Case Report

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대한영상의학회지

Pancreatic Schwannoma with Cystic Degeneration: A Case Report and Literature Review

낭성변화를 보이는 췌장의 신경초종: 증례 보고와 문헌고찰

Joon Suk Park, MD[1], Seon Jeong Min, MD[1]*, Hyunchul Kim, MD[2], Jung-Ah Choi, MD[1]

Departments of [1]Radiology, [2]Pathology, Hallym University Dongtan Sacred Heart Hospital, Hwaseong, Korea

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***Corresponding author**
Seon Jeong Min, MD
Department of Radiology,
Hallym University
Dongtan Sacred Heart Hospital,
7 Keunjaebong-gil,
Hwaseong 18450, Korea.

Tel 82-31-8086-2588
Fax 82-31-8086-2582
E-mail msj542@hallym.or.kr

ORCID iDs
Joon Suk Park
https://orcid.org/0000-0003-2848-1531
Seon Jeong Min
https://orcid.org/0000-0002-1647-5671
Hyunchul Kim
https://orcid.org/0000-0002-9322-725X
Jung-Ah Choi
https://orcid.org/0000-0002-0896-4766

Schwannomas originate from Schwann cells, and they are the most common benign neoplasms of the peripheral nerves. They can occur in most parts of the body but have a predilection for the head, the neck, and the flexor aspects of the extremities. Pancreatic schwannomas are uncommon, and only a few cases have been reported in the English literature. Approximately two-thirds of pancreatic schwannomas undergo cystic degeneration, and they should be considered in the differential diagnosis of solid pancreatic tumors with cystic changes to facilitate accurate diagnosis and optimal treatment. We report a case of a pathologically proven schwannoma in the pancreatic tail with multiple cystic and hemorrhagic changes followed by a review of relevant literature.

Index terms Schwannoma; Neurilemmoma; Pancreas; Pancreas Neoplasms

INTRODUCTION

Schwannomas, also known as neurilemmomas, are rare, benign encapsulated, slow growing tumors arising from Schwann cells that encase the peripheral nerves (1). They frequently involve the head and neck area, major nerve trunks, and flexor aspects of the upper and lower extremities (2). Deeply situated schwannomas are predominantly found in the retroperitoneum and posterior mediastinum, but rarely found in the trunk and gastrointestinal tract (2).

According to a PubMed database search, 77 cases of pancreatic schwannomas have been described in the English literature over the past 40 years. Given the rarity and its

overlapping appearance with other more common pancreatic neoplasm, preoperative diagnosis of pancreatic schwannoma is challenging. It becomes even more challenging when the tumor undergoes cystic degeneration, and it may mimic other cystic tumors or solid tumors with cystic change in pancreas, such as mucinous and serous cystic neoplasms, the solid pseudopapillary neoplasm of pancreas (SPN), pseudocysts and neuroendocrine tumors (NET) (2).

We discuss a patient with the schwannoma that underwent multiple cystic degenerations at the tail of pancreas. Due to cystic and hemorrhagic components of the tumor, a preoperative diagnosis was first assumed to be the more common pancreatic tumor with cystic change in middle aged female such as SPN or cystic NET. We describe the imaging findings of pancreatic schwannoma with multiple cystic and hemorrhagic degeneration on computed tomography (CT) and magnetic resonance imaging (MRI). The final diagnosis was confirmed by pathology following distal pancreatectomy.

