

## Case Report

# Solid pseudopapillary tumor of the pancreas: a case series of 11 children

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**Abstract:** This study aimed to analyze the clinical data, pathologic features and surgical management about solid pseudopapillary tumor of the pancreas (SPTP) in children. The clinical data and pathological findings from 11 children with SPTP were retrospectively analyzed, who were diagnosed and treated in three tertiary academic centers between January 2001 and December 2015. The 11 children consisted of 10 females and 1 male, of median age at operation of 11.4 years old (range from 3.7 to 16.3 years old). The clinical symptoms were non-specific, and mainly manifested upper abdominal pain or discomfort. The CT results of all patients were similar, and the feature of SPTP was solid or mixed solid and cystic. The neoplasm was localized in the pancreatic head/neck in 4 patients, and in the body in 3 patients, and in the tail in 4 patients, which were all confirmed during abdominal operations. The diameter of these lesions ranged from 2.8 to 17.3 cm, without abdominal cavity metastasis or hepatic metastases. All of the tumors were resected successfully, which included 2 pancreaticoduodenectomies (Whipple-Child), 4 local resections, 1 segmental pancreatectomy, 2 distal pancreatectomies, 2 spleen-preserving distal pancreatectomies. The follow-up of all cases ranged from 8 months to 14 years, and no tumor recurrence or metastasis was observed in these SPTP children. The results indicated that all tumors were positive for the immunohistochemical staining of  $\beta$ -catenin, progesterone receptor (PR), vimentin, and CD99. However, all the tumors were negative for the immunohistochemical staining of E-cadherin and cytokeratin 7 expression. CT examination combined with age and sex profile should be sufficient for children SPTP diagnosis. Surgery was the main treatment choice. Children with SPTP have an excellent prognosis after surgical excision. Surgery strategy should be determined according to preoperative CT examination, intraoperative findings of tumor location, capsule integrity, and invaded surrounding tissues. Immunohistochemical staining of E-cadherin/ $\beta$ -catenin, PR, vimentin, and CD99 may help the diagnosis, though their expressions were found to have the indistinct complex immunophenotypes.

**Keywords:** Solid pseudopapillary neoplasm, diagnosis, treatment, children, immunohistochemistry

### Introduction

Solid pseudopapillary tumor (SPTP) is one of the rare primary tumors of the pancreas, and this kind of tumor has a low malignant potential, which was first described by Frantz in 1959 [1]. Clinical symptoms of children SPTP are non-specific and mainly include upper abdominal pain and discomfort or vomit. The abdominal mass could be palpated during the abdominal physical examination. However, there were still reports about recurrent pancreatitis caused by pancreatic tail SPTP in children cases [2]. When SPTP size was small, the patient could be

asymptomatic, and generally, SPTP was occasionally found during the physical examination. And it could also be found accidentally for other reason, such as abdominal examination for patients who had abdominal closed injury [3]. When SPTP size was relatively larger, the patient could have the non-specific symptoms, such as abdominal pain, nausea, vomiting and so on.

Due to lack of specific clinical manifestations, the preoperative SPTP diagnosis was relatively difficult. The needle biopsy under the guide of B ultrasound, CT or endoscopic ultrasonography had been verified as the effective and feasible

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preoperative diagnosis methods [4-7]. CT examination played an important role in preoperative SPTP diagnosis for its non-invasive, quick and convenient characteristics [8]. Surgical resection of the primary tumor as far as possible is the main effective way of children SPTP treatment [9]. Even if the patients had metastasis, surgical resection of both primary and metastatic lesions could also result in good prognosis. In addition, it is important to differentiate patients with SPTP from other pancreatic neoplasms, which perform the various biologic behavior. The results of immunohistochemical staining could provide the valuable information to help in distinguishing SPTP patients from other pancreatic tumors, such as pancreas islet cell tumor, pancreatic ductal adenocarcinoma and so on. However, although many biomarkers have been applied in the immunohistochemical evaluation of SPTP patients, the single and specific antibody still has not been observed [10].

