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Case Report

Solid Pseudopapillary Neoplasm of the Pancreas: Report of Two Cases and Review of the Literature

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Abstract

Solid pseudopapillary neoplasm (SPN) of the pancreas is a rare low-grade malignant-potential epithelial tumor that predominantly affects young women aged 20–35 years with a mean age of 22 years. It is currently categorized in the World Health Organization classification under exocrine pancreatic tumor. Here, we present two cases of SPN with initial presentation of large intra-abdominal masses. Both patients underwent successful *en bloc* distal pancreatectomy and splenectomy. Local recurrence and distant metastasis were not detected at the follow up at 21 months and 9 years respectively. In summary, a large, well-encapsulated cystic mass in the pancreas of a young woman should raise suspicion of SPN. [*Tzu Chi Med J* 2009;21(1):81–84]

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1. Introduction

Solid pseudopapillary neoplasm (SPN) is an uncommon pancreatic neoplasm and accounts for only 5% of cystic pancreatic tumors and about 1–2% of exocrine pancreatic cancers [1,2]. SPN is a low-grade malignant-potential epithelial tumor, and predominantly affects young women aged 20–35 years [2]. In contrast to the almost invariable lethal behavior of pancreatic ductal adenocarcinomas, most SPNs, although often large in size, are usually well circumscribed, and complete surgical resection has been reported to cure more than 95% of patients with SPN limited to the pancreas [1]. Frantz first described this tumor in 1959 [3]. To date, more than 700 well-documented cases of pancreatic SPNs have been reported in the English literature [2]. Previously, this tumor was reported using different terms such as: papillary epithelial neoplasm [4], papillary cystic neoplasm [5], solid and papillary epithelial neoplasm [6], solid and cystic acinar tumor [7], papillary and solid neoplasm [8], papillary cystic epithelial neoplasm [9], papillary cystic carcinoma [10], solid and cystic papillary tumor [11], and Frantz's tumor [12], depending on the proportion of their various components. However, these terms do not exactly reflect what is observed either grossly or microscopically. Since being reclassified by World Health Organization in 1996, solid pseudopapillary neoplasm of the pancreas has been recognized as the internationally accepted nomenclature [13].

We present our experience of the surgical management of solid pseudopapillary neoplasms of the pancreas in two women who initial presented with large intra-abdominal masses.

2. Case reports

2.1. Case 1

A previously healthy woman aged 43 years was admitted to our institute because of intermittent vague epigastric pain for 1 month. She had postprandial vomiting and reported recent progressive increases in abdominal girth. On physical examination, one ill-defined, firm, non-tender mass was palpable in the left upper quadrant of her abdomen. Laboratory examinations demonstrated normal levels of pancreatic enzymes and normal liver function test results. The serum tumor markers such as alpha-fetoprotein, carcinoembryonic antigen, carbohydrate antigen 19-9 (CA19-9) and carbohydrate antigen 15-3 (CA15-3) were within reference ranges and her cancer antigen 125 level was 74.5 IU/mL. On a contrast-enhanced abdominal computed tomography (CT) scan, one 11-cm heterogeneous, lobulated retroperitoneal mass was identified near the pancreatic tail (Fig. 1). The tumor displaced the stomach anteriorly and was adherent to splenic flexure of the colon.

The patient underwent elective laparotomy and a large tumor measuring 15×12 cm in size with a smooth grayish white capsule was identified in the pancreatic tail. *En bloc* distal pancreatectomy and splenectomy were performed with a gross negative resection margin. During the operation, there was no evidence of distant metastasis to the liver, regional lymph nodes or the peritoneal cavity. The postoperative recovery went well and the patient was discharged on postoperative day 7. Neither local recurrence nor distant metastasis were detected from abdominal CT scans during a 21-month follow-up period.

Grossly, the grayish white pancreatic tumor was 15×14×10 cm in size. On cross section, the tumor showed focal hemorrhage and central necrosis (Fig. 2). Microscopically, the pancreatic tumor showed marked cellular proliferation in the solid areas that alternated with a pseudopapillary and cystic pattern (Fig. 3). Immunochemical stains disclosed that the neoplastic cells were reactive for CD10, vimentin, neuron-specific enolase (NSE), synaptophysin and progesterone receptor.

2.2. Case 2

A woman aged 43 years presented with postprandial abdominal fullness for 1 week. On physical examination, one ill-defined palpable mass was found in the left upper quadrant of her abdomen. All hematological, biochemical parameters and tumor markers were within references ranges.