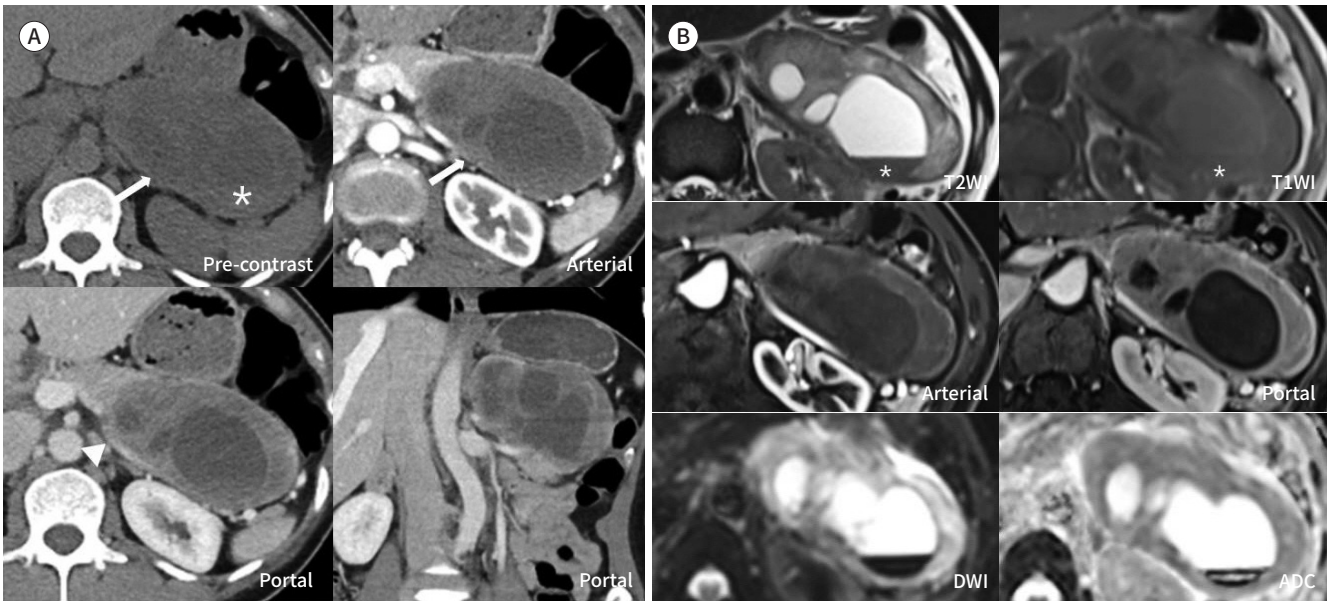
CASE REPORT

A 43-year-old female patient presented our hospital for an abdominal discomfort over 3 months. She had no history of medical record or symptoms associated with pancreatitis. On admission, physical examinations did not reveal significant abnormalities. The laboratory findings including amylase, tumor markers, liver function test or hemoglobin were within normal range.

The patient underwent a multiphase contrast-enhanced CT scan and a well-defined solid mass with multiple cystic and hemorrhagic changes was found in pancreatic tail (Fig. 1A). On pre-contrast image, the lesion appears as a low-attenuated area compared with the surrounding pancreas parenchyma. On contrast-enhanced CT images during the arterial and the portal venous phase, the lesion shows homogeneously enhancing well-circumscribed solid mass with cystic degeneration. The main solid peripheral portion enhances less intensely than the adjacent normal pancreas, while non-enhancing inner contents indicate the presence of cystic and hemorrhagic degeneration. On pre-contrast image, the heterogeneously high attenuating cyst with Hounsfield unit (HU) up to 40 HU at dependent level suggests the hemorrhagic component. The normal parenchyma of pancreas surrounds the tumor making sharp angles on each side of the mass, indicating that the tumor is within the pancreatic parenchyma.

On MRI, the lesion correlating with CT image similarly appears as a well-defined tumor with multiple cystic components. Relative to the normal pancreas, the main solid portion shows mild high signal intensity (SI) on axial T2-weighted image (T2WI) and mild low SI on axial T1-weighted image (T1WI) (Fig. 1B). After intravenous administration of contrast agent, gadobutrol (Gadovist®; Bayer-Healthcare, Berlin, Germany), the solid portion shows homogeneous enhancement, but less intensely in comparison with the remainder of the pancreas on arterial and portal phase. Non-enhancing multifocal areas of high SI on T2WI and low SI on T1WI suggest the presence of abundant cystic components. As shown on CT image, the cyst with fluid-fluid level containing low SI on both T1WI and T2WI at dependent level, and slightly high SI on T1WI and low SI on T2WI at upper level, suggests the presence of hemorrhagic components. There was no vascular encasement, metastasis or lymph node enlargement.

