

A Rare Case of Cystic Schwannoma of the Portal Triad

Dimitrios V. Avgerinos, MD,¹ Meshach Heenatigala, MD,² and Andrew Y. Lo, MD²

¹ Department of Cardiothoracic Surgery, New York Presbyterian–Weill Cornell Medical Center, ² Department of Surgery, Beth Israel Medical Center, Albert Einstein College of Medicine.

Schwannomas can occur anywhere throughout the body and have often been mistaken for more-sinister lesions, especially when found in relation to the pancreas. Clinical symptoms range from none to vague abdominal pain, back pain, anorexia, weight loss, vomiting, jaundice, and episodes of cholangitis and gastrointestinal bleeding. Preoperative diagnosis is difficult, and endoscopic ultrasound with fine-needle aspiration is often limited in specificity. Given the low statistical likelihood of schwannomas, therapy is usually targeted at the possibility of pancreatic cystadenoma/cystadenocarcinoma. Simple enucleation is usually the preferred treatment, and

diagnosis can be established at the time of operation by frozen section. Schwannomas can be malignant, but preoperative imaging and pathology can help establish the benign nature of most specimens. Patients typically do well with resolution of symptoms. Here we present the case of a patient with abdominal pain and a peripancreatic mass observed with computed tomography, who was found to have a cystic schwannoma extending from the portal triad. The mass was removed and the patient was discharged without complications.

INTRODUCTION

Schwannomas are often encountered within the cranial vault. Within the vault, they are most common at the vestibular branch of the eighth cranial nerve at the cerebellopontine angle, presenting with tinnitus and hearing loss (acoustic neuroma or vestibular schwannoma). Schwannomas can, however, be encountered in any peripheral nerve, most commonly occurring in the head and neck, flexor surfaces of the extremities, mediastinum, retroperitoneum, pelvis, and posterior spinal roots. Operative resection can be considered when pain, paresthesia, or weakness occurs; generally, only mild neurologic deficits occur. Magnetic resonance imaging (MRI) with gadolinium contrast can aid in diagnosis (Cowan and Thompson, 2009).

To date, at least 44 schwannomas anatomically related to the pancreas have been described in the English literature. There are also descriptions of the involvement of surrounding structures, including the portal vein (Todd et al., 1997; von Dobschuetz et al., 2004), splenic vein (Di Benedetto et al., 2007; Bui et al., 2004), and superior mesenteric artery (Todd et al., 1997; Bui et al., 2004). However, a search of the English literature has not revealed the description of a schwannoma of the portal triad itself. Here is a case of a cystic schwannoma anatomically connected to the portal triad.

CASE PRESENTATION

A 40-year-old female with hypertension and borderline diabetes controlled by diet presented with a cystic mass in the pancreas, observed with computed tomography (CT), that increased in size over one year. Her symptoms consisted of vague abdominal discomfort for two years and increasing discomfort for the six months prior to admission.

She originally had a CT scan one and a half years prior

to admission for mild abdominal pain and urinary tract infections that demonstrated a 3 x 3 cm cystic lesion in the head of the pancreas. Follow-up endoscopic ultrasound (EUS) with fine-needle aspiration (FNA) showed no malignant cells and a carcinoembryonic antigen (CEA) content of 0.3 ng/ml. Ninety-five percent of normals have CEA levels less than 5.0 ng/ml, with levels above 200 ng/ml considered significantly elevated. A follow-up CT scan at one year showed the lesion enlarged to 5.3 x 4.2 cm.

An MRI one month prior to presentation demonstrated a large cystic mass, which appeared to arise from the superior portion of the pancreas at the junction of its body and head. The mass appeared exophytic with hyperintensity on T2 signal with multiple thin and thick internal septations, some of which were enhanced with delayed postcontrast imaging. The mass displaced the portal vein and the common bile duct posteriorly and displaced the hepatic artery laterally. There was minimal proximal intrahepatic biliary duct dilatation. The radiologic differential diagnosis included mucinous cystadenoma/cystadenocarcinoma among other less likely possibilities. A second, much smaller mass in the distal pancreatic body/tail region, which was 15 x 13 mm, was also noted.

Physical exam was unremarkable and did not reveal any notable abdominal tenderness. The patient was sent for endoscopic retrograde cholangiopancreatography (ERCP) to determine if there was communication between the pancreatic duct and the cystic mass. No communication was seen, and at the time of ERCP, a long pancreatic stent was placed in the body of the pancreas to prevent leakage of the duct during surgical resection, with plans for its removal after the surgery. After the ERCP, the patient had abdominal pain, was febrile, and had a lipase of 3869 U/l and was thought to have pancreatitis secondary to the ERCP procedure. After improving, she was taken to the operating room one week later for resection of the cystic neoplasm and

► CASE REPORT

A Rare Case of Cystic Schwannoma of the Portal Triad

cholecystectomy. Depending on intraoperative findings, a possible Whipple's procedure was planned.

