Pancreatic head sparing surgery for solid pseudopapillary tumor in patients with agenesis of the dorsal pancreas

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Abstract

Background: This study aimed to clarify the feasibility and justification of pancreatic head sparing (PHS) enucleation for patients with agenesis of the dorsal pancreas (ADP) associated with a solid pseudopapillary tumor (SPT).

Methods: Data of the SPT patients with and without ADP, including clinical presentations, surgical options, and surgical and survival outcomes, were recruited for comparison.

Results: A total of 31 patients with SPTs were included, three of whom displayed ADP and underwent PHS enucleation. Surgical complications were comparable between the groups. Overall, the 5- and 10-year disease-free survival rates were 100% and 90%, respectively. The 20- and 25-year overall survival rates were 100% and 66.7%, respectively. Only one patient (3.2%) developed tumor recurrence 7.3 years after pancreatectomy for an SPT with lymph node involvement, and the patient survived 24.5 years after the initial operation. No tumor recurrence occurred in any patient with ADP after PHS enucleation.

Conclusion: PHS enucleation seems to be feasible and justifiable for SPT patients with ADP in terms of surgical and survival outcomes, and this approach could be recommended to avoid pancreatic insufficiency.

Keywords: Agenesis of dorsal pancreas; Enucleation; Pancreatic head sparing; Solid pseudopapillary tumor

1. INTRODUCTION

Agenesis of the dorsal pancreas (ADP) is an exceedingly rare congenital anomaly, with around 100 cases having been reported in the literature since the first case was described in 1911.[1] This condition is characterized by the partial or total loss of the body and tail of the pancreas. Most patients with this anomaly are asymptomatic but may develop diabetes mellitus (DM), pancreatitis, and other pancreatic diseases such as pancreatic tumors.[1–6] ADP is compatible with life and requires no special treatment; however, it is sometimes necessary to treat any accompanying diseases.[2]

Such a disease includes the solid pseudopapillary tumor (SPT) of the pancreas, which is a rare low-grade malignant neoplasm. This tumor was first described by Frantz in 1959 and was then further specified as a unique clinicopathological entity by Hamoudi in 1970.[7] Therefore, it was known as Franz's tumor or the Hamoudi tumor until the World Health Organization accepted and named it the SPT in 1996.[8] SPTs account for approximately 0.2% to 2.7% of primary nonendocrine tumors of the pancreas[8,9] and occur mostly in females (90%), with a female-to-male ratio of 20:1. Furthermore, 85% of these patients are under 30 years of age, with a median age of 26 and an average age of 28 years.[8–12] The standard treatment for these tumors is complete surgical resection to achieve a negative margin, usually accomplished using pancreaticoduodenectomy, distal pancreatectomy, or central pancreatectomy.[8,9,13,14] However, these options present a surgical dilemma when a pancreatic SPT is associated with ADP.

This report aimed to share our experiences of the long-term survival outcomes of ADP patients with SPT treated using pancreatic head sparing (PHS) surgery at our institute. The main purpose of this study was to clarify the feasibility and justification of PHS enucleation. We, therefore, compared the surgical outcomes of SPT patients with ADP, treated using PHS surgery, with those of patients without ADP who underwent pancreatic resections.

2. METHODS

2.1. Patient selection and study design

The data of patients with an SPT of the pancreas were retrieved from our computer database, prospectively compiled from January 1991 to February 2021. This study was approved by our institutional review board (IRB), Taipei Veterans General Hospital (IRB-TPEVGH No.: 2021-03-007AC). The study was carried out in accordance with our IRB guidelines and regulations. The informed consent was waived in this retrospective

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cohort study with anonymity of the data by our IRB. Among the patients with SPT of the pancreas, those displaying ADP were identified and described individually. All the data were compared between groups of SPT with ADP and without ADP, including demographics, operation types, surgical outcomes, pathology, and survival outcomes. To avoid severe pancreatic insufficiency, all SPT patients with ADP underwent PHS enucleation, instead of pancreaticoduodenectomy. The pancreatic duct was identified and closed by the suturing-ligation technique. This procedure was defined to enucleate the tumor with minimal resection of the pancreatic head parenchyma in order to preserve pancreatic function. The pancreatic endocrine function was assessed by periodic measurement of fasting blood sugar, serum hemoglobin A1c (HbA1c), and serum C-peptide. The pancreatic exocrine function was evaluated by the presence of steatorrhea.

