Case Report

Pancreatic Schwannoma: A Case Report and Review of Liturature

Wiboon Poosawang MD*,
Pravitpong Kiatkungwankai MD*

** Department of Surgery, Samutsakhon Hospital, Samutsakhon, Thailand*

Pancreatic schwannoma is an extremely rare neoplasm, derived from Schwann cells that line the nerve sheaths. It is also referred to as neurilemmoma. The authors report a case of a 46-year-old Thai female who presented with dyspepsia, weight loss and epigastric mass. An examination by ultrasonography and computed tomography (CT) scan revealed a septated cystic tumor in the pancreatic head, 5.8x5.5x5.3 cm in size. Pancreaticoduodenectomy was performed to remove this tumor. A microscopic examination identified proliferating spindle cells that are consistent with neurilemmoma (schwannoma). No complications were found after the operation. At 18-month follow-up, the patient remains asymptomatic and has no signs of recurrence.

Keywords: *Pancreatic schwannoma, Neurilemmoma, Pancreaticoduodenectomy*

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Primary schwannoma of the pancreas is an extremely rare tumor. Moriya et al[1] reviewed PubMed databases and searched for English-language articles published between 1980 and 2010 using a list of keywords, as well as references from review articles. Only 41 articles, including 47 cases, have been reported in the English literature. The mean age was 55.7 years (range 20 to 87 years), with 45% of patients being male. Mean tumor size was 6.2 cm (range 1 to 20 cm). Tumor location was the head (40%), head and body (6%), body (21%), body and tail (15%), tail (4%) and uncinate process (13%). Thirty-four percent of patients exhibited solid tumors and 60% of patients exhibited cystic tumors. Treatment included pancreaticoduodenectomy (32%), distal pancreatectomy (21%), enucleation (15%), unresectable (4%), refused operation (2%) and the detail of resection was not specified in 26% of patients.

After reviewing Thai literature in Thai index medicus, there was no previous report. This may be the first reported case in Thailand. This case of pancreatic schwannoma was presented with dyspepsia, weight loss and epigastric mass, which was successfully treated with pancreaticoduodenectomy (Whipple's operation).

Correspondence to:
Poosawang W, Department of Surgery, Samutsakhon Hospital, Samutsakhon 74000, Thailand.
Phone: 081-490-1820
E-mail: boon8281@hotmail.com

Case Report

A 46-year-old Thai female presented with chronic dyspepsia and 11 kilograms weight loss within three months. Sometimes, she could palpate epigastric mass without history of abdominal trauma, no nausea and vomiting and no alcoholism. Her mother died due to carcinoma of the stomach.

On physical examination, she had mild pale conjunctiva, no jaundice and no febrile. There was an ill-defined mass at the epigastric region, 6x8 cm in size, smooth surface without tenderness.

On laboratory tests, CBC showed Hb 8.59 gm%, Hct 27.2%, WBC 5.57 k/ul and normal LFT. Carcino-embryonic antigen (CEA) was 1.7 ng/mL (normal 0-5.0).

All other tumor markers and endocrine hormonal data were within the normal limit. Her family history was negative for findings of von Recklinghausen's disease.

Ultrasonography revealed cystic mass at pancreatic head. Abdominal CT with IV contrast medium revealed a large thin wall cystic lesion at pancreatic head area measuring 5.8x5.5x5.3 cm. There was thin enhancement of internal septation. The pancreatic body and tail appeared normal. Pancreatic duct was not dilated. These data suggested cystic pancreatic tumor, which mucinous cystadenoma of pancreas was considered (Fig. 1).

On exploratory laparotomy, a large cystic mass at the head of pancreas was discovered.

