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CASE REPORT

췌장의 장액낭샘종으로 오인된 췌장의 샘꽈리세포낭종 1예

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Pancreatic Acinar Cell Cystadenoma Mimicking Pancreatic Serous Cystadenoma

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Acinar cell cystadenoma, also known as an acinar cystic transformation of the pancreas, is an exceedingly rare but benign pancreatic lesion. A 51-year-old woman was transferred to Inje University Busan Paik Hospital because of an 8 cm-sized calcified, multiseptated, and multilocular cystic mass in the pancreatic tail observed during abdominal CT performed at another hospital. The patient did not complain of abdominal pain or other symptoms, and her laboratory findings were normal. MRI showed that the cyst was not connected to the main pancreatic duct. A pancreatic serous cystadenoma was suspected, and a laparoscopic distal pancreatectomy was performed. The resected mass was composed of variable sized multilocular cysts with incomplete septa and focally lined by epithelium with acinar differentiation. The patient was diagnosed with acinar cell cystadenoma and is currently being followed up regularly. No complications or recurrences have been observed. **(Korean J Gastroenterol 2021;78:138-143)**

Key Words: Acinar cells; Cysts; Cystadenoma; Pancreas

INTRODUCTION

Acinar cell neoplasms of the pancreas are quite rare and include acinar cell carcinoma, acinar cell cystadenocarcinoma, and acinar cell cystadenoma.[1] Among them, acinar cell cystadenoma, also known as an acinar cystic transformation of the pancreas, is a cystic lesion of the pancreas that is very uncommon, benign, and shows evidence of acinar differentiation.[1,2] This paper reports a case of acinar cell cystadenoma, which was misrecognized as a serous cystadenoma of the pancreas.

CASE REPORT

A 51-year-old woman with no significant medical history other than dyslipidemia was transferred to gastroenterology of Inje University Busan Paik Hospital after an 8 cm-sized calcified, multiseptated, and multilocular cystic mass was observed in the pancreatic tail on a CT scan performed in another hospital (Fig. 1).

There were no abdominal symptoms, such as pain, and the tumor marker test was negative at CEA 0.904 ng/mL (reference value ≤4.7 ng/mL), CA 19-9 12.8 U/mL (reference value ≤34 U/mL). The patient was neither a drinker nor a smoker and had no significant family history.

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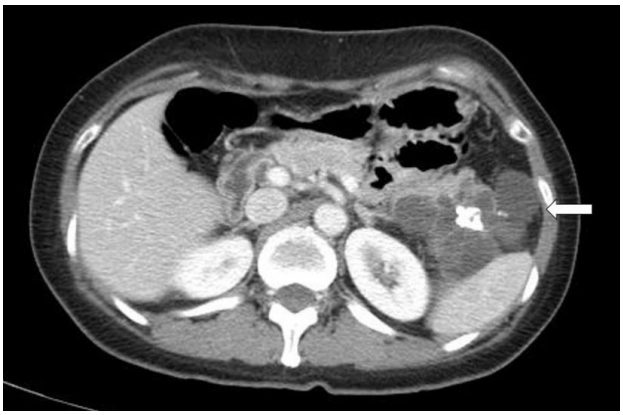

Fig. 1. Abdominal computed tomography revealed an 8 cm-sized calcified, multiseptated, and multilocular cystic mass in the pancreatic tail (arrow).

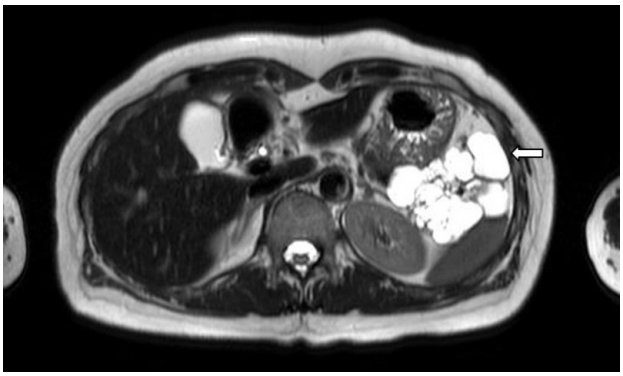

Fig. 2. Pancreatic magnetic resonance imaging (T2-weighted axial image) revealed an 8 cm-sized lobulated multilocular cyst in the pancreatic tail (arrow).

An 8 cm-sized lobulated and multilocular cyst was observed in the pancreatic tail, which was not connected to the main pancreatic duct on MRI performed after she was transferred (Fig. 2).

Serous cystadenoma of the pancreas as large as 8 cm was suspected, and as the patient was anxious, surgery was requested for postoperative confirmation.