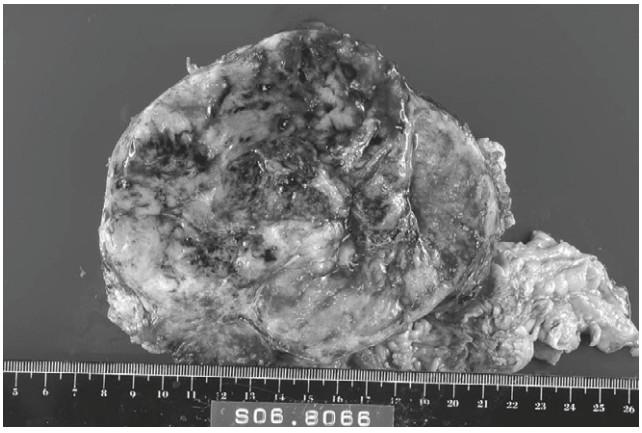

Fig. 2 — The pancreatic tail tumor is well circumscribed and shows focal hemorrhage on cross section.

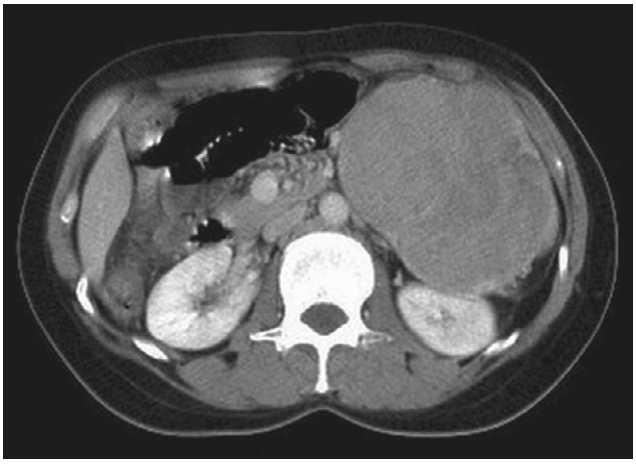

Fig. 1 — Abdominal computed tomography shows one heterogeneous, multiloculated and well-encapsulated huge mass near the pancreatic tail.

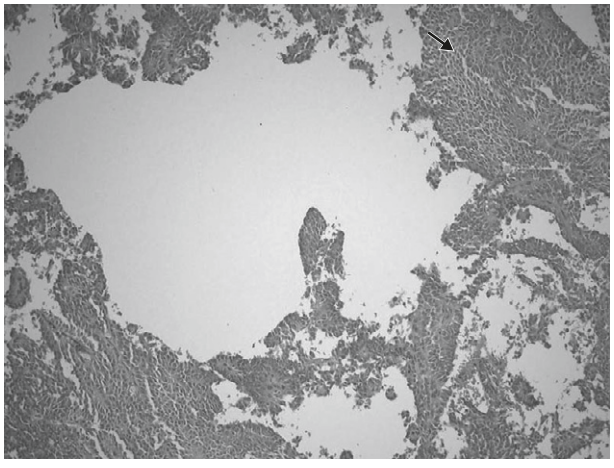

Fig. 3 — The tumor shows proliferation in solid areas that alternates between a pseudopapillary (arrow) and cystic pattern (hematoxylin & eosin, 40×).

Abdominal ultrasonography demonstrated one 8×6 cm multiloculated cystic mass with internal homogeneous echogenicity and septation formation in the left upper quadrant of her abdomen. Contrast-enhanced CT confirmed an 11×9 cm heterogeneous space occupying the mass, which contained cystic and solid components, in the tail of the pancreas. After contrast injection, the solid part of the tumor showed moderate to strong enhancement.

She underwent elective laparotomy and one 9-cm well-circumscribed mass was identified in the tail of the pancreas. *En bloc* resection of the tumor in addition to distal pancreatectomy and splenectomy was performed. No regional lymphadenopathy was found during the operation. The postoperative recovery was unremarkable and the patient was discharged on postoperative day 12. During a 9-year follow-up period, physical examination, abdominal CT and ultrasound did not reveal local recurrence or distant metastasis.

On gross examination, the pancreatic tumor was oval, 9×6×5 cm in size, and was surrounded by a grayish-white smooth fibrous capsule. On cross section, the tumor showed mainly cystic and partially solid components. Microscopically, the neoplastic cells exhibited histological architecture and features typical of a solid pseudopapillary neoplasm.

