Surgical treatment for solid pseudopapillary neoplasm of the pancreas in children: experience at a center

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ABSTRACT

Objective: To evaluate the surgical outcomes of solid pseudopapillary neoplasms (SPNs) of the pancreas in children at the Vietnam National Children's Hospital. **Methods:** This study was a descriptive case series that retrospectively analyzed 26 patients diagnosed with pancreatic SPNs who underwent surgical treatment at the National Children's Hospital between January 2021 and May 2024. **Results:** The average age of onset in children was 10.9 years (range 7-15 years), with a predominance in females (88.5%). The main clinical symptoms were epigastric pain and left hypochondrial pain (80.5%). Computed tomography (CT) revealed that the tumors were located in the head of the pancreas in 12 of 26 patients (46.2%) and in the body and tail of the pancreas in 14 of 26 patients (53.3%). The average tumor size was 5.6 cm (range 2-12 cm), with the majority having a mixed structure in 18 out of 26 patients (69.2%). All patients underwent open surgery with an intraoperative complication rate of 30.8%. The most common postoperative complication was exocrine pancreatic insufficiency, occurring in 45% of the cases. No cases of early recurrence or postoperative mortality were observed. **Conclusion:** SPNs are rare pancreatic tumors with low malignancy, minimal invasiveness, and low metastasis and recurrence rates. Therefore, surgery is a safe and effective treatment for children, with low complication rates.

Keywords: Solid pseudopapillary neoplasm of the pancreas, children.

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INTRODUCTION

Solid pseudopapillary neoplasm (SPN) of the pancreas is a rare exocrine pancreatic tumor that accounts for approximately 1% of all pancreatic tumors [1]. The first discovery of SPN was made by Gruber Frantz in 1927, who provided a detailed description of its pathology in 1959. SPN predominantly affects women of reproductive age and is rare in children. However, with advancements in medical science, the detection rate of SPN in children has been increasing. It is often difficult to diagnose because of its nonspecific clinical symptoms, and routine blood tests are not useful for diagnosis. Preoperative diagnosis relies on CT or MRI imaging, with histopathology being the gold standard. Surgery is the primary treatment option for this condition, and it offers long-term postoperative survival. Therefore, in this report, we aimed to evaluate the surgical outcomes of treating solid pseudopapillary

neoplasms of the pancreas in children at Vietnam National Children's Hospital.

METHODS

A descriptive case series study was conducted with 26 patients between January 2012 and May 2024.

Patient information was collected using the *following procedure.*

• All patients were selected based on histopathological results confirming SPN from the pathology department.

• Supplement of corresponding patient information from the clinical department.

• The medical records of each patient were retrieved from the medical record storage department.

• Ensure completeness of information in the records.

• Data were collected and analyzed using SPSS software (version 26.0).

Each patient selected for the study met the *following criteria.*

• The lesion was located in the pancreas, with histopathological confirmation of the SPN.

• Male and female patients of all ages.

• Surgical treatment was performed at Vietnam National Children's Hospital during the study period.

• Complete medical records with clinical and paraclinical results, surgical reports, and histopathological results.

Exclusion Criteria:

• Histopathological types not found in the pancreas.

• Patients without stored specimens.

• Patients without complete medical records, including histopathological results.

RESULTS

Among the 26 patients, the majority were female (23 of 26 cases, 88.5%). The average age at diagnosis in the children was 10.9 years (range, 7–15 years). Abdominal pain was observed in most patients (21 of 26, 80.8%). Less common symptoms included loss of appetite, weight loss, digestive disorders, palpable masses, or post-abdominal trauma. (Figure 1)

