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Cystic Schwannoma of the Pancreas: A Case Report[1]

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A pancreatic schwannoma is an extremely rare pancreatic neoplasm. This tumor can vary in size, and can be variably cystic such that lesion mimics the common cystic tumor of the pancreas. We report a case of a 73-year-old woman with a surgically proven cystic schwannoma of the pancreas. CT images showed the presence of a well-defined, cystic mass in the pancreatic head area, which contained solid, enhancing areas. Furthermore, multi-planar reformatted images provided additional information by demonstrating the intrapancreatic location of the mass. According to our experience, a cystic schwannoma of the pancreas should be considered in the differential diagnosis of a cystic tumor of the pancreas.

Index words : Neurilemmoma
Pancreatic cyst
Pancreatic neoplasms
Pancreas
Tomography, X-Ray computed

A schwannoma of the pancreas is an extremely rare neoplasm that is derived from Schwann cells of either autonomic sympathetic or parasympathetic fibers, which course through the pancreas via the vagus nerve (1, 2). They often show degenerative changes including cyst formation, calcification, hemorrhage, hyalinization, and a xanthomatous change (2 - 5). To the best of our knowledge, 26 cases of pancreatic schwannoma have been reported in the clinical literature; 75% of the reported tumors were partly or completely cystic in nature (1, 6). We describe here a case of cystic schwannoma of the pancreas. The cystic nature and location of the tumor was well demonstrated on CT images including multi-planar reformatted (MPR) images.

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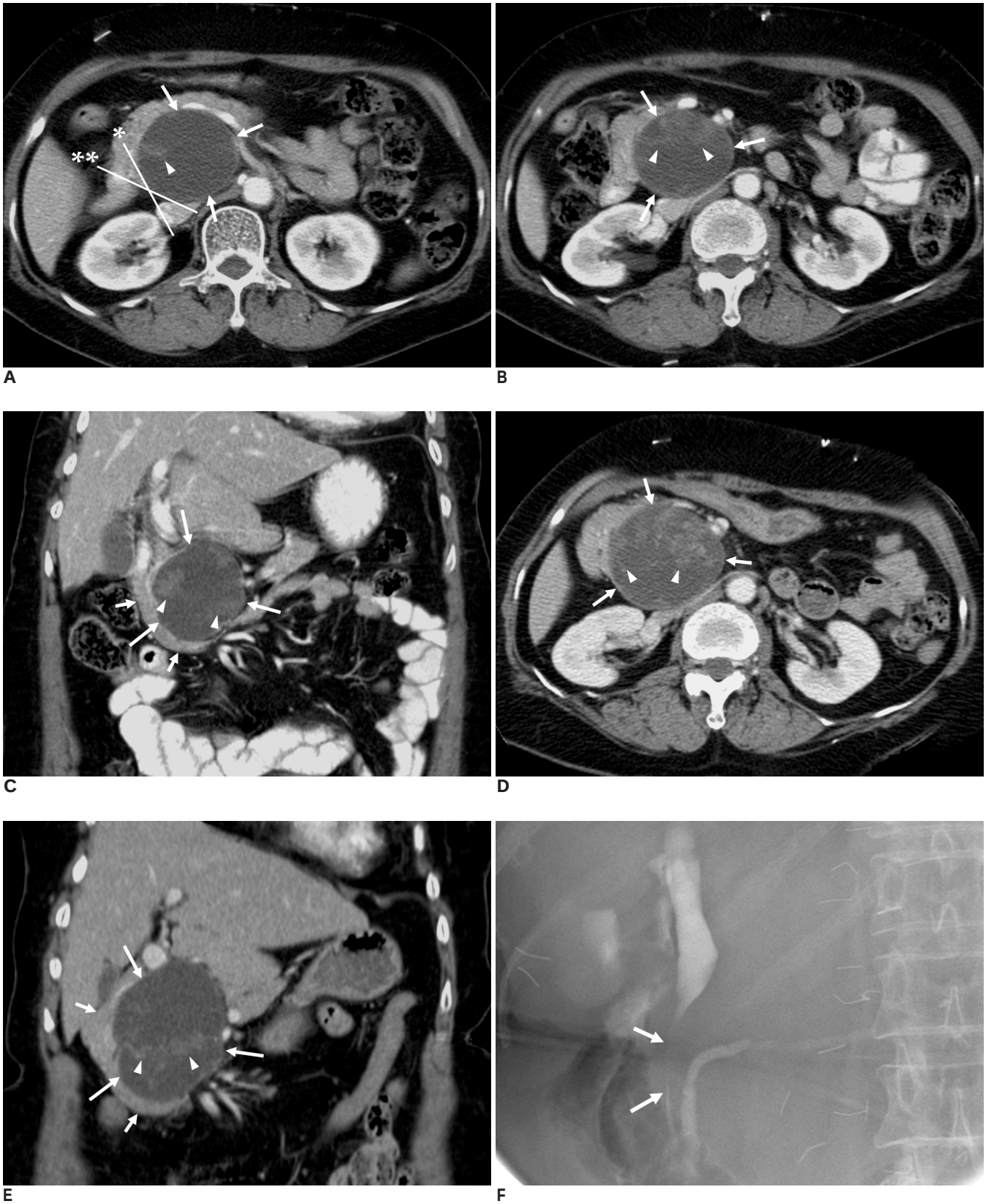
Case Report