3. Discussion

The precise incidence of pancreatic SPN in the Chinese population is not known because of its rarity. Lam et al reported eight SPNs during a 24-year period and this accounted for 2.5% of primary pancreatic tumors in Hong Kong Chinese [14]. There is a female preponderance of SPN with a female-to-male ratio of 9.78:1 [2], although rare cases have been reported in children and men [14]. The age at diagnosis ranged from 2 to 85 years with a mean age of 21.9 years [2]. Both of our cases were women and both were aged 43 years old at diagnosis.

The initial presentations of SPN are usually nonspecific. Upper abdominal pain is the most common symptom (46.5%), followed by a slowly enlarged, palpable, non-tender upper abdominal mass (34.8%). Asymptomatic cases are reported in 15.5% of cases [2], and the lesions are detected either after routine examination or after blunt abdominal injury. Because of these subtle symptoms, the tumor size can be quite large (mean diameter of 6.08 cm) at diagnosis with 83% of cases more than 5 cm in diameter [2]. The most common location of the SPN is the tail (35.9%) and the head (34%) of the pancreas [2]. Both of our cases had tumors larger than 5 cm located in the pancreatic tail. Thus a large cystic mass in the pancreas of a young woman should raise suspicion for SPN.

Accurate preoperative diagnosis of SPN is difficult because of the similarity of the findings among cystic lesions of the pancreas. The differential diagnosis includes disoncogenetic cysts, retention cysts, pseudocysts, hydatid cysts and any cystic or solid pancreatic lesions such as serous cystadenoma or cystadenocarcinoma, microcystic adenoma, lymphangioma, hemangioma, mucinous cystic neoplasms, cystic islet cell tumors, and acinar cell cystadenocarcinoma [2,14]. Imaging studies of SPNs that help with differentiation include abdominal ultrasound, endoscopic ultrasound, abdominal CT and magnetic resonance imaging (MRI) and, in some reports, fine needle aspiration biopsy (FNAB) or cytology. Characteristic abdominal ultrasound and CT demonstrate a large, round, well-encapsulated, heterogeneous mass with solid and cystic components and displacement of adjacent viscera. Although some image characteristics are suggestive of SPN, percutaneous fine needle cytology of the cystic wall can be used to obtain a possible preoperative histological diagnosis [15]. However, some authors have suggested that FNAB should be avoided because of the potential risk of tumor spillage, which may compromise surgical cure [16,17]. In our experience, FNAB is not necessary if the tumor is resectable according to the preoperative image evaluation.

SPN exhibits pathological features and is diagnosable based on the classic histological appearance of the neoplasm. Grossly, SPN commonly has a fibrous pseudocapsule surrounding the tumor, and is usually demarcated from the pancreas. There are variable combinations of solid hemorrhagic and cystic necrosis areas [1]. Microscopically, the neoplastic cell is quite uniform with a combination of solid, pseudopapillary or hemorrhagic pseudocystic structures in various proportions. Histological indicators predicting the aggressive behavior of SPNs include capsule thickness of more than 2 mm, high nuclear grade, prominent necrobiotic nests, capsule invasion into the surrounding normal pancreatic tissue and vascular invasion [1].

In contrast to conventional ductal adenocarcinomas, complete surgical resection of SPNs has been reported to provide a more than 95% cure rate [1]. Thus the majority of SPNs are considered to be of low malignant potential with only 10–15% of cases being malignant, showing local infiltration, recurrence or distant metastasis [18]. Complete surgical removal with *en bloc* resection of the involved organ is the treatment of choice. Distal pancreatectomy with or without splenic preservation can be performed for tumors in the body or tail of the pancreas, and a classic or pylorus-preserving pancreaticoduodenectomy for tumors of the pancreatic head [14]. Invasion to the portal vein or superior mesenteric vein or artery should not be included as a criterion for nonresectability of these pancreatic neoplasms [2]. From a surgical standpoint, because of its favorable tumor biology,

an aggressive approach to the occasional neoplasm in the proximal pancreas encasing the superior mesenteric vein and superior mesenteric artery may even be justified. Extensive lymph node dissection is not indicated when the disease is localized (19,20). Long-term survival is often the outcome, even with the presence of metastatic disease on initial presentation (2). Overall, the 2-year survival rate for SPNs (with metastasis or not) is 97% and the 5-year survival rate is about 95% (2). In our study, Case 2 was free of disease during the 9-year follow-up period after complete resection of the primary tumor.

In summary, a diagnosis of SPN should be considered in young women presenting with large and well-circumscribed pancreatic cystic masses. Despite the large size of SPNs and their ability for local invasion, their relatively low malignancy means that aggressive treatment with complete surgical resection of both the primary and metastatic lesions offers high cure rates.

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