Fig. 1. A 43-year-old female with a surgically proven pancreatic schwannoma with multiple cystic and hemorrhagic changes.
A. A pre-contrast CT image shows a low-attenuated mass (arrow) with cystic changes in the pancreatic tail. A cyst with high attenuating inner content of up to 40 HU at the dependent level (asterisk) and up to 20 HU at the upper level, which is indicative of a hemorrhagic cyst, is observed. On contrast-enhanced CT images, the arterial phase reveals a well-defined, homogeneously enhancing solid tumor with non-enhancing, cystic inner components (arrow). The portal venous phase shows persistent enhancement. The enhancement is less intense compared with the adjacent normal pancreas. The normal parenchyma surrounds the tumor on each margin (arrowhead), which indicates that the lesion is located within the intra-pancreatic area. The coronal reconstruction image shows the enhancing solid mass within the intra-pancreatic location.
B. The peripheral solid portion is characterized by a mild high SI on T2WI and a mild low SI on T1WI, relative to the normal adjacent pancreas. The inner cystic portions are characterized by a high SI on T2WI and a low SI on T1WI. As shown on the CT image, the cyst with fluid-fluid level is characterized by a low SI on T1WI and T2WI at the dependent level (asterisks) and an intermediate SI on T1WI and a high SI on T2WI at the upper level, which suggests an older hemorrhage at the dependent portion. On the dynamic MRI, the periphery shows enhancement, but it is less intense than that of the normal pancreas on the arterial and portal phase images. The lesion shows a diffusion restriction on the peripheral solid portion on DWI (b = 1000).
ADC = apparent diffusion coefficient, DWI = diffusion-weighted image, HU = Hounsfield unit, SI = signal intensity, T1WI = T1-weighted image, T2WI = T2-weighted image

