CASE SERIES

Clinicopathologic characteristics and surgical treatment of solid pseudopapillary tumor of the pancreas

Huang Y[1], Feng JF[2]

[1]Nursing Department, Zhejiang Cancer Hospital, Hangzhou, China
[2]Department of Oncological Surgery, Zhejiang Cancer Hospital, Hangzhou, China

Abstract

Background and aim: Solid pseudopapillary tumor (SPT) of the pancreas is a very rare neoplasm of low malignant potential that mostly affects young women. The aim of the present study is to report our experience in surgical treatment of SPT and review of the literature.

Material and methods: A retrospective review of three cases of SPT who were treated at our department during the last two years was performed. The clinicopathologic characteristics, surgical treatment, and prognosis are described in detail.

Results: Case 1 described an asymptomatic SPT in a pregnant woman. To the best of our knowledge, only one case of SPT in pregnancy has been reported in the literature. Case 2 described an SPT in the pancreatic tail causing splenic infarction, and a distal pancreatectomy combined with splenectomy was performed. Case 3 described an SPT in the pancreatic head, for which a pancreatoduodenectomy was successfully performed. All of the three patients were followed up for 10-22 months without recurrence or metastases after the initial surgery at the time of reporting.

Conclusions: At present, radical resection is the treatment of choice for SPT. Enucleation can be performed for tumors with complete amicula. Distal pancreatectomy combined with or without splenectomy can be performed for pancreatic body and/or tail tumor, and pancreatoduodenectomy for pancreatic head tumor. The prognosis of SPT is good.

Key words: solid pseudopapillary tumor, pancreatic neoplasm, treatment

Corresponding author: Ji-Feng Feng, MD, Department of Oncological Surgery, Zhejiang Cancer Hospital, No.38 Guangji Road, Hangzhou 310022, China, tel: +86-0571-88122038, e-mail: Fengjif@163.com

Introduction

Solid pseudopapillary tumor (SPT) of the pancreas, first reported by Frantz et al[1] in 1959, is an uncommon but distinct pancreatic neoplasm, accounting for 1%-2% of all pancreatic tumors[2-4]. The tumor has been given several different names according to its macroscopic and microscopic character until this name, solid pseudopapillary tumor of the pancreas, was defined by the World Health Organization (WHO) as unique tumor in 1996[5]. In this paper, we report our experience in surgical treatment of SPT and review the literature.

Case 1

A 26 year-old female in the 14th week of pregnancy was admitted to our department with abdominal mass accidently detected by ultrasonography (US) in prenatal care. US revealed a well-circumscribed inhomogenous mass (9.5 × 6.2 × 9.0 cm) with intact amicula (Figure 1A). Magnetic resonance imaging (MRI) was performated to verify a giant solid-cystic mass with T1- and T2- weighted images in the right of pancreatic head (Figure 1B-C). FIne needle aspiration biopsy (FNAB) showed that tumor cells were composed of papillary structures, with lots of neoplastic epithelial cells, polygonal in form. At laparotomy, successful tumor enucleation was performed. Microscopically, the pancreatic tumor showed marked cellular proliferation in the solid areas that alternated with a pseudopapillary and cystic pattern (Figure 1D). Immunohistological results revealed that the tumor cells were positive for Vimentin (Vim), Cytokeratin (CK), Synaptophysin (Syn), Neuron specific enolase (NSE), CD56, and CD10. On the 3rd postoperative day, pancreatic fistual occurred. US detected a local opaque dark area of fluid measuring about 52mm × 32mm in the upper middle abdomen (Figure 1E). Fetal position was good (Figure 1F). On the 25th postoperative day, the abdominal drainage tube was removed because the liquid less than 5ml. After 28 days of hospital stay, she was discharged in good general condition. On the 38th week, a healthy, mature girl with an Apgar score 9/10 was born with cesarean section.