The preoperative blood test results were in the normal range as follows: white blood cell count 5,770/mm^3 (reference value 4,000-10,000/mm^3), hemoglobin 13.9 g/dL (reference value 12.0-16.0 g/dL), total bilirubin 1.1 mg/dL (reference value 0.2-1.2 mg/dL), AST 21 U/L (reference value 13-33 U/L), ALT 16 U/L (reference value 6-27 U/L), ALP 200 U/L (reference value 115-359 U/L), urea nitrogen 19 mg/dL (reference value 8-22 mg/dL), creatinine 0.52 mg/dL (reference value 0.6-0.9 mg/dL), sodium 145 mmol/L (reference value 138-145 mmol/L), potassium 4.2 mmol/L (reference value 3.6-4.8 mmol/L), and chloride 105 mmol/L (reference value 101-108 mmol/L).

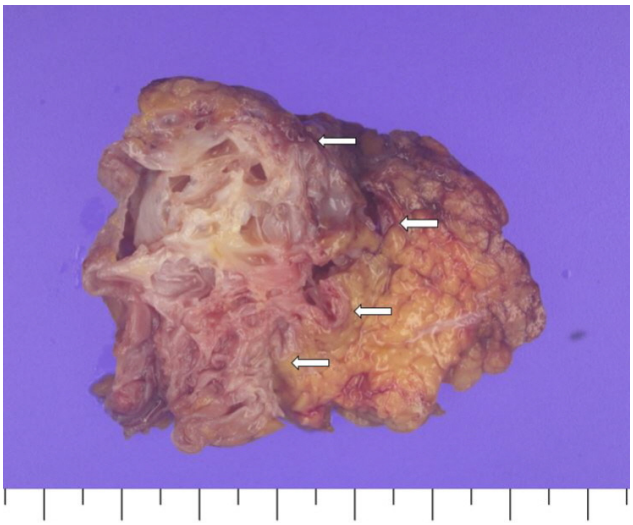

Fig. 3. Distal pancreatectomy specimen showed well delineated mutilocular cystic mass measuring 7.0×4.0×3.0 cm (arrows indicate mass boundaries).

Laparoscopic distal pancreatectomy was performed, and a 7.0×4.0×3.0 cm-sized multilocular cystic tumor was observed during a visual examination. The tumor was well distinguished from the normal pancreatic tissue, and the cyst wall was thin and translucent (Fig. 3).

Microscopically, the tumor was composed of variable sized multilocular cysts with incomplete septa. The cysts were interconnected and focally lined by epithelium with acinar differentiation (Fig. 4). The patient was diagnosed with acinar cell cystadenoma based on a histologic findings. The patient is currently being followed up regularly, and there have been no complications or recurrence since surgery.

DISCUSSION

Acinar cell cystadenoma is a benign cystic lesion of the pancreas that shows acinar cell differentiation defined as the generation of zymogen granules, including the pancreatic exocrine enzyme.[3] The condition is also known as an acinar cystic transformation of the pancreas and is very rare.

Klöppel[4] described an acinar cystic transformation in 2000, and Albores-Saavedra[5] proposed acinar cell cystadenoma to describe a 9 cm-sized multilobular pancreatic cyst found dur-

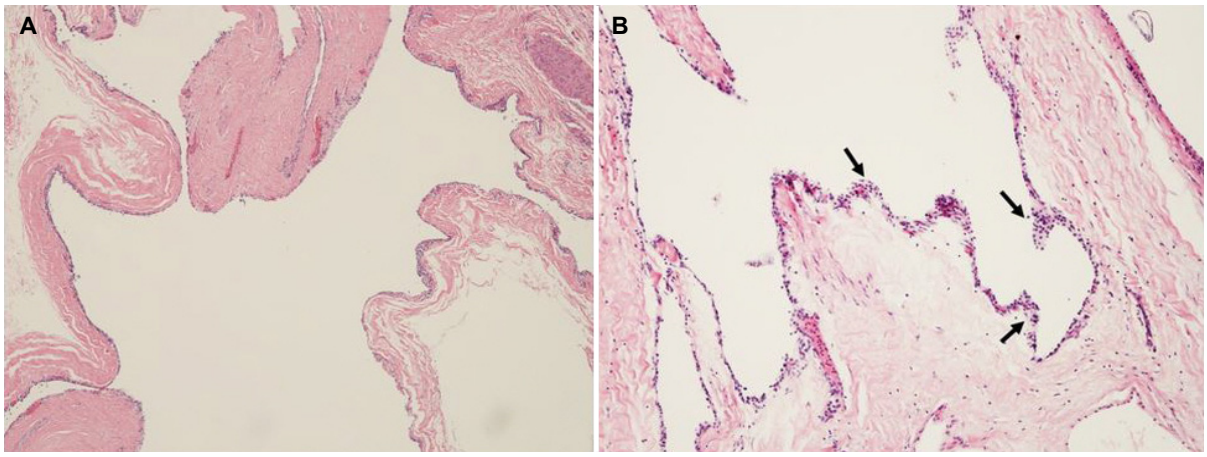

Fig. 4. (A) Microscopic findings revealed variable sized multilocular cysts with incomplete septa (H&E,×100), and (B) focally linned by epithelium with acinar differentiation (arrows) (H&E,×100).

