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CASE REPORT

Pancreatic Schwannoma. A Case Report and Review of the Literature

Trung D Bui[1], Tien Nguyen[1], Sergio Huerta[1], Mai Gu[2], David Hsiang[1]

[1]Department of General Surgery and [2]Department of Pathology, UCI Medical Center. Orange, CA, USA

ABSTRACT

Context Pancreatic schwannomas are rare neoplasms. These tumors vary in size and two thirds are partially cystic which grossly mimic pancreatic cystic lesions. Computed tomography and magnetic resonance imaging are the primary initial imaging modalities. Definitive diagnosis is typically made at the time of laparotomy following biopsy. Surgical resection is the mainstay of treatment.

Case report A 69-year-old woman presented with abdominal pain in the epigastric and left upper quadrant. The patient had no systemic symptom and laboratory results including tumor markers were negative. A CT scan of the abdomen showed a 5 cm mass arising from the head of the pancreas. Needle biopsy revealed a mass consistent with schwannoma. At laparotomy, a large pancreatic head mass was found to encase the superior mesenteric artery, and portal vein confluence. Frozen biopsy showed schwannoma. Curative resection was deferred due to extensive vascular involvement and favorable tumor biology. A gastrojejunostomy was performed and radiation therapy was instituted post-operatively.

Conclusions Only 24 cases of pancreatic schwannoma had been previously reported. Definitive diagnosis is obtained with routine histology. Most tumors are benign and surgical resection is curative. The role of radiation therapy in the management of unresectable tumors is still unclear.

INTRODUCTION

Schwannomas are uncommon neoplasms. They typically arise from neural crest cells which encapsulate the nerve sheath and are sometimes also referred as neurilemmomas. Schwannomas usually occur in the extremities, but can also be found in the trunk, head and neck, retroperitoneum, mediastinum, pelvis and rectum [1]. Benign schwannomas comprise 65% of all neurogenic tumors; however 10-15% of these tumors are malignant [2, 3].

Pancreatic schwannomas are even more unusual neoplasms that affect adults with an equal gender distribution [4]. These tumors vary considerably in size and approximately two thirds are reported to undergo degenerative changes including cyst formation, calcification, hemorrhage, hyalinization and xanthomatous infiltration [5]. As a result, they may radiographically mimic cystic pancreatic lesions (e.g. pancreatic cystadenomas and cystadenocarcinomas and pancreatic pseudocysts), making a preoperative diagnosis a clinical challenge.

In the present report, we discuss a case of pancreatic schwannoma and provide a pertinent review of the literature with an emphasis on the clinical presentation,