Case 2

A 18-year-old female was admitted to our department with abdominal pain for four days. US revealed a well-circumscribed inhomogenous mass measuring about 4.5 × 3.9 cm between the spleen and left kidney (Fig-

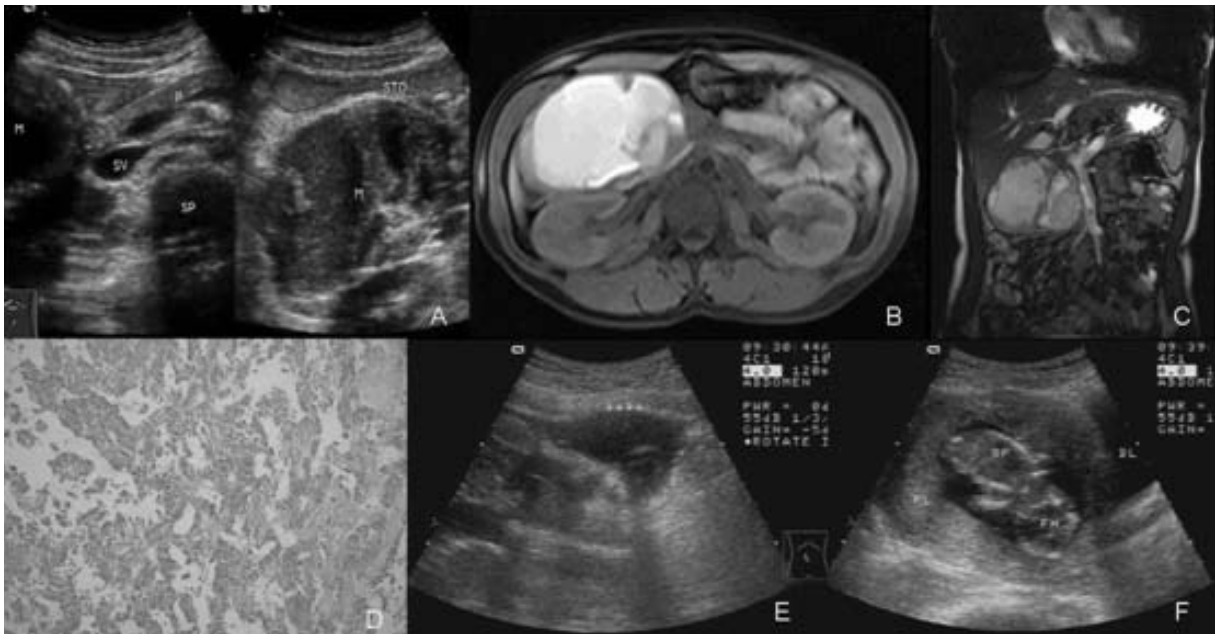

Figure 1: US revealed a well-circumscribed inhomogenous mass with intact amicula in the right of pancreatic head (A). MRI was performated to verify a giant solid-cystic mass with T1- and T2- weighted images in the right of pancreatic head (B-C). The pancreatic tumor showed marked cellular proliferation in the solid areas that alternated with a pseudopapillary and cystic pattern (D). US detected a local opaque dark area of fluid in the upper middle abdomen (E). Fetal position was good (F).

