

CASE REPORT

Pancreatic Schwannoma: A Case Report and Literature Review with Special Reference to Imaging Features

Shiji Suzuki¹, Satoshi Kaji¹, Nobusada Koike¹, Nobuhiko Harada¹,
Tsuneo Hayashi¹, Mamoru Suzuki¹, Fujio Hanyu¹, Shinichi Ban²

¹Hachioji Digestive Disease Hospital. Hachioji City, Tokyo, Japan. ²Department of Pathology, Saiseikai Kawaguchi General Hospital. Kawaguchi, Saitama, Japan

ABSTRACT

Context We report the imaging features of pancreatic schwannomas, a rare benign type of pancreatic tumor. **Case report** A 66-year-old woman was admitted to our hospital with a pancreatic tumor indicated in medical examinations. Computed tomography (CT), magnetic resonance imaging (MRI) and endoscopic ultrasonography (EUS) revealed a solid and cystic tumor, 3 cm in diameter, within the body of the pancreas. Contrast-enhanced CT, MRI and ultrasonography showed partial enhancement in the solid component. Endoscopic retrograde cholangiopancreatography (ERCP) and angiography showed no abnormal findings. A distal pancreatectomy together with a splenectomy and lymph node dissection were performed with a tentative diagnosis of mucinous cystic neoplasm of the pancreas. The cut surface of the resected pancreas showed a well-demarcated, pale yellow, solid tumor within the pancreas parenchyma. Histopathological examination of the tumor revealed proliferation of the spindle cells showing interlacing and palisading patterns. Immunohistochemically, these spindle cells were positive for S-100 protein and vimentin, and negative for alpha-smooth muscle actin, CD34, and cytokeratin. Thus the tumor was diagnosed as a pancreatic schwannoma. **Conclusion** CT and US can detect pancreatic schwannomas as solid and cystic masses, and MRI shows a relatively characteristic feature. Imaging procedures such as CT, MRI and US are able to differentiate a pancreatic tumor, such as a pancreatic schwannoma.

INTRODUCTION

Schwannomas are neurogenic neoplasms derived from Schwann cells of the sheaths of the peripheral nerves [1, 2]. They are basically soft tissue neoplasms, usually found in the head and neck, extremities, mediastinum and retroperitoneum [3, 4], but rarely found in the pancreas. Moreover, schwannomas often contain various patterns of solid and cystic components within a tumor. In this context, the preoperative diagnosis of a pancreatic schwannoma is difficult, and they are often confused with pancreatic cystic neoplasms, such as non-functioning endocrine neoplasms, solid pseudopapillary neoplasms and mucinous cystic neoplasms [5, 6]. We herein report a case of pancreatic schwannoma and an English language literature review with special reference to the imaging features of pancreatic schwannomas.

CASE REPORT

A 66-year-old Japanese woman was admitted to our hospital with a pancreatic tumor indicated in medical examinations. There was no remarkable past history of disease. No inherited factors or marked diseases in her family were noted. On admission, she had no complaints, and physical examinations and laboratory data revealed no noticeable abnormalities. Serum levels of CA 19-9 and s-pancreas-1 (pancreatic associated) antigen were within the normal range. Magnetic resonance imaging (MRI) revealed a mass, 3 cm in diameter, in the body of the pancreas, showing hypointensity on T1-weighted images, marked enhancement after gadolinium diethylenetriaminepenta acetic acid (Gd-DTPA) administration, and mixed hypo- and hyperintense patterns on T2-weighted images (Figure 1abc). Magnetic resonance cholangiopancreatography (MRCP) and endoscopic retrograde cholangiopancreatography (ERCP) showed no abnormal findings. The results of bacterial cultures of the pancreas juice were negative. Computed tomography (CT) revealed a 3 cm solid and cystic tumor within the body of the pancreas, with high resorption CT levels of the cystic components (Figure 1d). Contrast-enhanced CT showed partial enhancement of the solid components, and the main pancreatic duct was not involved in the tumor.

