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CASE REPORT

# Central pancreatectomy for pancreatic schwannoma: A case report and literature review

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## Abstract

Schwannomas are mesenchymal tumors originating from Schwann cells in peripheral nerve sheaths. Although the tumor can be located in any part of the human body, the most common locations are the head, neck, trunk and extremities. Pancreatic

schwannomas are rare. To our knowledge, only 64 cases of pancreatic schwannoma have been reported in the English literature over the past 40 years. In this paper, we present a pancreatic schwannoma in a 59-year-old female. Ultrasound, computed tomography and magnetic resonance imaging revealed the tumor located in the pancreatic body; however, accurate diagnosis was hard to obtain preoperatively and a pancreatic cystadenoma was preliminarily considered. During laparotomy, the mass was found in the body of the pancreas. An enlarged gallbladder with multiple stones was also observed. We performed central pancreatectomy, end-to-side pancreaticojejunostomy and cholecystectomy. Notably, central pancreatectomy has been reported in only one case prior to this report. The gross specimen showed a mass with a thin capsule, 1.6 cm × 1.1 cm × 1.1 cm in size. Microscopic examination showed that the tumor was mainly composed of spindle-shaped cells with palisading arrangement and no atypia, which is consistent with a benign tumor. Both hypercellular and hypocellular areas were visible. Immunohistochemical staining revealed strongly positive results for protein S-100. Finally, the tumor was diagnosed as a schwannoma of the pancreatic body. Postoperatively, the patient recovered well and left the hospital 6 d later. During the 53-mo follow-up period, the patient remained well and free of complications.

**Key words:** Pancreaticojejunostomy; Schwannoma; Pancreas; Central pancreatectomy; S-100



**Core tip:** Over the past 40 years, only 64 cases of pancreatic schwannomas have been reported in the English literature. It is a considerable challenge to obtain a precise preoperative diagnosis, despite the application of multiple imaging modalities. We present a patient with a pancreatic schwannoma and enlarged gallbladder with multiple stones. After central pancreatectomy, end-to-side pancreaticojejunostomy and cholecystectomy, the patient recovered quickly and had a good prognosis. In this study, we focused on the diagnosis and treatment of a pancreatic schwannoma and conducted a literature review to deepen the understanding of the subject.

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## INTRODUCTION

Schwannomas are tumors originating from the Schwann cells of peripheral nerve sheaths[1]. Most schwannomas show either monosomy 22 or loss of 22q material; however, the pathogenesis of the tumor remains unclear[2]. Schwannomas are generally encapsulated tumors, and more than 90% of them are benign[2]. Schwannomas occur in patients with no obvious gender difference and at all ages. However, patients between 20 and 50 years of age were most frequently reported. Surgery is the most common treatment for schwannomas, and patients usually have a good postoperative prognosis[3]. While almost every part of the human body can be involved, the most common locations are the head, neck, trunk and extremities[3]. Pancreatic schwannomas are extremely rare. To our knowledge, only 64 cases of pancreatic schwannomas have been reported in the English literature over the past 40 years[4-61]. Although most patients with pancreatic schwannomas were symptomatic, a considerable number of patients were asymptomatic with tumors that were found incidentally. This paper presents a case of pancreatic schwannoma in a 59-year-old female and a review of the literature.

## CASE REPORT

On January 7, 2012, a 59-year-old female was referred to our hospital because of a pancreatic mass found during a routine health examination. The abdomen was soft and nondistended without evidence of a palpable mass. Her family history was not significant. Abnormal laboratory results included: Unconjugated bilirubin, 2 μmol/L (normal, 3-14) and serum kalium, 3.42 mmol/L (normal, 3.5-5.2). Ultrasound revealed a 1.4 cm × 1.3 cm, well-defined cystic lesion in the pancreatic body (Figure 1A), as well as a 6.8 cm sized strong echo in the gallbladder (Figure 1B). An unenhanced computed tomography (CT) scan showed a 1.6 cm × 1.1 cm well-defined hypodense mass in the pancreatic body (Figure 2A). On the contrast-enhanced CT, the mass was not enhanced (Figure 2B). On magnetic resonance imaging (MRI), the mass in the pancreatic body and gallbladder appeared hypointense on T1 weighted images (Figure 3A). The mass in the pancreatic body appeared inhomogeneously hyperintense and the enlarged gallbladder appeared hyperintense on T2 weighted images (Figure 3B). We also performed endoscopic ultrasound-guided fine needle aspiration (EUS-FNA). However, the tumor sample was difficult to acquire, and the procedure was unsuccessful. According to these results, a pancreatic cystadenoma and an enlarged gallbladder with multiple stones were preliminarily considered.

