

## Mucinous Cystic Neoplasm of the Pancreas

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A rare case of mucinous cystic neoplasm of the pancreas in a 38-year-old woman is reported herein. Clinical course and physical findings showed the possibility of mucinous cystic neoplasm, particularly in young woman. Furthermore, an accurate preoperative diagnosis was made based on findings showing a large cyst with thin wall and mural nodules by abdominal ultrasonography and CT scan as well as elevation in serum levels of CA 19-9, CEA and elastase 1 by laboratory tests. A distal pancreatectomy was performed with splenectomy and lymph nodes dissection. Mucinous cystic neoplasm of the pancreas is usually recognized to be low malignant potential. In cystic diseases of the pancreas, differentiating mucinous cystic neoplasm from other cystic diseases, particularly pseudocyst is important. In this paper, we discussed the differentiation between mucinous cystic neoplasm and pseudocyst.

**Key words :** Mucinous cystic neoplasm, Cystadenoma, Cystadenocarcinoma, Pancreas

### INTRODUCTION

Detection of cystic lesions of the pancreas is increasing with recent advances in diagnostic imaging procedures. A large number of studies have dealt with the classification of cystic lesions of the pancreas<sup>1-4</sup>. Mostly, cystic lesions were divided into retention cyst (pseudocyst etc.), serous cystadenoma, mucinous cystic neoplasm (mucinous cystadenoma and cystadenocarcinoma), mucin-producing pancreatic carcinoma, and secondary cyst due to pancreatic carcinoma. In those lesions, it is reported that most of large cysts of the pancreas were pseudocyst or mucinous cystic neoplasm<sup>5,6</sup>. Usually, serous cystadenomas and pseudocysts are benign, but mucinous cystadenomas are thought to coexist with, or transform into, mucinous cystadenocarcinoma<sup>7</sup>. Mucinous cystadenomas should therefore be resected with complete local clearance, unlike the management of pseudocysts or serous cystadenomas. Accordingly, in the case of large cyst of the pancreas, differentiation of mucinous cystic neoplasm from pseudocyst is important because of the malignant potential of mucinous cystic neoplasm. In this report, we described an relatively younger woman with mucinous cystic neoplasm (mucinous cystadenocarcinoma) of the pancreas, which was diagnosed preoperatively by the diagnostic imaging procedures and others.

### CLINICAL COURSE

A 38-year-old woman complaining of dull pain in the upper abdomen that had been present for a few months. She denied weight loss, anorexia, and changes in bowel habits. There was no history of pancreatitis, abdominal trauma, or alcohol abuse.

On physical examination, a mobile tender mass was palpable in the left hypochondrium; it

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measured  $9 \times 10$  cm in size, with smooth rounded margin, and did not show respiratory movement. Bowel sounds were normal. The liver was not enlarged, nor were other abdominal masses noted.

The following routine laboratory tests on admission were all within normal limits: the hemogram, white cell count, coagulation studies, serum levels of electrolytes, albumin, calcium triglycerides, total bilirubin, alkaline phosphatase and transaminase. On the contrary, serum levels of amylase, carcinoembryonic antigen (CEA), carbohydrate antigen (CA19-9) concentration and elastase 1 elevated, with values being (and they were) 452 IU/L (normal 160 IU/L), 12 ng/ml (normal 5.0 ng/ml), 130 U/ml (normal 37 U/ml) and 460 ng/dl (normal 400 ng/dl), respectively.

