Surg Today (2004) 34:550–553
DOI 10.1007/s00595-004-2744-2

SURGERY TODAY

Melanotic Schwannoma of the Pancreas: Report of a Case

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Abstract
We report a case of pancreatic melanotic schwannoma mimicking an epithelial cystic neoplasm of the pancreas. A 67-year-old Japanese woman underwent routine ultrasonography, which showed a large cystic mass in the head of the pancreas. Contrast-enhanced computed tomography (CT) and magnetic resonance imaging (MRI) showed an inhomogeneous, round cystic mass, which was encapsulated, in the head of the pancreas. We performed pyrolus-preserving pancreatoduodenectomy under the tentative diagnosis of a cystic neoplasm of the pancreas. The histopathologic diagnosis was benign melanotic schwannoma. The patient is now well after 43 months of follow-up. We describe the CT and MRI findings in detail, and review the literature on previously reported cases of pancreatic schwannoma.

Key words Pancreas · Melanotic schwannoma · Cystic tumor

Introduction

Pancreatic schwannomas are rare neoplasms derived from nerve sheath cells. To our knowledge, only 15 cases of benign pancreatic schwannoma have been reported in the English literature.[1–11] Since imaging techniques often show these tumors as cystic lesions, clinical confusion with other cystic neoplasms of the pancreas, including serous cystadenoma and mucinous cystic neoplasms, can arise. Histologically, schwannomas consist of two components, hypercellular Antoni A areas and hypocellular Antoni B areas. The diversity of the findings on imaging can be attributed to many variables, which may characterize any given tumor. This case is remarkable in that it was histologically diagnosed as melanotic schwannoma, which is a rare form of schwannoma defined by widespread fine pigment granules in the cytoplasm. We present a case of pancreatic melanotic schwannoma mimicking a cystic neoplasm of the pancreas, followed by a review of the relevant literature.

Case Report

A 67-year-old Japanese woman was referred to us for further investigation of a tumor in the head of the pancreas, which was incidentally found by ultrasonography during a regular checkup. On admission, she was asymptomatic and all laboratory data were within normal limits. She had no history of clinical pancreatitis and there was no sign of von Recklinghausen's disease. Ultrasonography showed a predominantly cystic mass with solid components in the head of the pancreas (Fig. 1). Computed tomography (CT) showed a well-defined round low-density mass, 4×5 cm in diameter. After the injection of contrast medium, enhancement of multiple fine internal septa was seen (Fig. 2). Magnetic resonance imaging (MRI) showed a mass, with hypointensity on T1-weighted images and hyperintensity on T2-weighted images. Gadolinium-diethylenetriamine pentaacetate (DTPA)-enhanced T1-weighted images showed a mass composed of peripheral marked areas of enhancement with nonenhanced cystic necrotic areas. Magnetic resonance cholangiopancreatography (MRCP) showed a hyperintense mass in the pancreatic head with no dilatation of the main pancreatic duct. Endoscopic retrograde cholangiopancreatography was unsuccessful. Angiography showed a hypovascular mass in the head of the pancreas. Displacement of the superior mesenteric vein was seen, without encasement. We

Reprint requests to: Y. Ueda
Received: September 12, 2002 / Accepted: November 4, 2003

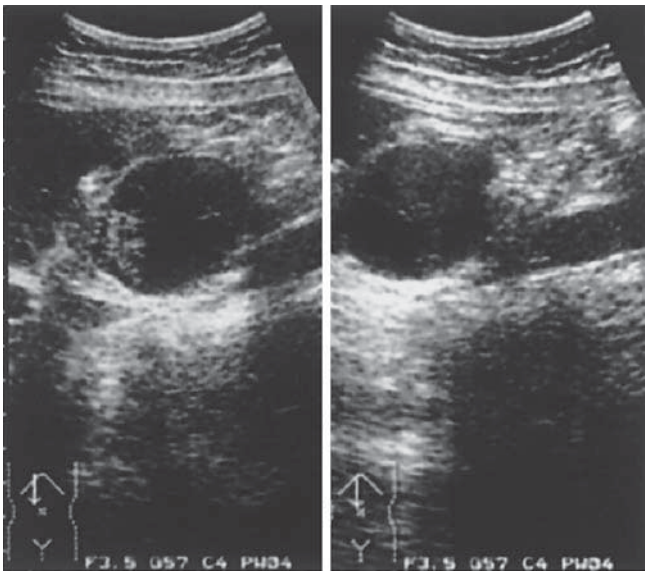

Fig. 1. Ultrasonography showed a predominantly cystic mass in the head of the pancreas

