Primary mesenchymal tumors of the pancreas in a single center over 15 years

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Abstract. In total, 95% of primary tumors in the pancreas are epithelial tumors; primary mesenchymal tumors at this site are extremely rare. At present, only one comprehensive study about these rare tumors has been performed. Another retrospective analysis of these rare tumors is performed in the present study, which, to the best of our knowledge, is the first to be performed in China. In the present study, 10 patients that underwent resection for primary mesenchymal tumors of the pancreas were identified in a 15-year period at the Chinese Academy of Medical Sciences and Peking Union Medical College, which accounted for 0.51% of the total surgically resected primary tumors of the pancreas at this hospital. Among the 10 patients, 7 patients (70%) were diagnosed with benign/borderline tumors, and the remaining 3 patients (30%) were diagnosed with malignant tumors. It was a unique finding of the present study that the preoperative diagnosis was frequently a misdiagnosis, in terms of the specific pathological diagnosis. Therefore, although primary mesenchymal tumors of the pancreas are extremely rare, they should be considered in order to make the correct preoperative diagnosis. Contrarily to a previous study, in the present study, the most common benign tumor was not desmoid tumor, but solitary fibrous tumors; the most frequent primary sarcoma was not undifferentiated/unclassified sarcoma either. In conclusion, the present study aids the understanding of these rare tumors; however, primary mesenchymal tumors of the pancreas require additional exploration in the future.

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Introduction

The predominant primary tumor that occurs in the pancreas is ductal adenocarcinoma (1). Stroma is scant in the normal pancreas, and therefore primary mesenchymal tumors of the pancreas are extremely rare (2).

Although numerous primary mesenchymal tumors of the pancreas have been reported, the majority of cases were reported as case reports (3-22). To the best of our knowledge, there has been only one comprehensive study of these tumors in a single institution (3). A lack of comprehensive studies from other large institutes to provide a comprehensive understanding of this type of tumor remains. In the present study, a thorough retrospective analysis of primary mesenchymal tumors of the pancreas in Peking Union Medical College Hospital (Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing, China) was performed and the relevant studies published in the English literature were reviewed.

According to the published studies, at present there have been 221 cases of primary mesenchymal tumors of the pancreas reported in the English literature (3). Among these, the most commonly reported primary benign/borderline mesenchymal tumors of the pancreas were schwannomas, followed by inflammatory myofibroblastic tumors, solid and cystic hamartomas, and solitary fibrous tumors. In addition, the most frequently reported primary malignant mesenchymal tumors of the pancreas were leiomyosarcomas, followed by Ewing sarcomas/primitive neuroectodermal tumors (PNETs), and undifferentiated/unclassified sarcomas.

However, the present study showed certain differences from the previously reported studies, with the most frequent benign/borderline cases being solitary fibrous tumors, and the malignant cases were three different types, with each occurring in only 1 case.

Materials and methods

Patient selection. The present study focused on primary mesenchymal tumors of the pancreas. Any mesenchymal tumors extending into the pancreas from surrounding organs, such as the tumors originating from the stomach, duodenum and bile duct, were excluded from the present analysis. The tumors were considered primary if the tumor was solitary or

the main body of the tumor was in the center of the pancreas, even if the tumor also involved the peripancreatic soft tissue.

Subsequent to approval by the Institutional Review Board of Peking Union Medical College Hospital (Beijing, China) and obtention of informed consent from the patients, all 1,944 pancreatic tumors surgically resected between January 2000 and December 2014 at the Department of Pathology of the Peking Union Medical College Hospital were reviewed. Among these tumors, upon excluding 1,927 cases that were epithelial tumors and lymphomas, there were only 10 cases that were ascertained to be primary pancreatic mesenchymal tumors. In addition, 1 case of metastatic primitive neuroectodermal tumor, 1 case of metastatic sclerosing epithelioid fibrosarcoma, 1 case of malignant gastrointestinal stromal tumor (GIST), 1 case of leiomyosarcoma, 1 case of malignant fibrohistiocytic tumor, 1 case of adenosarcoma and 1 case of schwannoma, which originated from the gastric, duodenum or retroperitoneum, were excluded.