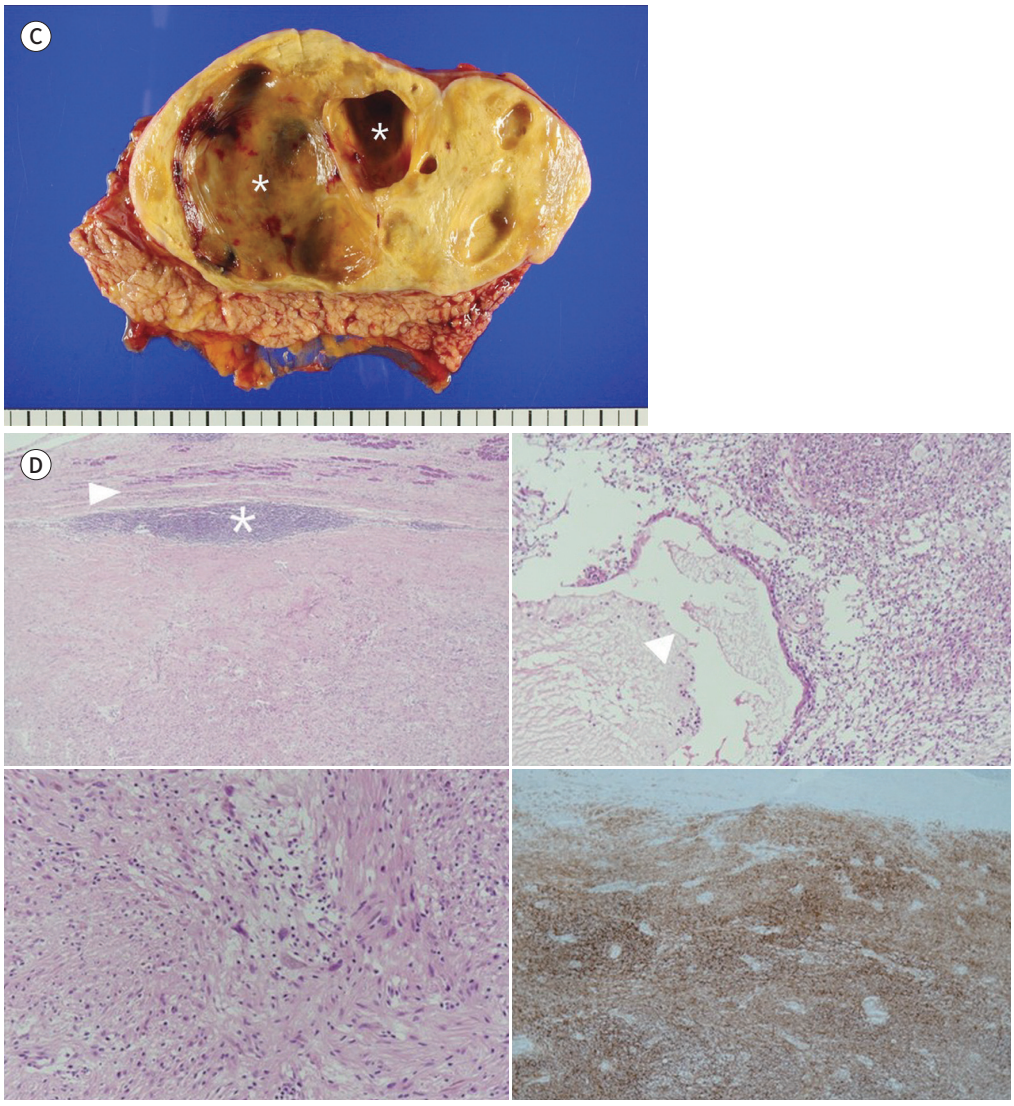


The patient underwent distal pancreatectomy, adjacent lymph node dissection and splenectomy. Macroscopically, the cut portion of distal pancreatectomy was consistent with image findings: a well-circumscribed yellow solid mass with approximately 11 cm in the largest diameter and with inner cystic and hemorrhagic components (Fig. 1C). On histopathologic analysis, well-demarcated tumor abutting pancreas with lymphoid aggregate was composed of spindle cells (Fig. 1D). Cystic change and xanthogranulomatous cells were also observed. On immunohistochemistry, the cells represented positive for S100, but negative for CD34, c-kit, smooth muscle actin, and Desmin. These findings were consistent with ancient schwannoma.

DISCUSSION

Schwannomas are benign peripheral nerve sheath tumors, which are entirely composed of the Schwann cells those normally wrap around the peripheral nerve (3). Pancreatic schwan-

Fig. 1. A 43-year-old female with a surgically proven pancreatic schwannoma with multiple cystic and hemorrhagic changes.
C. A gross cross-section specimen reveals a well-defined yellow mass with inner cysts. Hemorrhagic components (asterisks) are also observable.
D. The histopathological specimen includes a lymphoid aggregate (asterisk) that is composed of spindle cells and is adjacent to the normal pancreas (arrowhead) (upper left; H&E stain, × 40). Cystic change and xanthogranulomatous cells are observed (arrowhead) (upper right and lower left; H&E, × 100; H&E, × 200). Immunohistochemical staining shows that the lesion is positive for S100 (lower right; immunohistochemical stain, × 100). The pathology is consistent with an ancient schwannoma.
H&E = hematoxylin and eosin



nomas grow slowly which explains their benign potential and range of mass size from 2 to 20 cm (3). The head of pancreas is the most preferential location for schwannoma growth in pancreas, while the tail is the rarest location as in our case (4). About two thirds of pancreatic schwannomas present with cystic formation within the tumor (2).

