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【 CASE REPORT 】

Rare Case of Pancreatic Cystic Lymphangioma

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Abstract:
Pancreatic cystic lymphangioma is an extremely rare tumor. The characteristic imaging findings are poorly defined, and distinguishing between this disease and other pancreatic cyst-related tumors is very difficult. We herein report a case of a Japanese woman in her 50s with this lesion, located in the tail of the pancreas. Pancreatic cystic lymphangioma should therefore be considered in the differential diagnosis of pancreatic cystic lesions. Laparoscopic resection can be a useful, minimally invasive surgical approach for treating these cysts as well as for the treatment of benign or low-grade malignant tumors located in the pancreatic body or tail.

Key words: pancreatic cystic lymphangioma, rare tumor, laparoscopic resection

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Introduction

Cystic lymphangiomas are congenital benign tumors of the lymphatic system. Cystic lymphangioma usually occurs in the neck, axillary region, and rarely, in the mediastinum, and most frequently occurs in children and young adults. Pancreatic cystic lymphangiomas are extremely uncommon (1-4). Although abdominal ultrasound (US), computed tomography (CT) or magnetic resonance imaging (MRI) can incidentally reveal lymphangioma, the preoperative diagnosis is difficult, as conventional imaging studies cannot characterize these lesions (5-7).

We experienced a case of a pancreatic cystic lymphangioma located in the tail of the pancreas. Given this rare presentation of pancreatic cystic lymphangioma, we initially favored a diagnosis of intraductal papillary mucinous neoplasm (IPMN) or mucinous cystic neoplasm (MCN). We therefore adopted a conservative approach by resecting the lesion in the patient through laparoscopic resection. We identified a few reports of this type of cyst, but they included little evidence concerning the characteristic imaging findings to guide management. Laparoscopic resection can be a useful procedure with low invasiveness for the treatment of benign or low-grade malignant tumors located in the pancreatic body or tail (8).

Case Report

A Japanese woman in her 50s underwent abdominal ultrasonography during a medical checkup, which revealed the enlargement of a pancreatic tail cyst from 25 mm to 36 mm over the previous 1 year (1 year ago, Fig. 1A; this time, Fig. 1B). The patient was admitted to our hospital for the examination of the pancreatic tail cyst. She had no relevant family medical history. Her drinking history was social drinking. She was 159 cm tall and weighed 59 kg. Her blood pressure was 117/76 mmHg, and her pulse was 62 beats/min. She had neither anemia nor jaundice and did not have abdominal pain. No abnormalities were apparent on blood tests, including tumor markers. Abdominal US showed a cyst of about 36 mm in the pancreatic tail near the left kidney. Abdominal CT showed a pancreatic cystic lesion of about 36 mm that projected outward from the pancreatic tail (plain, Fig. 2A; contrast-enhanced CT image, Fig. 2B). On MRI, the cystic lesion that grew outward from the pancreatic tail showed a T1 low signal (Fig. 3A) and T2 high signal (Fig. 3B). Magnetic resonance cholangiopancreatography (MRCP) showed a cystic lesion in the pancreatic tail; no clear nodules were apparent within the cyst. Endoscopic ultrasound (EUS) (GF-UM 2000; Olympus, Tokyo, Japan) revealed a multilocular cystic lesion in the pancreatic tail with thin multiple septa, without a hypoechoic component or mural nodules (Fig. 4). Endoscopic retrograde cholan-

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giopancreatography (ERCP) including a pancreatic juice cytological diagnosis did not show any abnormalities.

We could not rule out branch-duct IPMN or MCN of the pancreatic tail. Because the lesion showed a tendency to enlarge, laparoscopic resection was performed. The surgical finding was a multilocular cystic lesion that grew toward the pancreas from the pancreatic tail (Fig. 5). Macroscopic findings demonstrated a multilocular cyst 35 mm in size with large and small thin septa (Fig. 6). The contents of the cyst resembled chyle. Microscopically, all sections showed a polycystic structure composed of ectatic lymphatics lined with endothelial cells. The cysts were separated by thin hypocellular septa. No cell atypia was found. The cysts had pancreas tissue in the border area and were therefore regarded as lesions that derived from the pancreas [Hematoxylin and Eosin (H&E) staining ×1, Fig. 7A; HE staining ×1, Fig. 7B; HE staining ×40, Fig. 7C]. Immunostaining results indicated that the cyst wall cells were CD31-positive (Fig. 8A) and D2-40-positive (Fig. 8B), which are characteristics of lymph duct endothelial cells, and CK-negative (Fig. 8C), suggesting they were not of epithelial origin.