After sufficient preparation and obtaining consent from the patient and her family members, a laparotomy was performed. A 1.5 × 1.0 cm mass surrounded by a thin fibrous capsule was found in the pancreatic body. An enlarged gallbladder with multiple stones was also found. We performed central pancreatectomy, end-

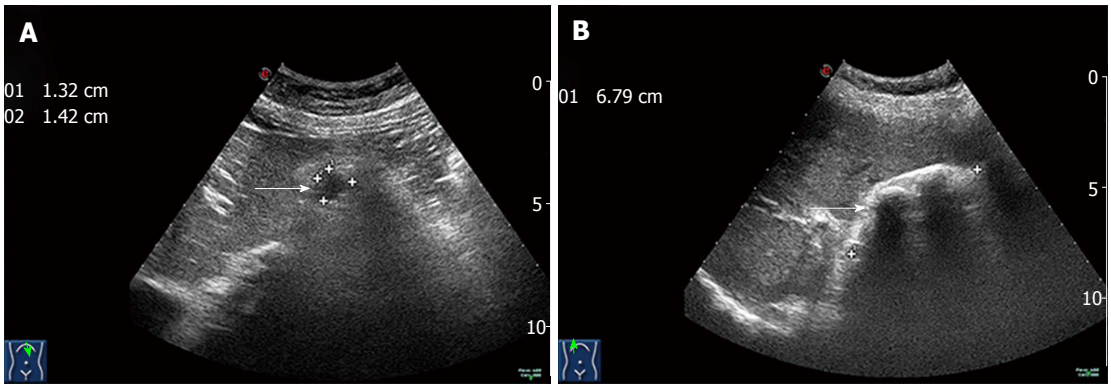


**Figure 1 Ultrasound findings.** A: Ultrasound revealed a 1.4 cm × 1.3 cm, well-defined cystic lesion (arrow) in the pancreatic body; B: A 6.8 cm sized strong echo (arrow) was shown in the gallbladder.

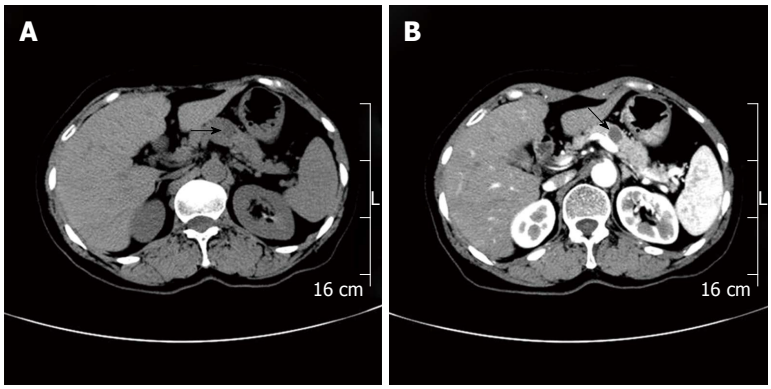


**Figure 2 Computed tomography findings.** A: An unenhanced CT scan showed a 1.6 cm × 1.1 cm well-defined hypodense mass (arrow) in the pancreatic body; B: On the contrast-enhanced CT, the mass (arrow) was not enhanced. CT: Computed tomography.

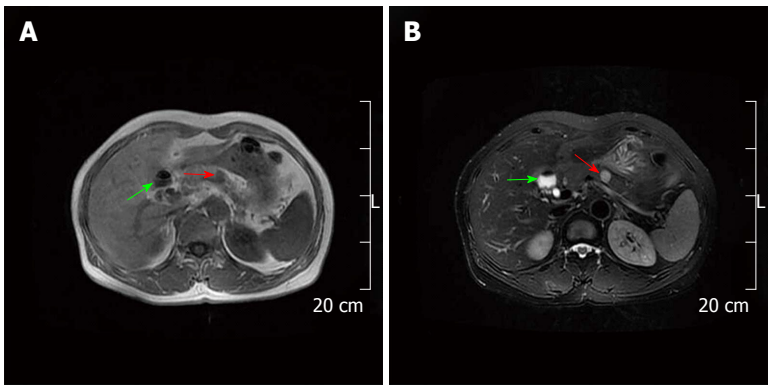


**Figure 3 Magnetic resonance imaging findings.** A: The mass in the pancreatic body (red arrow) and gallbladder (green arrow) appeared hypointense on T1 weighted images; B: The mass in the pancreatic body (red arrow) appeared inhomogeneously hyperintense and the enlarged gallbladder (green arrow) appeared hyperintense on T2 weighted images.

to-side pancreaticojejunostomy, cholecystectomy and inserted a pancreatic stent. Intraoperative frozen pathology revealed a schwannoma in the pancreatic body.