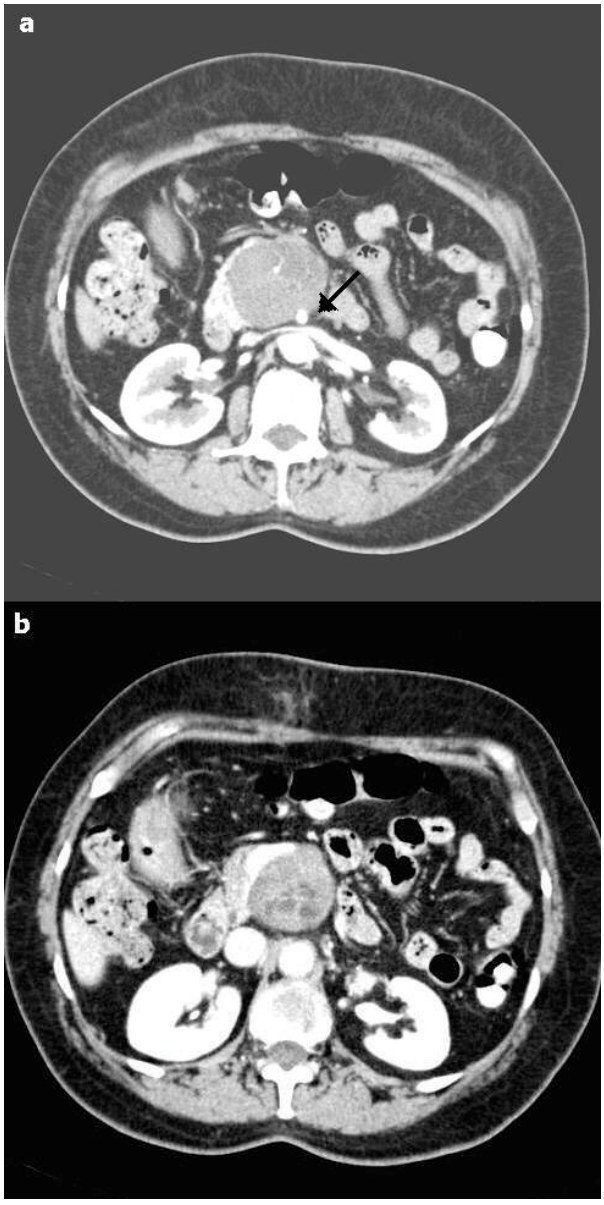

Figure 1. a. CT scan demonstrated a pancreatic head mass that abuts the superior mesenteric artery (black arrow). **b.** Loculated cystic degeneration can also be seen in the center of the tumor.

diagnostic modalities and treatment options in the management of this rare clinical entity.

CASE REPORT

A 69-year-old female presented to the hospital with progressive worsening left upper quadrant and epigastric pain for three weeks. Her pain was constant and ranged from moderate to severe and was associated with nausea and vomiting. The patient denied other constitutional symptoms such as weight loss or fever.

Her past medical history was unremarkable. The patient denied alcohol, tobacco or illegal drugs use. On physical examination, she was hemodynamically stable and afebrile. Her abdomen was soft, non-distended, without a palpable mass, but moderately tender to palpation in the epigastrium and left upper quadrant.

Laboratory tests demonstrated normal hemoglobin and hematocrit at 12.7 g/dL (reference range: 12.7-17.5 g/dL) and 37.3% (reference range: 39-49%), respectively. The chemistry panel, liver function tests and amylase were normal as were tumor markers cancer antigen 19-9 (CA 19-9: 10.7 U/mL; reference range: 5.0-37.0 U/mL) and carcinoembryonic antigen (CEA: 2.2 U/mL; reference range: 0-2.5 U/mL).

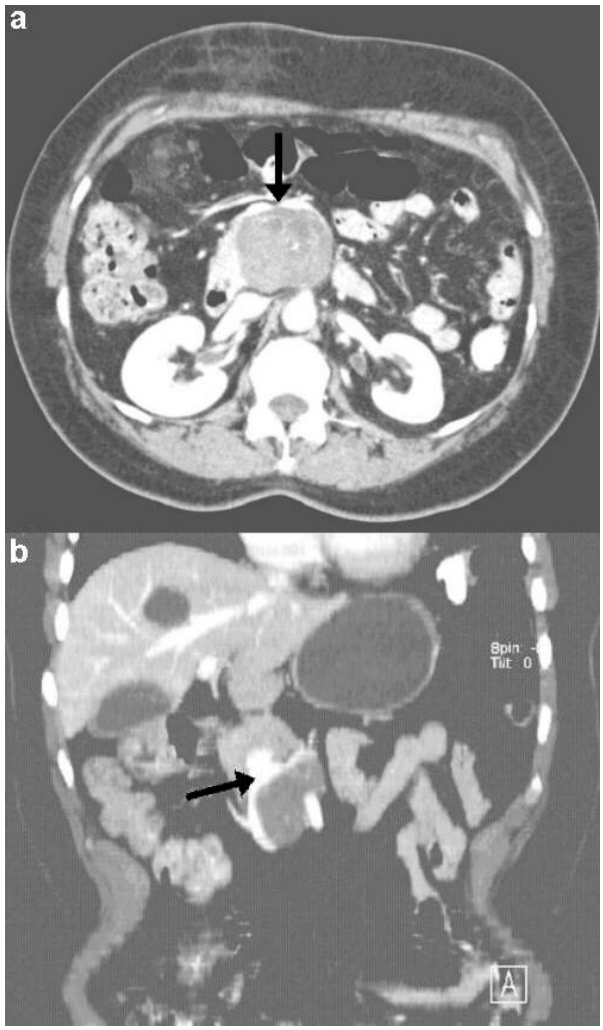

Figure 2. Delayed-phase CT scan (**a.**) with 3-D reconstruction (**b.**) demonstrated tumor abutment at the portal vein confluence (black arrows).