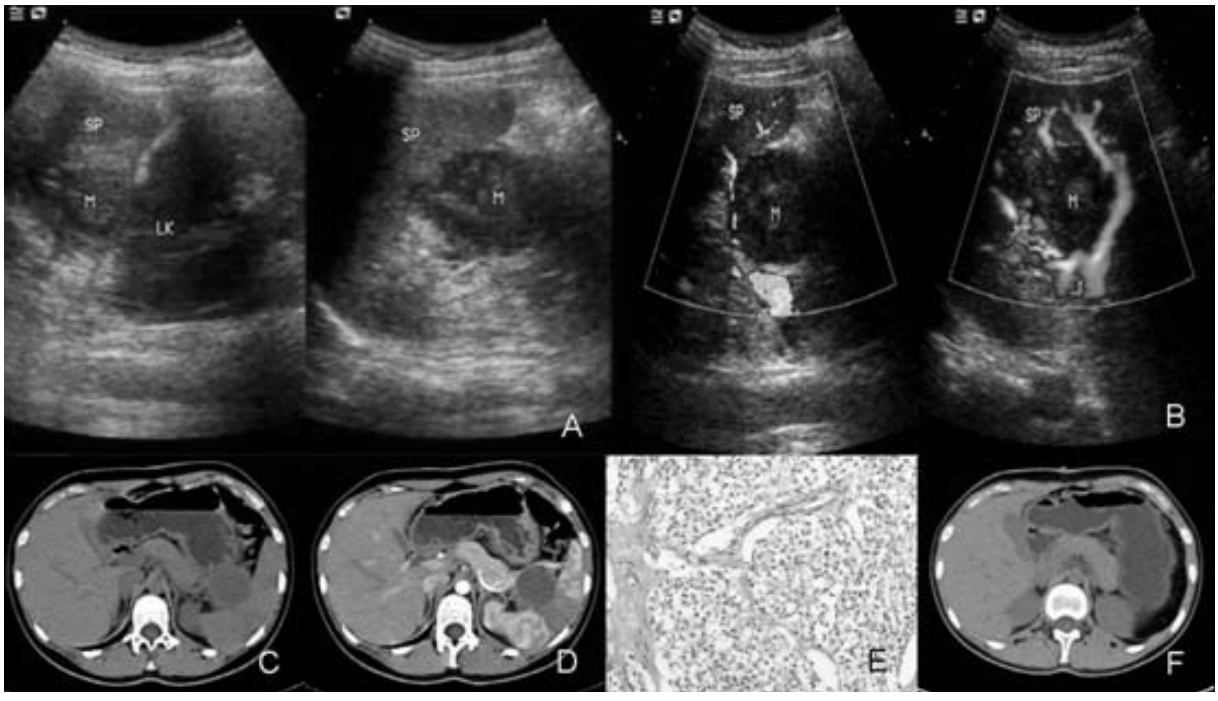

Figure 2: US revealed a well-circumscribed inhomogenous mass between the spleen and left kidney (A-B). CT demonstrated a predominantly cystic, well-encapsulated mass in the pancreatic tail causing splenic infarction (C). After contrast injection, the solid part of the tumor showed moderate to strong enhancement (D). The tumor showed proliferation in solid areas that alternates between a pseudopapillary and cystic pattern (E). The patient was followed up for 22 months without recurrence or metastases (F).

ure 2A-B). Computed tomography (CT) demonstrated a predominantly cystic, well-encapsulated mass with a CT value of 26 Hounsfield Units (HU), measuring about 4.2 ×3.8 cm, in the pancreatic tail, causing splenic infarction (Figure 2C). After contrast injection, the solid part of the tumor showed moderate to strong enhancement (Figure 2D). At laparotomy, a distal pancreatectomy combined with splenectomy was successfully performed. Microscopically, the tumor showed proliferation in solid areas that alternates between a pseudopapillary and cystic

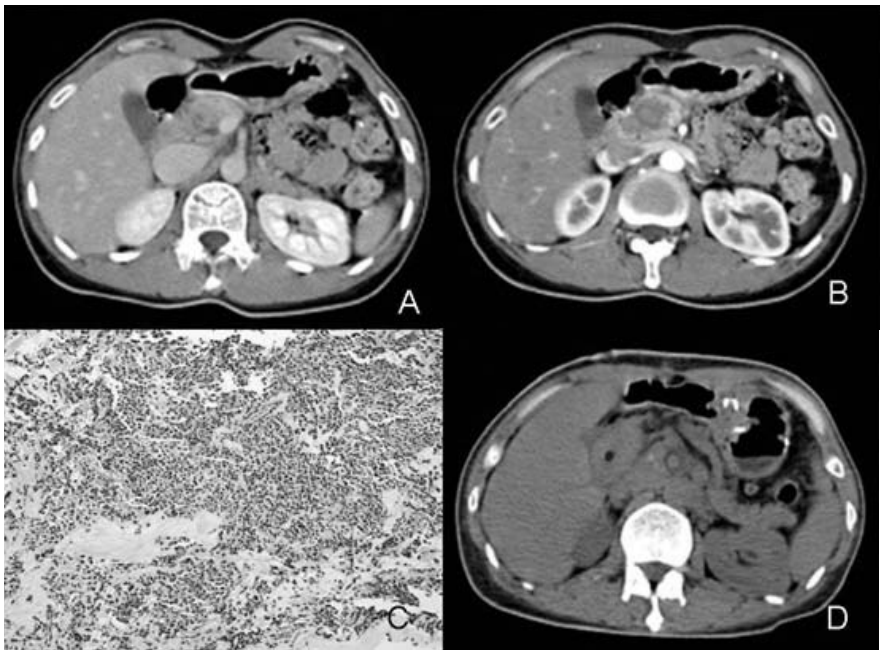

Figure 3: CT revealed a low-density mass in the pancreatic head (A-B). Solid areas consist of sheets and cords of round to ovoid cytologically bland cells arranged around a delicate fibrovascular septa (C). The patient was followed up for 10 months without recurrence or metastases (D).