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Abbreviations Gd-DTPA: gadolinium diethylenetriaminepenta acetic acid

Correspondence Shuji Suzuki

Hachioji Digestive Disease Hospital, 177-3 Yorozu-cho, Hachioji City, Tokyo 192-0904, Japan

Phone: +81-42.626.5111; Fax: +81-42.620.5735

E-mail: dr802@nifty.com

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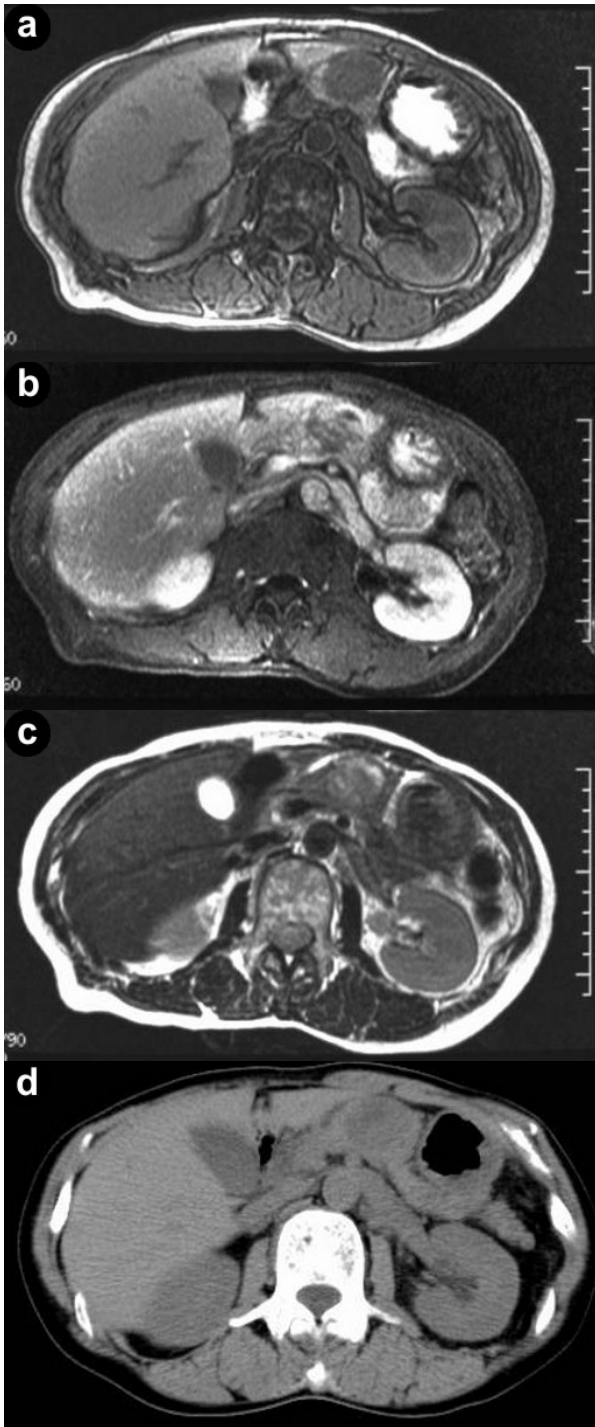


Figure 1a. A T1-weighted MRI indicating a hypointense tumor in the body of the pancreas. **b.** On T1-weighted images, Gd-DTPA administration showing marked tumor enhancement. **c.** A T2-weighted MRI indicating a mixed pattern with hypointensity and hyperintensity. **d.** Plain CT revealing a solid and cystic tumor within the body of the pancreas, with high resorption CT levels of cystic components.

Ultrasonography (US) and endoscopic ultrasonography (EUS) revealed a 3 cm solid and cystic tumor with low echoic margins in the body of the pancreas (Figure 2a). Contrast-enhanced US with Levovist® (Bayer Schering, Berlin, Germany) showed partial enhancement of the solid components (Figure 2b).

Angiography showed no abnormal findings. From these imaging findings, mucinous cystic neoplasms or acinar cell tumors were considered.