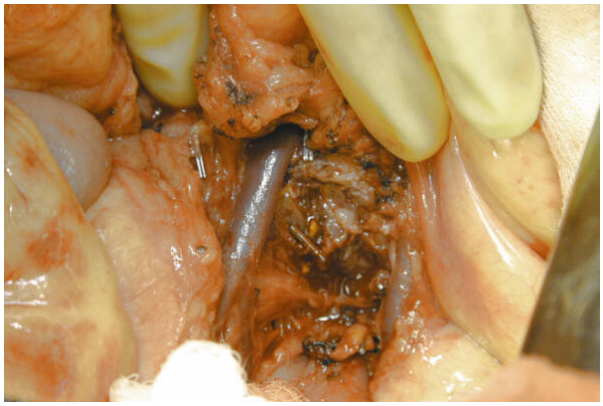

Figure 3. Tumor encasement of the portal vein confluence. The SMA was also involved but very fibrotic for safe mobilization.

Dynamic computed tomography (CT) scans with 3-dimenional reconstruction of the abdomen and pelvis demonstrated a 5.0x4.4 cm mass at the pancreatic head with abutment of the superior mesenteric artery and portal vein confluence (Figures 1 and 2). Endoscopic ultrasound (EUS) and fine needle aspiration (FNA) of the mass demonstrated characteristic spindle cells, whorling appearance and strong immunoperoxidase staining with S-100 protein which were consistent with schwannoma. Clinical staging with CT scans and EUS demonstrated no lymph node involvement or metastasis. The patient was subsequently taken to the operating room for tumor resection.

At laparotomy, a large retroperitoneal mass at the head of the pancreas was found to encase the superior mesenteric artery, and the portal vein confluence (Figure 3). No lymphadenopathy in the mesentery, peritoneal implants or liver lesion was found. Frozen biopsy confirmed benign schwannoma (Figures 4 and 5). Due to extensive vascular involvement and favorable tumor biology, the tumor was not completely resected. A gastrojejunostomy was then performed.

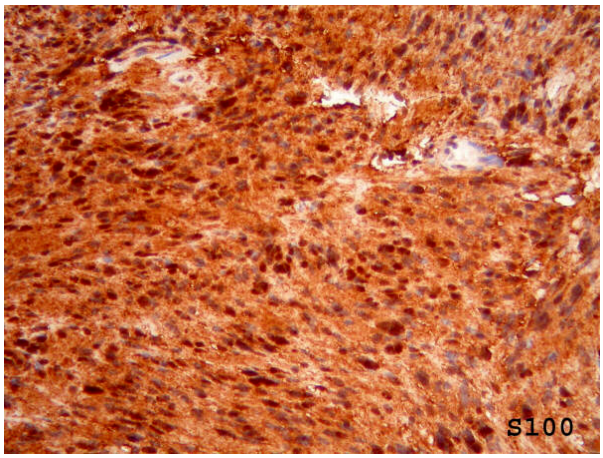

Figure 4. Immunohistochemical stain with S100 protein antibody shows that the tumor cells are diffusely and strongly positive (Avidin-biotin-peroxidase complex method, 10x).

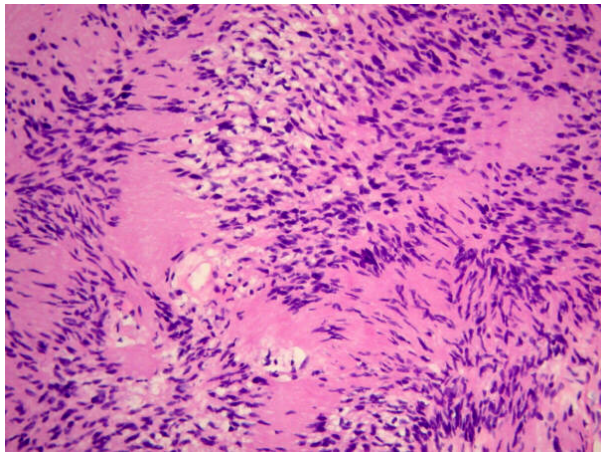

Figure 5. The histologic section from the biopsy of this pancreatic tumor reveals a long spindle cell proliferation with eosinophilic neurofibliary background. The nuclei are slender, slightly wavy, and with pointed ends. Antoni A and B areas are both evident. In this field of hypercellular area, palisading nuclei are arranged in a palisading fashion (Verocay body). (Hematoxylin and Eosin stain, 10x).

The patient's post-operative course was unremarkable. She was discharged home on post-operative day number five and was scheduled for radiation therapy and CT scan surveillance. Curative resection will be offered if adjuvant therapy results in safe tumor resection.