A 73-year-old woman, who had a two-year history of abdominal pain, was admitted to our institution due to progressive worsening of abdominal pain. The patient denied having other constitutional symptoms such as weight loss or fever. On the past medical history, the patient visited our institution two years prior due to epigastric pain, and underwent a three-phase helical CT scan in which a well-defined, cystic mass was found in the pancreatic head area. The mass measured 5.4 cm in the maximum diameter (Fig. 1A - C). However, the patient refused surgery because of her age and general weakness.

A physical examination was unremarkable, and labo-

ratory tests including measurements of the level of tumor markers and pancreatic enzymes, were negative. The patient underwent a repeated three-phase helical CT scan, and the same cystic mass was found that now measured up to 7.2 cm in the maximum diameter (Fig. 1D, E). Even with the size increment, there was no evidence of peripancreatic or vascular involvement as well as the presence of a distant metastasis as seen on CT



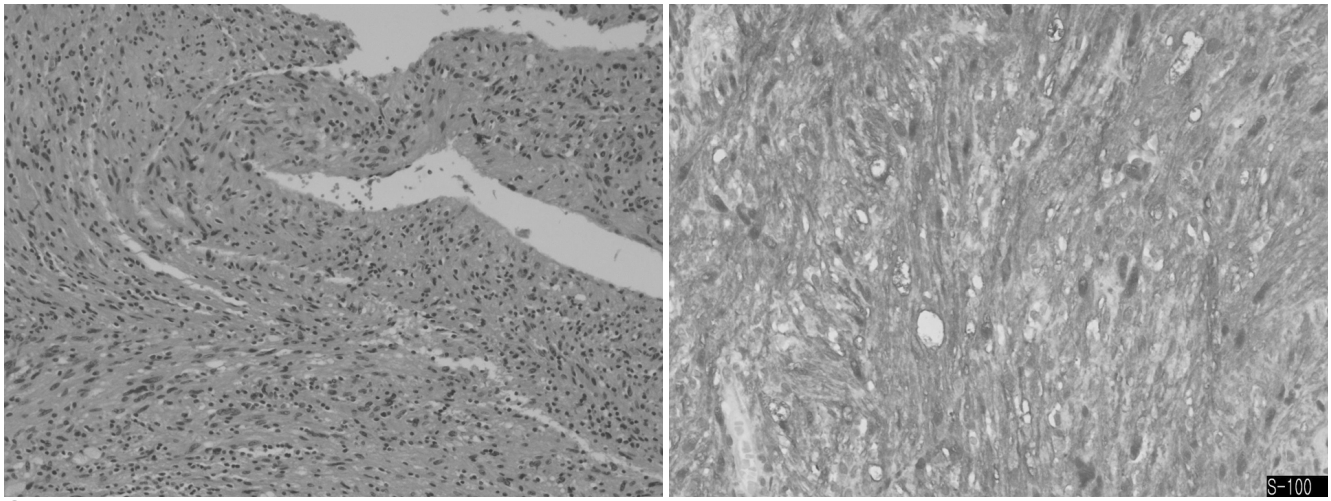


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Fig. 1. A 73-year-old woman with a surgically proven cystic schwannoma of the pancreas.
A-C. CT images obtained two years before surgery. (**A**) A contrast-enhanced CT image obtained during the portal venous phase shows a 5.4 cm, well-defined, cystic mass (arrows), which is mainly situated between the pancreas and the inferior vena cava. Note the enhancing area (arrowhead) within the mass. The pancreatic tissue adjacent to the mass is effaced so that the angle between the inner margin (*) and outer margin (**) of the pancreatic head adjacent to the mass is acute. This finding suggests that the mass is intrapancreatic in location. (**B**) A contrast-enhanced CT image obtained during the portal venous phase at a level lower than (**A**) show the same mass (arrows), which also effaces the pancreatic head. Note the more prominent enhancing areas (arrowheads). (**C**) An MPR image in the coronal plane shows the same mass (long arrows) with enhancing areas (arrowheads). Note that one-half of the mass is embedded in the pancreatic head (short arrows), representing that it is intrapancreatic in location. (**D, E**) CT images obtained immediately before surgery. (**D**) Contrast-enhanced CT images obtained during the portal venous phase at the same level as (**B**) show the interval increase in the size of the same mass (arrows), which now measures 7.1 cm in diameter with more prominent enhancing areas (arrowheads). (**E**) An MPR image in the coronal plane shows the same mass (long arrows) with enhancing areas (arrowheads). One-half of the mass is embedded in the pancreatic head (short arrows).
F. ERCP shows smooth narrowing (arrows) of the intrapancreatic portion of the common bile duct due to extrinsic compression of the mass. The pancreatic duct is mildly dilated duct without definite obstruction.
G. A photomicrograph of the histopathological specimen shows the presence of cigar-shaped spindle cells with a palisading pattern, representative of an Antoni type A area (hematoxylin and eosin staining, ×100). The thickened septa and excrescences as well as the cyst wall were mostly composed of this type of spindle cells. However, some areas were composed of an Antoni type B area. (**H**) A photomicrograph of the histopathological specimen shows that the tumor cells are strongly positive for S-100 protein by immunohistochemical staining (×400).

imaging.

Both helical CT scans were performed with the same protocol, which included the use of pre-contrast CT images and post-contrast helical CT images obtained during the arterial and portal venous phases. Both three-phase helical CT scans also utilized reconstructed multiplanar reformatted (MPR) images that were analyzed on a three-dimensional workstation. On axial images of both CT scans, the cystic mass was found between the pancreatic body and the inferior vena cava, and was apparently situated outside the pancreas. Nevertheless, the cystic mass was attached to the pancreatic head, and the parenchyma of the pancreatic head adjacent to the mass was effaced as shown in Figure 1A, suggesting that the mass was intrapancreatic in location. The intrapancreatic location of the mass was further elucidated by the MPR images in that one-half of the cystic mass was embedded in the pancreatic parenchyma, as seen in the coronal and sagittal planes (Fig. 1C, E). The cystic mass contained enhancing areas that were different in extent between the two CT scans (compare Fig. 1B and 1D). In addition, the extent of the enhancing areas was better depicted on the MPR images (Fig. 1C, E). There was no calcification within the mass as seen on the pre-contrast CT images.

