lymphoid tissue underneath. The reports so far classi-
fied cysts as guided by Adsay et al: “LECs are characterized
microscopically by stratified squamous epithelium surrounded
by a band of mature lymphoid tissue with intervening well-
formed germinal centers.” Adsay et al also added the following:
“The second type of squamous-lined cyst in the pancreas is the
epidermoid cyst arising in intrapancreatic accessory spleen.”
These investigators did not mention lymphoid-rich cysts in the
accessory spleen. Some of the cases epidermoid cysts described
by Truong et al in 1987 are now thought to be LECs. Owing to
the unclear classification criteria, it is possible that other cases
reported as epidermoid cysts are also LECs.

In the present case, the lumen epithelium of the multilocular
cyst consisted of mature squamous material with developed sub-
epithelial lymphoid tissue. In addition, white and the red pulp
were detected in the pancreatic accessory spleen; hence, the cyst
was diagnosed as a splenic LEC. There are three theories re-
garding the pathogenesis of pancreatic LECs. The first theory
suggests that LECs originate in the misplaced branchial cleft
tissue because of the histologic resemblance. The second sug-
gests that squamous metaplasia in an obstructed pancreatic duct,
which subsequently protrudes into a peripancreatic lymph node,
gives rise to LECs. The third links LECs to cyst development
from an ectopic pancreas in a peripancreatic lymph node. At
present, a consensus has not been reached. Tateyama et al sum-
marized the findings of their immunohistochemical study as
follows: “The cytokeratin phenotypes of the epithelial lining of
LEC were similar to those of branchial cleft cysts. In addition to
the cytokeratin pattern, the presence of some islets and ducts in
the fibrous wall of the LEC might support the second hypothesis.”
In the present case, the LEC was found in the accessory spleen
in the pancreas, which is consistent with the second hypothesis.
However, further consideration and discussion are required.
4 | CONCLUSION

This study is the first report of an LEC in the intrapancreatic accessory spleen. The diagnostic criteria for LECs are ambiguous, and the difference between LECs and epidermoid cysts is unclear. It is necessary to consider LECs from a pathological point of view.

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CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

SH: drafted the manuscript. MH, HN, and TI: supervised the preparation of the manuscript and reviewed and modified the manuscript. YM: contributed to the surgery and reviewed and modified the manuscript. RY: contributed to the pathological diagnosis and reviewed and modified the manuscript. TN: contributed to the pathological diagnosis and TI reviewed and modified the manuscript. All authors read and approved the final manuscript.

ETHICAL APPROVAL

This manuscript has not been published elsewhere, and this treatment strategy has been approved by the appropriate ethics review board.

DATA AVAILABILITY STATEMENT

This manuscript contains all the necessary data in the text.

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