2.2. Study aims

The primary aim of this study was to clarify the feasibility and justification of PHS enucleation for SPT patients with ADP by comparing surgical outcomes of these patients and the SPT patients without ADP treated with pancreatic resections. The secondary aim was to evaluate the pancreatic endocrine and exocrine functions after PHS enucleation for SPT patients with ADP and also to share our experience of survival outcomes with long-term follow-up for SPT patients after treatment at our institute.

2.3. Study definitions

Postoperative pancreatic fistula was defined as grades B and C based on the new definition by the International Study Group for Pancreatic Fistula in 2016.[15] Delayed gastric emptying, postpancreatectomy hemorrhage, and chyle leak were identified and classified using the standardized criteria proposed by the International Study Group of Pancreatic Surgery.[16–18] Postoperative complications were graded according to the Clavien-Dindo classification.[19] Resection status was organized into three categories based on the resection margin: R0, a resection with no gross or microscopic evidence of cancer at the resection margin and defined as a margin >0 mm, instead of 1 mm, as indicated by the National Comprehensive Cancer Network; R1, a resection with grossly negative but microscopically positive cancer at the resection margin; and R2, a resection with grossly positive cancer at the resection margin.[20] Surgical mortality was defined as death within 90 days of surgery, including during the same hospitalization period subsequent to the operation or at readmission.

2.4. Statistics

Statistical analyses were performed using Statistical Product and Service Solutions (SPSS) version 21.0 software (SPSS Inc., IBM, Armonk, NY, USA). All continuous data were presented as median (range) and mean (standard deviation [SD]), and frequencies were presented when appropriate to the type of data. Mean values of continuous variables were compared with a two-tailed Student's *t* test. Nonparametric statistical tests were used for variables that do not follow normal distribution. Categorical variables were presented as numbers and percentages and were compared using Pearson's χ^2 test or Fisher's exact test contingency tables. To determine the subset of factors that provided independent information on survival time, a Cox proportional hazard regression model was developed. For all analyses, $p < 0.05$ was considered statistically significant.

3. RESULTS

We observed three cases of SPTs with ADP at our institute (Table 1). All were young females aged between 23 and 30 years. The tumor size ranged from 2.7 to 7.6 cm. All three of these patients underwent PHS enucleation and recovered uneventfully (Fig. 1). No evidence of recurrence has been noted in any of the patients so far, even in the one R1 resection patient, who has been alive for more than 6 years. They have all been experiencing a normal life without any impairment of their pancreatic endocrine and exocrine functions.

No significant differences regarding gender and age were observed between the groups with and without ADP (Table 2). Females were predominant, at 87.1% of the total SPT patients and 100% of the ADP group. The median age was 32 years old, ranging from 8 to 60 years, with a mean age of 33.2 + 11.4 years. The tumor size ranged from 2.2 to 15 cm, with a median of 3.2 cm in the group with ADP and 5 cm in those without ADP. The pancreatic head (45.2%) was the most common site of SPT, followed by the pancreatic tail (32.3%), the body (19.4%), and the neck (3.2%). Most SPT patients displayed no symptoms; however, epigastric pain was the most common symptom, presented in 35.5% of overall patients, 66.7% of those with ADP, and 32.1% of patients without ADP.

PHS enucleation was performed for all three SPT patients with ADP; otherwise, pancreaticoduodenectomy, localized to the pancreatic head, was conducted for those without ADP (Table 3). Blood loss during the operation was similar between the SPT patients with and without ADP, with a median of 120 cc lost during PHS enucleation in the second group. No surgical mortality occurred in our series. Surgical morbidity was comparable between the patients with ADP, who underwent PHS enucleation, and those without ADP, who received pancreatic resection with pancreaticoduodenectomy, distal pancreatectomy, or central pancreatectomy, at 33.3% and 32.1%, respectively. With regard to surgical complications, including severity based on the Clavien-Dindo classification, postoperative pancreatic fistula, postpancreatectomy hemorrhage, intra-abdominal abscess, chyle leakage, and postoperative hospital stay, no significant differences were observed between the two groups. No postoperative de novo DM, follow-up with normal blood glucose, c-peptide and hemoglobin A1c (HbA1c), and no steatorrhea developed in any of these three SPT patients with ADP following PHS enucleation; however, postoperative de novo DM did occur in 11.5% (3/26) of the SPT patients without ADP following pancreatic resection.