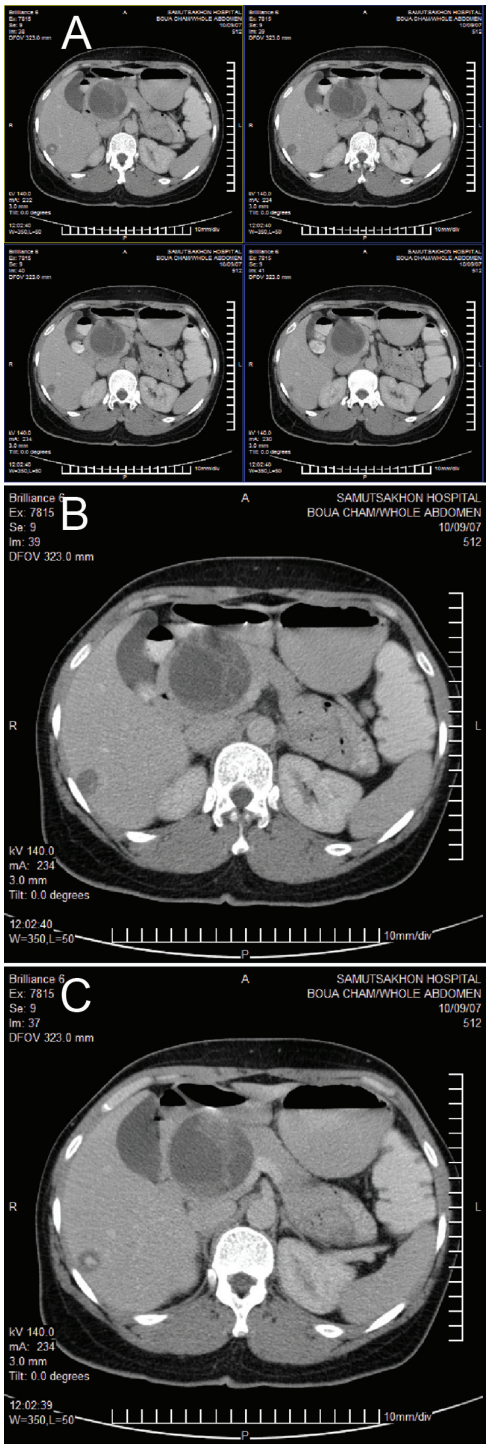

Fig. 1 Abdominal CT with IV contrast medium revealed a large thin wall cystic lesion at pancreatic head area measuring 5.8x5.5x5.3 cm in size (A). There was thin enhancement of internal septation (B, C). The pancreatic body and tail appeared normal. Pancreatic duct was not dilated.

There were no mesenteric lymphadenopathy and normal liver contour. The mass was removed with pancreaticoduodenectomy.

On pathlogical findings, the head of pancreas was measured 8x6x5 cm. The cut surface of the pancreas showed a cystic yellow light brown mass measuring 5.5x5 cm. The mass bulged to the duodenal wall and did not extend to the ampulla area. The remaining pancreatic tissue was lobulated and light brown. On microscopic findings, the cystic mass of the pancreas was well circumscribed and spindle cytoplasm setting in loose to myxoid background. Multiple cystic degenerations were noted. There was no evidence of malignancy. These suggested neurilemmoma or schwannoma with cystic degeneration (Fig. 2).

The patient's postoperative course was uneventful. At 18-month follow-up, the patient had no specific complaints and no sign of recurrence.

Discussion

Schwannomas are benign spindle cell tumors derived from Schwann cells that line the nerve sheaths. Sometimes were also referred to as neurilemmomas. The most common locations in descending order of frequency are the lower extremity, upper extremity, trunk, head and neck, retroperitoneum, mediastinum and pelvis[2-4]. Rarely, schwannomas can involve the abdomen (e.g., the stomach, small bowel, liver, appendix, large bowel, lesser sac and fallopian tube).

Schwann cells can give rise to two types of tumors-schwannomas and neurofibromas. The distinction of these tumors is important because neurofibromas are associated with von Recklinghausen's disease and can undergo malignant change more frequently[4-6].

Pancreatic schwannomas arise from either autonomic sympathetic or parasympathetic fibers, both of which course through the pancreas as branches of the vagus nerve[5,6]. These tumors are even more unusual neoplasms that affect adults with 45% of patients being male. The mean age is 55.7 years (20-87)[1]. They vary considerably in size, from 1.0 to 20.0 cm in maximal diameter (mean 6.2 cm). Approximately two thirds are reported to undergo degenerative changes including cyst formation, calcification, hemorrhage, hyalinization and xanthomatous infiltration[7]. The majority of tumors are located in the head and body of the pancreas, but a few have presented in the tail[1,8,9].

Non-specific abdominal pain was the most commonly reported symptom but weight loss,