SPTP has lower malignant potential, and tumor cell doubling time reaches up to 765 days [11]. Because the children are in the stage of growth and development, the continuity of the digestive tract, spleen immune function, and internal and external secretory function of the pancreas play the important roles in the process of children growth and development. The long-term postoperative quality of life should be considered, and it is recommended that the extent of surgical resection should be decreased as far as possible, in order to sustain the continuity of the digestive tract, retain spleen and more normal pancreatic tissue [12]. Children SPTP is rare in clinical practice, its incidence is about 0.01/100,000 person/year. However, it accounts for above 50% of all primary pancreatic tumors in children [13]. Due to lack of specific clinical manifestations and lower preoperative diagnostic rate, it was usually misdiagnosed as pancreatoblastoma. Then the scope of surgical resection was expanded, which increases surgery related complications. Otherwise, the patients also could be misdiagnosed as pancreatic pseudocyst, and underwent the surgery treatment of 'Cholecystoduodenostomy' Y operation. And they are required to receive secondary radical surgery. Therefore, it had significant clinical significance for pediatric surgeons to improve the understanding of this disease. Here, we reviewed clinical data and pathological findings

of 11 children diagnosed and treated in our hospital between January 2001 and December 2015.

### Materials and methods

#### *Subjects*

After Institutional Review Board approval, we conducted this retrospective chart review. In total, eleven children with the pathology proven diagnosis of SPTP surgically treated at Department of General Surgery in Children's Hospital of Shanghai Jiaotong University, Tongji Medical College of Huazhong University of Science and Technology, and Wuhan Children's Hospital were enrolled in this study between January 2001 and December 2015. SPTP was diagnosed based on its gross and microscopic appearance as well as on immunohistochemical staining. Tumors were assumed to be malignant if there was extrapancreatic or pancreatic parenchymal invasion, lymph node involvement, perineural or vascular invasion, or distant metastases.

#### *Immunohistochemical assays*

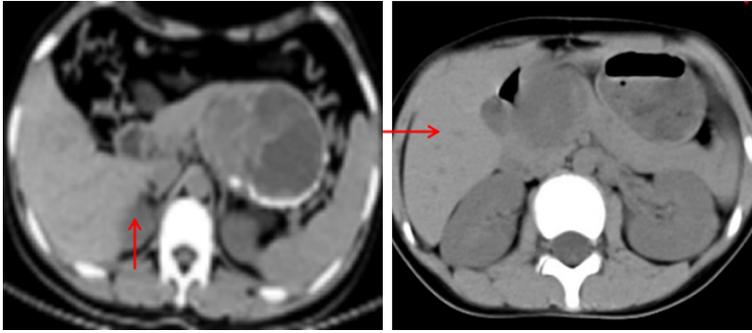
Immunohistochemical assays were performed using an automated Benchmark platform (Ventana Medical Systems, Tucson, AZ, USA) according to the manufacturer's recommendations. Four-micrometer-thick sections were immunostained with antibodies to chromogranin-A, synaptophysin, progesterone receptor (PR), c-kit,  $\beta$ -catenin, cytokeratin (CK), CK7, CD10, E-cadherin, CD99 and vimentin using an UltraView universal DAB detection kit (Ventana Medical Systems). All immunohistochemically stained slides were evaluated by a single pathologist. For all antibodies, staining of more than 5% of the tumor cells was regarded as positive. For PR, cells showing nuclear staining were considered positive. For  $\beta$ -catenin, cells showing cytoplasmic/nuclear staining were considered positive. Cells were considered positive for E-cadherin, CK, and CK7 when the cytoplasmic membrane was stained and were considered positive for c-kit when cytoplasmic staining with or without nuclear staining was observed. Tumor cells were considered positive for chromogranin-A and synaptophysin when cytoplasmic granular staining was observed and were considered positive for CD99 when a paranuclear dot-like

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**Table 1.** The clinical baseline data of 11 SPTP children