All patients underwent an R0 resection, except for one patient in the ADP group who displayed an R1 status (Table 4). Perineural invasion was noted in three patients without ADP, but none of them experienced recurrence. Lymph node involvement was found in one patient without ADP, who developed

Table 1

Agenesis of the dorsal pancreas associated with solid pseudopapillary tumor

Case	Gender	Age, y/o	Symptom	Duration, mo	Tumor size, cm	Treatment	Radicality of resection	Follow-up
1	Female	28	Epigastric pain	3	2.7	PHS Enucleation	R1	Aliver for 75 mo
2	Female	30	Epigastric pain	3	6	PHS Enucleation	R0	Aliver for 38mo
3	Female	23	No	0	7.6	PHS Enucleation	R0	Aliver for 22 mo

PHS = pancreas head sparing.

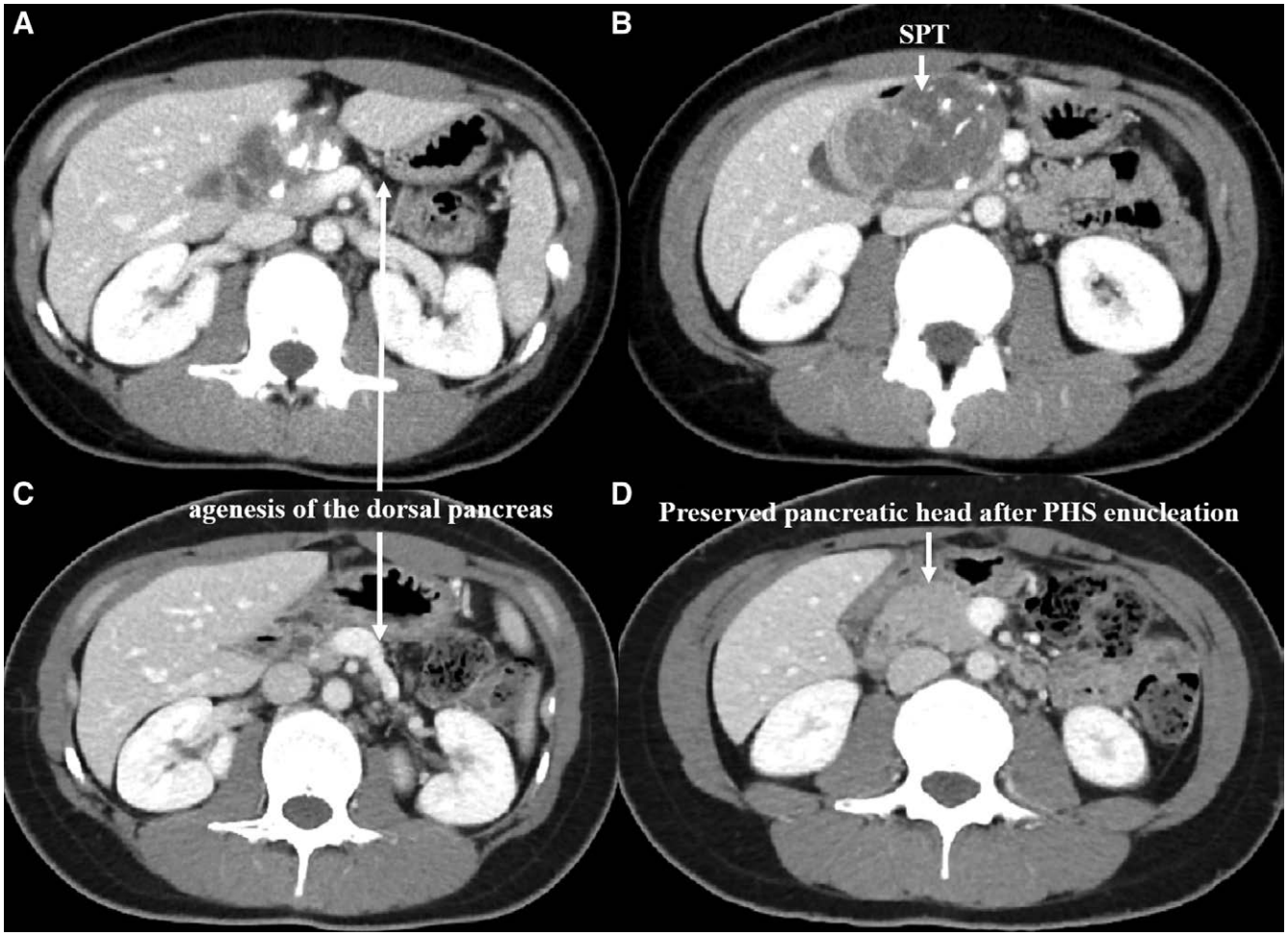

Fig. 1 1A and 1B, Solid pseudopapillary tumors in one (case 2) of the patients with agenesis of the dorsal pancreas before pancreatic head sparing enucleation; 2A and 2B, preserved pancreatic head parenchyma without evidence of tumor recurrence 2 years after PHS enucleation for the same patient.