An open procedure with a right subcostal incision was performed. Intraoperatively, a 6 x 7 cm cystic mass was seen sitting on the portal triad, displacing the portal vein posteriorly, the hepatic artery medially, and the common bile duct laterally. The mass was able to be enucleated and did not involve the pancreas. Frozen section intraoperatively found the mass to be a schwannoma with cystic degeneration. The surgery was then completed without further resection. The pathology showed a tan-pink cystic structure with average wall-thickness of 0.3 cm, with spindle cells, atypical cells, degenerative changes, and S-100 immunoreactivity suggestive of benign cystic schwannoma. The gallbladder was negative for significant histopathological changes and was 8.9 x 3.7 x 3.0 cm without calculi.

On postoperative day three, the patient underwent an upper endoscopy for stent removal without complications. The patient was later discharged from the hospital in good health.

DISCUSSION

Schwannomas, or neurilemmomas, are slow-growing tumors of the peripheral nerves. They are the most common peripheral nerve tumors. Previous authors have noted that the tumors, when occurring around the pancreas, likely arise from the epineurium of the sympathetic or parasympathetic fibers that course along with the vagus nerve through the pancreas (Tafe and Suriawinata, 2008). Similarly, our patient's schwannoma likely arose from autonomic sympathetic or parasympathetic fibers on the portal triad vessels.

While neurofibromas have nerve fascicles running through the tumor, schwannomas have nerve fascicles running alongside the tumor. Characteristics of Schwann cells that aid in identification include the presence of the S-100 antigen and the potential for melanocytic differentiation; other indicatory antigens include vimentin, CD56 and, occasionally, CD57, and glial fibrillary acidic protein (Gupta et al., 2009). Schwannomas are composed histologically of Antoni A and Antoni B areas. Antoni A regions are hypercellular and composed of tightly packed spindle cells with occasional nuclear palisading and Verocay bodies. Antoni B regions are hypocellular with degenerative changes; tumor cells are more spread out in a myxoid stroma (Gupta et al., 2009). This explains the solid and cystic appearance of schwannomas on CT scan, corresponding to the presence of Antoni A and B areas, respectively (Wu et al., 2005; Ferrozzi et al., 1995).

The tumors tend to be firm and gray, with some reports describing variegated tan-yellow-to-red (Tafe and Suriawinata, 2008), and pale yellow cut surfaces (Morita et al., 1999). Degenerative changes described include

cyst formation, calcification, hemorrhage, hyalinization, and xanthomatous infiltration (Lee et al., 2001). Smaller tumors tend to be solid while larger tumors tend to exhibit the aforementioned degenerative characteristics (Bui et al., 2004; Burd et al., 1992). Malignant changes can occur but are rare in schwannomas; local recurrence, though, can follow incomplete resection. Malignant peripheral nerve sheath tumor is also known as malignant schwannoma but is actually a malignant sarcoma that is locally invasive with potential for metastatic spread. Almost two-thirds develop from neurofibromas in the setting of neurofibromatosis type I, with the remainder arising de novo or from external beam radiation. Schwannomas can occur sporadically but are also associated with neurofibromatosis type II.

Case reports in the literature of schwannomas within or adjacent to the pancreas demonstrate that patients tend to have vague complaints of generalized abdominal or epigastric pain. Many patients also have been asymptomatic; other reported symptoms include pain radiation to the back, anorexia, vomiting, weight loss, jaundice, gastrointestinal bleeding secondary to erosion into the bowel wall, and recurrent cholangitis (Bui et al., 2004; Wu et al., 2005; Walsh and Brandspigel, 1989; Møller Pedersen et al., 1982; Eggermont et al., 1987; Tofigh et al., 2008). Physical exam has been either unrevealing or demonstrative of epigastric tenderness and presence of a mass in larger cysts (Yu and Sun, 2006).

A lack of response to GERD treatment or clinical suspicion tends to lead to abdominal CT scan, which demonstrates similar findings to schwannomas elsewhere in the body: heterogeneity with hypodense solid areas and cystic necrotic areas, encapsulation, and clear demarcation (Yu and Sun, 2006; Feldman et al., 1997). The degenerative cystic components give rise to a radiologic differential diagnosis that includes pancreatic pseudocysts, neuroendocrine tumors, cystadenoma, cystadenocarcinoma, intraductal papillary mucinous tumors, lymphangiomas, von Hippel-Lindau disease, hydatid cysts (Bui et al., 2004; Ferrozzi et al., 1995; Urban et al., 1992; Tan et al., 2003), and choledochal cysts.