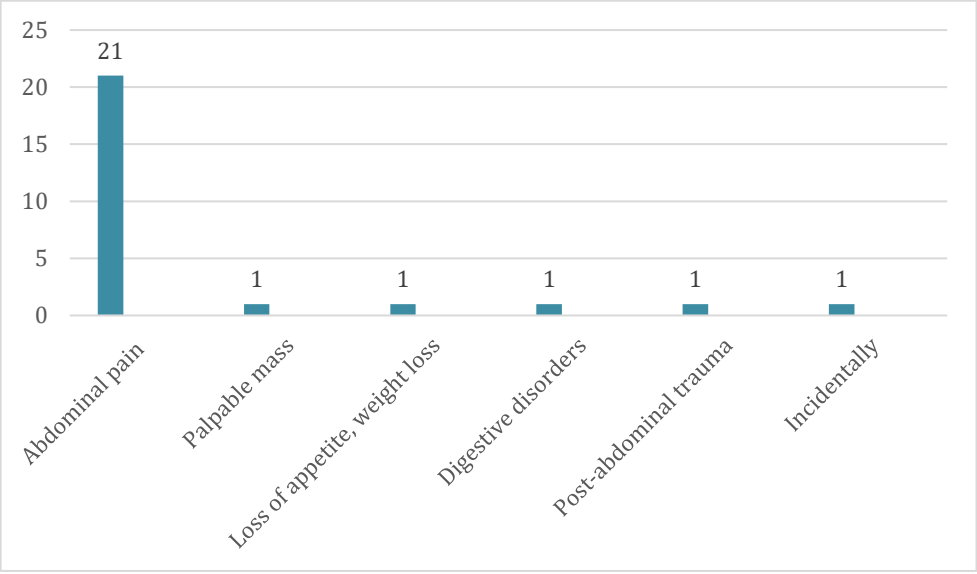

Figure 1. *Clinical symptoms*

CT scans showed that 12 of 26 patients (46.2%) had tumors located in the head of the pancreas, and 14 of 26 patients (53.8%) had tumors in the body and tail of the pancreas. The average tumor size was 5.6 cm (range: 2–12 cm). The tumor structure on CT was solid in 7 out of 26 patients (27%), cystic in 1 out of 26 patients (3.8%) and mixed in 18 out of 26 patients (69.2%). Most tumors exhibited irregular contrast enhancement due to necrosis and intratumoral hemorrhage. (Table 1)

Table 1. *Paraclinical features*

Paraclinical features			**N**	**%**
CT	Location	Head of the pancreas	12	46,2
		Body and tail of the pancreas	14	53,8
	Tumor	Solid	7	27
		Cystic	1	3,8
		Mixed structure	18	69,2
	Contrast enhancement	Intense	0	0
		Mild	12	46,2
		Uneven	14	53,8
	Size	< 3cm	5	19,3
		3-10cm	18	69,2
		> 10cm	3	11,5
AFP	Normal		26	100
Serum glucose	Normal (3,3-5,5 mmol/L)		26	100
Serum P-Amylase	Normal (< 53 U/L)		26	100
Serum Lipase	Normal (7-39 U/L)		26	100

All patients underwent open surgery. Among them, 18 patients experienced no complications during surgery. Early complications: No patient experienced bleeding, pancreatic fistula, biliary fistula, residual abscess, or death immediately after surgery. Late complications: One patient developed acute pancreatitis in the second week after surgery. All the patients were evaluated for pancreatic function after surgery. None of the patients had endocrine pancreatic insufficiency; however, 20 of 26 patients had their exocrine pancreatic function assessed, and exocrine pancreatic insufficiency was noted in 9 of 20 patients (45%). (Table 2)

Table 2. *Surgical Characteristics*

Surgical Characteristics	**N**	**%**

SPN at the Head of the pancreas	Pancreaticoduodenectomy		1	3,8
	Duodenum-preserving pancreatic head resection		11	42,3
SPN at the Body and tail of the pancreas	Central pancreatectomy		3	11,5
	Distal pancreatectomy	with Spleen Preservation	8	30,8
		with Splenectomy	3	11,5
Intraoperative complications	Portal Vein Injury		1	3,8
	Common Bile Duct Injury		1	3,8
	Pancreatic Duct Injury		3	11,5
	Splenic Vascular Bundle Injury		3	11,5
Postoperative complications	Exocrine pancreatic insufficiency		9/20	45
	Acute Pancreatitis		2	7,7

DISCUSSIONS

SPN of the pancreas is a rare exocrine pancreatic tumor, accounting for approximately 1% of pancreatic tumors [1]. The tumor was first discovered by Gruber Frantz in 1927, and in 1959, Frantz detailed the pathology of the lesion.

SPN have been referred to by various names in the literature, including Gruber Frantz tumors, papillary and solid tumors, papillary cystic tumors, solid cystic tumors, epithelial papillary tumors, and even pancreatic endocrine tumors, due to their histological appearance. The exact pathogenesis of SPN remains unclear. Two primary hypotheses exist regarding its origin: one suggests that SPNs arise from pluripotent pancreatic cells, whereas the other proposes an origin from female genital ridge cells [2]. Some studies suggest a higher prevalence in women of reproductive age and a potential genetic predisposition [3][4]. Although it is less common in children than in adults, the detection rate in pediatric patients has increased with advances in medical technology.

The clinical symptoms of SPN are often nonspecific. Patients may present with abdominal pain, nausea, vomiting, a palpable abdominal mass, or other symptoms due to tumor compression of the adjacent organs. Maimaijiang et al. reported that among 18 pediatric patients diagnosed with SPN, 50% were asymptomatic, 38.89% experienced abdominal pain, 22.22% had a palpable abdominal mass, and 11.11% presented with trauma [1].