A plane abdominal roentgenogram and gastrointestinal tract series demonstrated the displacement of the transverse colon by a space-occupying lesion in the upper abdomen (Fig. 1). Ultrasonography and CT scan of the abdomen showed a large and unilocular cyst with thin wall in the tail of the pancreas (Fig. 2-A, B). The abdominal CT scan demonstrated a  $7.5 \times 8.5$  cm single cyst with mural nodules in the region (Fig. 2-B). Visceral angiography showed shifting of the inferior pancreaticoduodenal artery and hypovascularity of the cyst, but was otherwise unremarkable (Fig. 2-C). An endoscopic retrograde cholangiopancreatogram (ERCP) showed displacement of the main pancreatic duct by the cyst and communication between the main duct and cyst (Fig. 2-D). However, dilatation of the main pancreatic duct was not observed. The papilla of Vater was normal. A preoperative diagnosis of mucinous cystic neoplasm (mucinous cystadenoma or cystadenocarcinoma) of the pancreas was made on the basis of clinical course, specific image findings (ultrasonography, CT and ERCP) and elevated tumor markers described above.

Laparotomy was performed under general anesthesia. The primary lesion was identified as a spherical smooth mass was found arising from the tail of the pancreas, but not fixed to any adjoining structures (Fig. 3-A). The head of the pancreas was free from the mass. A distal pancreatectomy was done with splenectomy and lymph nodes dissection. The abdomen was closed with drainage.

### Pathological Findings

The distal pancreatectomy specimen consisted of spleen, omental fat and the pancreas (Fig. 3-B). The mass measured approximately 10 cm in diameter and, on sectioning, 1.2 L of dark mucinous fluid was evacuated from cystic cavity (Fig. 3-B). Levels of CEA and CA 19-9 in the fluid



Fig. 1 Gastrointestinal tract series on barium studies. The transverse colon is compressed by the mass in the upper abdomen (arrow).