The patient also underwent endoscopic retrograde cholangiopancreatography (ERCP) where smooth narrowing of the intrapancreatic portion of the common bile duct suggestive of extrinsic compression by the mass was seen (Fig. 1F). With these imaging findings, we suggested that the differential diagnoses of this cystic mass could be a mucinous cystic neoplasm or a solitary pseudopapillary tumor.

A pylorus-preserving pancreatoduodenectomy was

performed, and a cystic mass was identified in the head of the pancreas without peripancreatic extension or vascular involvement. There was no lymphadenopathy or peritoneal seeding seen at surgery. The cut section of the pancreas revealed a well-demarcated, multi-locular cyst with thickened septa and excrescences along the cyst wall, with solid portions that corresponded to the enhancing areas seen on CT images. Microscopically, the thickened septa and excrescences as well as the cyst wall were mostly composed of cigar-shaped spindle cells with a palisading pattern, representing Antoni type A areas (Fig. 1G). Some areas showed areas of sparse cellularity, representing Antoni type B areas. There was no cellular atypia. These spindle cells showed a strong positivity for S-100 protein based on immunohistochemical staining, suggesting that the cells were of Schwann cell origin (Fig. 1H). The histopathological findings were compatible with a pancreatic schwannoma with cystic change.

After surgery, abdominal pain and fever developed due to a leakage at the anastomotic site, but eventually the patient recovered after supportive treatment.

Discussion

Schwannomas are benign spindle cell tumors derived from Schwann cells that line the nerve sheaths; schwannomas are also denoted as neurilemmomas. Microscopically, schwannomas are composed of two different components, designated as Antoni type A and B (1, 2, 4 - 7). Antoni type A areas are highly cellular and consist of spindle cells that often have a palisade or organoid arrangement. Mitotic figures are usually absent or are extremely scant (2). Antoni type B areas are relatively hypocellular, and tumor cells that are separated by abundant edematous fluid that may form cystic spaces (7). In addition, cystic degeneration can be induced by vascular thrombosis and subsequent necrosis (4, 5). Besides cyst formation, additional degenerative changes can exist such as calcification, hemorrhage, hyalinization and a xanthomatous change (2, 3). These degenerative tumors are called ancient schwannomas, in which marked atypia can be seen, but are of no clinical significance (2, 3). Schwannomas usually occur in the extremities, but the lesions can also be found in the trunk, head and neck, retroperitoneum, mediastinum, pelvis and rectum (8). Schwannomas of the pancreas are extremely rare, and are known to arise from Schwann cells of either autonomic sympathetic or parasympathetic fibers, both of which course through the pancreas via the vagus nerve (1, 2). These pancreatic schwannomas are reported to affect older adults with an equal ratio of men to women, contrary to schwannomas of non-pancreatic origin that usually occur in young to middle-aged adults and affect women twice as often as men (2, 7, 9). The tumor size of a pancreatic schwannoma varies considerably, ranging from 1.5 cm to 20 cm in diameter, and half of the masses are less than 5 cm in diameter(1). Furthermore, two-thirds of the lesions are located in the head as in the present case (1, 6). Pancreatic schwannomas have also been described as showing degenerative changes, as with cases of non-pancreatic origin (2, 3). Among 24 pancreatic schwannomas reported in the clinical literature, to the best of our knowledge, five cases (21%) were completely solid, while eleven (46%) cases were completely cystic at pathology (1, 6).