DISCUSSION

Only 24 cases of pancreatic schwannoma have been reported in the English literature (Table 1) [6]. These tumors vary considerably in size ranging from 1.5 to 20.0 cm in diameter with the majority of the tumors located in the head (38%) and body (25%) of the pancreas. Schwannomas of the pancreas arise from Schwann cells encasing peripheral nerves and can be benign or malignant. Generally, pancreatic schwannomas have slow growth rates and usually originate from

Table 1. Reported cases of pancreatic schwannoma.

Case	Study	Year	Gender, age (yrs)	Size (cm)	Location	Gross	Histology	Treatment	Follow-up (months)
1	Paranjape [16]	2004	F 77	3.5	Head	Solid	Benign	Enucleation	3 A/W
2	Tan [6]	2003	M 46	2.2	Head	Cystic/solid	Benign	Whipple	N/A
3	Almo [17]	2001	F 73	3.0	Head	Cystic	Benign	Whipple	17 A/W
4	Almo [17]	2001	F 47	5.5	Head	Solid	Benign	Whipple	14 A/W
5	Lee [5]	2001	F 63	10	Tail	Cystic	Benign	DP+S	6 A/W
6	Brown [4]	1998	M 52	5.5	Body	Cystic	Benign	Resection**	N/A
7	Brown [4]	1998	M 69	6.0	Head	Cystic	Benign	Whipple	N/A
8	Hsiao [18]	1998	F 70	17.0	Body/tail	Cystic	Benign	Resection**	24 A/W
9	Feldman [11]	1997	M 63	2.5	Body	Solid	Benign	Enucleation	N/A
10	Feldman [11]	1997	F 54	2.0	Uncinate	N/A	Benign	Enucleation	20 A/W
11	Todd [19]	1997	F 46	12.0	Head	Solid/Cystic	Benign	Excision**	N/A
12	Ferrozzi [12]	1995	M 47	3.5	Body	Inhomogeneous	Benign	PP	48 A/W
13	Ferrozzi [12]	1995	M 63	N/A	Body	Cystic	Benign	N/A	N/A
14	Ferrozzi [12]	1995	F 68	N/A	Body/head	Cystic	Benign	N/A	6 A/W6
15	Sugiyama [20]	1995	M 41	1.5	Uncinate	Cystic	Benign	Whipple	NA
16	Steven [21]	1994	M 59	4.0	Uncinate	Solid	Benign	DP	10 A/W
17	Melato [22]	1993	M 87	20.0	Body/tail	Cystic	Benign	Resection**	N/A
18	David [23]	1993	M 46	6.0	Uncinate	Partially cystic	Benign	DP	N/A
19	Urban [7]	1992	F 56	4.0	Body	Cystic	Benign	DP	N/A
20	Burd [24]	1992	M 73	2.0	Body	Solid	Benign	N/A	N/A
21*	Coombs RJ [1]	1990	F 74	7.0	Head	Necrotic center	Malignant	Excision**	N/A
22*	Walsh [10]	1989	F 35	N/A	Head	N/A	Malignant	Whipple	2 A/W
23	Eggermont [9]	1987	F 40	10.0	Head	Central necrosis	Malignant	Whipple	9 A/W
24	Moller-Pederson [8]	1982	M 60	15-20	Body/head	Cystic	Malignant	Unresectable	4 A/W

*Associated with von Recklinghausen's disease
**No specific operation documented
A/W: alive and well
N/A: not available
DP: Distal pancreatectomy
DP+S: Distal pancreatectomy and splenectomy
PP: Partial pancreatectomy

the peripheral epineurium of either autonomic sympathetic or parasympathetic fibers, which embryologically course through the pancreas via the vagus nerve [7].

As depicted in Table 1, more than half of the reported schwannomas are cystic and 83% are benign. Malignant pancreatic schwannomas, albeit uncommon, have been reported in four cases [1, 8, 9, 10]. Malignant transformation is associated with von Recklinghausen's disease in 8% of cases [1, 10].