Macroscopically, the mass in the pancreatic body measured 1.6 cm × 1.1 cm × 1.1 cm. Microscopically, the tumor, surrounded by a thin capsule, was mainly composed of spindle-shaped cells with palisading arrangement and no atypia, which was consistent with a benign schwannoma. Both hypercellular and hypocellular areas were visible (Figure 4). Immunohistochemical staining was strongly positive for protein S-100 (Figure 5) and negative for SMA, CD34 and CD117. The final diagnosis was a schwannoma of the

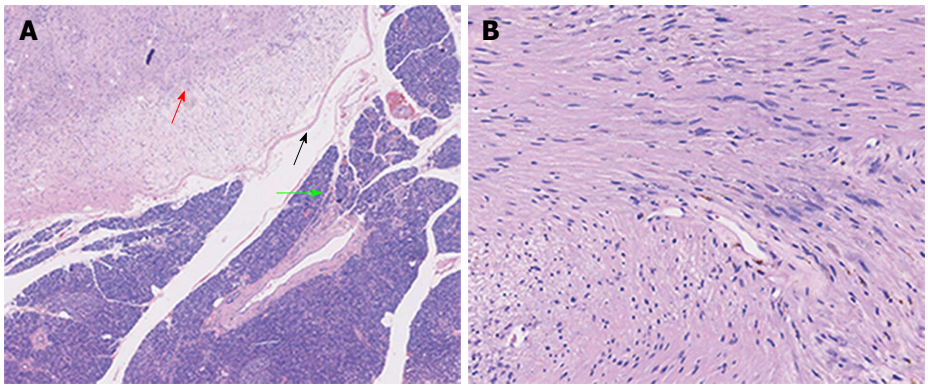


**Figure 4 Microscopic examination.** A: A thin capsule (black arrow) was found between the tumor (red arrow) and pancreatic (green arrow) tissues (HE, 40 ×). B: The tumor was mainly composed of spindle-shaped cells with palisading arrangement and no atypia, which is consistent with a benign schwannoma. Both hypercellular and hypocellular areas were visible (HE, 200 ×). HE: Hematoxylin and eosin.

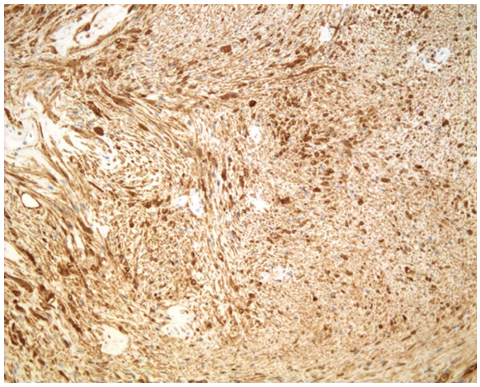

**Figure 5 Immunohistochemical staining.** The tumor revealed strongly positive staining for S-100 (HE, 200 ×). HE: Hematoxylin and eosin.

pancreatic body. Postoperatively, the patient recovered well and left the hospital 6 d later with no evidence of postoperative pancreatic fistula. During the follow-up period of 53 mo, the patient remained well without any complications.

## DISCUSSION

Schwannomas are neoplasms that originate from the Schwann cells of nerve sheaths[62]. More than 90% of schwannomas are benign and manifest in approximately 5% of cases as benign soft-tissue neoplasms[63]. Schwannomas can occur in patients of any age with no obvious gender difference. However, they are most commonly found in patients aged between 20 years and 50 years. Although nearly any part of the human body can be involved, the head, neck and extremities are the most common sites[64]. In the abdominal cavity, the retroperitoneum (6% of primary retroperitoneal tumors)[65] and stomach[66] are the most common sites involved. Schwannomas of the pancreas are rare. Table 1 summarizes the important available clinicopathological characteristics of the 64 cases reported in the English literature over the past 40 years[4-61] and the case presented in our study. Continuous variables were summarized as the mean ± SD and range. The Student's *t* test was used for comparisons of continuous variables. Statistical analyses were conducted using SPSS version 20.0 for Windows (SPSS Inc., Chicago, IL, United States). All tests for significance were two-sided, and $P$ values < 0.05 were considered statistically significant.