The patient was diagnosed with pancreatic cystic lymphangioma. The postoperative course was good, and there have been no postoperative recurrences in the 418-day follow-up period, during which time the patient has been followed in the outpatient department.

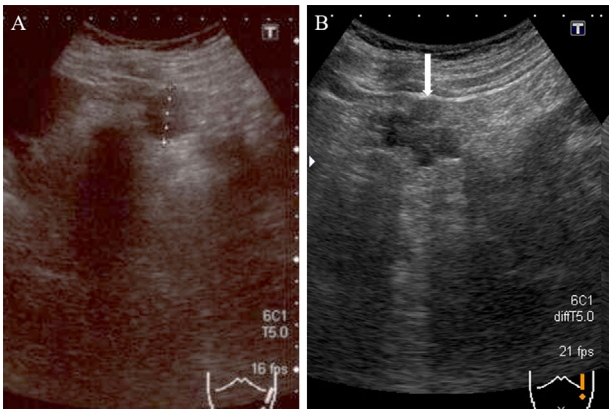

Figure 1. **A Japanese woman in her 50s underwent abdominal ultrasonography during a medical checkup, which revealed enlargement of a pancreatic tail cyst from 25 mm to 36 mm over 1 year. (A) One year ago, (B) this time (arrow).**

Discussion

Lymphangiomas typically arise in the head, neck, and ax-

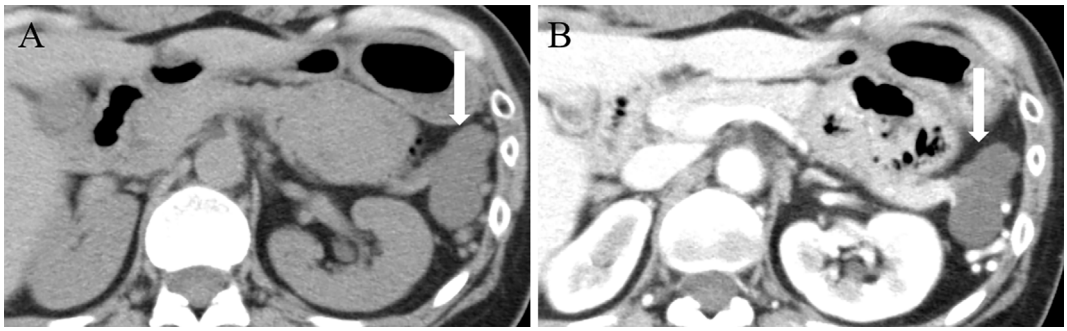

Figure 2. **Abdominal computed tomography (CT) showed a pancreatic cystic lesion of about 36 mm that projected outward from the pancreatic tail. (A) Plain (arrow), (B) contrast-enhanced CT image (arrow).**