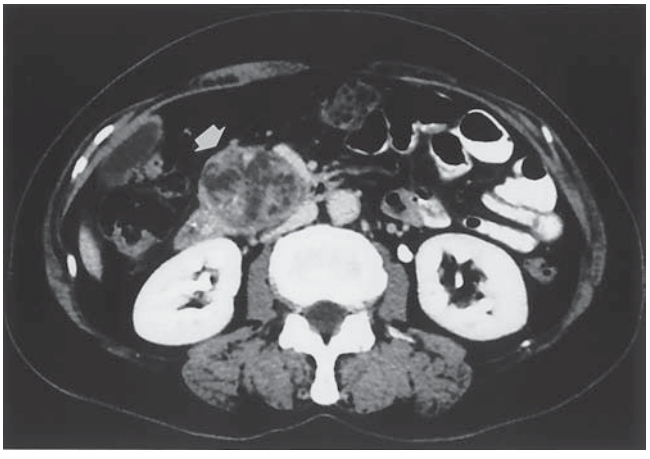

Fig. 2. Contrast-enhanced computed tomography showed a round mass (*arrow*) with multiple fine internal septa

performed surgery under the tentative diagnosis of a cystic neoplasm of the pancreas. Laparotomy revealed an encapsulated, noninvasive tumor in the pancreatic head and uncinate process. We could not separate the tumor from the pancreas, and a pyrolus-preserving pancreatoduodenectomy was performed. The cut surface of the tumor revealed a $5 \times 4 \times 4$-cm encapsulated mass with multiple cystic degeneration, and pale yellow or black-brown areas (Fig. 3). Microscopically, the tumor was composed of compact areas of spindle cells arranged in fascicular or whorled patterns associated with prominent secondary changes, such as cystic change, hyalinization, and hemosiderin deposits (Fig. 4). The tumor cells were positive for S-100 and negative for smooth muscle actin, CD-34, and HMB-45 (melanocytic marker). Many of the tumor cells had fine granular brown pigment in their cytoplasm. This pigment stained positively for Fontana-Masson (Fig. 5) and negatively for iron and periodic acid-Schiff (PAS). The tumor was therefore histologically diagnosed as melanotic schwannoma.

