Original Article

Surgical Outcomes of Solid Pseudopapillary Neoplasm of the Pancreas: A Single Institution's Experience of 16 Cases

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Abstract

Introduction: Solid-pseudopapillary tumor (SPT) of the pancreas is a rare tumor, generally characterized by a well-encapsulated mass. The aim of the current study was to analyse the clinicopathological characteristics and treatment outcomes of patients with SPT. In this study, we report our clinical experience with 16 cases of SPTs.

Methods: Sixteen patients who underwent surgery for pathologically confirmed SPT were included. Data of the patients were reviewed from the prospectively recorded database. Patients' demographics, laboratory values, clinical presentation, radiological imaging findings, surgical treatment, perioperative complications, pathological features, post-operative course, and long-term survival were collected and analyzed. Statistical analyses were performed using the computer program Statistical Package for Social Sciences (SPSS) 16.0 for Windows.

Results: The tumors ranged from 2 to 11 cm in diameter and were located in the head in ten patients (62.5%), the neck in two patients (12.5%), and the body or tail in four patients (25%). All patients were women whose ages ranged from 21 to 79 years (mean age was 41.62 ± 15.08). Patients had resection margins free of tumor resections and there were no preoperative or postoperative mortalities. There was no recurrence or metastasis after the surgical resection. All patients were alive at a mean follow-up of 49.06 ± 29.53 months (range 6 to 99).

Conclusion: SPT is a rare pancreatic neoplasm with a low malignant potential, and is common in young women. If SPT is diagnosed before surgery, complete surgical resection, generally enucleation is the most effective therapy for SPT.

Keywords: Pancreas, single institution's experience, solid-pseudopapillary tumor, surgery, treatment

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Introduction

Solid-pseudopapillary tumor (SPT) of the pancreas is a rare tumor and characterized by a well- encapsulated mass. SPT was first described by Dr. Frantz in 1959,[1] and was defined by the World Health Organization (WHO) in 1996 as "solid pseudopapillary tumours" of the pancreas. SPT is an uncommon neoplasm of the pancreas accounting for 1% – 2% of all pancreatic tumors.[2] It occurs predominantly in young females, with a male to female ration of 1: 9.5.[3] Most SPTs are limited to the pancreas, and complete surgical intervention results in long term survival.[1,4] A 5-year survival rate is higher than 95%.[5] Although SPTs are considered as tumors of low malignant potential, patients with SPT occasionally present with liver metastasis or invasion into adjacent organs. In this study, we report our clinical experience with 16 cases of SPTs. The aim of the current study was to analyse the clinicopathological characteristics and treatment outcomes of patients with SPT.

Methods

Between July 2006 and August 2014, data of sixteen patients who underwent surgery for pathologically confirmed SPT, were reviewed from the prospectively recorded database. Patients' demographics, laboratory values, clinical presentation, radiological imaging findings, surgical treatment, perioperative complications, pathological features, post-operative course, and long-term survival were collected and analyzed. We obtained information on follow-up from outpatient notes and outpatient records combined with telephone interviews. All statistical analyses were performed using the computer program Statistical Package for Social Sciences (SPSS) 16.0 for Windows.

Results

Patient characteristics

All patients were women whose ages ranged from 21 to 79 years (mean age was 41.62 ± 15.08). The most common clinical presentation was abdominal pain (86.6%). Other symptoms included back pain (26.6%), nausea and emesis (20%). Four patients had exprienced abdominal pain together with back pain and two patients had exprienced abdominal pain associated with nausea and emesis. Two tumors were found incidentally. The tumors ranged from 2 to 11 cm in diameter (mean diameter of the tumor was 5.64 ± 3.16 cm) and were located in the head in ten patients (62.5%), the neck in two patients (12.5%), and the body or tail in four patients (25%). The clinical features of the patients are summarized in Table 1.