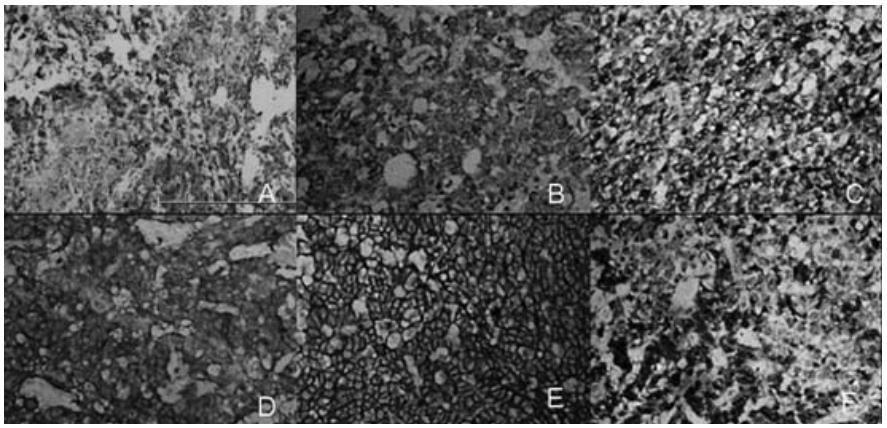

Figure 4: Immunohistological results revealed the tumor cells were positive for Vim (A), CK (B), Syn (C), NSE (D), CD56 (E), and CD10 (F) (200×).

pattern (Figure 2E). The immunohistological results revealed that the tumor cells were positive for Vim, Syn, CD56, and CD10. She was followed up for 22 months without recurrence or metastases after the initial surgery at the time of reporting (Figure 2F).

Case 3

A 38-year-old female was admitted to our department with abdominal pain for two months. Abdominal CT revealed a low-density mass with a CT value of 26 HU, measuring about 1.9×3.2 cm, in the pancreatic head (Figure 3A-B). At exploration, the tumor was located in the pancreatic head. A pancreatoduodenectomy was successfully performed. The histologic appearance varied in different regions of the tumor. Solid areas consisted of sheets and cords of round to ovoid cytologically bland cells arranged around a delicate fibrovascular septa (Figure 3C). Immunohistological results revealed that the tumor cells were positive for Vim, Syn, NSE, CD56, and CD10. She was followed up for 10 months without recurrence or metastases (Figure 3D).

Disscussion

The cellular origin of SPT is unclear and might involve ductal cells, acinar cells, endocrine cells or multipotential stem cells.[3] The pathogenesis of SPT has not been revealed yet, however, the disease is generally considered to have a benign course with low malignant potential, usually affecting young women in their second or third decade of life[3,4]. There is a female preponderance of SPT with a female-to-male ratio of 9.78:1, although rare cases have been reported in men[2-4,6]. Tien et al[7] showed that there were no gender-specific trends in expression of sex hormone receptor protein or clinicopathologic characteristics.

SPT was classified according to the WHO criteria as either an SPT with an uncertain potential for malignancy or as a solid pseudopapillary carcinoma (SPC)[8].Criteria that could distinguish potentially malignant tumors, classified as SPC, included the following: 1) perineural invasion, 2) angioinvasion, 3) deep invasion into the surrounding tissue, and 4) distant metastases. Postoperatively, patients were further classified using the American Joint Commit-

tee on Cancer (AJCC)/International Union Against Cancer (UICC) tumor node metastasis (TNM) classification system: R0 (no residual tumor), R1 (microscopic residual tumor), or R2 (macroscopic residual tumor) [9].

The malignant potential of SPT is reported to be 10%-15%.[4] The most common sites of metastases are the liver, regional lymph nodes, mesentery, omentum, and peritoneum[10]. Local invasion may involve adjacent organs, including the duodenum, spleen, portal vein, superior mesenteric vein, and bile duct; lymph node metastasis also has been reported [11,12].Washington[13] showed that the clinicopathologic characteristics of SPT, including diffuse growth, venous invasion, nuclear pleomorphism, mitotic rate, necrosis and dedifferentiation, are related to its aggressive behavior or metastatic potential. Yang et al[14] showed the high proliferative index assessed by immunohistochemical staining for Ki-67 may predict poor outcome of malignant SPT.

The initial presentations of SPT are usually nonspecific. It is a non-functional, slow-growing neoplasm that very often reaches considerable size before the first symptoms appear[4,6,15,16]. SPT often demonstrates peripheral artery enhancement and central calcification. Papavramidis et al[4] summarized 718 SPT patients in the literature, showing upper abdominal pain is the most common symptom (46.5%), followed by a slowly enlarged, palpable, and non-tender abdominal mass (34.8%). Asymptomatic cases are reported in 15.5%. Differential diagnosis includes disoncogenetic cysts, pseudocysts, hydatid cysts, and cystic tumors, such as cystadenoma, cystadenocarcinoma, microcystic adenoma, lymphangioma, various forms of sarcomas, cystic islet cell tumors, and acinar cell cystadenocarcinomas.