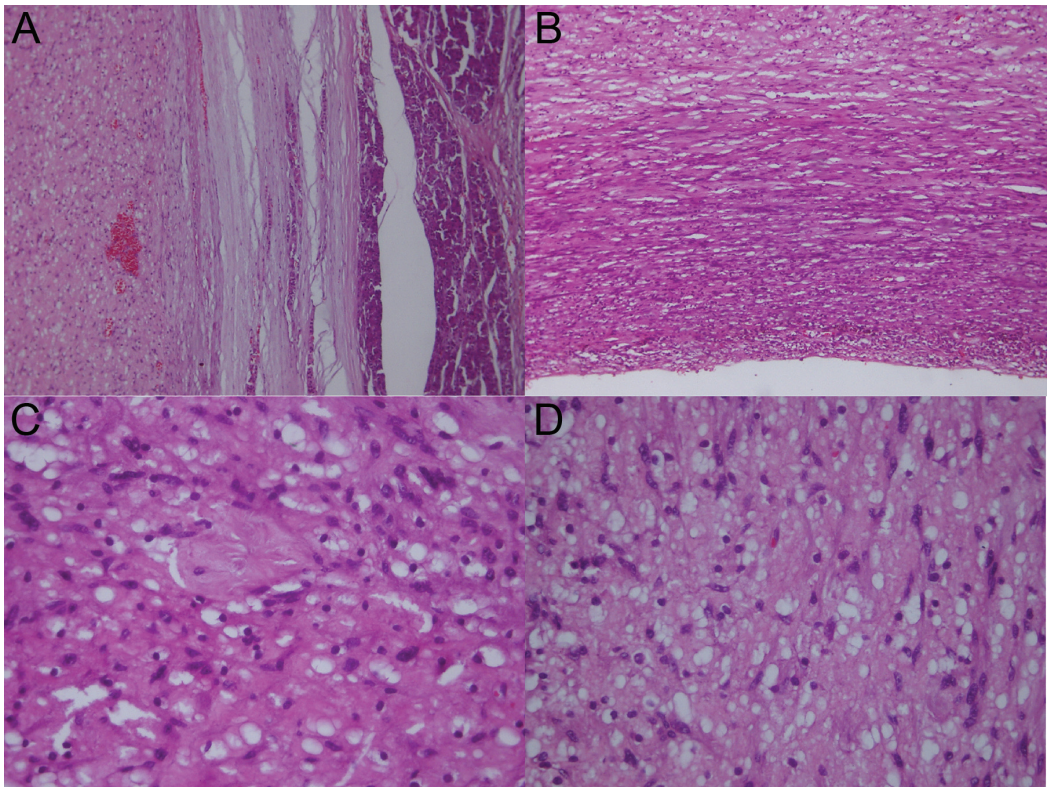

Fig. 2 On microscopic findings, the cystic mass of the pancreas was well circumscribed with well capsulation (A) and spindle cytoplasm setting in loose to myxoid background (B).There was no evidence of malignancy (C, D).

jaundice and gastrointestinal bleeding had also been reported[1,6,10].

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 which typically similar to other cystic pancreatic lesions including neuroendocrine tumors, cystadenoma, cystadenocarcinoma, intraductal papillary mucinous tumor, lymphangiomas and pancreatic pseudocysts. These may lead to missed diagnosis, which is most likely caused by lack of awareness of intrapancreatic schwannoma[11].

The most characteristic feature on CT scan was the presence of an area of low density and/or cystic images reflecting the Antoni B component or degenerative cystic areas of the schwannoma, The contrast-enhanced CT scan 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[12].

MRI is also helpful in characterizing schwannomas by their typical encapsulation, hypointensity on T1-weighted images and hyperintensity on T2-weighted images. It may reveal vascular involvement that can further characterize a lesion with malignant potential[5]. In addition, MRI may differentiate pancreatic schwannoma from adenocarcinoma by characteristic of hyperintensity on T2-weighted images and marked enhancement of the lesion in comparison with the remainder of the gland[5].

Fine needle aspiration (FNA) may establish diagnosis of pancreatic schwannoma pre-operatively but a recent study demonstrated that FNA correctly diagnosed only one out of eight histologically proven schwannomas[13].

Microscopically, schwannomas have two different patterns, highly cellular composing of spindle cells arranged in a palisading fashion without mitotic figure (Antoni A area) and a loose hypocellular component with degenerative changes (Antoni B). Their immunostaining for the S-100 protein is uniformly positive[11,14].

Most pancreatic schwannomas can be treated with simple enucleation depending on their size and location. A review of the treatment showed that the

most common operation was pancreaticoduodenectomy (32%), followed by distal pancreatectomy (21%) and enucleation (15%)[1]. Was it over treatment? The answer is how to establish the correct preoperative diagnosis. An intraoperative frozen section should be performed, as it helps to establish the diagnosis of a benign neoplasm/schwannoma and obviates more radical resection. However large tumor in the head of pancreas or that involving the ampulla, or tumor involving the splenic hilum may require a more radical resection than simple enucleation[4].

In the presented case, pancreaticoduodenectomy was performed due to large size and its location in the head of pancreas.

Conclusion

Pancreatic schwannoma is extremely rare. Preoperative diagnosis is difficult but certain imaging features should raise the suspicion of this diagnosis. The possibility of pancreatic schwannoma should be considered in the differential diagnosis of cystic lesions of the pancreas. Definitive diagnosis is obtained by routine histology. The treatment of choice is surgical excision, simple enucleation or more radical resection such distal pancreatectomy or pancreaticoduodenectomy, which depends on location, size and histopathology.

Potential conflicts of interest

None.

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***Schwannoma* ของตับอ่อน: รายงานผู้ป่วย 1 ราย และทบทวนวรรณกรรม**

วิบูลย์ ภู่สว่าง, ประวิทย์พงษ์ เกียรติกังวาฬไกล

เนื้องอก schwannoma หรือ neurilemmoma ของตับอ่อนพบได้น้อยมาก มีต้นกำเนิดมาจาก Schwann cell ของเยื่อหุ้มเส้นประสาท ผู้นิพนธ์ได้รายงานผู้ป่วยหญิงไทย อายุ 46 ปี มาด้วยอาการท้องอืด น้ำหนักลด และคลำได้ก้อนบริเวณลิ้นปี่ จากการตรวจร่างกาย คลื่นความถี่สูงและคอมพิวเตอร์ช่องท้อง พบมีก้อนถุงน้ำขนาด 5.8x5.5x5.3 ซม. ได้ทำการรักษาโดยการผ่าตัด ผลการตรวจทางพยาธิวิทยา รายงานเป็น neurilemmoma หรือ schwannoma ผลการผ่าตัดและการติดตามผลการรักษาช่วงเวลา 18 เดือน ยังไม่พบอาการผิดปกติใด ๆ