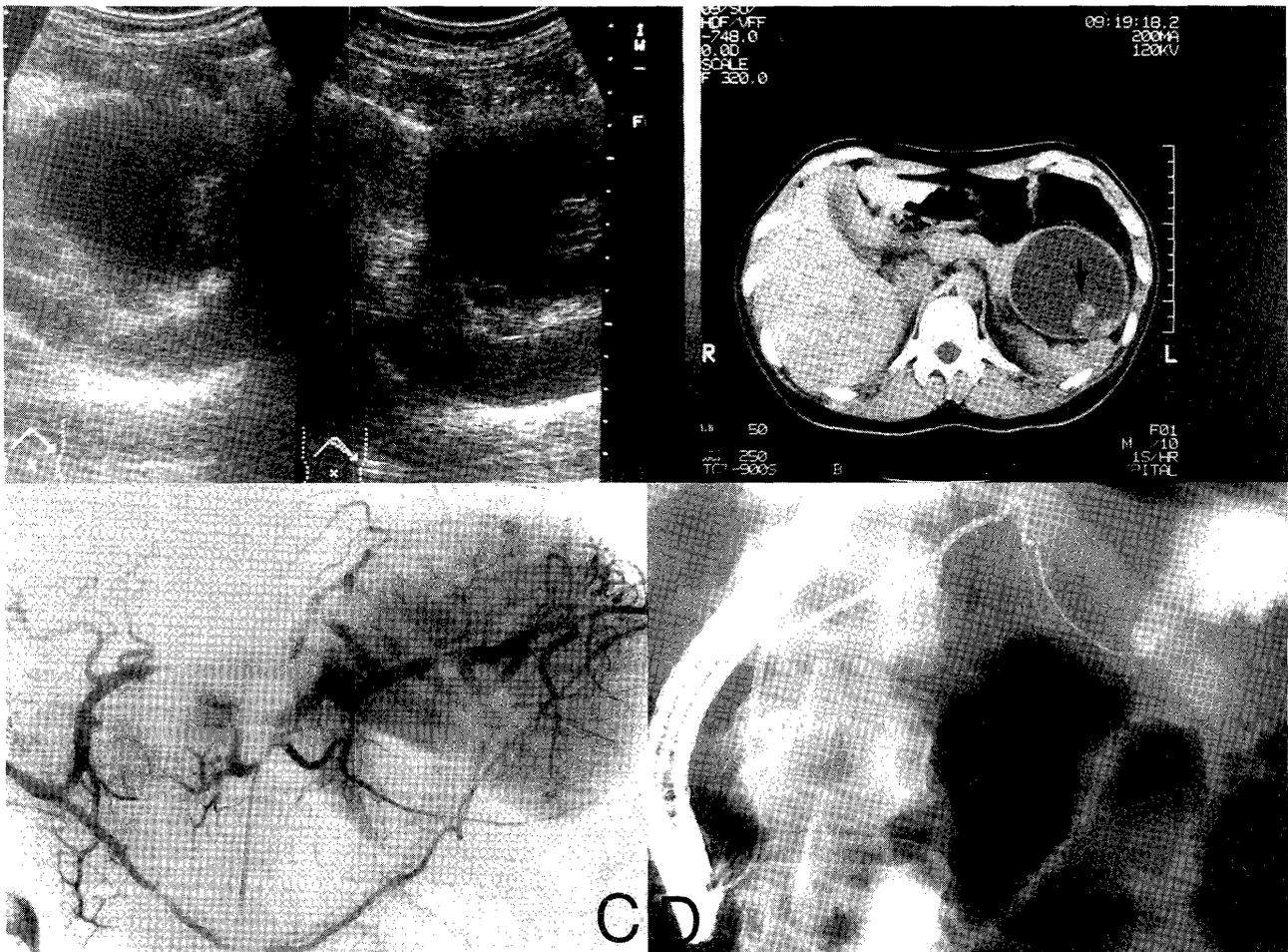


Fig. 2 Abdominal ultrasonography (A) and CT scan (B) show unilocular cyst with thin wall in the tail of the pancreas. The cystic lesion contains mural nodule (arrow) on CT scan. (C): The hypovascular mass and shifting of the inferior pancreaticoduodenal artery on celiac angiography. (D): ERCP shows displacement of the main pancreatic duct, communication with the cyst.

were elevated as high as 4300 ng/ml and 7700 U/ml, respectively. The cyst communicated with the main pancreatic duct (Fig. 3-C). The mass was unilocular, and the thin and irregular cyst wall consisted of excrescent projects (Fig. 3-D).

Microscopically, the cystic wall showed papillary structures composed of double layers of columnar cells with dysplasia, and lined by mucus-producing columnar epithelia. Pathologic examination of the surgical specimen showed a cystadenocarcinoma of the pancreas with margins free. Dissected lymph nodes were free of morphologic abnormality.

## DISCUSSION

Most cystic lesions of the pancreas are pseudocysts, and uncommon are mucinous cystic neoplasms<sup>5,6</sup>. About 100 cases of mucinous cystic neoplasms of the pancreas have been reported since first described by LICHTENSTEIN in 1934<sup>8</sup>, which occur mainly in adolescent and young adult females. Mucinous cystic neoplasms of the pancreas were divided into mucinous cystadenoma

