Case Report

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Solid-pseudopapillary neoplasm of the pancreas in a 12-year-old female: case report

Kundan Kumar Yadav[a,*], Krishna Yadav[b], Binaya Adhikhari[c], Pratibha Yadav[d], Prashant Yadav[d]

Introduction and importance: A solid-pseudopapillary tumor of the pancreas is a rare solid neoplasm. It is an uncommon exocrine tumor that accounts for 1–3% of exocrine pancreatic tumors.

Case presentation: The authors have reported two cases of solid-pseudopapillary neoplasm of the pancreas with a median age of 12 years, both in females with abdominal pain.

Clinical discussion: Both cases are in females with different treatment modalities. The patient underwent distal pancreatectomy in the first case and enucleation of mass from the distal pancreas in the second case because the size of the tumor is 5 cm and does not involve the bile duct and major pancreatic duct.

Conclusion: The authors discussed two cases of solid-pseudopapillary neoplasm of the pancreas both in a 12-year-old female—both the patients presented with abdominal pain. The patient underwent a different treatment. The diagnosis was confirmed by radiological investigation through non-contrast and contrast CT and supported by histopathology findings.

Keywords: SPN

Introduction

A solid-pseudopapillary tumor of the pancreas is a rare neoplasm first reported by Frenzz in 1959. It was formally named SPN by the WHO in 1996. SPN is a slow-growing, low-grade malignant tumor with a strong female predominance[9]. Metastasis rates range from 10 to 15%. A histologically rare case exhibiting high-grade malignant transformation has been reported, indicating that this disease presents more aggressive behaviors in the same patient[2]. According to the 2019 WHO classification, it is a low-grade malignant tumor and can occur anywhere in the pancreas[1].

Solid-pseudopapillary neoplasm is an uncommon exocrine tumor that accounts for 1–3% of exocrine pancreatic tumors. SPN has an annual incidence rate of 0.2/1 000 000 in children, which is considered a very rare tumor[5]. These tumors generally occur in young females during the second and fourth decades of life. Patients usually present with nonspecific symptoms such as abdominal pain or abdominal mass. Click or tap here to enter text. It is critical to have a proper diagnosis of this tumor since a well-planned excision usually results in a complete cure[3]. The most common imaging appearance of SPN is of well-defined, encapsulated, and large heterogenous tumors, consisting of solid and cystic composites, due to various degrees of intralesional hemorrhage and necrosis[6]. The treatment of choice is a complete surgical resection, which depends on the location of the tumor.

Surgery is the mainstay of treatment if feasible, and performing a limited pancreatectomy is very important to avoid metabolic and endocrine complications (Kumar *et al.*, 2023)[7].

The recurrence rate of this tumor is 2.6–3.5%. prognosis is extremely favorable if followed by a complete resection[4].

HIGHLIGHTS

- SCARE guideline is uploaded as per protocol.
- A separate table and figure is uploaded.
- All necessary comment is uploaded as per your advice.

Case 1 presentation

Twelve-year-old female presented to our institution with a chief complaint of abdominal pain for 3 months, which is acute in onset, and the site of the pain was localized to epigastric regions, which is severe enough to limit her daily activities. She didn't complain of fever, Nausea, or vomiting. Appetite was normal. Bladder and bowel habits are normal. She didn't complain of any surgery in the past, and there has been no medical illness in the past. On

[a]Maharajgunj Medical Campus, Institute of Medicine, Tribhuvan University, Kathmandu, Nepal, [b]Department of Surgery, Maharajgunj Medical Campus, Institute of Medicine, Tribhuvan university, Kathmandu, Nepal, [c]Department of Radiology, Maharajgunj Medical Campus, Institute of Medicine, Tribhuvan university, Kathmandu, Nepal and [d]Kathmandu Medical College and Teaching Hospital, Kathmandu, Nepal

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*Corresponding author. Address: Maharajgunj Medical Campus, Institute of Medicine, Kathmandu, Nepal. Maharajgunj, 44600, Nepal. Tel.: +977 986 429 8610, +977 981 783 9675. E-mail: ykundankumar2055@gmail.com (K. K. Yadav).

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examination General condition is fair and on general examination, there is no pallor, icterus, clubbing, cyanosis, edema, lymphadenopathy, or dehydration. On per abdominal examination, the abdomen is soft, non-tender, and bowel sound is present.

Figure 1A shows the resected specimen macroscopic view. The resected mass was 7×4.7×4 cm in size consisting of part of the pancreas. There is a solid cystic mass measuring 4.5×4×3.5 cm in size. Figure 1B shows the intraoperative mass shown while doing a distal pancreatectomy. Figure 2A shows magnification (× 100, *Hand E*). Tumor cells arranged in sheets, papillae, and nests(represented by a small arrow with a larger head) with areas of hemorrhage (represented by an arrowhead).Figure 2B shows magnification (×400, H and E) Tumors show moderate nuclear pleomorphism with a high N: C ratio (represented by an arrow), where granular chromatin with occasional grooving. Figure 3A shows on-contrast CT abdomen axial image shows an oval heterogenous solid cystic mass in the pancreatic tail. No calcification or fat attenuating areas within the lesion. And Figure 3(B) shows the parenchymal phase of the post-contrast CECT image of the abdomen shows mild enhancement in the solid areas and no enhancement in the cystic areas of the lesion in the pancreatic tail.