A distal pancreatectomy together with a splenectomy and lymph node dissection were performed. The cut surface of the resected pancreas showed a well-demarcated, pale yellow, solid tumor within the pancreas parenchyma. The tumor was 3x3x3 cm in size, and composed of a mixture of solid areas and myxomatous and/or hemorrhagic areas (Figure 3a). Histopathological examination of the tumor revealed proliferation of the spindle cells showing interlacing and palisading patterns (Figure 3b). Focal hypocellular areas showing edematous and myxomatous degeneration, hemorrhage and hemosiderin deposition, and dilated hyalinized vessels were also noted (Figure 3c). The proliferating cells showed minimal pleomorphic nuclei, but no mitotic figures were found. Immunohistochemically, the spindle cells were positive for S-100 protein and vimentin, and negative for alpha-smooth muscle actin, CD34 and cytokeratin. Based on these findings, the tumor was diagnosed as a pancreatic schwannoma of mixed Antoni A and Antoni B types. No remarkable findings were noted in the spleen and the dissected lymph nodes. At a 24-month follow-up after surgery, the patient is doing well without any recurrent disease.

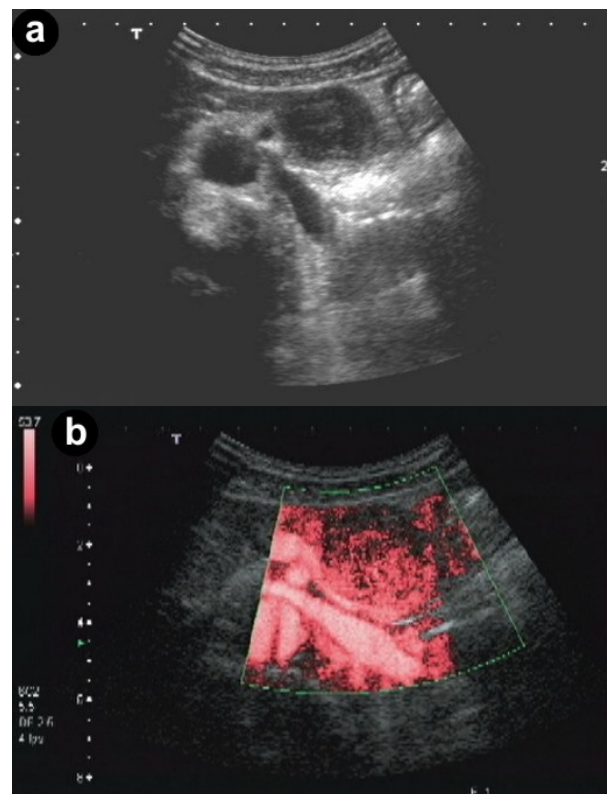


Figure 2. a. US reveals a solid and cystic tumor with low echoic margin in the body of the pancreas. **b.** Contrast-enhanced US with Levovist® (Bayer Schering, Berlin, Germany) shows partial enhancement at the solid component.

DISCUSSION

In 1910, Verocay reported a schwannoma as a true neoplasm which originated from Schwann cells, and which did not contain neuroganglion cells [7, 8]. Since then, schwannomas have been described in almost every location of the body [9, 10]. However, the pancreas is an extremely unusual site of origin for these tumors. In a PubMed search of English publications from 1982 to 2008, only 34 cases of pancreatic schwannoma were found [1, 2, 4, 5, 6, 9, 10, 11, 12, 13,