Preoperative examination and diagnosis

Preoperative tumor markers (α-fetoprotein, carbohydrate antigen 19-9 and carcinoembryonic antigen) and other laboratory

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Table 1. Clinicopathological Data of the Resected Patients with Pancreas Solid Pseudopapillary Neoplasm

Characteristics (n = 16)	Total (%)
Age (mean ± SD)	41,62 ± 15,08
Gender	
Male	0
Female	16 (100%)
Symptom	
Abdominal pain	13 (86.6%)
Back pain	4 (26.6%)
Nausea and emesis	3 (20%)
Tumor Size (cm) (mean ± SD)	5.64 ± 3.16 cm
Location	
Head of pancreas	10 (62.5%)
Distal pancreas	4 (25%)
Neck	2 (12.5%)
Operation	
Enucleation	3 (18.75%)
Whipple	4 (25%)
Whipple & liver metastasectomy	3 (18.75%)
Distal pancreatectomy & splenectomy	4 (25%)
Central pancreatectomy	1 (6.25%)
Total pancreatectomy & splenectomy & SMV excision	1 (6.25%)
Tumor nature	
Solid	6 (37.5%)
Mix	10 (62.5%)

values were in normal limits. Radiologic investigations included abdominal ultrasonography (n = 13), computed tomography (CT, n = 16), magnetic resonance imaging (MRI, n = 6) and endosonography (EUS). CT scans showed a well- circumscribed, heterogeneous (cystic or solid) mass with lack of central enhancement in most cases. The most common location was the head of the pancreas (62.5%).

Surgical data

Of the 16 patients treated with surgery, seven patients underwent pancreaticoduodenectomy. Partial liver resection was performed for suspected metastasis in three of these seven patients. Types of operations are listed in Table 2.

Type of surgery was selected according to the site and the appearance of the tumor. All of the patients had R0 resections and there were no perioperative mortalities. Total surgery time ranged from 120 to 660 minutes (mean 343.75 ± 138.79 minutes), (Table 2).

Histopathological features

The majority of tumors were in the head of the pancreas (10/16, 62.5%) and the mean diameter was 5.64 cm (range 2 – 11). All tumors were encapsulated (16/16, 100%). Resection margins were free of tumor (R0) in all patients. Capsular invasion was found in one patient. Mitotic activity was low or undetectable in all patients. None of the patients had evidence of perineural, vascular invasion and lymph node involvement. In the one patient who had capsular invasion, superior mesenteric vein invasion with desmoplastic reaction was observed additionally. This patient underwent en-bloc resection including total pancreatectomy and splenectomy. Three patients had suspected liver metastases according to preoperative evaluation. However, postoperative histopathologic evaluation revealed focal nodular hyperplasia in these patients.

Follow-up

Postsurgical complications occurred in three patients. One patient had wound infection four days after surgery. Two patients had pancreatic leakage and were treated with percutaneous drainage catheter placement. The mean postsurgical hospital stay was 15.87 ± 11.18 days (range 6 to 45 days). Most patients were followed up initially at 3- to 6-month intervals and later annually. There was no recurrence or metastasis after surgical resection. All patients were alive at a mean follow-up of 49.06 ± 29.53 months (range 6 to 99). All of the patients were alive at the end of the study. Pelvic osteosarcoma developed in one patient and treatment is still ongoing.

Discussion

SPT is a rare pancreatic neoplasm with a low malignant potential. SPT is common in young women, especially aged between 20 to 40 years.[2,5,6,7] and is rarely seen in older males and females,, with a 5-year survival rate of 95% – 97% after a complete resection.[8,9] In this study, all of our patients were female; the youngest patient was 21 years old, and the oldest patient was a 79-year old woman. There was a considerable number of patients with no symptoms in whom SPT was found accidentally.[10] Patients most commonly present with nonspecific symptoms such as discomfort or abdominal pain. Due to nonspecific symptoms and silent onset leading to a delay in diagnosis, SPTs are generally large tu-

Table 2. Operative Procedures in 16 Patients with Pancreas Solid Pseudopapillary Neoplasm