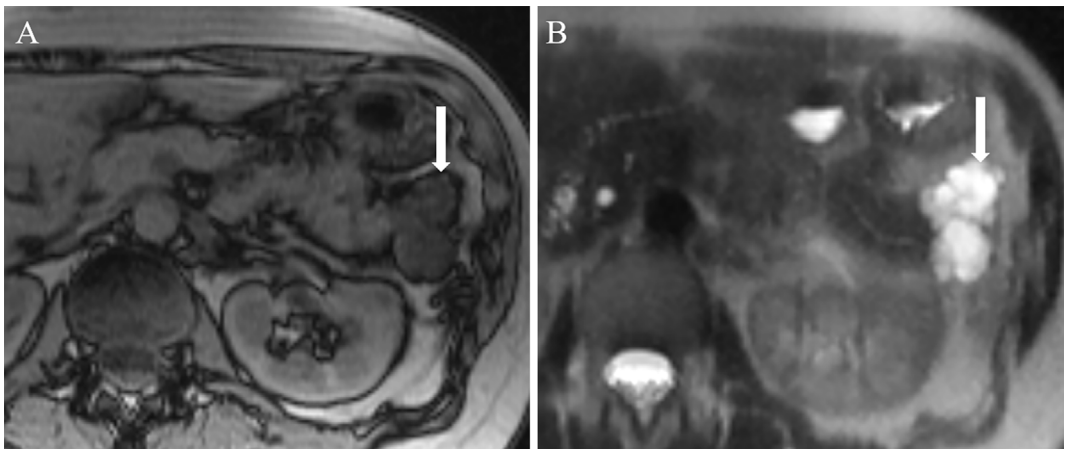

Figure 3. **On magnetic resonance imaging (MRI), the cystic lesion that grew outward from the pancreatic tail showed a T1 low signal (arrow) (A) and T2 high signal (arrow) (B).**

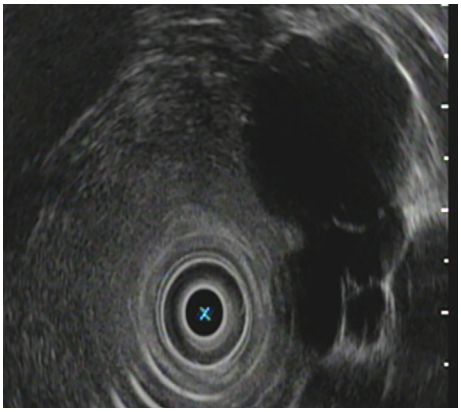

Figure 4. **Endoscopic ultrasound (EUS) revealed a multilocular cystic lesion in the pancreatic tail with multiple thin septa without a hypoechoic component or mural nodules.**

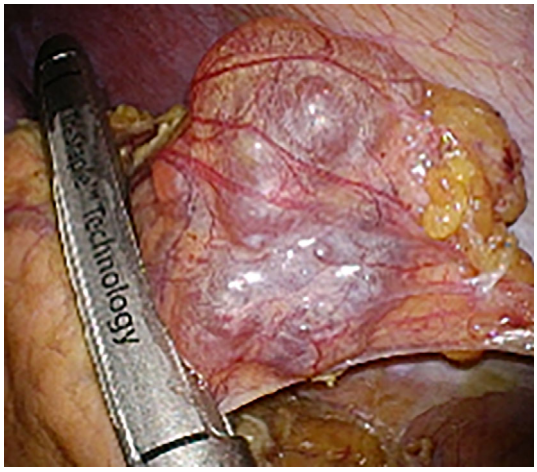

Figure 5. **The surgical finding was a multilocular cystic lesion that grew toward the pancreas from the pancreatic tail.**

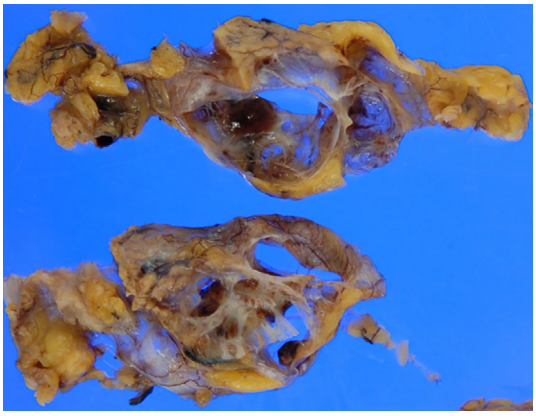

Figure 6. **Macroscopic findings demonstrated a multilocular cyst 35 mm in size with large and small thin septa.**

illary regions (95%). They are benign cystic tumors (1-4) and are classified into three types: simple capillary, cavernous, and cystic (5, 9). While the etiology of lymphangiomas is not completely clear, the most popular theory suggests that lymphangiomas may be the result of congenital malformations of the lymphatic channels or of lymphatic obstruction secondary to inflammation (5, 10). Pancreatic cystic lymphangiomas comprise an extremely rare subgroup and account for <1% of all lymphangiomas and only 0.2% of pancreatic lesions. The first reported case of pancreatic lymphangioma was published by Koch in 1913 (1). Since then, to our knowledge, 82 cases have been reported in the English medical literature (11, 12). Pancreatic cystic lymphangiomas are composed of dilated cystic spaces lined by flattened endothelium containing abundant lymphoid tissue and smooth muscle in the wall of the cyst.