Case no.	Age (years old)	Gender	Symptom duration	Symptom	Mass	Tumor diameter (cm)	Site	Shape	Intraoperative capsule	Surgical strategy	Lymph nodes involved	Follow up (months)	Last follow up
1	11.2	F	7 days	Abdominal pain	Unpalpable	5.2	Pancreatic body	Mixed solid and cystic	Complete	Local resection	0/2	158	Normal
2	12.3	F	2 months	Abdominal pain	Palpable	12.3	Pancreatic head/neck	Mixed solid and cystic	Incomplete	Pancreaticoduodenectomyies (Whipple-Child)	0/6	134	Moderate malnutrition
3	14.4	F	3 days	Abdominal mass	Palpable	8.5	Pancreatic tail	Mixed solid and cystic	Complete	Dstal Pancreatospfenectomy	0/3	125	Increased platelet
4	3.7	F	1 day	No symptom	Unpalpable	2.8	Pancreatic head	Solid	Complete	Local resection	0/2	114	Normal
5	9.6	M	12 days	Abdominal pain	Palpable	4.8	Pancreatic body	Mixed solid and cystic	Complete	Segmental pancreatectomy	0/3	92	Normal
6	10.2	F	1.2 days	Abdominal pain	Palpable	16.4	Pancreatic head	Mixed solid and cystic	Complete	Local resection	0/5	88	Normal
7	16.3	F	6 months	Abdominal pain	Palpable	11.2	Pancreatic head	Mixed solid and cystic	Incomplete	Pancreaticoduodenectomyies (Whipple-Child)	0/8	78	Mild malnutrition
8	12.6	F	4 months	Abdominal pain	Palpable	14.4	Pancreatic tail	Mixed solid and cystic	Complete	Spleen-preserving distal pancreatectomies	0/3	67	Normal
9	11.4	F	18 days	Abdominal pain	Palpable	9.2	Pancreatic body	Mixed solid and cystic	Complete	Local resection	0/5	53	Normal
10	8.5	F	5 days	No symptom	Palpable	5.6	Pancreatic tail	Mixed solid and cystic	Complete	Spleen-preserving distal pancreatectomies	0/4	32	Normal
11	13.2	F	8 days	Abdominal pain	Palpable	17.3	Pancreatic tail	Solid	Complete	Dstal Pancreatospfenectomy	0/4	8	Increased platelet

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**Figure 1.** The plain CT results showed that low or equal density mixed solid and cystic mass with clear edges could be observed, and part of wall had shown calcification.

pattern was present. Cells were regarded as positive for vimentin and CD10 when cytoplasmic staining was observed.

### *Preoperative biochemical and imaging examinations*

All the children underwent the serum tumor biomarker test, such as  $\alpha$ -FP, CEA, CA199, CA125 and so on before the surgery. And they also underwent the examinations of liver and kidney function, fasting blood glucose and urine amylase. All the patients had preoperative abdominal CT examination (plain + enhanced).

### *Treatment*

All the children underwent treatment of laparotomy operation. No children had received chemotherapy and radiotherapy before and after the surgery.

### *Follow-up*

Follow-up included clinical examination, routine laboratory tests, abdominal US, and CT or MRI every 3 months. The SPTP children were followed up for a mean duration of 86 months, (range 8 to 158 months) and all 11 children were alive with no evidence of disease recurrence or metastasis.

### *Statistical analysis*

Normality of the distribution of continuous variables was determined using the Kolmogorov-Smirnov test. Continuous data were presented as mean  $\pm$  SD or median (range). Categorical data were presented as n (%). Statistical analysis was performed using the Statistical Program

for Social Sciences (SPSS) software 17.0 (SPSS Inc., Chicago, IL).

## **Results**

### *Clinical data*

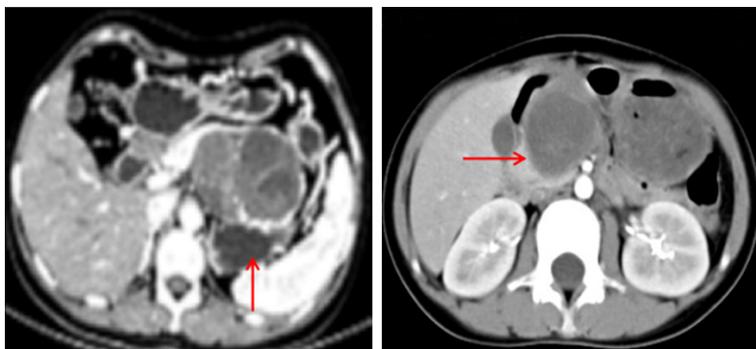
The baseline data of the SPTP children were listed in **Table 1**. The 11 cases included 10 females and 1 male patients, and the median age was 11.4 years old (range from 3.7 to 16.3 years old). About clinical manifestations, 8 cases (8/11, 72.7%) had symptoms and signs of abdominal pain and discomfort, and vomiting, 2 cases found abdominal masses during physical examinations. And 1 child was found by abdominal CT occasionally during the examination for abdominal closed injury. No children had signs of jaundice (skin and eyes). And no children had the medical history of pancreatitis occurrence and abdominal trauma. All of the tumors were resected successfully, which included 2 pancreaticoduodenectomies (Whipple-Child), 4 local resections, 1 segmental pancreatectomy, 2 distal pancreatectomies, 2 spleen-preserving distal pancreatectomies.