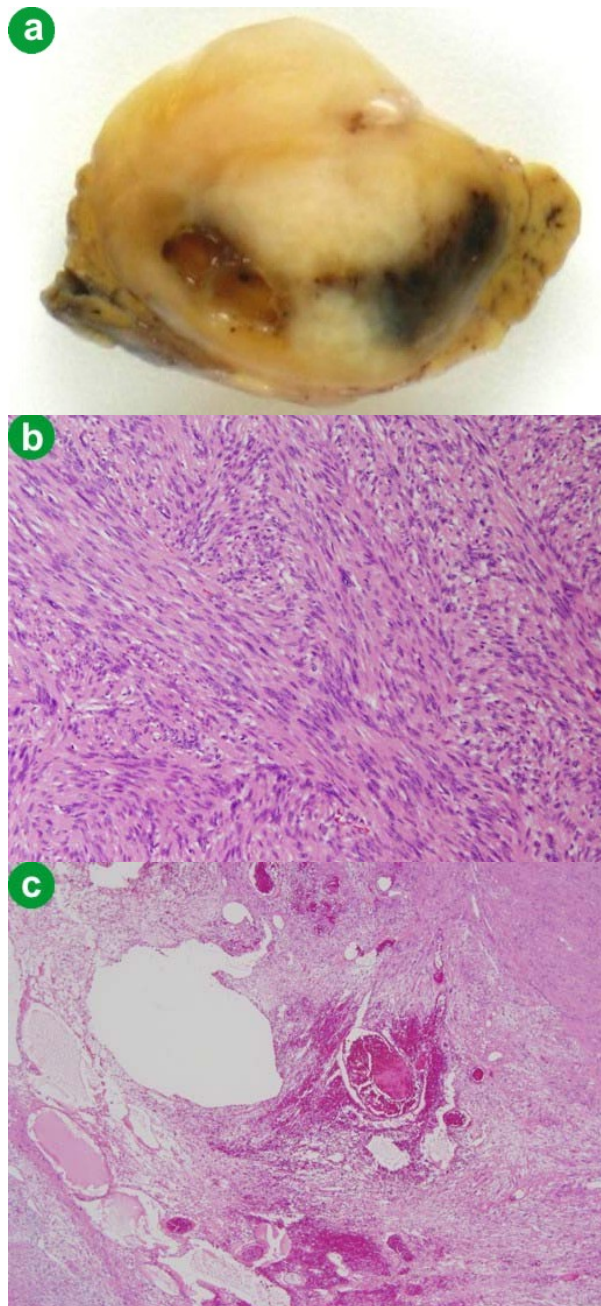


Figure 3. a. The cut surface of the resected tumor is composed of a mixture of solid areas and myxomatous and/or hemorrhagic areas. b. Histopathology of the resected specimen shows proliferation of the spindle cells in interlacing and palisading patterns (Antoni A) (H&E, original magnification x100). c. Edematous degeneration areas with hemorrhage, hemosiderin deposition and hyalinization of the dilated vascular walls (Antoni B) (H&E, original magnification x100).

Table 1. Imaging features among 35 cases of the pancreatic schwannoma. This Table summarizes the findings of 35 cases including 34 cases of the literature review and the present case.

| CT | |
|--|------------|
| - Well-defined, low density or cystic mass | 18 (72.0%) |
| - Solid and cystic mass | 6 (24.0%) |
| - Solid mass | 1 (4.0%) |
| Not available | 10 |
| MRI | |
| - T1-weighted images: hypointensity | 8 (88.9%) |
| - T1-weighted images: hyperintensity | 1 (11.1%) |
| Not available | 26 |
| - T2-weighted images: hyperintensity | 8 (88.9%) |
| - T2-weighted images: mixed | 1 (11.1%) |
| Not available | 26 |
| US | |
| - Cystic and low echoic mass | 12 (63.2%) |
| - Cystic and solid mass | 4 (21.1%) |
| - Solid mass | 3 (15.8%) |
| Not available | 16 |
| Angiography | |
| - Hypervascular patterns | 4 (57.2%) |
| - Hypovascular patterns | 3 (42.8%) |
| Not available | 28 |

14, 15, 16, 17, 18, 19, 20, 21, 22, 23, 24, 25, 26, 27, 28, 29, 30].