Other diagnostic modalities include abdominal ultrasound if the mass is large or palpable. EUS with FNA is an option (Sugiyama et al., 1995), although retrieving a sufficient quantity of cells has been a problem, as schwannomas tend to be hypocellular and have large areas of stroma. Even adequate specimens may yield nonspecific cytology (Yu et al., 1999). Clinicians in one case specifically avoided performing EUS-guided FNA given that the presence of a mass observed with CT and symptoms of vague abdominal discomfort would warrant surgical removal regardless of the FNA findings (Mummadi et al., 2009). If FNA is deemed useful, however, options include CT guidance, FNA under vision intraoperatively, and FNA using high negative suction pressure (Bui et al., 2004; Hirabayashi et al., 2008; Larghi et al., 2005).

A Rare Case of Cystic Schwannoma of the Portal Triad

MRI can also be helpful with characteristic T1 hypointensity and T2 hyperintensity, which can help distinguish schwannomas from adenocarcinomas (Bui et al., 2004; Wu et al., 2005; Feldman et al., 1997; Novellas et al., 2005).

Changes suggestive of malignancy include faster growth, lack of demarcation and presence of invasion, contrast enhancement, irregular borders, and vascular thrombosis (Ferrozzi et al., 1995); hypervascularity on angiography has not been found to be helpful, as even most benign tumors are hypervascular, correlating with the tumor's Antoni A component (Morita et al., 1999).

Rarely is the entity of schwannoma included in the preoperative differential diagnosis. Definitive preoperative diagnosis is indeed difficult, and many such masses are treated with surgical resection. If the mass is well circumscribed and does not have infiltrative properties on preoperative imaging, simple enucleation with intraoperative frozen section finalizing the diagnosis is the recommended approach. Because of diagnostic uncertainty and the possibility of pancreatic neoplasm or schwannoma with malignant changes, infiltrative tumors without clear demarcation have been removed en bloc, including with such procedures as distal pancreatectomy, concurrent splenectomy, omentectomy, and Whipple's procedure (Gupta et al., 2009). Patients typically do well and are without complaints at a year's follow-up. If involvement of vascular structures prevents surgical resection, radiotherapy is a possible option, although details of management are not well established (Bui et al., 2004).

CONCLUSION

In summary, we present here the case of a 40-year-old female who presented with vague abdominal pain for two years and was found to have a large cystic mass near the pancreas. The preoperative diagnostic concern was that this was a pancreatic cystadenoma/cystadenocarcinoma. Intraoperatively, the mass was found to arise from the portal triad, and by frozen section, it was concluded that the lesion was a benign cystic schwannoma. Though rare, schwannomas near or within the pancreas have been reported and are often mistaken for entities that are more pathologically worrisome. Preoperative diagnosis remains difficult, but simple enucleation remains the standard treatment with excellent prognosis.

Corresponding Author: Dimitrios V. Avgerinos, MD (dva9001@nyp.org).

Author Contributions: DA and MH performed data collection and manuscript preparation. AL edited the manuscript.

Conflict of Interest Disclosures: The authors have completed and submitted the ICMJE Form for Disclosure of Potential Conflicts of Interest. No conflicts were noted.

Supplementary Materials: Supplementary materials may be found online at www.einstein.yu.edu/ejbm.