recurrence later. The follow-up period ranged from 3.4 to 361.7 months, with a median of 67.7 months and a mean of 91.9 + 89.6 months. Overall, the 5- and 10-year disease-free survival rates were 100% and 90%, respectively. The overall 20- and 25-year survival rates were 100% and 66.7%, respectively. As mentioned above, only one patient (3.2%) presented tumor recurrence 7.3 years after distal pancreatectomy and splenectomy for an SPT with lymph node involvement. Resection of the recurrent SPT, with celiac axis resection and liver wedge resection, was performed. However, the tumor recurred 5.5 years after the second operation. The patient underwent a third operation, with resection of the recurrent SPT and multiple liver wedge resections. Thereafter, palliative chemotherapy and multiple radiofrequency ablations were applied until the patient died of the disease 24.5 years after the initial operation. No tumor recurrence after PHS enucleation in the patients with SPTs and ADP was observed during the follow-up period ranging from 10 to 63 months.

4. DISCUSSION

4.1. Solid pseudopapillary tumor in patients with agenesis of the dorsal pancreas reported in the world

SPT of the pancreas with ADP is extremely rare, with only two cases having been reported in the literature prior to this study,[21] Herein, we describe another three cases, making a total of five reported thus far. More than 10 ADP cases have been associated with pancreatic neoplasia, although only about 100 occurrences of ADP are recorded in the literature,[1–6] The mechanism of tumorigenesis in ADP patients is unclear, but chronic pancreatitis caused by the ADP itself could be one of the risk factors for neoplasia,[2] Thus, the association of tumorigenesis with ADP indicates that every ADP patient should be observed with a focus on the early detection of pancreatic neoplasia,[4,5,22,23] According to literature reports and our own study, the clinical features of SPTs in patients with ADP are similar to those observed in patients without ADP,[12,13,21,24,25] This tumor usually occurs in young females, with less than 10% of cases reported in males.[11,26] Our series also agrees with previous research in terms of the most common localization of SPTs[27] and the most frequent symptom[10,12,14,21,25] being the pancreas head and epigastric pain, respectively.

4.2. Treatment options of the solid pseudopapillary tumor

With regard to the treatment of this tumor, chemotherapy and radiotherapy do not play a significant role. Therefore, surgery remains the only curative option and is the standard of treatment for SPT, even with metastasis or local recurrence.[12,14,27] The aim of surgery is to achieve complete (R0) resection, while preserving as much of the pancreatic tissue as possible.[13] Resection approaches include pancreaticoduodenectomy, central pancreatectomy, and distal pancreatectomy, depending on the location of the tumor.[8,9] Traditionally, enucleation is not seen as an option, but the application of this procedure for SPTs is still

Table 2

Demographics for patients with solid pseudopapillary tumor of the pancreas

Demographics	Total	With ADP	Without ADP	*p*
Patients	31	3 (9.7%)	28 (90.3%)	
Sex				1.00
Female	27 (87.1%)	3 (100%)	24 (85.7%)	
Age, year old				0.334
Median	32	28	34	
Range	8-60	23-30	8-60	
Mean + SD	33.2 + 11.4	27.0 + 3.6	33.8 + 11.8	
Preoperative BMI, kg/m^2	n = 26	n = 3	n = 23	0.260
Median	21.2	19.9	22.3	
Range	15.9–34.5	19.0–21.1	15.9–34.5	
Mean + SD	22.7 + 4.3	19.97 + 1.07	23.02 + 4.49	
Preoperative DM	2 (6.5%)	0	2 (7.1%)	1.00
Tumor size, cm	n = 28	n = 3	n = 25	0.568
Median	4.7	3.2	5.0	
Range	2.2–15.0	2.7 – 8.0	2.2–15.0	
Mean + SD	5.7 + 3.4	4.6 + 2.9	5.8 + 3.5	
Location of SPT				0.258
Head	14 (45.2%)	3 (100%)	11 (39.3%)	
Neck	1 (3.2%)	0	1 (3.6%)	
Body	6 (19.4%)	0	6 (21.4%)	
Tail	10 (32.3%)	0	10 (35.7%)	
Symptom				1.000
No	15 (48.4%)	1 (33.3%)	14 (50%)	
Epigastric pain	11 (35.5%)	2 (66.7%)	9 (32.1%)	
Abdominal mass	4 (12.9%)	0	4 (14.3%)	
Nausea/vomiting	1 (3.2%)	0	1 (3.6%)	
Other	3 (10.0%)	0	3 (10.7%)	
Duration, month	n = 28	n = 3	n = 25	0.608
Median	0	0	0	
Range	0–36.0	0–3.0	0 -36.0	
Mean + SD	3.0 + 7.1	1.0 + 1.7	3.3 + 7.4	