ing the autopsy of a 58-year-old woman in 2002. The etiology of acinar cell cystadenoma is unclear; it is more prevalent in women and occurs across different age groups.[2,6] It can occur in any part of the pancreas but tends to be found more commonly in the head. It is mostly localized but may exist as diffuse lesions in approximately 10% of patients.[2,7] Acinar cell cystadenoma is diagnosed based on a combination of the clinical features, radiological features, and histopathological findings. Clinically, abdominal pain is the most common symptom, but it is often discovered incidentally during an imaging test.[2,8]

The imaging features are not specific, but Delavaud et al.[9] suggested that they are more closely related to acinar cell cystadenoma than to branch duct intraductal papillary mucinous neoplasia based on the following imaging findings: 1) five or more cysts, 2) clustered small, peripheral cysts, 3) presence of cyst calcification, and 4) absence of communication with the main pancreatic duct. The modality showed 100% sensitivity and 60% specificity when at least one criterion is satisfied, 100% sensitivity and 85% specificity when at least two criteria are satisfied, 85% sensitivity and 100% specificity when at least three criteria are satisfied, and 60% sensitivity and 100% specificity when all four criteria are satisfied. This case satisfied three of the imaging findings: five or more cysts, presence of cyst calcification, and absence of communication with the main pancreatic duct.

Although EUS-guided fine needle aspiration (FNA) is used to diagnose approximately 75% of pancreatic cystic tumor patients, it may show false-negative results because of small specimen size, sampling error, and lack of preservation of the tissue architecture. EUS-guided FNA may be helpful in a differential diagnosis before surgery, but the diagnosis is mostly confirmed based on the histopathological testing of the surgical specimen.[3]

Histopathologically, the cyst wall consists of cells with acinar cell differentiation that lack nuclear mitotic figures, distinct cellular atypia, necrosis, and infiltrative growth.[3] The presence of intracellular eosinophilic zymogen granules is an excellent pathological indicator that the lesion may be an acinar cell cystadenoma.[10]

Acinar cell cystadenoma and serous cystadenoma are difficult to distinguish, especially those with a multilocular and microcystic pattern, as in the present case. Acinar cell cystadenoma is characterized by interconnected and dilated acinar epithelium. The cysts are lined by 1-2 layer flatten or cuboidal epithelium with acinar differentiation.

In immunohistochemical staining, cells in the cyst wall differ from the normal cells. They are positive for acinar cell differentiation markers, such as trypsin and chymotrypsin, and also for CK7, which is negative in normal acinar cells.[11]

Caution should be taken when distinguishing acinar cell cystadenoma from acinar cell cystadenocarcinoma during a differential diagnosis. Acinar cell cystadenoma can be distinguished from acinar cell cystadenocarcinoma because it shows very low positivity in Ki-67 staining, no dysplasia, and no infiltration into the surrounding tissues.[2]

A surgical resection is recommended to exclude other cystic neoplasms associated with malignant tumors, prevent local

Table 1. Clinical Features of the Reported Cases of Pancreatic Acinar Cell Cystadenoma