Biochemical blood tests, including those for glucose, amylase, and lipase, are generally not useful for diagnosing SPN. Tumor markers are usually not elevated, and tumors are typically not associated with syndromes caused by excessive hormone secretion [1]. In our study, most of the patients had normal blood biochemical markers and AFP levels.

On ultrasound, SPNs appear as heterogeneous hypoechoic masses with solid and cystic components and crescent-shaped calcifications [5]. Preoperative diagnosis is primarily based on CT or magnetic resonance imaging (MRI) of the abdomen. CT scans typically show large, well-circumscribed tumors with heterogeneous densities due to hemorrhage and necrosis within the tumor. Differential diagnosis of SPN includes pancreatic pseudocysts, pancreatic adenocarcinoma, mucinous cystic neoplasms, serous cystadenomas, acinar cell carcinomas, and hemangiomas [6]. CT imaging helps assess the relationship between the tumor and surrounding structures and evaluates lymph node involvement and distant metastases. MRI offers higher soft tissue resolution and is superior in evaluating the relationship between the tumor and biliary and pancreatic ducts [7]. Endoscopic ultrasound (EUS) with biopsy is an invasive diagnostic procedure with relatively low complication rates, showing sensitivity and specificity of 56%-71% and 45%-97%, respectively [8][9]. However, few studies have evaluated EUS as a staging tool for pancreatic cystic tumors.

Surgical resection remains the primary treatment for SPN. Preoperative comprehensive assessment is crucial, and surgeons should aim for radical resection while preserving the pancreatic function as much as possible.

SPNs have a low malignancy potential, limited invasiveness, and low recurrence and metastasis rates. Even with metastasis or recurrence post-surgery, patients still have treatment options [11][12]. Central pancreatectomy is indicated for tumors located in the neck or body of the pancreas and has been shown to reduce the risk of exocrine pancreatic insufficiency compared with distal pancreatectomy [1].

For tumors in the tail of the pancreas, spleen-preserving distal pancreatectomy is the preferred choice. The spleen is an important immune organ, and its preservation is crucial for children. In cases where the tumor significantly adheres to the splenic vein, splenectomy may be necessary. Maimaijiang (2022) recommends preserving the splenic artery and spleen whenever possible [1].

For tumors in the pancreatic head, less invasive procedures, such as pylorus-preserving pancreaticoduodenectomy or tumor enucleation with duodenal preservation, can be considered. Lymph node metastasis is rare in SPN, and extensive lymph node dissection is usually unnecessary even when metastasis is present [1]. In our study, one patient with lymph node metastasis underwent primary tumor resection and lymphadenectomy without extensive lymph node dissection, showing no recurrence or metastasis at 14 months post-surgery.

All the patients underwent open tumor resection with pancreatic and duodenal preservation. Intraoperative complications occurred in four patients, with pancreatic duct injury in three and common bile duct injury in one. No postoperative bleeding, bile leaks, pancreatic fistulas, or residual abscesses were observed. According to Beger et al. [13], the overall complication rates are

similar between duodenum-preserving pancreatic head resection and traditional pancreaticoduodenectomy, with significantly lower rates of severe complications, such as pancreatic fistulas and pancreatic insufficiency in the former group.

Radical resection (R0) is considered the definitive treatment for SPN. The role of chemotherapy in SPN is not well established, and adjuvant chemotherapy is generally not required after complete resection. Chemotherapy is reserved for cases with distant metastasis or unresectable tumors [1][7].

The prognosis for SPN is generally favorable. Kato and colleagues [14] reported long-term survival (>10 years) in cases with unresectable tumors, likely due to the slow doubling time of the tumor (765 days). Incomplete resection, large tumor size, intraoperative tumor rupture, and male gender are considered risk factors for recurrence [15][16]. Metastasis and recurrence did not preclude surgical intervention. Current reports indicate an approximate 2% recurrence rate for SPN, which is significantly lower than those reports (5-15%) [3]. The average time to recurrence was 41 months, with most recurrences occurring within the first five years, although 25% occurred after five years. Long-term follow-up is necessary to detect local recurrence or distant metastasis [3].

The limitations of our study include the relatively short average follow-up period and small sample size, necessitating a longer follow-up to evaluate recurrence, metastasis, and postoperative pancreatic function.

CONCLUSIONS

SPN is a rare tumor predominantly found in young females. Its clinical manifestations are nonspecific, often leading to misdiagnosis and oversight. SPN of the pancreas are characterized by low malignancy potential, limited invasiveness, and low rates of metastasis and recurrence. Therefore, surgical treatment is considered a safe and effective method for children with low complication rates.

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CONFLICT OF INTERESTS

The authors declare that there is no conflict of interest regarding the publication of this article.

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None.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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