The clinicopathological variables, which included the age and gender of the patients, symptoms at presentation, pre-operative diagnosis, type of surgical interventions, tumor location, tumor size and final pathological diagnosis, were retrieved from clinical records and the pathological files. The follow-up information, including the recurrence and survival time, was obtained by telephone interview or clinical records.

Pathological review. All available hematoxylin and eosin and immunohistochemical staining slides were reviewed by 3 independent pathologists and a diagnosis for each case was reached by consensus. The immunohistochemical markers were stained using an automated immunohistochemistry/ *in situ* hybridization staining instrument (Ventana Medical Systems, Inc.; Roche Diagnostics, Indianapolis, IN, USA) according to the manufacturer's protocol.

Statistical analysis. Statistical analyses were performed using SPSS (version 17; SPSS, Inc., Chicago, IL, USA). The means and standard deviations (SDs) of the clinicopathological variables were compared using the unpaired Student's t-test or Mann-Whitney U test. Cumulative survival rates were calculated by Kaplan-Meier analysis. P<0.05 was considered to indicate a statistically significant difference.

Results

Clinicopathological variables. The clinicopathological variables of the 10 primary mesenchymal tumors of the pancreas are summarized in Table I. The patients consisted of 5 males and 5 females, with a male-to-female ratio of 1:1. The mean age was 52.4 years (SD, 10.6; range, 37-70 years). In total, 4 tumors were benign (40%), 3 tumors were borderline (30%) and 3 tumors were malignant (30%). The 7 benign/borderline tumors included 2 cases of solitary fibrous tumors, 1 case of fibromatosis, 1 case of ganglioneuroma, 1 case of myofibroblastoma accompanied by serous cystadenoma, 1 case of schwannoma and 1 case of uncertain malignant potential gastrointestinal stromal tumor. The 3 malignant cases consisted of 1 case of malignant gastrointestinal stromal tumor, 1 case of malignant solitary fibrous tumor and 1 case of undifferentiated pleomorphic sarcoma. One-half

of these patients (5/10; 50%) were asymptomatic and their tumors were identified incidentally at check-ups. The other 5 patients presented with various symptoms, such as mild or acute abdominal pain, nausea, vomiting, weight loss or jaundice. Among these patients, 1 patient had a history of surgical resection of benign tumors; patient 9 had previously undergone 2 procedures for meningioma. There were 4 patients with hypertension and 1 patient had diabetes. All primary lesions were classified as centrally located in the pancreatic parenchyma; however, they may infiltrate the peripancreatic soft tissue, retroperitoneum or gastroduodenal wall. There were 3 tumors (30%) that arose in the head of the pancreas and the remaining 7 tumors (70%) were located in the body or tail of the pancreas. In terms of morphology, 1 tumor (10%) was cystic, 2 tumors (20%) had a cystic-solid-like appearance and 7 tumors (70%) were solid. The mean tumor size was 5.18 cm (SD, 2.4 cm; range, 3-10 cm). In total, 9 patients were followed up and 1 patient was lost to follow-up. The follow-up period ranged between 7 and 113 months. Almost all the preoperative diagnoses were misdiagnoses in terms of the specific terminology of these rare tumors (Table I). However, in terms of the identification of benign/borderline or malignant tumors, 5 patients (50%) were correctly diagnosed preoperatively, and only 1 of the 3 patients with malignant tumors was misdiagnosed preoperatively. Overall, 7 patients received intraoperative frozen section diagnosis, and only patient 9 was misdiagnosed with a neuroendocrine tumor. The correct diagnosis rate was 86%.