The results of serum tumor biomarker, including  $\alpha$ -FP, CEA, CA199, CA125, were normal in all the children. And the values of liver and kidney function, fasting blood glucose and urine amylase were also normal in all the children before the surgery.

### *Preoperative CT imaging features*

All the children underwent preoperative abdominal CT examination. The CT results of 9 children showed that low or equal density mixed solid and cystic mass with clear edges could be observed, and part of wall had shown calcification in plain scan. The solid part of tumor showed mild enhancement, and the cystic part of tumor did not show enhancement, the capsule showed obvious enhancement during CT enhanced scan. There were two cases found to be solid characteristics. Four cases had the tumors in pancreatic head/neck four cases had the tumors in pancreatic tails, and the other three cases had the tumor in pancreatic body.

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**Figure 2.** The enhanced CT results showed that the solid part of pancreatic head and tail showed mild enhancement, and the cystic part of tumor did not show enhancement.



**Figure 3.** The CT 3D reconstruction result of SPTP child showed that the stomach was oppressed by mixed solid and cystic pancreatic head.

Eight children had the exophytic tumors, and the other three children had the tumors mainly located in pancreas parenchyma. None of the tumors had evidence of surrounding organs involved, and pancreatic duct dilatation were also not observed in all cases. The CT images were shown in **Figures 1-3**.

### *Immunohistochemical staining*

Postoperative pathology results showed that the neoplasms shapes of 9 cases were mixed solid and cystic and 2 cases were solid. Under the observation of microscopy, the neoplasms consisted of pseudopapillary tissues, and tumor cells arranged surrounding the blood vessels.

**Table 2** showed the immunohistochemical results in detail. Cytokeratin profiles (CK, CK7)

staining showed the features of ducts and ductular cells in the region of pancreas. Chromogranin A and synaptophysin should be considered as neuroendocrine biomarkers. All 11 SPTP children underwent the assessment of immunohistochemical analysis. All the samples were positive for the expression of vimentin,  $\beta$ -catenin, PR, and CD99. CD99, which were stained as intracytoplasmic paranuclear dot-like patterns. However, all of them were negative for the staining of E-cadherin and CK7.

All these 11 children diagnosed as SPTP indicated the characteristic cytoplasmic/nuclear immunoreactivity of  $\beta$ -catenin and loss of membranous E-cadherin. In addition, 10 children (90.9%) showed positive expression of chromogranin A staining, 9 (81.8%) children were positive for CK, each of 7 patients (63.5%) could be observed positive for CD-10 and c-kit, respectively, and only 4 (36.4%) children were found to be positive for synaptophysin expression. Ten patients (91%) were positive for chromogranin A, 9 (82%) were positive for CK, 7 (64%) each were positive for c-kit and CD10, and 4 (36%) were positive for synaptophysin.