REFERENCES

- Bui, T.D., Nguyen, T., Huerta, S., Gu, M., and Hsiang, D. (2004). Pancreatic schwannoma: A case report and review of the literature. *JOP* 5:520-526.
- Burd, D.A., Tyagi, G., and Bader, D.A. (1992). Benign schwannoma of the pancreas. *Am J Roentgenol* 159:675.
- Cowan, J.A. and Thompson, B. (2009). Neurosurgery. In: CURRENT Diagnosis and Treatment: Surgery. 13th ed. Doherty, G.M. (ed.), McGraw-Hill.
- Di Benedetto, F., Spaggiari, M., De Ruvo, N., Masetti, M., Montalti, R., Quintini, C., Ballarin, R., Di Sandro, S., Costantini, M., and Gerunda, G.E. (2007). Pancreatic schwannoma of the body involving the splenic vein: Case report and review of the literature. *Eur J Surg Oncol* 33:926-928.
- Eggermont, A., Vuzevski, V., Huisman, M., De Jong, K., and Jeekel, J. (1987). Solitary malignant schwannoma of the pancreas: Report of a case and ultrastructural examination. *J Surg Oncol* 36:21-25.
- Feldman, L., Philpotts, L.E., Reinhold, C., Duguid, W.P., and Rosenberg, L. (1997). Pancreatic schwannoma: Report of two cases and review of the literature. *Pancreas* 15:99-105.
- Ferrozzi, F., Bova, D., and Garlaschi, G. (1995). Pancreatic schwannoma: Report of three cases. *Clin Radiol* 50:492-495.
- Gupta, A., Subhas, G., Mittal, V.K., and Jacobs, M.J. (2009). Pancreatic schwannoma: Literature review. *J Surg Educ* 66:168-173.
- Hirabayashi, K., Yasuda, M., Umamura, S., Itoh, H., Itoh, J., Yazawa, N., Imaizumi, T., and Osamura, R.Y. (2008). Cytological features of the cystic fluid of pancreatic schwannoma with cystic degeneration: A case report. *JOP* 9:203-208.
- Larghi, A., Noffsinger, A., Dye, C.E., Hart, J., and Waxman, I. (2005). EUS-guided fine needle tissue acquisition by using high negative pressure suction for the evaluation of solid masses: A pilot study. *Gastrointest Endosc* 62:768-774.
- Lee, J.S., Kim, H.S., Jung, J.J., Han, S.W., and Kim, Y.B. (2001). Ancient schwannoma of the pancreas mimicking a cystic tumor. *Virchows Arch* 439:697-699.
- Møller Pedersen, V., Hede, A., and Graem, N. (1982). A solitary malignant schwannoma mimicking a pancreatic pseudocyst: A case report. *Acta Chir Scand* 148:697-698.
- Morita, S., Okuda, J., Sumiyoshi, K., Taketani, M., Moriguchi, A., Katsu, K., and Tanigawa, N. (1999). Pancreatic schwannoma: Report of a case. *Surg Today* 29:1093-1097.
- Mummadi, R.R., Nealon, W.H., Artifon, E.L.A., Fleming, J.B., and Bhutani, M.S. (2009). Pancreatic schwannoma presenting as a cystic lesion. *Gastrointest Endosc* 69:341.
- Novellas, S., Chevallier, P., Saint Paul, M.C., Gugenheim, J., and Bruneton, J.N. (2005). MRI features of a pancreatic schwannoma. *Clin Imaging* 29:434-436.
- Sugiyama, M., Kimura, W., Kuroda, A., and Muto, T. (1995). Schwannoma arising from peripancreatic nerve plexus. *Am J Roentgenol* 165:232.
- Tafe, L.J. and Suriawinata, A.A. (2008). Cystic pancreatic schwannoma in a 46-year-old man. *Ann Diagn Pathol* 12:296-300.
- Tan, G., Vitellas, K., Morrison, C., and Frankel, W.L. (2003). Cystic schwannoma of the pancreas. *Ann Diagn Pathol* 7:285-291.
- Todd, K.E., Lewis, M.P., Gloor, B., Kusske, A.M., Ashley, S.W., and Reber, H.A. (1997). Management decisions for unusual periampullary tumors. *Am Surg* 63:927-932.
- Tofigh, A.M., Hashemi, M., Honar, B.N., and Solhjoo, F. (2008). Rare presentation of pancreatic schwannoma: A case report. *J Med Case Rep* 2:268.
- Urban, B.A., Fishman, E.K., Hruban, R.H., and Cameron, J.L. (1992). CT findings in cystic schwannoma of the pancreas. *J Comput Assist Tomogr* 16:492-493.
- von Dobschuetz, E., Walch, A., Werner, M., Hopt, U.T., and Adam, U. (2004). Giant ancient schwannoma of pancreatic head treated by extended pancreatoduodenectomy. *Pancreatol* 4:505-508.
- Walsh, M.M. and Brandspigel, K. (1989). Gastrointestinal bleeding due to pancreatic schwannoma complicating von Recklinghausen's disease. *Gastroenterology* 97:1550-1551.
- Wu, Y.L., Yan, H.C., Chen, L.R., Chen, J., Gao, S.L., and Li, J.T. (2005). Pancreatic benign schwannoma treated by simple enucleation: Case report and review of literature. *Pancreas* 31:286-288.
- Yu, G.H., Sack, M.J., Baloch, Z., and Gupta, P.K. (1999). Difficulties in the fine needle aspiration (FNA) diagnosis of schwannoma. *Cytopathology* 10:186-194.
- Yu, R.S. and Sun, J.Z. (2006). Pancreatic schwannoma: CT findings. *Abdom Imaging* 31:103-105.