Case no.	Age (years)	Sex	Size (cm)	Location	Abdominal pain	Treatment
1	39	F	2.5	Entire pancreas	Present	Total pancreatectomy
2	57	M	5.7	Body	Present	Distal pancreatectomy
3	27	F	15.0	Entire pancreas	Present	Total pancreatectomy
4	52	F	6.0	Head	Present	Pancreaticoduodenectomy
5	59	F	1.8	Head	Present	Pancreaticoduodenectomy
6	53	F	9.3	Head	Present	Pancreaticoduodenectomy
7	36	F	4.0	Head	Present	Pancreaticoduodenectomy
8	18	F	10.0	Head	Present	Pancreaticoduodenectomy
9	27	F	7.5	Tail	Present	Laparoscopic distal pancreatectomy
10	41	F	6	Head	Present	Pancreaticoduodenectomy
11	52	F	5.1	Body and tail	Present	Distal pancreatectomy
12	39	M	17.8	Tail	Present	Distal pancreatectomy
13	67	F	9.7	Head	Present	Pancreaticoduodenectomy
14	33	M	6.5	Neck	Present	Pancreaticoduodenectomy
15	56	F	14.2	Tail	Present	Distal pancreatectomy
16	62	M	5.3	Neck and body	Absent	Central pancreatectomy
17	48	F	19.7	Body and tail	Present	Distal pancreatectomy
18	65	F	3.0	Body	Present	Distal pancreatectomy
19	33	F	10	Head	Present	Pancreaticoduodenectomy
20	46	F	4 (head); 10 (tail)	Head and tail	Present	Internal drainage
21	16	F	7.5	Head	Present	Pancreaticoduodenectomy
22	44	F	0.1-1.5	Entire pancreas	Absent	Total pancreatectomy
23	47	F	2.5 (head); 0.5 (tail)	Head and tail	Present	Pancreaticoduodenectomy
24	39	F	4	Head	Present	Pancreaticoduodenectomy
25	49	F	0.5	Tail	Absent	Distal pancreatectomy
26	57	M	0.5	Tail	Absent	Distal pancreatectomy
27	66	M	0.2	Head	Absent	Distal pancreatectomy
28	61	M	0.2	Head	Absent	Distal pancreatectomy
29	52	M	5	Head and body	Absent	Distal pancreatectomy
30	9	M	11.7	Entire pancreas	Absent	Laparoscopic biopsy
31	52	M	5	Body	Present	Pancreaticoduodenectomy
32	58	F	9	Body and tail	Absent	Autopsy
33	25	M	6	Head	Present	Pancreaticoduodenectomy
34	54	F	1.5	Body and tail	Absent	Laparoscopic distal pancreatectomy
35	50	F	7.5	Head and neck	Absent	Pancreaticoduodenectomy
36	33	M	1.7	Tail	NI	NI
37	38	M	2.4	Head	NI	NI
38	46	F	3	Body	NI	NI
39	33	M	4.6	Head	NI	NI
40	63	F	2	Head	NI	NI
41	19	F	3.6	Tail	Present	Distal pancreatectomy
42	25	M	NI	Entire pancreas	Present	Total pancreatectomy
43	46	F	NI	Entire pancreas	Absent	Total pancreatectomy
44	62	F	NI	Head	Absent	Pancreaticoduodenectomy
45	61	M	NI	Entire pancreas	Absent	Total pancreatectomy
46	52	M	5	Body	Present	Distal pancreatectomy

Table 1. Continued

Case no.	Age (years)	Sex	Size (cm)	Location	Abdominal pain	Treatment
47	40	F	4	Head	Present	Pancreaticoduodenectomy
48	52	M	5	Head and body	Absent	Pancreaticoduodenectomy
49	13	F	4.6	Body and tail	NI	NI
50	47	F	1.1	Tail	NI	NI
51	65	F	2.1	Head	NI	NI
52	42	F	3.0	Head	NI	NI
53	77	M	2.5	Neck	NI	NI
54	59	F	0.5	Tail	NI	NI
55	37	M	3.0	Head, body, and tail	Present	Distal pancreatectomy
56	40	F	4.5	Entire pancreas	NI	Pancreaticoduodenectomy
57	22	F	6.3	Tail	Present	Distal pancreatectomy
58	14	M	2.6	Tail	Present	Laparoscopic distal pancreatectomy
59	42	F	Up to 2	Head and body	Absent	Pancreaticoduodenectomy
60	23	F	6	Head	Present	Pancreaticoduodenectomy
61	65	M	6.9	Head	Present	Pancreaticoduodenectomy
62	25	F	2.9	Body	Present	Central pancreatectomy
63	68	M	3.5	Tail	Absent	Laparoscopic distal pancreatectomy
64	71	F	5.1	Head and neck	Absent	Pancreaticoduodenectomy
65	67	F	5	Head	Absent	Pancreaticoduodenectomy
66	59	F	3.2	Head and neck	Absent	Pancreaticoduodenectomy
67	57	F	1.5	Tail	Present	Distal pancreatectomy
68	53	F	5.8	Head	Absent	Pancreaticoduodenectomy
69	25	F	6.2	Head	Present	Pancreaticoduodenectomy
70	51	F	7.0	Tail	Absent	Laparoscopic distal pancreatectomy

Patient 70 is our additional experience.
F, female; M, male; NI, not identified.

expansion or malignant transformation of the cyst, and relieve the symptoms.[8] The prognosis is good, and malignant transformations or recurrences have not been reported.[3]

The PubMed database was searched from 2000 until the present using a keyword search for “acinar cell cystadenoma” and “acinar cystadenoma” and 69 patients with acinar cell cystadenoma were identified (Table 1).[1,3,5-8,10,12-23]

This was the case of an 8 cm-sized cystic tumor in the pancreatic tail that was detected accidentally during an abdominal imaging test in a 51-year-old woman without any symptoms. The diagnosis of acinar cell cystadenoma was made after surgery, while serous cystadenoma of the pancreas was suspected from the image. Although only a few cases are being reported as acinar cell cystadenoma, which is a very rare disease, it should be considered in a differential diagnosis of cystic tumors of the pancreas. Here, the authors report one case of acinar cell cystadenoma that was misrecognized as a serous cystadenoma of the pancreas.

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