Case report

Solid pseudopapillary neoplasms of the pancreas in children

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Solid pseudopapillary neoplasms (SPNs) are very rare pancreatic tumors in children. We present clinical presentation, tumor characteristic, surgical management and outcome of SPNs at our institute. There were four patients (3 males, 1 female) undergoing pancreatectomy for SPNs with median age of 11.5 years (10 - 14 years). Presenting symptoms were abdominal pain (n = 4), palpable abdominal mass (n = 2) and vomiting (n = 2). All patients had tumor located in the pancreatic body, and in two of them, the tumor also involved the pancreatic tail. Three patients underwent distal pancreatectomy and one patient underwent central pancreatectomy. The median maximal diameter of tumor was 7.5 cm (3.5 - 12.5 cm). Neither tumor involving surgical margins in all patients nor distant metastasis was detected. There has been no recurrence at the median follow-up period of 32.6 months (1 - 57 months). Although SPNs are rare, they should be considered in the differential diagnosis of pancreatic tumor even in male children. In children, these tumors are usually resectable and pancreatectomy with negative margin provides good prognosis.

Keywords: Solid pseudopapillary neoplasms of the pancreas, children, pancreatectomy.

Solid pseudopapillary neoplasms (SPNs) are uncommon pancreatic tumor that usually occurs in young females. [1 – 4] Although SPNs in children are very rare, it accounts for 22.0% – 68.0% of pancreatic tumors in children and adolescents. [5 – 8] The tumor has low malignant potential and complete surgical resection provided favorable prognosis. We report case series of this rare tumor in children.

Case report

There were four patients with SPNs. All of them underwent pancreatectomy. Three were males and one female. Their median age at diagnosis was 11.5 years (10 - 14 years). All patients presented with abdominal pain (n = 4). The other presenting symptoms were palpable abdominal mass (n = 2) and vomiting (n = 2). All patients had tumors located in the pancreatic body and two of the tumors also involved pancreatic tail. None of them had distant metastasis at presentation. Three patients underwent distal pancreatectomy and one patient underwent central pancreatectomy. The median maximum diameter of the tumors was 7.5 cm (3.5 - 12.5 cm). Pathology reports confirmed SPNs with no tumor involving surgical margins in all patients. There has been no recurrence or distant metastasis at median follow-up period of 32.6 months (1 - 57 months). Details of individual patients are shown in Table 1.

Case 1

A 13-year-old boy presented with 1-year history of palpable abdominal mass with abdominal pain. Computed tomography (CT) scan revealed a mixed solid and cystic mass at the tail of the pancreas. On surgical exploration, a well-circumscribed $8.0 \times 7.0 \times 6.0$ cm^3 tumor was found at the body and tail of the pancreas. He underwent distal pancreatectomy. Pinkish white tumor contained dark brown material with foci of hemorrhage and necrosis was revealed on cut surface (Figure 1). Pathology report confirmed of SPNs.

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Table 1. Clinical presentation, tumor characteristics, surgical procedure and outcome.

Case	Age (years)	Sex	Clinical presentation	Tumor location	Maximum diameter of tumor (cm)	Margin	Surgical procedure, year of operation	Follow-up time (months)	Recurrence	Metastasis
1	13	Male	Abdominal pain, palpable mass	Body, tail	8.0	Negative	Distal pancreatectomy, 2007	56	No	No
2	10	Male	Abdominal pain	Body	3.5	Negative	Distal pancreatectomy, 2011	57	No	No
3	14	Female	Abdominal pain, palpable mass, vomiting	Body, tail	12.5	Negative	Distal pancreatectomy with splenectomy, 2013	1	No	No
4	10	Male	Abdominal pain, vomiting	Body	7.0	Negative	Central pancreatectomy, 2015	9	No	No

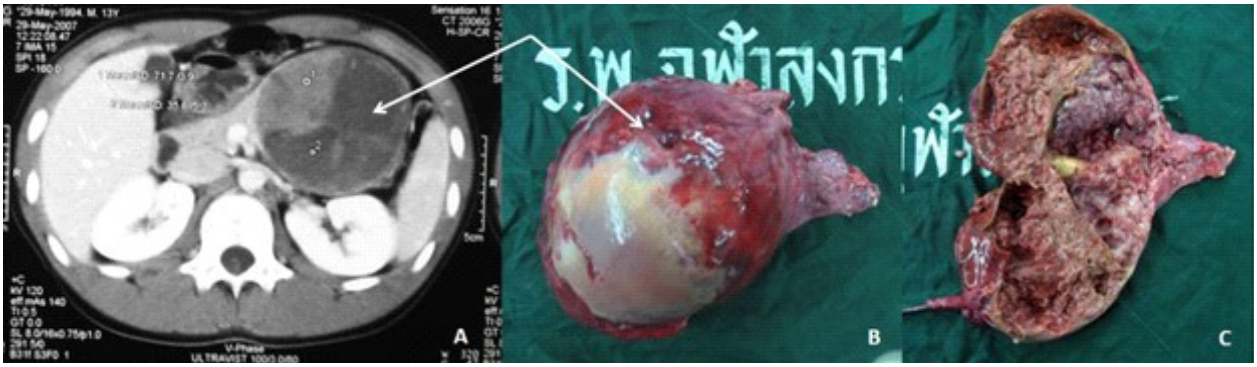

Figure 1. Computed tomography (CT) scan revealed mixed solid and cystic mass at tail of pancreas (A). Pinkish white tumor contained dark brown material with foci of hemorrhage and necrosis (B and C).