Precise preoperative diagnosis of a pancreatic schwannoma is challenging because the clinical symptoms and radiological characteristics of schwannomas are nonspecific. Definitive diagnosis can be achieved only based on the combined results of histopathological and immunohistochemical examinations of surgical specimens. Microscopically, pancreatic schwannomas are encapsulated tumors that consist of hypercellular (Antoni type A area) and hypocellular areas (Antoni type B area) with varying amounts of these histological components[22]. The hypercellular area consists of closely packed spindle cells with occasional nuclear palisading as well as Verocay bodies. The hypocellular area is composed of loosely arranged tumor cells and abundant myxoid stroma[6]. Occasionally these may become cystic, hemorrhagic and calcified[18]. More than 90% of pancreatic schwannomas are benign. However, malignant pancreatic schwannomas have been reported in 5 cases (7.69%)[22,57,59-61]. Immunohistochemically, schwannomas show strongly positive staining for S-100 and negative staining for desmin, smooth muscle myosin, SMA, CD34 and CD117[16,67].

Accurate diagnosis of a pancreatic schwannoma prior to operation is nearly impossible. US, CT and MRI can be performed to establish a probable diagnosis. A pancreatic schwannoma usually appears as a well-defined hypodense lesion on US and shows no echoic enhancement with color Doppler. On unenhanced CT scan, schwannomas are usually well-defined hypodense lesions with encapsulation and/or cystic degeneration. Schwannomas with high Antoni A areas show high density and have a heterogeneous appearance due to high cellularity and increased lipid content. Antoni B areas of schwannomas appear cystic and show low

**Table 1 Summary of clinicopathological data from all 65 cases of pancreatic schwannoma**

| | *n* (%) or mean ± SD (range) |
|---|---|
| Age (yr) (*n* = 64) | |
| Mean | 55.22 ± 15.26 (20-87) |
| Sex (male/female), (male %) (*n* = 64) | 29/35 (45.31) |
| Symptoms[1] (*n* = 64) | |
| Asymptomatic | 24 (37.50) |
| Symptomatic | |
| Abdominal pain | 28 (43.75) |
| Weight loss | 8 (12.50) |
| Back pain | 4 (6.26) |
| Nausea/vomiting | 3 (4.69) |
| Anorexia | 2 (3.13) |
| Abdominal mass | 1 (1.56) |
| Anemia | 2 (3.13) |
| Melena | 2 (3.13) |
| Jaundice | 2 (3.13) |
| Abdominal discomfort | 1 (1.56) |
| Location (*n* = 65) | |
| Head | 26 (40.00) |
| Head/body | 3 (4.62) |
| Body | 15 (23.08) |
| Body/tail | 7 (10.78) |
| Tail | 7 (10.78) |
| Uncinate process | 7 (10.78) |
| Mean size (cm) | 5.83 ± 4.59 (1-20) |
| Benign | 56 (5.27 ± 3.95 ) (1-20) |
| Malignant | 4 (13.75 ± 6.24) (7-20) |
| Operation (*n* = 65) | |
| PD[2] | 20 (30.77) |
| PPPD | 2 (3.08) |
| DP[3] | 16 (24.62) |
| Enucleation | 9 (13.85) |
| Central pancreatectomy | 2 (3.08) |
| Unresectable | 2 (3.08) |
| Refused | 1 (1.54) |
| Not specified | 13 (20.00) |
| Histology (*n* = 65) | |
| Malignant | 5 (7.69) |
| Benign | 59 (90.77) |
| Not specified | 1 (1.54) |
| Nature of tumor (*n* = 65) | |
| Solid | 17 (26.15) |
| Cystic | 28 (43.08) |
| Solid and cystic | 14 (21.54) |
| Not specified | 6 (9.23) |
| Mean follow-up months (*n* = 29) | 20.59 ± 17.76 (3-66) |
| Died of disease | 0 |

[1]Some patients had several symptoms; [2]One patient underwent resection of portal vein; [3]One patient underwent resection of transverse colon. M: Male; F: Female; NA: Not available; DP: Distal pancreatectomy; PD: Pancreaticoduodenectomy; PPPD: Pylorus preserving pancreaticoduodenectomy.

density due to loose stroma and low cellularity[10]. On contrast-enhanced CT, the Antoni A areas are usually enhanced, whereas the Antoni B areas are unenhanced[7]. On MRI, a typical schwannoma appears hypointense in T1-weighted images and appears inhomogeneously hyperintense in T2-weighted images[16]. EUS-FNA may greatly contribute to precise preoperative diagnosis. In a case reported by Li *et al*[28], a pancreatic schwannoma was accurately diagnosed preoperatively by EUS-FNA. In the present study, we also attempted EUS-FNA. However, a sample of the tumor was difficult to acquire because of the small tumor size and the procedure was unsuccessful.