No	Age (year)	Localization	Tumor diameter (mm)	Preop FNA	Type of surgery	Surgery time (min)	Hospital stay (day)	Survival (month)
1	79	Body&Tail	90	FNA (-)	Distal pancreatectomy &splenectomy	180	8	99
2	41	Head	55	FNA (+) SPT (+)	Whipple	420	14	99
3	35	Body&Tail	73	FNA (-)	Distal pancreatectomy & splenectomy	360	6	83
4	35	Head	30	FNA (+) SPT (+)	Enucleation	120	7	70
5	68	Body&Tail	110	FNA (-)	Enucleation	120	45	65
6	35	Head	103	FNA (+) SPT (+)	Whipple & partial liver resection	660	11	63
7	42	Head	85	FNA (-)	Total pancreatectomy & splenectomy & parsiyel SMV excision	420	20	61
8	47	Head	26	FNA (-)	Whipple & partial liver resection	420	19	43
9	34	Head	26	FNA(+) SPT (+)	Whipple	420	14	42
10	47	Head	20	FNA (+) SPT (+)	Whipple & partial liver resection	450	40	40
11	22	Body&Tail	72	FNA (-)	Laparoscopic distal pancreatectomy&splenectomy	300	8	39
12	35	Neck	24	FNA (+) SPT (+)	Distal pancreatectomy & splenectomy	210	14	27
13	45	Head	30	FNA (+) SPT (+)	Whipple	420	14	20
14	30	Neck	31	FNA (+) SPT (+)	Central pancreatectomy & pankreaticogastrostomy	330	14	14
15	21	Head	40	FNA (+) SPT (+)	Whipple	360	8	14
16	50	Head	88	FNA (+) SPT (+)	Enucleation	310	12	6

FNA: Fine Needle Aspiration; SPT: Solid Pseudopapillary Tumor; FNA (+) means: FNA was done; SPT (+) means Solid Pseudopapillary Tumor was detected with FNA; Survival time: The time to begining to end of the study. (All of the patients were alive at the end of the study and there was no reccurrens).

mors at the time of presentation.[11,12] In this study, the mean diameter of the tumour was 5.43 ± 3.16 cm (range 2 – 11 cm).

Tumor markers were not significant clinical factors to predict SPTs.[11] Usually, the serum tumor markers, such as alfa-fetoprotein, carcinoembryonic antigen, and CA 19.9 are within normal ranges.[13] In our study, preoperative laboratory values and serum tumor markers were within normal ranges.

Abdominal US, CT, MRI, and EUS are imaging studies that are used to detect SPTs. Abdominal examination with Doppler ultrasonography (USG) can be very helpful by measuring low blood flow around the tumour.[14] However, abdomen USG and Doppler USG are not specific in the diagnosis of SPT. Computed tomography is the dominant imaging modality, accounting for almost 50% of the imaging procedures. Abdomen CT and MRI scans usually show a well-encapsulated complex mass with both solid and cystic components, sometimes with calcification. Dynamic contrast-enhanced abdomen CT can demonstrate less enhanced tumor, typical cystic spaces in the center, and enhanced solid areas at its surroundings.[15] The characteristic features of SPN on CT or MRI are a well-defined and capsule formation of the tumor, low-attenuation mass with peripheral enhancement and complex cystic components with areas of necrosis and internal hemorrhage.[16]

When compared with MRI, CT has inherent limitations in showing certain tissue characteristics, such as hemorrhage, cystic degeneration or the integrity of the tumour capsule that would be suggestive of SPN. Due to its superior contrast resolution, MRI scans display a capsule and intratumoural hemorrhage, better than CT scans. In Yu's study, MRI was much better than CT in detecting the cyst or solid components of the tumor.[17] If MRI shows an encapsulated mass with solid and cystic components as well as hemorrhage without apparent internal septum, SPT of the pancreas should be extremely suspected.[18] PET may not add additional information for diagnosis.[19]

On Endosonography, it is often an echo-poor mass occasionally with mixed echogenicity. Endosonography can provide fine needle aspiration biopsy (FNA) with the probability of preoperative pathologic diagnosis, particularly to allow differentiation of SPT from other pancreatic neoplasms with similar radiographic appearance.[20] In our study, we performed FNA for ten patients. Histopatologically, SPT was diagnosed in all of these patients. FNA was not performed in the other six patients due to the site of the lesion and suspected diagnosis of adenocarcinoma (three patients). Preoperative imaging studies and FNA- biopsy revealed the diagnosis of SPT before the surgery in 13/16 patients.