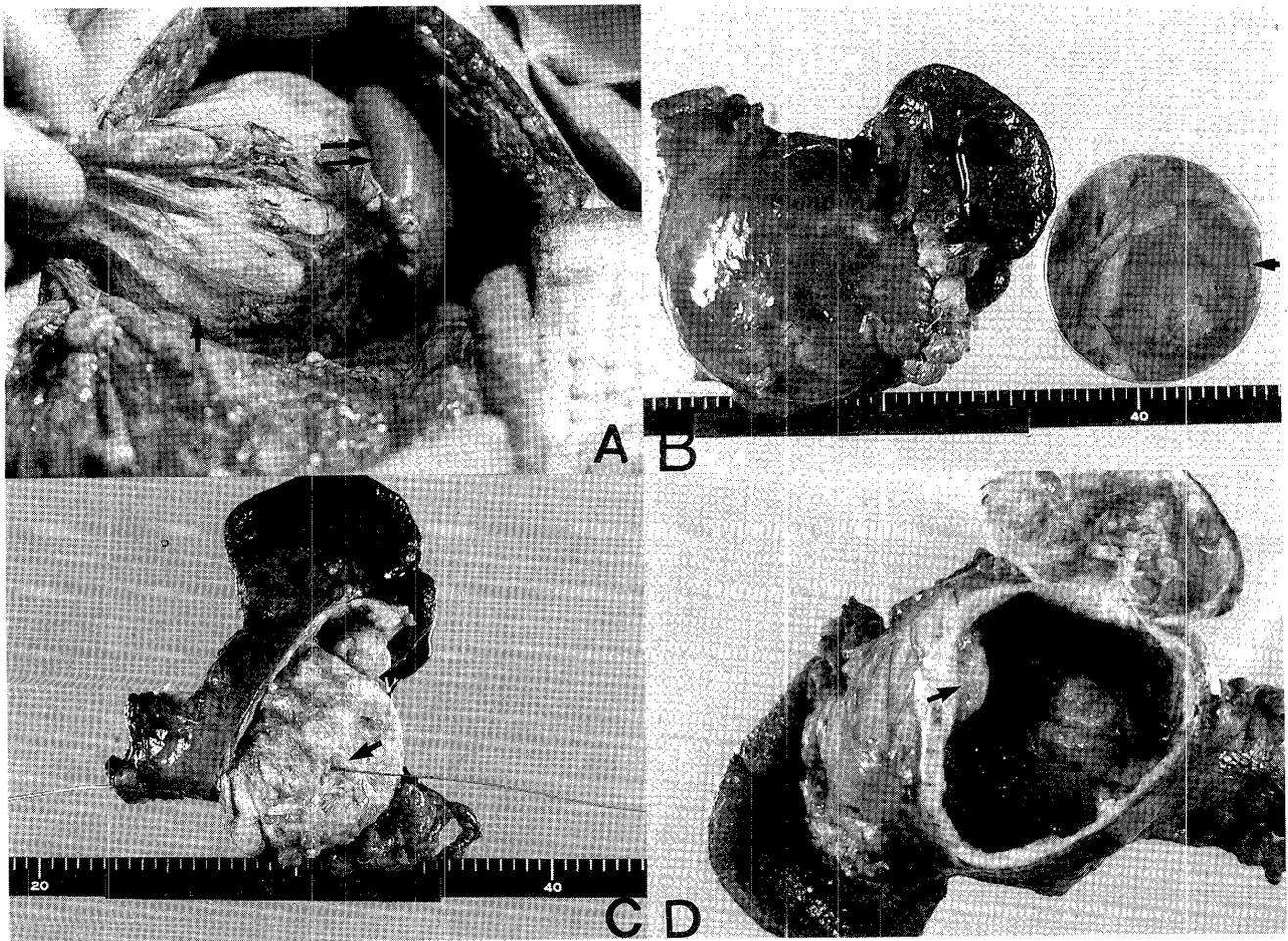


Fig. 3 (A): Exploratory laparotomy reveals a spherical smooth cyst in the tail of the pancreas (arrow) and spleen (double arrows). (B): The resected specimen reveals unilocular cyst with sticky mucus. Mucus in culture dish (arrow). (C): The pin shows the communication between the main pancreatic duct and the cyst cavity (arrow). (D): Excrescent project arising the wall (arrow).

(covertly or potentially malignant) and cystadenocarcinoma. Serous cystadenoma in the pancreas seldom was found, and mostly benign and microcystic adenoma<sup>9,10</sup>.

Pseudocysts are inflammatory or traumatic origin, and occasionally will resolve spontaneously. Excision is usually infeasible, except for small pseudocysts located distally in the tail of the pancreas, and therefore, most are treated by drainage. In contrast, mucinous cystic neoplasms of the pancreas should be excised totally whenever possible because of the malignant potential of the disease. Therefore, differentiating mucinous cystic neoplasm from pseudocyst is important when a large cyst is detected in the pancreas<sup>8</sup>.

Clinical course, ultrasonography, CT, ERCP, angiography, tumor marker measurement and fine-needle aspiration cytologic study are useful in differentiation between pseudocyst, mucinous cystic neoplasm and others<sup>11~14</sup>.