Surgery is the optimal treatment for pancreatic schwannoma. As tumors could be located in different parts of the pancreas, surgical methods vary accordingly. In the present case, laparotomy permitted discovery of the mass in the body of the pancreas. An enlarged gallbladder with multiple stones was also found. We performed central pancreatectomy, end-to-side pancreaticojejunostomy, cholecystectomy and inserted a pancreatic stent. To date, central pancreatectomy had been reported in only one case[5] prior to this study. Compared with traditional distal pancreatectomy and splenectomy for tumors in the body or tail of the pancreas, central pancreatectomy can not only completely resect the tumor, but also preserve the distal pancreas and spleen, which is beneficial to patients. Following complete tumor excision, patients with pancreatic schwannomas generally have a good prognosis.

In conclusion, a schwannoma of the pancreas is rare. To our knowledge, only 64 cases of pancreatic schwannoma have been reported in the English literature over the past 40 years. Precise preoperative diagnosis is challenging despite the application of multiple imaging modalities. Surgery is the most effective treatment for pancreatic schwannoma. As tumors could be located in different parts of the pancreas, surgical methods vary accordingly. Following complete tumor removal, patients with pancreatic schwannomas generally have a good prognosis.

## COMMENTS

### *Case characteristics*

A 59-year-old female was referred to our hospital because of a pancreatic mass found during a routine health examination.

### *Clinical diagnosis*

The abdomen was soft and nondistended without evidence of a palpable mass.

### *Differential diagnosis*

Differential diagnoses included intraductal papillary mucinous neoplasm, mucinous cystic neoplasm, solid pseudopapillary tumor, pancreatic endocrine tumor or pancreatic ductal adenocarcinoma.

### *Laboratory diagnosis*

Abnormal laboratory results included: Unconjugated bilirubin, 2 μmol/L (normal, 3-14) and serum kalium 3.42 mmol/L (normal, 3.5-5.2).

### *Imaging diagnosis*

Ultrasound revealed a 1.4 cm × 1.3 cm, well-defined cystic lesion in the pancreatic body, as well as a 6.8 cm sized strong echo in the gallbladder. An unenhanced computed tomography (CT) scan showed a 1.6 cm × 1.1 cm well-defined hypodense mass in the pancreatic body. On the contrast-enhanced CT, the mass was not enhanced. On magnetic resonance imaging, the mass in the pancreatic body and gallbladder appeared hypointense on T1 weighted images. The mass in the pancreatic body appeared inhomogeneously hyperintense and the enlarged gallbladder appeared hyperintense on T2 weighted images. We also performed Endoscopic ultrasound-guided fine needle aspiration. However,

the tumor sample was difficult to acquire, and the procedure was unsuccessful. According to these results, a pancreatic cystadenoma and an enlarged gallbladder with multiple stones were preliminarily considered.

### Pathological diagnosis

Microscopic examination revealed a tumor composed mainly of spindle-shaped cells with palisading arrangement and no atypia, which is consistent with a benign tumor. Both hypercellular and hypocellular areas were visible. Immunohistochemical staining was strongly positive for protein S-100. Finally, the tumor was diagnosed as a schwannoma of the pancreatic body.

### Treatment

The patient underwent central pancreatectomy, end-to-side pancreaticojejunostomy and cholecystectomy.

### Related reports

Schwannoma of the pancreas is rare. Over the past 40 years, only 64 cases of pancreatic schwannomas have been reported in the English literature.

### Experiences and lessons

Precise preoperative diagnosis is challenging despite the application of multiple imaging modalities. Surgery is the most effective treatment for pancreatic schwannoma. As tumors could be located in different parts of the pancreas, surgical approach varies accordingly. Following complete tumor removal, patients with pancreatic schwannomas generally have a good prognosis.

### Peer review

This manuscript is an interesting surgical case report, good literature review, and well written. This study highlights the diagnosis and treatment of a rare pancreatic schwannoma and presents a literature review to deepen the understanding of the subject. The information included is worthwhile to the reader.

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