Comparison between benign/borderline and malignant primary mesenchymal tumors of the pancreas. Comparisons were performed between the benign, borderline and malignant primary mesenchymal tumors of the pancreas. All 4 benign tumors were located in the body/tail of the pancreas, whereas 66.7% of the borderline tumors were located in the body/tail of the pancreas and 33% of the malignant tumors were at this location (P=0.19). In total, 75% of the benign tumors were identified incidentally; however, only 2 borderline and malignant tumors (33%) were found incidentally (P=0.51). In total, 75% of patients with benign tumor were female, while 67% patients who had the borderline and malignant tumor was male (P=0.42). There was also no significant difference in patients' age, tumor size, or tumor appearance between these benign, borderline and malignant mesenchymal tumors.

Comparison between the survival status of patients. In the present study, only 2 patients succumbed to the disease, and there was 1 patient lost to follow-up. The survival status was compared with patient age, survival time, tumor size, gender, tumor location, symptoms, macroscopy of the tumor and classification as benign/borderline or malignant by unpaired Student's t-test or Mann-Whitney U test, which demonstrated that patients who succumbed were older than patients who survived, with the tumor location more likely to be at the head of the pancreas (Table II; P≤0.05). There were no significant differences between the tumor size, symptoms, macroscopy of the tumors and patient gender (P>0.05). The Kaplan-Meier analysis showed that the benign/borderline tumors had a more favorable prognosis than malignant tumors (P=0.02; Fig. 1).

Table I. Clinicopathological characteristics of patients with primary mesenchymal tumor of the pancreas.

Patient	Gender	Age, years	Location	Size, cm	Pre-D	Prognosis	Treatment	Symptoms	Immunohistochemistry	Pathological diagnosis	Macroscopy
1	Female	37	Body/tail	4.5x4.0x3.0	DCq	A	DPS	Acute abdominal pain	Vimentin(+), S-100(±), CD34(-),CD117(-), SMA(-), Ki-67 index <1%	Fibromatosis	Solid
2	Male	43	Body/tail	4.0x3.0x3.0	Castleman	A	Excision	Abdominal discomfort	CD34 (focally+), CD117(-), desmin(-), S-100(-), SMA(-)	Solitary fibrous tumor	Solid
3	Female	46	Body/tail	2.6x2.0x1.7	MCN	A	DP	Incidentally detected	CD117(-), CD34(-), DOG-1(-), desmin (focally+), Ki-67 index <1%, S-100 (focally+), SMA(+), caldesmon(-)	Myofibroblstoma, SCN	Cystic
4	Male	46	Body/tail	4.0x3.0x2.5	CP	A	DPS	Mild abdominal pain	AE1/AE3(-), CD34(+), Bcl-2(+), CD99(+), CD117(-), SMA(+), CD31(-), CD21(-), CD35(-), CD68 (focally+), caldesmon(-), DOG-1(-), desmin(-), S-100(-), ALK(-), Ki-67 index <1%	Solitary fibrous tumor	Solid
5	Male	46	Head	10.0x3.0-8.0 x6.0	SCN	A	DPS/CTx	Incidentally detected	SMA(-), desmin(-), CD117(+), CD34(++), S100(+)	Malignant GIST	Cystic-solid
6	Male	52	Body/tail	4.0x3.5x3.0	NET	A	Excision	Incidentally detected	S-100(+), SMA(-), CD34(-), vimentin(+)	Schwannoma	Cystic-solid
7	Female	60	Head	3.6x3.6x3.0	NET	A	Whipple	Mild abdominal Pain	CD34(+), vimentin(+), CD117(-), CgA(-), SMA(-), S-100(-), desmin(-)	GIST	Solid
8	Female	61	Body/tail	6.6x4.0x2.5	DC	N	DP	Incidentally detected	NSE(+), S-100(+)	Ganglioeuroma	Solid
9	Male	63	Body/tail	3.0x3.0x3.0	NET	D	DPS	Incidentally detected	SMA(+), NSE(+), CD99(+), CgA(-), EMA(-), F-VIII(-), CD34(+),vimentin(+), Syn(-), AE1/AE3(-), gastrin(-), VIP(-), somatostatin(-), Ki-67 index >30%	Malignant solitary firous tumor	Solid

Table I. Continued.