ADP = agenesis of the dorsal pancreas; BMI = body mass index; DM = diabetes mellitus; SPT = solid pseudopapillary tumor.

Table 3

Surgical outcomes for solid pseudopapillary tumor of the pancreas

	Total	With ADP	Without ADP	*p*
Patients, n	31	3 (9.7%)	28 (90.3%)	
Operation type				<0.001
Pancreaticoduodenectomy	11 (35.5%)	0	11 (39.3%)	
Central pancreatectomy	1 (3.2%)	0	1 (3.6%)	
Distal pancreatectomy	16 (51.6%)	0	16 (57.1%)	
Enucleation	3 (9.7%)	3 (100%)	0	
Operation time, h				0.291
Median	4.5	3.0	5.4	
Range	0–11.6	2.0–4.2	0–11.6	
Mean + SD	4.5 + 2.5	3.1 + 1.1	4.7 + 2.6	
Blood loss, cc	n = 28	n = 3	n = 25	0.646
Median	140	120	150	
Range	0–4600	20–300	0–4600	
Mean + SD	371 + 874	147 + 142	398 + 922	
Surgical mortality	0	0	0	1.000
Surgical morbidity	10 (32.3%)	1 (33.3%)	9 (32.1%)	1.000
Postoperative complications				0.896
No complication	20 (64.5%)	2 (66.7%)	18 (64.3%)	
Clavien-Dindo I	7 (22.6%)	1 (33.3%)	6 (21.4%)	
Clavien-Dindo II	3 (9.7%)	0	3 10.7%)	
Clavien-Dindo III	1 (3.2%)	0	1 (3.6%)	
POPF, ISGPF grade B and C	2 (6.5%)	1 (33.3%)	1 (3.6%)	0.187
Postpancreatectomy hemorrhage	1 (3.2%)	0	1 (3.6%)	1.000
Intraabdominal abscess	2 (6.5%)	0	2 (7.1%)	1.000
Chyle leakage	4 (12.9%)	0	4 (14.3%)	1.000
Postoperative de novo DM	3/29 (10.3%)	0	3/26 (11.5%)	1.000
Postoperative steatorrhea	0	0	0	NA
Postoperative hospital stay, d				0.301
Median	11	8	12	
Range	5–35	8–10	5–35	
Mean + SD	13.6 + 8.6	8.7 + 1.2	14.1 + 8.9	

ADP = agenesis of the dorsal pancreas; POPF = postoperative pancreatic fistula; ISGP = International Study Group of Pancreatic Fistula; ISGPS = International Study Group of Pancreatic Surgery; DM = diabetes mellitus.

under debate. Tjaden et al[28] advocate an oncological resection because limited surgery may be associated with an increased risk of recurrence and could significantly impair long-term outcomes. In contrast, Liu et al[25] promote the use of function-preserving surgery if possible, based on a study of 243 consecutive patients. Wang et al[29] also state that enucleation for SPT appears to be feasible and safe for preserving the exocrine and endocrine functions of the pancreas, based on a study of 110 SPTs, including 31 patients who underwent enucleation. In the case of SPTs with ADP, the oncological resection approach would be pancreaticoduodenectomy. This would inevitably result in postoperative pancreatic exocrine insufficiency and exogenous insulin dependence due to the lack of the body and tail of the pancreas. Thus, all three of our ADP patients underwent PHS enucleation to preserve the pancreatic head parenchyma as much as possible. Postoperatively, the DM profiles such as blood glucose, serum c-peptide, and HbA1c were all within normal limits and no steatorrhea occurred. They have all experienced a normal life without impairment of pancreatic endocrine and exocrine functions. Moreover, no evidence of recurrence has been noted so far, even in a patient displaying an R1 resection.