Pancreatic cystic lymphangiomas are usually large lesions with an average diameter of 12.3 cm (13). Pancreatic lymphangioma are usually encountered in young females and are often located in the body and tail of the pancreas (3, 14, 15). The female preponderance can be partially explained by the positive effects of oral contraceptives, hyperprogesteronemia, and pregnancy in promoting the growth of lymphangioma (16, 17).

Patients are primarily asymptomatic. A number of cases reported asymptomatic lymphangiomas that were diagnosed incidentally during a workup for unrelated diseases (18). There have been no reported cases of malignant transformation of pancreatic/peripancreatic lymphangiomas, so a conservative approach and expectant follow-up remains the best plan for asymptomatic patients (19). Ten cases of cystic pancreatic lymphangiomas diagnosed only with endoscopic ultrasound-guided fine needle were reported and followed. In one case, complete aspiration of the cyst contents was performed, but symptoms recurred after four months. Surgical resection was ultimately performed (20).

However, when complicated, anemia, pain, hemorrhaging, nausea, hydronephrosis, and/or infection may also be present (14). If patients have symptoms that necessitate therapeutic intervention, surgical resection is the best choice. Lymphangioma rarely but occasionally recur for young patients, so complete resection is required to reduce the risk of recurrence. However, because it is a benign tumor, caution must be exercised during excessively aggressive surgeries. Recurrence can still occur even after complete resection, although the rate is extremely low (approximately 7%) (21).

A laparoscopic approach is reported to be a safe and effective alternative for the treatment of pancreatic cystic lymphangioma. This minimally invasive technique also improves patient comfort with little postoperative pain, rapid recovery after surgery, and minimal scar formation. Laparoscopic excision should therefore be considered as a therapeutic option, especially for treating uncomplicated pancreatic cystic lymphangiomas (18, 21).

Histologically, pancreatic cystic lymphangiomas consist of interconnecting cysts separated by thin septa lined by thin and flat epithelial cells containing serous, serosanguineous, or chylous fluid (22, 23). Immunohistochemistry can be used to increase the accuracy of the diagnosis. Since the endothelial cells of the outer membrane of pancreatic cystic lymphangiomas do not normally undergo epithelial differen-

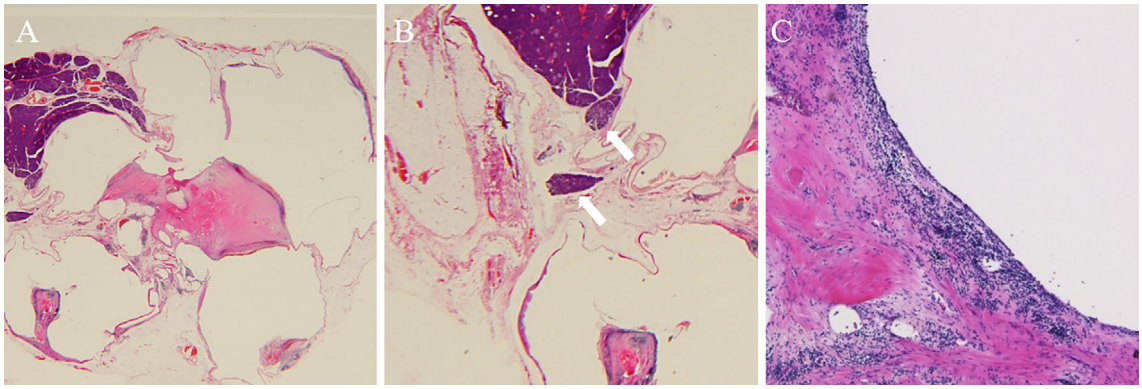

Figure 7. **Microscopically, all of the sections showed a polycystic structure composed of ectatic lymphatics lined with endothelial cells. The cysts were separated by thin hypocellular septa. No cell atypia was found. The cysts had pancreas tissue in the border area and were therefore regarded as lesions derived from the pancreas. (A) Hematoxylin and Eosin (H&E) staining ×1, (B) H&E staining ×1, and (C) H&E staining ×40.**