Patient	Gender	Age, years	Location	Size, cm	Pre-D	Prognosis	Treatment	Symptoms	Immunohistochemistry	Pathological diagnosis	Macroscopy
10	Female	70	Head	6.0x4.0x3.0	Invasive IPMN	D	Whipple	Anorexia	AE1/AE3(-), CAM5.2(-), CEA(-), CK19(-), CK7(-), CK8(-), CgA(-), Syn(-),vimentin(+), p53(+), CD56 (NK-1)(±)	Polymorphic sarcoma	Solid

A, alive; CP, chronic pancreatis; CTx, chemotherapy; D, death; DC, ductal adenocarcinoma; DP, distal pancreatectomy; DPS, distal pancreatectomy and splenectomy; GIST, gastrointestinal stromal tumor; IPMN, intraductal papillary mucinous neoplasm; LN, lymph node; MCN, mucinous cystic neoplasm; N, not available/not performed; NET, neuroendocrine tumor; PPPD, pylorus-preserving pancreaticoduodenectomy; pre-D, preoperative diagnosis; SCN, serous cystic neoplasm; SPN, solid pseudopapillary neoplasm; CD, cluster of differentiation; SMA, smmoth muscle actin; DOG-1, discovered on GIST-1; Bcl, B-cell lymphoma; ALK, anaplastic lymphoma kinase; CgA, chromogranin A; NSE, euron-specific enolase; EMA, epithelial membrane antigen; F, factor; Syn, synuclein; VIP, vasoactive intestinal polypeptide; CEA, carcinoembryonic antigen; CK, cytokeratin; NK-1, neurokinin 1.
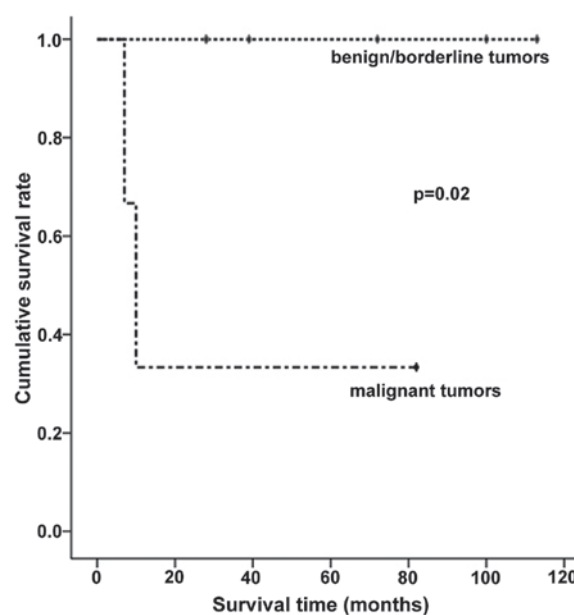

Figure 1. Kaplan-Meier survival analysis of patients with primary benign/borderline and malignant pancreatic mesenchymal tumors.