4.3. Prognosis and survival outcomes of the solid pseudopapillary tumor

According to the World Health Organization, an SPT is regarded as either having an uncertain potential for malignancy or being a solid pseudopapillary carcinoma.[27] Recurrence is seen with 4.5% to 15% of these tumors.[10,12,26] Factors that have been observed to predict SPT recurrence after resection include tumor size >5 cm, lymphovascular invasion, deep infiltration into the surrounding tissue, lymph node metastasis, tumor rupture, synchronous metastasis, and positive margin; however, some series have reported recurrence without any risk factors.[30] In this study, no significant risk factors were identified to predict tumor recurrence and survival outcomes. Only one SPT patient (3.2%) without ADP presented tumor recurrence in our series, and lymph node involvement may have been a possible contributing factor. Therefore, every SPT should be regarded as having malignant potential.

This patient survived a total of 24.5 years, including 7.3 disease-free years after the initial operation, 5.5 years with the disease following the second operation for local recurrence and distant metastasis, and another 11.7 years with the disease after the third palliative operation and chemotherapy and multiple radiofrequency ablations. In our series, the 5- and 10-year disease-free survival rates were 100% and 90%, respectively, and 20- and 25-year overall survival rates were 100% and 60%, respectively. The reason to describe the unique case of recurrence within the control group is to emphasize the very indolent clinical course. This unique experience has also been observed in other series reported in the literature.[8,9,13,25] Given that most SPTs are biologically indolent and have a favorable prognosis in the normal pancreatic parenchyma, PHS enucleation for SPTs

Table 4

Pathology and survival outcomes for solid pseudopapillary tumor of the pancreas

	Total	With ADP	Without ADP	*p*
Patients, n	31	3 (9.7%)	28 (90.3%)	
Radicality of resection				0.097
R0	30 (96.8%)	2 (66.7%)	28 (100%)	
R1	1 (3.2%)	1 (33.3%)	0	
R2	0	0	0	
Lymphovascular invasion	0	0	0	1.000
Perineural invasion	3 (9.7%)	0	3 (10.7%)	1.000
Lymph node involvement	1 (3.6%)	0	1 (4%)	0.782
Recurrence	1 (3.2%)	0	1 (3.6%)	1.000
Follow-up period				
Median	67.7	25.8	72.8	
Range	3.4–361.7	9.5–62.3	3.4–361.7	
Mean + SD	91.9 + 89.6	32.5 + 27.0	98.3 + 91.8	
Disease-free survival, y				1.000
1	100%	100%	100%	
5	100%	100%	100%	
10	90.0%	N/A	90.0%	
20	90.0%	N/A	90.0%	
Overall survival, y				1.000
1	100%	100%	100%	
5	100%	100%	100%	
10	100%	N/A	100%	
20	100%	N/A	100%	
25	66.7%	N/A	66.7%	

ADP = agenesis of the dorsal pancreas; N/A= not available; R0 = curative resection without gross and microscopic cancer at the resection margin; R1 = palliative resection without gross, but with microscopic cancer at the resection margin; R2 = palliative resection with gross cancer at the resection margin.

with ADP seems to be justified, as shown by our preliminary experience. Nevertheless, there have been reports of recurrence more than 10 years after the initial operation. Therefore, long-term follow-up periods are necessary to establish PHS enucleation as viable for the treatment of this extremely rare disease.[21]

Limitations of this study are the small sample size and selection bias for both study and control groups. Given that ADP associated with SPT is exceedingly rare but interesting, it is worthy to share the precious experience of PHS enucleation for these patients. However, it would be impossible to clarify the justification of PHS enucleation in terms of oncological outcomes and preservation of pancreatic functions without the experience and comparison of the control group of SPT without ADP, although it is hard, if not meaningless, to undergo a statistical analysis among so few cases. Besides, it would be inevitable to have many biases to include different types of pancreatic surgery for the control group consisting of SPT without ADP at all locations in the pancreas.

In conclusion, the association of tumorigenesis with ADP suggests that every congenital ADP patient should be observed with a focus on the early detection of pancreatic neoplasia. With regard to surgical and survival outcomes, PHS enucleation is feasible and justifiable for SPTs in patients with ADP and could thus be recommended to avoid pancreatic insufficiency. Nevertheless, further reports with larger sample sizes and long-term follow-up periods are needed to reach a reliable conclusion.

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