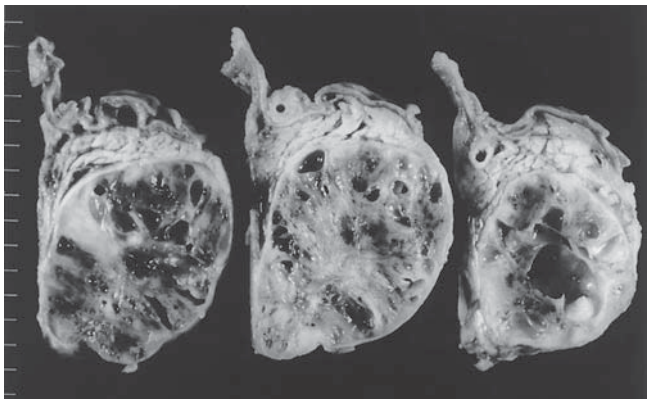

Fig. 3. Cut surface of the tumor showing an encapsulated and multicystic mass

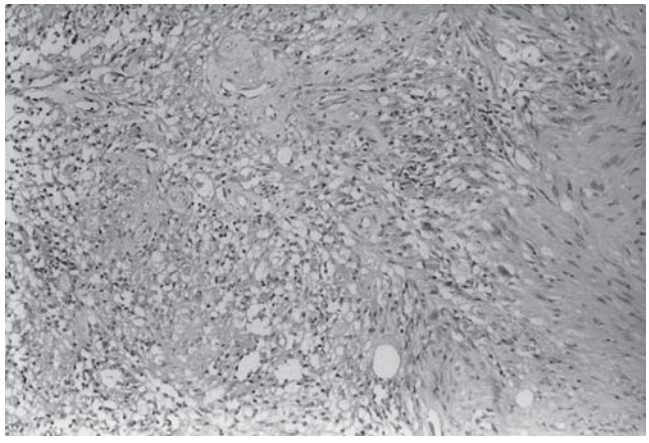

Fig. 4. Photomicrograph showing spindle cells arranged in fascicular or whorled patterns with prominent secondary changes (H&E, ×100)

The hospital stay was extended because it took a long time for delayed gastric emptying to resolve. The patient was finally discharged on postoperative day 72 and remains well 43 months after her operation, without any signs of recurrence.

Discussion

Schwannomas are encapsulated benign tumors of the nerve sheath, commonly occurring in the soft tissue of

the head and neck, the flexor surfaces of the upper and lower extremities, and the trunk. They can also occur in deep structures such as the posterior mediastinum and the retroperitoneum. Peripheral schwannomas are usually less than 5 cm in diameter, but those in the mediastinum or retroperitoneum tend to grow larger.[12] Pancreatic involvement is rare. To the best of our knowledge, only 15 cases of benign pancreatic schwannoma have been previously reported in the English literature. The clinical features of theses 15 cases and our own are summarized in Table 1. The patients ranged in age from 41 to 87 years (mean 61 years), with a nearly equal sex distribution of nine (56%) men and seven (44%) women. The tumor size ranged from 1.5 to 20 cm (mean 6.5 cm), with the pancreatic head involved in seven patients (44%), and the body and tail in nine patients (56%).

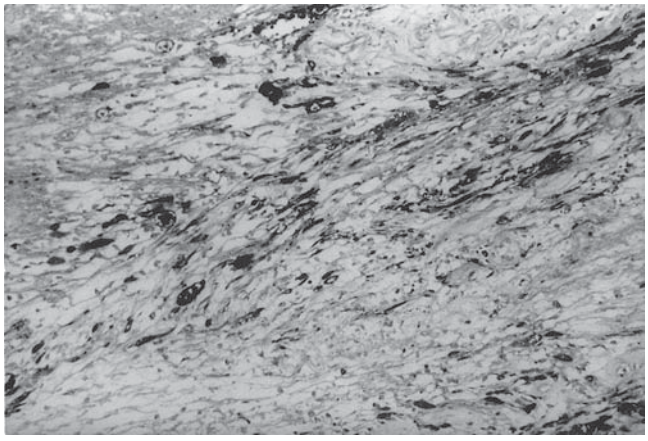

Fig. 5. Fontana-Masson stain revealed cytoplasmic melanin granules (×100)

Schwannomas can be benign or malignant, although malignant transformation of a benign schwannoma is extremely rare.[13] Schwannomas usually occur as solitary lesions, but are occasionally multiple when associated with von Recklinghausen's disease.[13] The prognosis of schwannomas associated with von Recklinghausen's disease is generally less favorable.[3] Two cases of malignant pancreatic schwannoma associated with von Recklinghausen's disease were reported,[14,15] but none of the benign pancreatic schwannomas were associated with von Recklinghausen's disease. Immunohistochemically, S-100 protein is strongly demonstrated by most schwannoma cells, in contrast to neurofibroma cells, which variably express the antigen.[13]

Computed tomography and MRI provide useful complementary information about the size, location, and extent of these tumors. Although three cases of small solid pancreatic schwannomas were reported,[3,6,8] well-defined round masses with multiple low-attenuation cystic necrotic areas are the most characteristic CT finding.[12] Moreover, the cystic changes appear to be somewhat radial.[12] These heterogeneities result from various amounts of the two microscopic components of schwannoma, namely, a highly ordered cellular component (Antoni A areas) and a loose myxoid component (Antoni B areas).[16,17] In tumors that are predominantly or exclusively composed of Antoni A areas, CT shows inhomogeneous hypodense solid masses with contrast enhancement. The hypodensity is created by the high lipid content of the tumor and the contrast enhancement is derived from the reticular vascular component of Antoni A areas.[8,16] This occasion-