Pseudocysts are often associated with an episode of clinically apparent acute or chronic pancreatitis, whereas mucinous cystic neoplasms rarely arise in the setting of pancreatitis. In this case, the patient did not have a history of pancreatitis or antecedent factors such as alcoholism, gallstones, or trauma. Serum amylase is elevated in approximately 65% of the patients with

pancreatic pseudocyst, but normal in the majority of those with mucinous cystic neoplasm of the pancreas<sup>15,16</sup>). There have been, however, a few exceptional patients, as in the present case, with mucinous cystic neoplasm who showed an increase in serum amylase level.

WARSHA and RUTLEDGE suggested that amylase level of the cyst fluid was uniformly low in mucinous cystic neoplasm, and that this measurement was useful in differentiating such lesions from pseudocysts, in which amylase level of the fluid was elevated. Actually, in the present case, amylase level was low in the cystic fluid, but was high in serum.

Tumor markers such as CEA, CA 19-9 and elastase 1 are useful for differentiating between malignant and benign cysts of the pancreas<sup>14</sup>). In the recent works, especially, CA 19-9 level was significantly higher in the fluid in malignant pancreatic cyst than in those from patients with pseudocyst<sup>17</sup>). In the present case, both serum and cystic fluid CA 19-9 levels were elevated.

It has been suggested that communication between the pancreatic duct and the cystic cavity is indicative of a pseudocyst, rather than a mucinous cystic neoplasm<sup>12</sup>). Moreover, the duct communicating with pseudocyst showed obstruction or other abnormalities. However, in a few reported cases, as in our case, ERCP and pathological findings disclosed communication between a mucinous cystic neoplasm and the pancreatic duct.

Ultrasonography and CT scan are the first-choice of diagnostic imaging techniques for pancreatic cysts. Mucinous cystic neoplasms are more likely to be separated or multilocular, by CT scan or ultrasonography, than pseudocysts. The reported prevalence of multiloculation in mucinous cystic neoplasm varied from 33% to 100%<sup>18~22</sup>). Thus the presence of multiple loculi in a pancreatic cyst favors the diagnosis of cystic neoplasm, but is not diagnostic in some cases such as ours. Pseudocysts tend to be thick walled, opaque, and adherent to adjacent viscera, especially the stomach. On the other hand, mucinous cystic neoplasms usually have a thin wall as in the present case. Intraluminal projects and useful features for differentiating between malignant and benign cysts of the pancreas<sup>23</sup>).

Findings on angiography such as displacement, compression of the adjacent vessels, encasement and hypervascularity, are not valid for mucinous cystic neoplasms. However, angiography is valuable for determining the organ of origin when the mass is excessively large<sup>1,23</sup>).

Fine-needle aspiration cytologic study under ultrasonography or CT scan is becoming widely accepted in the diagnosis of pancreatic neoplasm, both pre- and intraoperatively<sup>24,25</sup>). Aspirated fluid facilitates chemical analysis and cytological examination, and is useful for differentiation between pseudocyst and mucinous cystic neoplasm. It is reported that cytologically, mucinous cystic neoplasms showed monomorphic cells forming a perivascular papillary pattern. On the other hand, biopsy for frozen section of the cyst wall usually enables the differentiation. If an epithelial lining is present, mucinous cystic neoplasm should be presumed. Most authors have reported that ultrasonography and CT scan were unable to differentiate mucinous cystadenoma from mucinous cystadenocarcinoma unless local invasion or distant metastases were present. In such a case, fine-needle aspiration cytologic study is useful to differentiate the two. In our case, however, we did not perform fine-needle aspiration for some reasons.

An accurate preoperative diagnosis of pancreatic cyst is important. Especially, distinguishing mucinous cystadenoma of the pancreas from pseudocyst is important because of the malignant potential of the former disease. In this case, we reviewed the preoperative differential diagnosis in various aspects. Clinical course, findings of ultrasonography and CT scan, and tumor markers are reliable diagnostic intervention for mucinous cystic neoplasm of the pancreas.

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