### *Postoperative follow-up and long term prognosis*

In this study, no patient died during the operation. All the 11 cases were cured and discharged. One patient who underwent segmental pancreatectomy had signs of pancreatic leakage at the fifth day after surgery. After the conservative treatment of adequate intraperitoneal drainage, pancreatic secretion inhibition, and anti-infection medicines, the patient recovered and discharged smoothly. The postoperative follow-up of 11 SPTP children ranged from 8 months to 14 years (158 months). The height, weight, nutritional status and tumor markers, blood glucose, glucose tolerance test, coagulation, liver and kidney function, and CT or ultrasound imaging were examined regularly during the followed-up period. Among all the children, 2 cases underwent pancreaticoduodenectomy (Whipple-Child) surgery had

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**Table 2.** Immunohistochemical staining results of 11 SPTP children

Case No	β-catenin (PR)	Progesterone receptor	Vime-ntin	CD99	E-cadherin	Cytokeratin 7	Chromogranin A	Synaptophysin	C-kit	CK	CD10
1	+	+	+	+	-	-	+	-	+	+	-
2	+	+	+	+	-	-	+	-	+	+	+
3	+	+	+	+	-	-	+	+	-	+	-
4	+	+	+	+	-	-	+	-	+	-	+
5	+	+	+	+	-	-	-	+	-	+	+
6	+	+	+	+	-	-	+	-	+	+	-
7	+	+	+	+	-	-	+	-	+	-	+
8	+	+	+	+	-	-	+	-	-	+	-
9	+	+	+	+	-	-	+	+	-	+	+
10	+	+	+	+	-	-	+	+	+	+	+
11	+	+	+	+	-	-	+	-	+	+	+

growth retardation of height and weight. Compared with children with same age, the 2 cases had mild to moderate malnutrition. The other children had normal growth and development. In addition, 2 children who underwent distal pancreatectomy surgery with elevated blood platelet, and their platelet level returned to normal after the administration of oral dipyridamole tablets. No patient was observed abnormal change of serum tumor biomarker, and all the patients had no recurrence and metastasis during the follow-up period.

### Discussion

The imaging characteristics of SPTP children in this study mainly manifested as below: The size of lesions was larger, usually more than 5 cm. Generally, the tumor showed the shape of circular or oval, with integrate capsule, smooth and uniform wall. The results of CT plain scanning showed cystic and solid mass with uneven density. The calcification could be observed in part of wall. The solid part could be observed mild enhancement by enhanced CT scan. No common bile duct and pancreatic duct dilatation, enlarged retroperitoneal lymph node and distant metastasis was observed in any SPTP children, and all the children had no surrounding organs and tissues involved. For space occupying lesion in pancreas, especially in female children with atypical clinical symptoms and signs, SPTP should be considered after exclusion of other pancreatic mass lesion possibility. The preoperative imaging diagnosis rate was 18.2% (2/11) in this study, which was similar to the result about 23.5%, which was reported by Law [14]. Maybe it should be considered that the low children SPTP incidence

maybe related with our lack of knowledge about the disease.

The results of Jani' study showed that under the guide of preoperative ultrasound guided endoscopic fine-needle aspiration for 28 patients, 21 cases were diagnosed as SPTP definitely [5]. Compared with postoperative pathological result, the accuracy rate reached up to 75%. However, Matsubayashi considered that though the complications of preoperative ultrasound guided endoscopic fine-needle pancreas aspiration were fewer, once the complication developed, the consequences maybe very serious [6]. In addition, the operator should be required to have certain professional skills. Levy also considered that the procedure of ultrasound guided endoscopic fine-needle pancreas aspiration could result in tumor dissemination, traumatic pancreatic leakage and pathological false negative results of puncture tissue and so on, which could exert the unfavorable influence on treatment and prognosis of SPTP patients [7]. Considering the above mentioned factors and poor compliance of children, ultrasound guided endoscopic fine-needle pancreas aspiration were not applied in the SPTP children in this study.

The exact cellular origin of SPTP still remains unclear [15]. Many attempts have been attempts to find the related immunoprofile of precursor cell types for SPTP patients, including Acinar, ductal, endocrine, and multipotential stem cells and so on [10, 16-18]. Actually, SPTP samples have been observed to express the biomarkers of exocrine, endocrine, mesenchymal, and even epithelial cell to some extent. Patients with SPTP performed the SPTs of the