Papavramidis, et al.[5] reported the locations of SPT as the tail (35.9%), the head (34%), the body (14.8%) and the neck of the pancreas (1.01%), respectively. Yu, et al.[14] found that most of the tumours were distributed in the pancreatic head (39.8%), with the tail (24.1%), body and tail (19.5%), body (11.2%), and neck (3.6%). Yin, et al.[21] reported that the location of the SPT in the pancreas were 35.4% in the head, 4.8% in the neck, 18.3% in the body, and 20.7% in the tail. In our study, the most common site of the tumour were pancreatic head in 10 patients (62.5%), neck in two patients (12.5%) and body or tail in four patients (25%).

Pancreatic SPT was reported to be mostly solitary[22] and approximately 15% – 20% of the cases showed malignant behaviour in the previously reported literature.[23] Definitive histopathologic criteria for malignancy is not well established. However, size >

5 cm; high proliferation rate; pancreatic parenchymal infiltration; invasion of vascular, lymphatic, perineural and surrounding structures; nuclear atypia and distant metastases may predict aggressive behaviour.[23] Patients with incomplete resection and atypical histopatology and older male patients may have a higher risk of recurrence and mortality.[24] In our study, tumor size was greater than 5 cm in eight patients and pancreatic parenchymal and capsule infiltration was found in one patient as a predictive factor for aggressive behaviour, however there was no recurrence or metastasis after surgical resection. All our patients were alive at a mean follow-up of 49.06 ± 29.53 months (range 6 to 99).

Although enucleation is an adequate treatment in the majority of patients with SPT, patients in our study required Whipple operation rather than enucleation. The selection of the surgical strategy should be made according to intraoperative pathological frozen section, tumor position, and invasion of surrounding structures.[25] Although most tumors are large, a minimized resection like enucleation can achieve a favorable curative effect; due to nonaggressive behavior of the tumor, presence of a dense capsule, and excellent prognosis.[25] However, due to the location of the tumor and involvement of the surrounding structures, distal pancreatectomy or pancreaticoduodenectomy can be necessary.[26] Intra-operative frozen section study may help to identify the resection margins.[27] In our study, intraoperative frozen section study was valuable for determining the negative margins and the extent of resection in all of the operations except total pancreatectomy and enucleations.

Extensive lymphadenectomy is not required as nodal metastases are very rare.[28] In our study, lymph node or distant metastases were found in none of the patients. In a previous study, it was reported that 17 patients underwent vascular resection and reconstruction without any mortality.[29] In our study, the infiltrated superior mesenteric vein was reconstructed with end-to end anastomosis after en-bloc resection and the patient had a good survival. The histopathological examination of the resected specimen revealed that inferior mesenteric vein infiltration was due to benign desmoplastic reaction of the tumor.

Timely resection for SPT provides a long-term survival. In a previous study, the overall recurrence rate was observed in 10% – 15% of patients after complete resection.[30] In our series, no recurrence or metastasis was found during a mean follow-up of 49.06 ± 29.53 months (range 6 to 99).

In conclusion, pancreatic mass with female sex, normal tumor markers and big tumor size, a well-defined capsule formation of the tumor, complex cystic components with areas of necrosis, calcification and internal hemorrhage on CT or MRI should be considered as a preliminary diagnosis of SPT. FNA should be performed preoperatively to avoid an extensive surgery and lymphadenectomy. If SPT is diagnosed before surgery, complete surgical resection (R0), generally enucleation is the most effective therapy for SPT. Most importantly, when the SPT is suspected, timely resection provides long-term survival.

Conflicts of interest

We declare that we have no conflict of interest and patient consent obtained.

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