Microscopically, a schwannoma can be subdivided into two components, namely Antoni A and Antoni B. Antoni type A area is an organized hypercellular component with presence of

Verocay bodies, consisting of palisading nuclei that are surrounding nucleus-free cytoplasmic processes (4). On the other hand, Antoni type B area is a hypocellular component where the tumor cells are separated by abundant edematous fluid that may form cystic spaces (4). Inadequate blood supply to the center of the tumor sometimes causes secondary degenerative changes such as cyst formation, calcification, hemorrhage, and hyalinization (5). These degenerative tumors are called ancient schwannoma and explain the polymorphism of the tumors as they appear at imaging (5).

On CT images, the areas of Antoni type A present as a solid, enhancing mass, while that of Antoni type B show a cystic, non-enhancing mass (6). The lesion also can be inhomogeneous when the confluent areas of Antoni type A and B are intermingled within the tumor (6). In our case, the solid area of tumor on CT correlated to Antoni A area on histologic finding, and multiple cystic and hemorrhagic components of tumor on CT correlated to Antoni B area with cysts and xanthomatous cells on histologic finding and also correlated to hemorrhagic cysts on macroscopic finding.

On MRI, a typical schwannoma has been described as a mass with low SI on T1WI and with inhomogeneous high SI on T2WI due to alternating Antoni A and B areas and secondary degenerative changes. Contrast enhanced T1-weighted MR images can show cystic necrotic areas and well-enhanced peripheral and intervening solid areas of the mass (5).

In our case, the patient had well encapsulated solid mass with multiple cystic and hemorrhagic changes in pancreatic tail at middle age female adult. Kim et al. (6) reported a case of multi-locular cystic form of schwannoma at body of pancreas, while we report a case of solid form of schwannoma with multiple cystic and hemorrhagic degenerations. In our case, the tumor underwent degenerative changes, which was pathologically proved as ancient schwannoma. The solid mass presented multiple inner cystic and hemorrhagic degenerations on CT image, and this polymorphism correlated with the pathologic diagnosis.

Given cystic formations within solid mass, well-circumscribed morphology, patient's age and pancreatic tail location, the more common tumors such as cystic NET or SPN were the primary suspicions.

NETs typically have a rich vascular supply and therefore enhance avidly during the arterial phase, enhancing rapidly and intensely than the normal pancreas (7). Small tumors less than 2 cm lesions may show homogeneous enhancement, whereas larger lesions show heterogeneous enhancement. Appreciating the enhancement pattern as a key distinguishing feature, in our case, solid portion of pancreas schwannoma does not enhance intensely than normal pancreas and we could exclude NET.

On the other hand, due the fragile vascular network of tumor, internal hemorrhagic and cystic degeneration is the hallmark of SPN which commonly occurs in the pancreatic tail (7). It typically shows peripheral heterogeneous enhancement during the arterial phase and progressive nonuniform enhancement thereafter, with enhancement generally being less than that of the normal pancreas. Image finding alone may not be sufficient to diagnose pancreatic schwannoma over SPN as in our case. The pathologic confirmation, however, made final diagnosis.

Another differential diagnosis may include pancreas ductal adenocarcinoma with necrotic change. According to Ma et al. (2) who has updated 40 year review of pancreatic schwanno-

mas yielding 68 cases, no pancreatic schwannoma shows associated dilatation of the main pancreatic duct or common bile duct. The presence of ductal dilatation rather favors ductal adenocarcinoma. In our case, no secondary signs of adenocarcinoma were shown such as mass effect, an abnormal convex contour of the pancreas, ductal obstruction, or vascular invasion.

In addition, for the solid form of schwannoma carcinoma, consideration for differential diagnosis include islet cell tumors, or other benign tumors such SPN (8). Despite the fact that the final diagnosis depends on histopathologic testing, well-defined margin without ductal dilatation favors solid pancreatic schwannoma over pancreatic adenocarcinoma. Moreover, due to their similar appearances, image finding alone may not be sufficient to distinguish islet cell tumors or SPN comprise from pancreatic schwannoma.