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pancreas were found to indistinct complex immunoprofile, with 93% positive for neuron-specific enolase and 0% for chromogranin A [15], which was not consistent with pancreatic neuroendocrine cell types. Our finding of SPTP children's immunoprofiles were similar to those previous reported studies [15, 17, 18], with 36.4% and 91% of the tumors positive expression for the neuroendocrine biomarkers synaptophysin and chromogranin A, respectively. Currently, the positive expression of intracytoplasmic paranuclear dotlike CD99 has been found to differentiate SPTP from other pancreatic tumors [10], which was also consistent with our findings. The process of SPTP pathogenesis is still obscure. It has also been observed that Wnt signaling is associated with a  $\beta$ -catenin (CTNNB1) mutation, which may play the crucial role in SPTP tumorigenesis. The above mentioned mechanism could result in diffuse cytoplasmic and aberrant nuclear expression of  $\beta$ -catenin and the lack of membranous E-cadherin [19, 20]. Moreover, due to its female predominance, the hormonal influence on its pathogenesis has also been proposed.

Surgical removal of the tumor is the main effective mean for children SPTP treatment [9, 13]. No children were observed distant metastases Tanaka Y, during preoperative CT examination and intraoperative exploration. The tumors were all resected completely. The tumors were located in pancreas head/neck in 4 cases, and 2 patients underwent local resection due to exophytic growth of intraoperative finding, and the other 2 children underwent Child surgery, due to incomplete capsule and surrounding organs invaded. However, compared with peer children, the children who underwent Child surgery had growth and development retardation of height and weight, showing appearance of mild to moderate malnutrition. Maybe it was associated with excessive organ resection and change of children intestinal continuity. Snajdauf observed 6 SPTP children of pancreatic head, who underwent duodenum-preserving resection of pancreatic head (DPRPH) for pancreatic head benign tumor [21]. The follow-up time ranged from 6 to 16 months. The growth and development of these children did not show significant difference, compared with peer children. And no tumor recurrence or metastasis was observed. It indicated that DPRPH surgery for pancreatic head tumor of

children SPTP was feasible. And the procedure of pancreaticoduodenectomy could exert a large burden for the children in the status of growth and development. If the tumor did not invade surrounding organs and tissues, then the surgical strategy of duodenum-preserving resection of DPRPH for pancreatic head benign tumor was recommended. However, Xiao YH suggested that, when the tumor showed infiltrative growth, had the performances of uneven surface and rich vessels like appearance, and the tumor infiltrated the duodenum, mesenteric artery and vein, and splenic vein, then the potential of malignant tumors should be considered, and the patients were recommended to receive extensive radical resection or volume reduction surgery [22].

Four cases had tumor which were located in pancreatic tail, two of them received distal pancreatectomy with spleen and splenic vessels preservation. One patient received distal pancreatectomy and splenectomy due to limited knowledge about this disease. One patient was suspected to have splenic vein infiltrated during the operation, and underwent distal pancreatectomy and splenectomy. Two patients who underwent splenectomy had increased blood platelet 2 weeks after the surgery, and the platelet level returned to normal after administration of oral dipyridamole tablets. And no deep vein thrombosis and pulmonary embolism was observed during follow-up period. And the patients did not occur overwhelming post-splenectomy infection, due to the treatment of oral penicillin for two months. Nakamura considered that spleen should be reserved as far as possible, when tumor did not infiltrate the splenic artery and vein [23]. When necessary, the patient could receive spleen partial splenectomy, in order to maintain the immune function and prevent overwhelming post-splenectomy infection. Three cases had tumors located in pancreatic body. Two cases received local tumor resection. One patient had tumor which was located in pancreatic parenchymatous tissue, and the tumor was adjacent to pancreatic duct and vessels. Considering that pancreas and its ducts could be damaged during intraoperative tumor exposure, which might result in severe postoperative complications such as pancreatic leakage and so on, the patient received the segmental pancreatectomy and pancreaticojejunostomy.