Histological and immunohistochemical characteristics. The diagnostic criteria for the soft tissue or other organs (23) were used to diagnose the present mesenchymal tumors of the pancreas. Therefore, the histological features of these tumors in the pancreas were consistent with those in the soft tissue or other organs. Representative images of these tumors are shown in Fig. 2. The antibodies used were as follows: Anti-vimentin (monoclonal mouse anti-human; 1:200; V9; Dako, Glostrup, Denmark); anti-S-100 (polyclonal rabbit anti-human; prediluted; ZSGB-Bio, Beijing, China); anti-cluster of differentiation (CD)34 (monoclonal mouse anti-human; 1:50; QBEnd 10; Dako); anti-CD117 (monoclonal rabbit anti-human; prediluted; EP10; ZSGB-Bio); anti-smooth muscle actin (SMA) (monoclonal mouse anti-human; 1:100; 1A4; Dako); anti-desmin (monoclonal mouse anti-human; 1:100; D33; Dako); anti-CD99 (monoclonal mouse anti-human; 1:100; 12E7; Dako); anti-discovered on GIST-1 (monoclonal mouse anti-human; 1:100; DOG1.1; Zeta Corporation, Sierra Madre, CA, USA); anti-Ki-67 (monoclonal mouse anti-human; 1,1:100; MIB; Dako); anti-caldesmon (monoclonal rabbit anti-human; prediluted; EP19; ZSGB-Bio); anti-chromogranin A (monoclonal rabbit anti-human; prediluted; EP38; ZSGB-Bio); anti-neuron-specific enolase (monoclonal mouse anti-human; prediluted; 5E2; ZSGB-Bio); anti-cytokeratin (CK) (monoclonal mouse anti-human; 1:200; AE1/AE3; Dako); anti-B-cell lymphoma-2 (monoclonal mouse anti-human; 1:50; 124; Dako); anti-CD31 (monoclonal mouse anti-human; 1:50; JC70A; Dako); anti-CD21 (monoclonal mouse anti-human; 1:50; 1F8; Dako); anti-CD35 (monoclonal mouse anti-human; 1:20; Ber-MAC-DRC; Dako); anti-CD68 (monoclonal mouse anti-human; 1:200; PG-M1; Dako); anti-anaplastic lymphoma kinase (monoclonal rabbit anti-human; prediluted; D5F3; Ventana Medical Systems, Inc.; Roche Diagnostics); anti-epithelial membrane antigen (monoclonal mouse anti-human; 1:100; E29; Dako); anti-factor-VIII (monoclonal mouse anti-human; prediluted; OTI9F3; ZSGB-Bio); anti-synuclein (monoclonal rabbit anti-human; prediluted; EP158; ZSGB-Bio);

Table II. Comparisons between the survial status of patients with primary mesenchymal tumor of the pancreas.

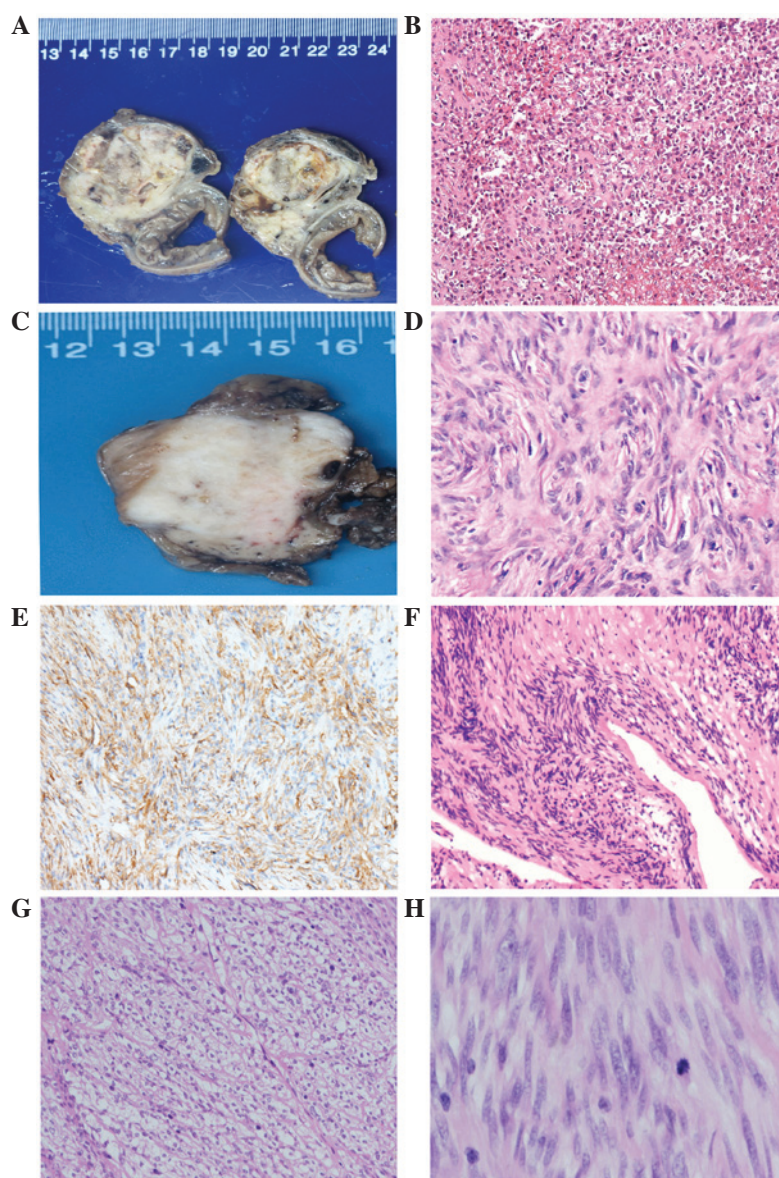| | Survival status | | |
Variable	Alive	Dead	P-value
Mean age ± SD, years	47.1±1.2	69.5±4.9	0.010
Mean survival time ± SD, months	66.0±34.8	8.5±2.1	0.005
Mean tumor size ± SD, cm	5.2±2.7	4.5±2.1	0.750
Male, %	50.0	57.0	0.860
Tumor located in the head, %	14.3	100.0	0.030
Incidental symptoms, %	42.9	50.0	0.860
Cyst, %	14.3	0.0	0.410
Benign/borderline, %	85.7	0.0	0.030