On CT images, schwannomas predominantly composed of Antoni type A areas usually show a solid, enhancing mass (5, 6). Schwannomas with a prevalence of Antoni type B areas mostly show a cystic, non-enhancing mass, and the lesions are thought to result from one or more of the histopathological features such as hypocellularity, cystic degeneration and a xanthomatous change (4 - 6). Moreover, a schwannoma may be inhomogeneous in appearance when the confluent areas of Antoni type A and B are intermingled within the tumor (4). In cases of cystic schwannomas of the pancreas, the CT findings are the same as the above-mentioned CT findings of Antoni type B schwannomas, and the tumors are usually depicted as a well defined, cystic or hypoattenuating masses with encapsulation, although in some cases, they may show hyperattenuation when there are areas of Antoni type A. Similarly, the CT findings of our case appeared to be mainly cystic with a part of enhancing areas that were demonstrated as thickened septa and excrescences on the macroscopic specimen, and were composed of Antoni type A and B areas as seen microscopically. The extent of the enhancing areas was different between the two CT scans, and could be explained as the solid portions became necrotic and then cystic during the period between the two CT scans.

Recently, MPR images have been widely used as an adjunct to axial CT images in order to enhance the diagnostic confidence for physicians and radiologists (10 - 12). As described above, the effacement of pancreatic parenchyma seen on the axial images might suggest that our case was intrapancreatic in location, but was not confirmative as a large, extrapancreatic mass can cause

such finding. The MPR images demonstrated that the cystic mass in the present case was in large part surrounded by the pancreas, further assuring that it was intrapancreatic.

A variety of pancreatic tumors can be manifested as a cystic mass, including a cystadenoma, cystadenocarcinoma, intraductal papillary mucinous tumor, pseudocyst and lymphangioma (1, 2, 7). Some solid tumors are frequently seen as a cystic mass such as a solid and pseudo-papillary tumor and neuroendocrine tumor (7). Our case suggests that a cystic schwannoma can be added to the differential diagnosis of cystic tumors of the pancreas, even though it is extremely rare.

Malignant transformation of a schwannoma is exceptionally rare, considering that only four cases of malignant schwannomas of the pancreas have been reported (1, 2). Moreover, all malignant schwannomas that have been reported were larger than 7 cm in diameter (1). Interestingly, the tumor size of our case increased from 5.4 cm to 7.1 cm during a two-year period. Despite these findings, our case was not believed to be a malignant schwannoma, since the growth rate was not as sufficiently high as a malignant mass. There was no evidence of malignant transformation from the histopathological findings.

The preoperative diagnosis of a cystic schwannoma of the pancreas is difficult as the differential diagnosis of cystic tumors of the pancreas is broad. Definitive diagnosis of a cystic schwannoma of the pancreas can only be established by a histopathological examination. Conclusively, we experienced a case of a cystic schwannoma of the pancreas, of which the CT findings were similar to the CT findings of other cystic tumors of the pancreas. Therefore, we believe that a cystic schwannoma is an important clinical entity to include in the differential diagnosis of cystic tumors of the pancreas.

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췌장의 낭성 신경집종: 1예 보고[1]

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췌장의 신경집종(schwannoma)는 매우 드문 췌장종양으로 다양한 크기와 다양한 정도의 낭성 변화를 보인다. 따라서 췌장에서 흔히 생기는 낭성 종양들과 감별이 어렵다. 저자들은 수술로 확인된 췌장의 신경집종을 가진 73세 여자 환자를 경험하였는데 CT 소견은 췌장 두부에 경계가 분명한 낭성 종괴가 보였고 내부에 조영증강되는 고형 부분이 있었다. 또한, 다면재구성영상에서 이 종괴가 췌장 내에 있음을 보여주었다. 따라서 췌장의 신경집종도 췌장의 낭성 종양의 감별진단에 포함해야 할 것으로 생각된다.