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In this study, four cases underwent local tumor resection, and 1 case received segmental pancreatectomy according to the preoperative CT examination results and intraoperative findings. The case who underwent segmental pancreatectomy had postoperative pancreatic leakage (inactive pancreatic trypsin), and other four children did not have any complications. During the follow up period, no recurrence or metastasis was observed. It was indicated that SPTP children should conduct local tumor resection or segmental pancreatectomy when intraoperative findings showed that tumor capsule was intact and no surrounding tissue and organs were obviously invaded, in order to preserve more pancreas tissues. The results of Li's study showed that, compared with patients who underwent standard pancreaticoduodenectomy, the surgical time, hospital time and postoperative complication rate of patients who underwent local tumor resection combined with segmental pancreatectomy were all decreased ( $p < 0.05$ ) [24]. However, the long term prognosis did not show significant difference between two groups ( $p > 0.05$ ). We believe that as long as the tumor capsule was intact and tumor did not invade surrounding organs and blood vessels, it was recommended the patient to select the surgical strategy with small trauma, on the basis of radical resection assurance. Both the local tumor resection and segmental pancreatectomy could conserve the endocrine and exocrine function of the pancreas. However, due to the larger tumor size, the wound after local tumor resection was relatively larger, and it should avoid part of small pancreatic ducts branches damaged. If necessary, the suspected small pancreatic ducts branches should be ligated appropriately and then the wound was sutured, in order to prevent the occurrence of pancreatic leakage. During the procedures of segmental pancreatectomy and distal pancreaticojejunostomy, it was recommended to ligate proximal main pancreatic duct, conduct pancreatic section transfixion and double U pancreaticojejunostomy application during distal pancreaticojejunostomy, in order to prevent the occurrence of pancreatic fistula. The number of lymph node dissection for 11 SPTP children in this study ranged from 2 to 8, and the pathological results showed chronic inflammatory changes. The patients were followed up for 8-14 years (158 months). And no tumor recurrence and metas-

tasis was observed in any patient. This indicated that extensive lymph node dissection and extended resection is not necessary for SPTP children. However, due to the limitation of small sample size and relatively short follow up time, the result still should be validated by long term follow up data.

In this study, all the children did not have tumor recurrence and metastasis during the follow up period. Hwang reported 4 recurrence cases of SPTP children after surgery treatment [25]. Compared with the SPTP children without recurrence, the author considered that the more solid components accounted for tumor volume, the higher rate of tumor recurrence potential could be. Tang retrospectively analyzed the imaging data of 100 SPTP cases, the results showed that enlarged peripheral lymph nodes were associated with SPTP malignancy [26]. Lee considered that when SPTP patients had other organ metastasis or postoperative recurrence, the combination resection of primary lesion and metastatic lesion or secondary resection of recurrence lesion could also achieve good effects [27]. The effects of chemotherapy and radiotherapy were relatively limited. However, Kante reported a female SPTP child, who could not receive surgical resection treatment due to huge size of tumor. After administration of gemcitabine, the tumor size was decreased, and then the lesion was successfully removed [28]. Few patients had been reported to receive radiotherapy, and few of them had perform corresponding treatment response [29].

In recent years, with the advance and maturity of laparoscopic techniques, the unique advantages of minimally invasive could develop the new avenue of children SPTP treatment. Sokolov treated 2 SPTP children by the application of laparoscopic surgery and achieved good treatment effects [30]. The 2 children followed up 6 months and 2 years, respectively, and no tumor recurrence and metastasis was observed. Petrosyan also believed that, for SPTP children whose lesions were located in the body and tail of the pancreas, the surgery strategy of laparoscopic distal pancreatectomy was feasible and safe [31]. However, Fais considered that laparoscopic biopsy or resection of the tumor could result in intra-abdominal diffusion of tumor cells due to injected gas, which

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was not very suitable for SPTP children [32]. Therefore, the efficacy and safety of laparoscopic surgery in the treatment of SPTP children were still obscure, which were lack of long term follow up validation, and still need further clinical research.

### Conclusion

In short, the incidence of children SPTP was low, and SPTP was a kind of tumor with low potential malignancy that usually affects young females. CT examination was the important means for the preoperative diagnosis of SPTP. And surgical removal is the main treatment strategy. Surgery strategy should be determined according to preoperative CT examination, intraoperative findings of tumor location, capsule integrity, and invaded surrounding tissues. Immunohistochemically, cytoplasmic/nuclear immunoreactivity of  $\beta$ -catenin and loss of membranous E-cadherin, PR, vimentin, and CD99 would help establish the diagnosis of children SPTP, though their expressions were found to have the indistinct complex immunophenotypes.

### Disclosure of conflict of interest

None.

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