SD, standard deviation.

Figure 2. Representative images of benign/borderline and malignant mesenchymal tumors of the pancreas. (A and B) Undifferentiated pleomorphic sarcoma (case 10). (A) Macroscopy of the tumor showed a solid mass. (B) Microscopically, the tumor was composed of giant cells with evident cytological atypia. (C-E) Solitary fibrous tumor (case 4). (C) Macroscopy of the tumor showed a tan-white solid mass. (D) The tumor was composed of spindle cells with a combination of hypocellular and hypercellular areas. (E) Cluster of differentiation 117 was diffusely stained in tumor cells diffusely. (F) Schwannoma (case 6). (G and H) Malignant gastrointestinal stromal tumor (case 5). (G) Epithelioid tumor cells of the tumor. (H) Brisk mitosis in the center of the tumor.

anti-gastrin (polyclonal rabbit anti-human; prediluted; ZSGB-Bio); anti-vasoactive intestinal polypeptide (monoclonal mouse anti-human; 1:100; H-6; Santa Cruz Biotechnology, Inc., Dallas, TX, USA); anti-somatostatin (monoclonal rabbit anti-human; prediluted; EP130; ZSGB-Bio); anti-CK CAM5.2 (monoclonal mouse anti-human; prediluted; CAM5.2 ZSGB-Bio); anti-carcinoembryonic antigen (monoclonal mouse anti-human; 1:150; II-7; Dako); anti-CK19 (monoclonal mouse anti-human; 1:100; RCK108; Dako); anti-CK7 (monoclonal rabbit anti-human; prediluted; EP16; ZSGB-Bio); anti-CK8 (monoclonal mouse anti-human; 1:100; C8/144B; Dako); anti-p53 (monoclonal mouse anti-human; prediluted; MX008; Fuzhou Maixin Biotech Co., Ltd., Fuzhou, China); and anti-CD56 (neurokinin-1) (monoclonal mouse anti-human; 1:100; 1B6; Novocastra; Leica Microsystems, Inc., Buffalo Grove, IL, USA).

The malignant undifferentiated pleomorphic sarcoma demonstrated no specific pattern and differentiation, consisting of numerous bizarre giant tumor cells and a large area of necrosis (Fig. 2A and B). The immunophenotype was diffusely positive for vimentin, whereas the epithelial markers were all negative.

The most common type of tumor in the present study was solitary fibrous tumors, which accounted for 3 cases, including 1 malignant tumor. The solitary fibrous tumors showed a combination of hypocellular and hypercellular areas, which were separated from each other by thick bands of hyalinized collagen and branching hemangiopericytoma-like vessels (Fig. 2C and D). The malignant tumor was hypercellular, showed marked focal cytological atypia and necrosis, with obvious mitosis. The Ki-67 index was >30%. All solitary fibrous tumors diffusely expressed CD34 and/or CD117 (Fig. 2E).