In conclusion, this case report suggests that the caution needs to be applied when diagnosing the solid tumor with cystic change in pancreas. This case particularly shows the solid form of schwannoma with multiple cystic and hemorrhagic components. As in our case, preoperative diagnosis of pancreatic schwannoma is challenging, because pancreatic tail itself is an unusual location, and moreover, cystic degeneration makes it even more challenging due to its similar appearance with other pancreatic tumors with cystic change. Imaging alone may not be sufficient for definite diagnosis of pancreatic schwannoma with cystic and hemorrhagic components, but it deserves attention as the differential diagnosis of solid tumor with cystic change in pancreas.

Author Contributions

Conceptualization, M.S.J.; data curation, M.S.J.; investigation, P.J.S., M.S.J.; project administration, M.S.J.; resources, M.S.J.; supervision, M.S.J.; writing—original draft, all authors; and writing—review & editing, all authors.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

REFERENCES

1. Ciledag N, Arda K, Aksoy M. Pancreatic schwannoma: a case report and review of the literature. *Oncol Lett* 2014;8:2741-2743
2. Ma Y, Shen B, Jia Y, Luo Y, Tian Y, Dong Z, et al. Pancreatic schwannoma: a case report and an updated 40-year review of the literature yielding 68 cases. *BMC Cancer* 2017;17:853
3. Manning MA, Srivastava A, Paal EE, Gould CF, Mortele KJ. Nonepithelial neoplasms of the pancreas: radiologic-pathologic correlation, part 1--benign tumors: from the radiologic pathology archives. *Radiographics* 2016;36:123-141
4. Moriya T, Kimura W, Hirai I, Takeshita A, Tezuka K, Watanabe T, et al. Pancreatic schwannoma: case report and an updated 30-year review of the literature yielding 47 cases. *World J Gastroenterol* 2012;18:1538-1544
5. Rha SE, Byun JY, Jung SE, Chun HJ, Lee HG, Lee JM. Neurogenic tumors in the abdomen: tumor types and imaging characteristics. *Radiographics* 2003;23:29-43
6. Kim HJ, Lee WJ, Kim SM, Jang KT. Cystic schwannoma of the pancreas: a case report. *J Korean Radiol Soc* 2008;58:509-514
7. Low G, Panu A, Millo N, Leen E. Multimodality imaging of neoplastic and nonneoplastic solid lesions of the pancreas. *Radiographics* 2011;31:993-1015
8. Yu RS, Sun JZ. Pancreatic schwannoma: CT findings. *Abdom Imaging* 2006;31:103-105

대한영상의학회지

낭성변화를 보이는 췌장의 신경초종: 증례 보고와 문헌고찰

박준석[1] · 민선정[1*] · 김현철[2] · 최정아[1]

신경초종은 슈반세포에서 기원하는 양성 종양이며 말초신경집에서 생기는 가장 흔한 종양이다. 신경초종은 신체 어디에서나 발견될 수 있으나, 보통 머리와 목, 그리고 사지의 굽힘 면에서 흔하게 보일 수 있는 종양이다. 췌장은 신경초종이 드물게 생기는 위치이며 적은 수의 증례가 보고되었다. 보고된 신경초종 증례의 약 3분의 2 정도에서 낭성변화를 동반한다. 따라서 적절한 진단과 치료를 위해서는 췌장에서 발견되는 낭성변화를 동반한 고형 종양의 경우 신경초종이 감별진단에 포함되어야 한다. 이에 저자들은 병리학적으로 진단된 낭성변화를 동반한 췌장의 신경초종을 문헌고찰과 함께 보고하고자 한다.

한림대학교 동탄성심병원 [1]영상의학과, [2]병리과