Case 2

A 10-year-old boy presented with 1-month of on-and-off abdominal pain. On physical examination, no abdominal mass was detected. Serum amylase was 138 U/L and ultrasonography revealed a 3.6 × 4.0 cm^2 well-defined hypovascular heteroechoic solid mass with few small internal cystic component at the pancreatic body. A well-circumscribed tumor size of 3.5 × 3.5 × 2.5 cm^3 was found during the operation (Figure 2), and the patient underwent distal pancreatectomy with pathology report confirmed of SPNs. The patient discharged from hospital without any symptom of abdominal pain.

Case 3

A 14-year-old girl presented with 1-year dyspepsia and 1-month progressive abdominal pain and vomiting. On physical examination, a large palpable abdominal mass at left upper abdomen was detected. CT scan revealed huge heterogeneous mass that involved pancreatic body and tail. Intraoperative tumor size was 12.5 × 10.0 × 6.5 cm^3 (Figure 3). There was iatrogenic intraoperatively rupture of the tumor capsule. The patient underwent distal pancreatectomy with splenectomy. The duration of hospitalization was 10 days. After 1-month follow up, the patient was lost to follow-up.

Case 4

A 10-year-old boy presented with 1-week epigastrium pain and vomiting. Palpable abdominal mass was detected on the epigastrium. Laboratory test showed anemia. Ultrasonography revealed large a heterogeneous mass at the pancreatic body. CT scan revealed a 7.0 × 6.0 × 5.0 cm^3 irregular thick-walled cystic lesion. On surgical exploration, encapsulated 7.0 × 5.5 × 5.0 cm^3 tumor with papillary-like friable content was located at the pancreatic body (Figure 4). Central pancreatectomy with pancreaticojejunostomy was performed with no post-operative complication.

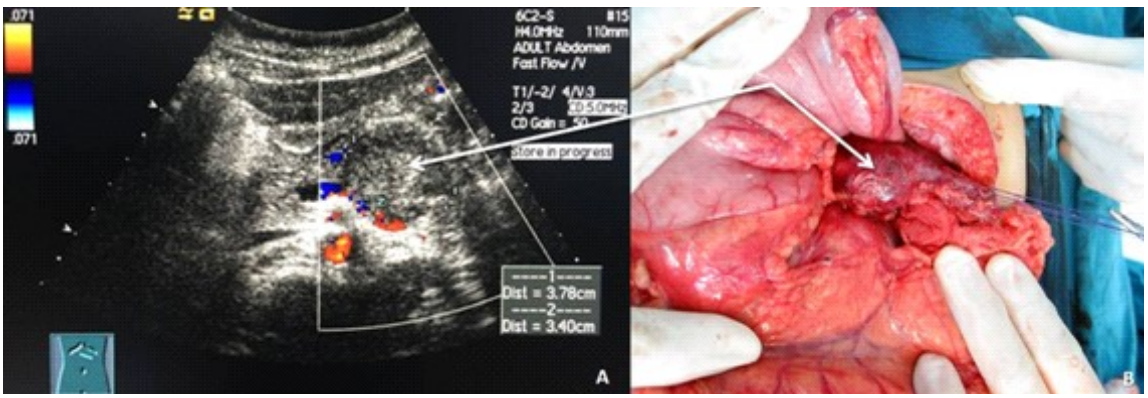

Figure 2. Ultrasonography revealed well-defined hypovascular heteroechoic solid mass with few small internal cystic components at pancreatic body (A). Well-circumscribed tumor size 3.5 × 3.5 × 2.5 cm^3 at the pancreatic body (B).

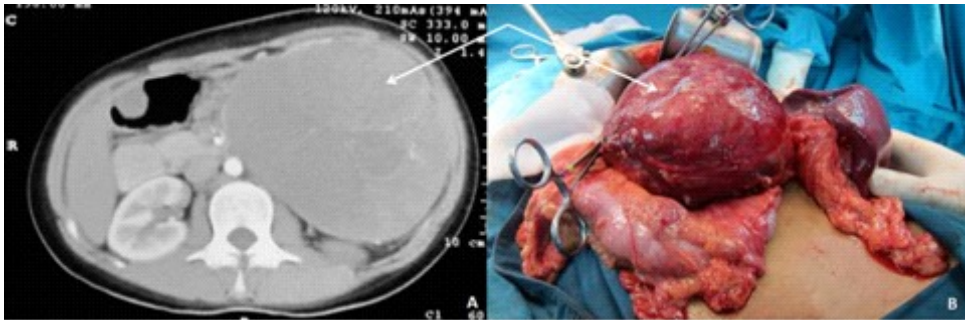

Figure 3. CT scan reveals huge heterogeneous mass that involves pancreatic body and tail (A). Large tumor size $12.5 \times 10.0 \times 6.5$ cm^3 involved pancreatic body and tail (B).