Table 1. Reported cases of benign pancreatic schwannoma

Case	First author[Ref.]	Year	Age (years)	Sex	Size (cm)	Location	Appearance	Treatment
1	Liessi[1]	1990	75	F	7	Head	NS	Biopsy
2	Urban[2]	1992	56	F	3 × 4	Body	Cystic	DPS
3	Burd[3]	1992	73	M	2	Body/tail	Solid	Resection
4	Melato[4]	1993	87	M	20	Body/tail	Cystic	DPS
5	David[5]	1993	46	M	6	Head	Cystic	Resection
6	Sugiyama[6]	1995	41	M	1.5	Head	Solid	Resection
7	Ferrozzi[7]	1995	47	M	3.5	Body	NS	Resection
8	Ferrozzi[7]	1995	63	M	NS	Body	Cystic	Resection
9	Ferrozzi[7]	1995	68	F	NS	Head/body	Cystic	NS
10	Feldman[8]	1997	63	M	2	Body	Cystic	Enucleation
11	Feldman[8]	1997	54	F	2	Head	Solid	Enucleation
12	Hsiao[9]	1998	70	F	17 × 17	Body/tail	Cystic	DPS
13	Brown[10]	1998	52	M	5.5	Body	Cystic	DP
14	Brown[10]	1998	69	M	6	Head	Cystic	Whipple
15	Morita[11]	1999	50	F	10	Body/tail	Cystic	DPS
16	Present case		67	F	4 × 5	Head	Cystic	PpPD

NS, not specified; DPS, distal pancreatosplenectomy; DP, distal pancreatectomy; PpPD, pylorus-preserving pancreatoduodenectomy; Whipple, Whipple's procedure

ally gives rise to a complex appearance, which may appear multiseptated. When the tumor is predominantly composed of Antoni B areas, CT shows homogeneous cystic masses without significant contrast enhancement.[8,16] Since deep tumors tend to grow larger, they are more likely to show secondary degenerative changes such as cyst formation, calcification, hemorrhage, and hyalinization,[13] and are known as "ancient" schwannomas. Because of their cystic appearance, pancreatic schwannomas are often misdiagnosed as pseudocysts or other epithelial cystic neoplasms of the pancreas, such as microcystic serous adenoma or mucinous macrocystic tumor. Pancreatic schwannomas should be considered in the differential diagnosis of cystic neoplasms of the pancreas, although the diagnosis can only be confirmed by microscopic examination.

On MRI, schwannomas are seen as masses of low signal intensity on T1-weighted images and of high signal intensity on T2-weighted images.[12] The T1-weighted MR images characteristic of melanotic schwannoma demonstrate a high signal intensity caused by the large amounts of melanin in the tumor.[1] The lesion in our patient was hypointense on the T1-weighted MR images.

Melanotic schwannoma, which commonly arises from the sympathetic nervous system, is very different from classic schwannoma.[13] More than 50% of patients with this tumor have signs of Carney's syndrome, such as myxomas, spotty pigmentation, and endocrine hyperactivity producing Cushing's syndrome.[13] Some report expressed concern about malignant potential with a capacity to metastasize.[1] The tumor is typically black-brown on gross examination, with microscopic coarse or fine intracytoplasmic melanin granules, which stain positively for Fontana and negatively for iron and PAS.[10]

Complete local excision is thought to be adequate surgical treatment for schwannomas, although radical operations such as pancreatoduodenectomy were performed in some patients because a definite preoperative diagnosis could not be established. Intraoperative frozen sectioning may be helpful in this regard.[8]

In conclusion, we reported a case of pancreatic schwannoma mimicking an epithelial cystic neoplasm of the pancreas on imaging. Thus, the possibility of pancreatic schwannoma should be considered in the differential diagnosis of cystic lesions of the pancreas.

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