The least rare tumor type was schwannoma, which showed the conventional appearance of spindle cells forming the compact area that alternated with loosely arranged foci of Antoni B areas, and one area exhibited cyst formation (Fig. 2F). The tumor classified as fibromatosis had abundant collagen matrix and hypocelluarity, with the fibroblastic cells being plump, Vimentin-positive and CD34-negative, with a Ki-67 index <1%. The ganglioneuroma consisted of large ganglion cells and spindled Schwann cells, with S100 being positive for these cells. The inflammatory myofibroblastic tumor was composed of loosely arranged plump or spindled myofibroblasts in an edematous background, with an infiltrate of lymphocytes, eosinophils and plasma cells. Immunohistochemistry showed that desmin and SMA were focally positive.

There were 2 cases of extragastrointestinal stromal tumor in the present study, with 1 being malignant and both being spindle cell tumors. The tumor cells had moderate cellularity, and certain areas had nuclear palisading. The malignant tumor was 10 cm in its maximum diameter, with 5 mitosis per 50 high power fields and had epithelioid area and necrotic areas (Fig. 2G and H). CD34 and/or CD117 were expressed in the tumors of case 5 and case 7 (Table I).

Treatments and prognosis. All 10 patients underwent radical resection or enucleation, consisting of the 2 patients with benign tumors undergoing mass excision, 2 Whipple procedures, 2 of distal pancreatectomies and 4 distal/segmental pancreatectomies plus splenectomies. There was no significant difference

between these different operational methods when compared with the survival status or World Health Organization classification groups.

Postoperative chemotherapy was only administered to the patient diagnosed with malignant GIST, who was treated using Gleevc (400mg, qd). The other two patients (cases 9 and 10) with malignant tumors did not receive chemotherapy due to intolerance.

The prognosis of patients with benign/borderline mesenchymal tumors was good, with no patients developing recurrence or metastasis. However, the prognosis of the patients with malignant tumors was poor, as case 9 developed multiple metastases within 6 months and succumbed to the disease 10 months after surgery, while case 10 developed metastases within 4 months and succumbed to the disease 7 months after surgery.

Discussion

Primary mesenchymal tumors of the pancreas are extremely rare, and at present only 221 cases have been reported in the English literature (3).

In the only other comprehensive single-institution study of primary mesenchymal tumors of the pancreas, performed by Kim *et al* (3), only 0.3% (20/7,129) of patients were confirmed to have pancreatic primary mesenchymal tumors (3). The rate of the occurrence in the present study was 0.5%, which is higher than the incidence reported by Kim *et al* (3). This difference may be due to biopsy cases not being included in the present study, since a number of these patients may not receive surgery or may not be subjected to surgery at Peking Union Medical College Hospital. With the exception of the two comprehensive studies of primary mesenchymal tumors of the pancreas, including the present current study, previously reported cases of mesenchymal tumors of the pancreas have been published as individual case reports, so a definite general occurrence rate could not be calculated.

According to the aforementioned studies, totaling >200 cases, the most common benign tumors were schwannomas (51 cases) (3-6), followed by inflammatory myofibroblastic tumors (34 cases) (3,7), solid and cystic hamartomas (28 cases) (3,8,9), fibromatosis (16 cases) (3,10,11), solitary fibrous tumors (15 cases) (3,12), cavernous hemangiomas (12 cases) (3,13) and 3 cases of angiomyolipoma (3,14,15). The most commonly reported primary sarcomas of the pancreas were leiomyosarcoma (41 cases) (3,16), followed by Ewing sarcomas/PNET (16 cases) (3), undifferentiated/unclassified sarcomas (15 cases) (3,17), and atypical lipomatous tumor/well-differentiated liposarcoma (2 cases) (3).