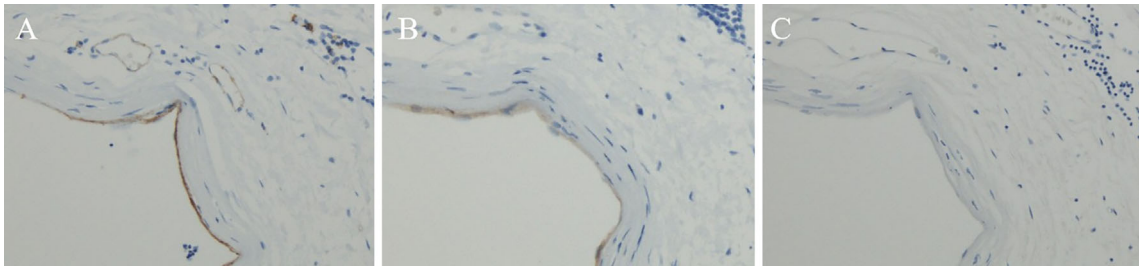

Figure 8. **Immunostaining results indicated that the cyst wall cells were CD31-positive (A) and D2-40-positive (B), which are characteristics of lymph duct endothelial cells, and CK-negative (C), suggesting they were not of epithelial origin.**

tiation, staining agents such as Periodic Acid-Schiff (PAS) stain, Mucicarmine, Alcian blue, and anti-cytokeratin are unsuitable. However, staining with immunohistochemistry reagents such as Factor VIII-R Ag, CD31, CD34, and D2-40 is helpful in the diagnosis of the disease (3, 24).

Pancreatic cystic lymphangiomas have a broad differential diagnosis, including pseudocysts, simple cysts, serous cystadenomas, MCNs, and IPMNs. Pancreatic cystic lymphangioma may clinically present as a palpable abdominal mass and cause diagnostic confusion with respect to other retroperitoneal cystic tumors, including those arising from the kidney, and pancreas. Although abdominal US, CT and MRI also incidentally reveal lymphangioma, the preoperative diagnosis is very difficult, as conventional imaging studies cannot fully characterize this lesion (5-7). CT scans show well circumscribed, encapsulated, water-dense, polycystic tumors within septa that are hardly distinguishable from cystadenomas or other cystic neoplasms. MRI may not provide a better morphological characterization of the mass than CT (7). We also were unable to diagnose the present case preoperatively.

However, some authors advocate the usefulness of EUS and EUS-fine-needle aspiration (FNA) in the preoperative workup of suspected lymphangiomas. In EUS imaging, a pancreatic lymphangioma usually appears as a well-defined, multi-cystic cavity with thin septae (20). The chylous appearance of the aspirated fluid with elevated triglycerides confirms the diagnosis of lymphangioma (6, 22, 25). However as a cautionary note, EUS-FNA of a pancreatic cyst may result in complications such a bleeding and infection (20). In our patient, we did not perform EUS-FNA since we did not suspect lymphangioma due to suspicion of IPMN or MCN.

To prevent unnecessary surgical intervention, it is important to recognize pancreatic cystic lymphangioma as a differential diagnosis of pancreatic cyst, as there are few specific imaging findings that can confirm the diagnosis of pancreatic cystic lymphangioma, contributing to the difficulty in distinguishing this entity from other cystic neoplasms of the pancreas. We chose to perform surgery with informed consent that would include adequate resection in the event the lesion was MCN or IPMN.

In conclusion, we encountered a case of pancreatic cystic lymphangioma after laparoscopic resection. This procedure has the advantage of being a minimally invasive approach for the treatment of benign or low-grade malignant tumors located in the pancreatic body or tail (8). Although these lesions are rare, it is important to consider this disease in the differential diagnosis of pancreatic cystic lesions.

The authors state that they have no Conflict of Interest (COI).

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