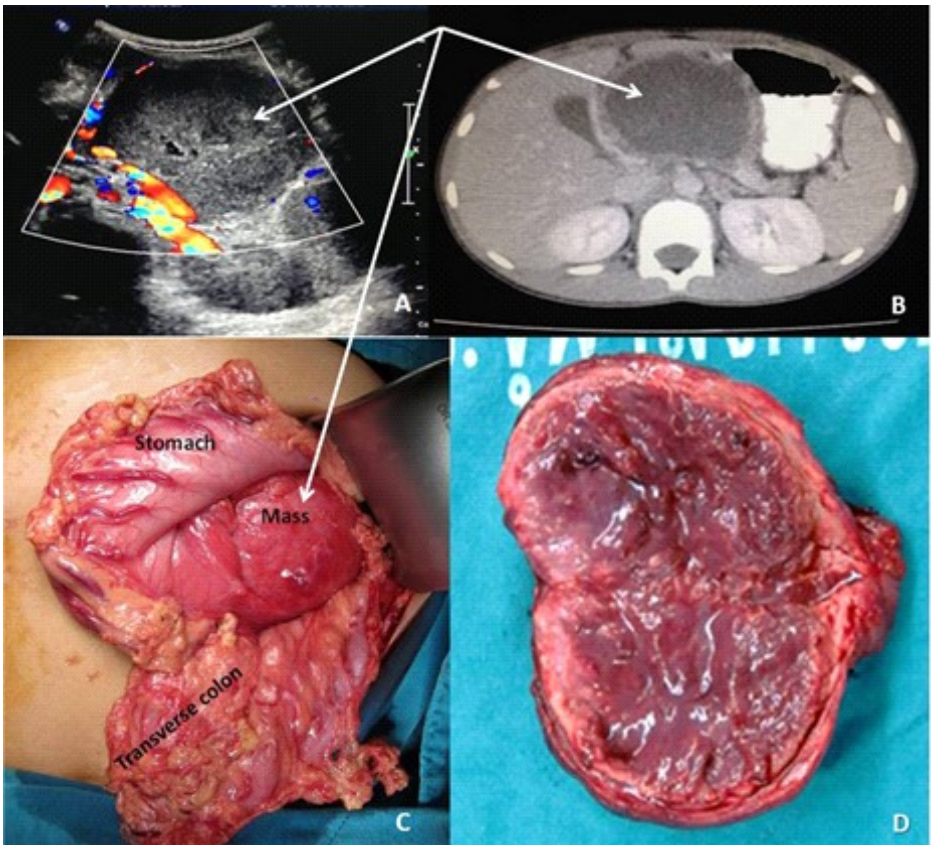

Figure 4. Ultrasonography reveals large heterogeneous mass at pancreatic body (A). CT scan reveals irregular thick walled cystic lesion (B). Encapsulated tumor was located at the pancreatic body (C). Tumor with papillary-like friable contents (D).

Discussion

Most cases of SPNs in children were female with median age at presentation about 12 - 15 years and the most common clinical presentation was abdominal pain. [7, 9–14] Similar to adults, SPNs in children can have malignant behavior. [1 – 3, 8 – 12, 15 - 16] There was the study reported that the high proportion of solid component in these tumors related to the malignancy potential. [12] In addition, malignant SPNs have high risk of recurrence. [1, 12] Surgical resection is recommended for SPNs due to excellent prognosis. [9 – 16] One study reported the good outcome of female children with malignant SPNs even there were positive margins after resection; therefore, radical resection may not be required in these patients. [16]

Although SPNs in male children was very rare, surprisingly, most of patients in our series were male (3/4 patients). There were some studies reported that SPNs in the male could have more aggressive behavior than in the female. [16 – 18] Therefore, SPNs should be considered in the differential diagnosis even in male children with pancreatic tumor. The attempt of complete surgical resection and close follow-up for recurrence and metastasis may be necessary in these patients. In children, there was the report of metastasis malignant SPNs to liver and omentum [12]; therefore, abdominal imaging such as CT scan or MRI might be mandatory in follow-up. Diagnosis of SPNs in children can be delayed due to non-specific presenting symptom, one of our patients was misdiagnosed and treated as dyspepsia for years and presented at our institute with a palpable huge abdominal mass. Furthermore, SPNs could be misdiagnosed as pancreatic pseudo-cyst due to uncertain history of abdominal trauma and cystic component of the mass; in this case, cross-sectional imaging of mixed solid and cystic pancreatic lesion suggests the diagnosis of SPNs.

Conclusion

SPNs are rare tumors in children but they should be considered in the differential diagnosis of pancreatic tumor even in males. In children, these tumors are usually benign but could have malignant behavior especially in male children, complete surgical resection with negative margin should be attempted in these patients. We report that pancreatectomy for SPNs in children is safe and provides good prognosis.

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