The preoperative diagnosis of these rare mesenchymal tumors may be challenging since mesenchymal tumors of the pancreas rarely have the typical findings of ductal adenocarcinomas (3). In the present study, only 50% of cases were diagnosed correctly as malignant or benign/borderline tumors prior to surgery. In addition, almost all the patients were wrongly diagnosed as a definite tumor type. However, the correct rate may be increased to 86% when intraoperative frozen section diagnosis is used. These results indicate that additional attention should be paid to these rare mesenchymal tumors in the clinic in order to make the correct diagnosis.

According to these previously reported studies, the most common benign tumor was schwannoma, and the most frequently reported primary sarcoma was leiomyosarcoma (3-17). The two comprehensive studies of primary mesenchymal tumors of the pancreas performed by Kim *et al* (3) and the present study identified certain different results. In the study by Kim *et al*, the most common benign tumor was desmoid tumor and the most frequently primary sarcoma was undifferentiated/unclassified sarcoma (3). In the present study, the most common benign/borderline tumor was solitary fibrous tumor and the most common sarcoma was neither leiomyosarcoma nor undifferentiated/unclassified sarcoma, as the 3 malignant tumors in the present study were of different types. However, among these malignant sarcomas, 1 was undifferentiated/unclassified sarcoma.

Incorporating the only two comprehensive studies performed in single institutions, the study by Kim *et al* (3) and the present study, it was found that desmoid tumor was the most common benign tumor type, accounting for 4 patients in the study by Kim *et al* (3) and 1 patient in the present study, followed by solitary fibrous tumor, accounting for 2 patients in the study by Kim *et al* (3) and 2 patients in the present study. The most common sarcoma was undifferentiated/unclassified sarcoma, accounting for 3 patients in the study by Kim *et al* (3) and 1 patient in the present study. The unique cases in the present study were 1 patient with ganglioneuroma and 2 patients with pancreatic extragastrointestinal stromal tumors. Ganglioneuroma is extremely rare in the pancreas, with only 2 other cases previously reported (18,19). However, by contrast, pancreatic GIST were numerous, as 32 cases have been reported (20-22). There were no diagnoses of leiomyosarcoma, Ewing sarcoma, liposarcoma, cavernous hemangioma, hamartoma or angiomyolipoma in the present study, in agreement with previous studies (3,8,9,13-16). Therefore, the true prevalence of the primary mesenchymal tumor types in the pancreas requires additional comprehensive studies to be elucidated.

The characteristics of the benign, borderline and malignant primary mesenchymal tumors of the pancreas were alike to the ductal adenocarcinoma. The present results showed that although there was no significant difference among the tumor types in terms of the age and gender of the patients, tumor location, tumor size and macroscopic appearance of the tumors, there was a notable tendency in them, with all 4 benign tumors being located in the body/tail of the pancreas, whereas 66.7% of the borderline tumors were located in this area and 33% of the malignant tumors were at this location. Almost 70% of malignant tumors arose in the head of the pancreas. In addition, 75% of benign tumors were identified incidentally, and 33% of borderline and malignant tumors were identified incidentally. In total, 75% of patients with benign tumors were female, while 67% of patients with borderline and malignant tumors were male. All these characteristics were similar to the characteristics of patients with pancreatic ductal adenocarcinomas to a certain extent, since the incidence of ductal adenocarcinoma is ~50% higher in men than in women and indolent tumors tends to be non-symptomatic (1).

The prognosis of the patients with malignant sarcoma were poor, and all 4 patients with undifferentiated sarcoma in the 2 comprehensive studies all succumbed; however, the patients with benign/borderline tumors had a good prognosis, with only 1 patient succumbing in the study by Kim *et al* (3) and no patients succumbing in the present study. It was also found that a poor prognosis was associated with an older age and the location of the tumor in the head of the pancreas. There was no significant association with the tumor size and the patient gender.

In summary, primary mesenchymal tumors of the pancreas are extremely rare; careful and comprehensive evaluation is required to make a correct pre-operative diagnosis. In addition, due to the rarity of comprehensive analyses of cases, additional studies of this type are required to improve the understanding of these rare neoplasms.

Acknowledgements

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