Non-specific abdominal pain was the most commonly reported symptoms but weight loss, jaundice and gastrointestinal bleeding had been reported [8, 9, 10]. The pre-operative diagnosis of schwannoma presents a clinical challenge and CT scan is often the initial study of choice to establish a pancreatic lesion. CT findings in pancreatic schwannomas, however, are typically similar to non-pancreatic schwannomas and usually demonstrate well-defined and hypodense tumors with encapsulation and cystic degeneration [11]. Degenerative changes with cyst formation pose a formidable diagnostic challenge because they mimic the whole spectrum of cystic pancreatic lesions including: neuroendocrine tumors, cystadenoma, cystadenocarcinoma, intraductal papillary mucinous tumor,

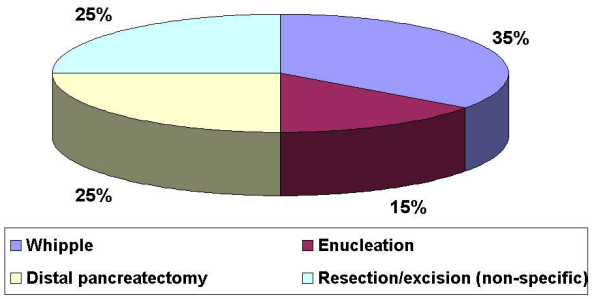

Figure 6. Operative management of pancreatic schwannomas.

lymphangiomas and pancreatic pseudocysts [6].

Once a pancreatic lesion has been determined, magnetic resonance imaging (MRI) is helpful in characterizing schwannomas by their typical encapsulation, hypointensity on T1-weighted images and hyperintensity on T2 [11, 12]. In addition to variable signal intensities, MRI may reveal vascular involvement which can further characterize a lesion with malignant potential [11]. MRI may also differentiate pancreatic schwannoma from adenocarcinoma by the characteristic hyperintensity on T2 weighted images and marked enhancement of the lesion in comparison with the remainder of the gland [11]. However, further differentiation from islet cell tumors cannot be established by this modality [11].

Further characterization of a pancreatic mass may be established by EUS-guided FNA. However, the diagnosis of pancreatic schwannoma by FNA may be complicated by inadequate amount of specimen or by inherent specimen collecting techniques. A recent study demonstrated that FNA correctly diagnosed only one out of eight histologically proven schwannomas [13].

Although pre-operative radiographic imaging may help narrow the differential diagnosis, pancreatic schwannoma is usually diagnosed at the time of laparotomy, followed by histological analysis [4, 6, 11]. Microscopically, schwannomas stain strongly positive for S-100 protein, vimentin and CD56 while negative for other tumor markers including cytokeratin AE1/AE3, desmin, smooth muscle myosin, CD34 and CD117 (Figure 4) [6]. On histological analysis, schwannomas have two different patterns which are designated as Antoni A and B. Antoni A refers to areas that are highly cellular and composed of spindle cells arranged in a palisading fashion without mitotic figures, while Antoni B areas are typically hypocellular with degenerative changes (Figure 5) [6].

Since malignant transformation of pancreatic schwannomas is uncommon, simple enucleation is usually sufficient even though oncologic resections (Whipple's procedure and distal pancreatectomy) had been reported (Figure 6). To date, no documented recurrent case has been reported for either mode of resection. In the present case, the tumor was found to encase the superior mesenteric artery as well as the portal vein confluence with severe associated inflammation and fibrosis of the surrounding tissues. Curative surgical resection was thus deemed high risk.

Review of the literature shows one reported case of unresectable tumor, but no report of previous treatment with chemoradiation therapy [8]. However, radiation therapy has been shown to decrease tumor growth and regression in neurogenic schwannomas [14, 15]. Because schwannomas share similar biological characteristics, it is conceivable that radiation therapy would have similar effects in pancreatic schwannomas as with neurogenic schwannomas. However, the role of radiation therapy in the management of pancreatic schwannomas is still unclear. Surgical excision with close follow up and surveillance remain the mainstay of treatment.

CONCLUSION

Pancreatic schwannoma, albeit rare, is an important clinical entity to include in the differential diagnosis of pancreatic lesions, especially when cyst formation was demonstrated on radiologic imaging. Pre-operative diagnosis is difficult but CT and MRI help raise the suspicion of this diagnosis. Definitive diagnosis, however, requires histological examination. Simple enucleation is usually adequate for benign tumors while malignant tumors require standard oncologic

resection. The role of radiation therapy in the management of unresectable tumors is still unclear and therapeutic effectiveness remains to be seen.

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Correspondence
Trung D Bui
8400 Edinger Avenue #K108
Huntington Beach, CA 92647
USA
Phone: +1-714.847.1996
Fax: +1-714.456.7207
E-mail address: buitd@uci.edu

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