Accurate preoperative diagnosis of SPT is difficult because of the similarity of the findings among cystic lesions of the pancreas[4,6]. As part of the general investigation, US shows a well-circumscribed inhomogenous mass in the epigastrium. Following US, CT usually shows heterogeneous enhancement with progressive central filling and late enhancement of the capsule[4,17]. If MRI reveals an encapsulated mass with solid and cystic components as well as hemorrhage without obvious internal septum, SPT should be highly suspected[18]. Although some image characteristics are suggestive of SPT, FNAB can be used to obtain a possible preoperative histological diagnosis[19]. However, some researchers have suggested that FNAB should be avoided because of the potential risk of tumor spillage[20,21]. In our case series, FNAB was performed in one case for the patient with pregnancy. We concluded that the FNAB was safe in SPT patient with pregnancy.

Microscopically, the growth pattern of the tumor cells is remarkably uniform, with a combination of solid, pseudopapillary, or hemorrhagic pseudocystic structures in various proportions[22]. The tumor contains a mixture of solid, cystic, and pseudopapillary patterns in various proportions. In immunohistological results, the tumor cells are diffusely positive for Vim in all tumors, most cases express diffuse positive staining for NSE, some of which are focally positive for CK and SYN, and few positive for S-100 protein[3,4,22]. Notohara et al[23] found that SPT exhibited unique immunohistochemical features with expression of CD56, CD10, and these results are diagnostically useful. In our case series, the immunohistological results were shown in Figure 4. Based on these histological findings, the final diagnosis of SPT was confirmed. Recently, immunoreactivity for ß-catenin is found in the cytoplasm and the nuclei of almost all tumor cells in the majority of SPT[4,24,25]. Loss of membrane staining and/or nuclear staining for E-cadherin is seen in 100% of cases of SPT of the pancreas[26].

At present, radical resection is the treatment of choice for SPT even with metastasis or local extension[2-4,16,27]. Local resection or enucleation can be performed for small tumors with complete amicula. Distal pancreatectomy combined with or without splenectomy can be performed for pancreatic body and/or tail tumor, and pancreatoduodenectomy for pancreatic head tumor. Complete surgical excision is curative in greater than 95% of patients with SPT limited to the pancreas[28]. The low grade of malignancy of this tumor, and because the mass is usually surrounded by a dense fibrous capsule, led some surgeons, especially for children, to perform simple enucleation of the neoplasm[6,29,30]. Invasion to the portal vein or superior mesenteric artery should not be included as a criterion for nonresectability of these pancreatic neoplasms[4,28]. For the metastases, there is also general consensus that surgical debulking should be performed[2,4,28,30]. In general, SPT can be removed laparoscopically because they are generally benign and have thick fibrous capsules. However, the decision to perform laparoscopic surgery should be made carefully to avoid the risk of rupture[31,32]. The role of chemotherapy and radiotherapy in treatment of SPT is poorly defined at present, since only few reports are available on them[33,34].

The prognosis of SPT patients even with local recurrence and metastasis or invasion is good. It has been reported that the overall 5-year survival rate of SPT patients is about 95%[4]. Due to the favorable prognosis and long survival rate of SPT patients with local recurrence or metastasis, it is difficult to identify the predictive factors for their survival time. Recurrence, local invasion, and limited metastasis are not the contraindications for resection, and some patients with unresectable SPT may also have a long survival time[4,16,27,35]. In our case series, all of the three patients were followed up for 10-22 months, without recurrence or metastases after the initial surgery at the time of reporting. Kim et al[36] reported that most SPT patients who developed recurrence had metastases at the first operation, tumor rupture, or adjacent organ invasion. Because recurrence was rare, however, statistically meaningful risk factors associated with recurrence could not be determined.

In conclusion, radical resection is the treatment of choice for SPT. Enucleation can be performed for tumors with complete amicula in pregnancy. Distal pancreatectomy combined with or without splenectomy can be performed for pancreatic body and/or tail tumor, and pancreatoduodenectomy for pancreatic head tumor. The prognosis of SPT is good.

Conflict of interest

The authors declare no conflicts of interest.

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