These 34 cases together with our case were studied. There were 16 (45.7%) men and 19 (54.3%) women, ranging in age from 40 to 87 years (mean 59.8 years). The locations in the pancreas included 14 cases in the head (40.0%), 8 in the body (22.9%), 4 in the body and tail (11.4%), 3 in the head and body (8.6%), 2 in the tail (5.7%), and 4 with unspecified locations (11.4%). The average size of these tumors was 6.7 cm in diameter (range: 1-17.5 cm). Gross appearances of these cases included 15 cases of a cystic mass (42.9%), 9 of a solid mass (25.7%), 7 of a cystic and solid mass (20.0%), 2 solid with a necrotic center (5.7%) and 2 unspecified (5.7%). A pancreatic schwannoma with von Recklinghausen's disease was confirmed in 2 (8.0%) out of 25 cases.

Microscopically, schwannomas generally consist of two alternating components: an organized cellular component consisting of long bipolar cells which often form a palisading arrangement and/or Verocay bodies (Antoni A area) and a loose hypocellular, degenerative component (Antoni B area) [31]. Most of the pancreatic schwannomas reported also had Antoni A and Antoni B areas in various proportions as was the case in the present tumor. The degenerative changes gave rise grossly to a frequent cystic component.

The imaging features of the pancreatic schwannomas reported and the present tumor are summarized in Table 1. The most characteristic feature on CT was the presence of a low density and/or cystic image in various degrees within the tumor. The low density and/or cystic images would reflect the Antoni B component or the degenerative cystic areas of the schwannoma. Contrast-enhanced CT showed the difference between the Antoni A and the Antoni B areas based on their vascularity, i.e., well-enhanced areas corresponding to Antoni A, and unenhanced areas

corresponding to Antoni B. Therefore, the CT findings of these tumors correlated quite well with the pathological features [1, 14].

The MRI findings usually showed hypointensity on T1-weighted images and hyperintensity on T2-weighted images [6, 9], and most tumors were gradually enhanced on T1-weighted images after Gd-DTPA administration. However, the present case was unusual, showing a mixed hypointensity and hyperintensity pattern on T2-weighted images. This can be attributed to the fact that the present tumor included a fair amount of both Antoni A and Antoni B areas, conceivably with the hypointense areas corresponding to the Antoni B areas, and the hyperintense areas to the Antoni A areas. The solid and cystic pattern observed on CT was also seen with US, in which hypoechoic and/or cystic findings were noted in a high percentage of cases. However, the details of the solid components were revealed more clearly by US as compared to CT or MRI. Moreover, in the present case, contrast-enhanced US with Levovist® (Bayer Schering, Berlin, Germany) showed partial enhancement at the solid component. On angiography, pancreatic schwannomas were reported to be either hypervascular or hypovascular, but this was not found in the present case. We feel that angiography, since it lacked enhancement, was unable to recognize minute vascularity when compared to the other imaging modalities. The usefulness of angiography in imaging diagnosis appears to be limited.

Consequently, CT and US can detect pancreatic schwannomas as solid and cystic masses, and MRI shows relatively characteristic features. However, other pancreatic tumors, such as a non-functioning endocrine neoplasm, solid pseudopapillary neoplasm or mucinous cystic neoplasm, often share those imaging features, and differential diagnoses should always be considered. The treatment of choice for pancreatic schwannomas is surgical resection. This tumor was reported to be malignant in 4 (11.4%) of the 35 cases reviewed in the literature [16, 17, 18, 19], but malignant potential does not appear to be high with regard to lymph node and distant metastases. All the reported cases of pancreatic schwannoma were treated by surgical resection. Radical surgical resections, such as a pylorus-preserving pancreaticoduodenectomy and a distal pancreatectomy, were performed in 62.9% of cases, and limited surgical resections, such as enucleation and excision were performed in 25.7%. The high frequency of radical surgical resections might reflect the difficulty in making a differential diagnosis between pancreatic schwannomas and other pancreatic neoplasms which require radical surgical resections.

In conclusion, imaging procedures, such as CT, MRI, and US, are able to differentiate a pancreatic tumor as a pancreatic schwannoma.

Conflict of interest The authors have no potential conflicts of interest

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