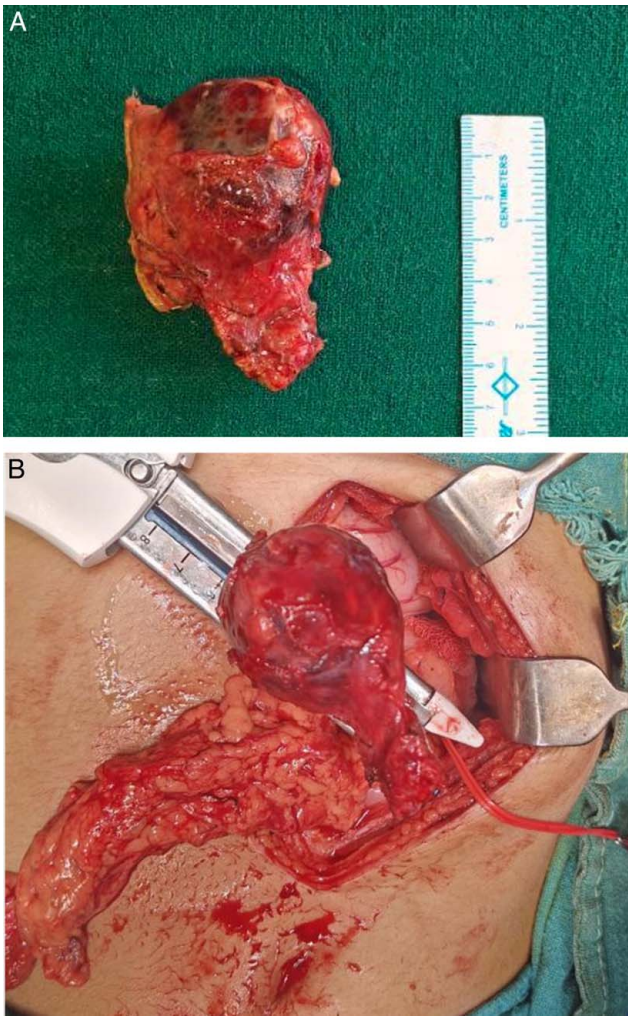

Figure 1. (A) Resected specimen macroscopic view. The resected mass was 7×4.7×4 cm in size consisting of part of the pancreas. There is a solid cystic mass measuring 4.5×4×3.5 cm in size. (B) The intraoperative mass showed while doing distal pancreatectomy.

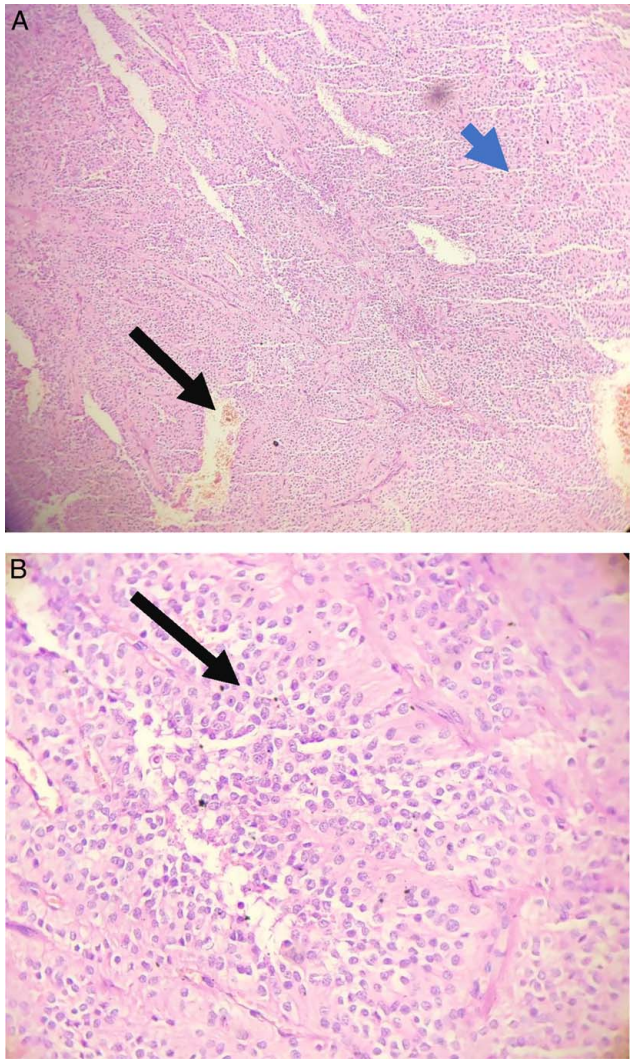

Figure 2. (A) Magnification (× 100, *Hand E*). tumor cells arranged in sheets, papillae, and nests(represented by small arrow with larger head) with areas of hemorrhage(represented by an arrowhead). (B) Magnification (×4oo, H and E). tumors show moderate nuclear pleomorphism with a high N: C ratio (represented by an arrow), where granular chromatin with occasional grooving. H and E, hematoxylin and eosin.

Postoperatively, the patient is stable. Pancreatic amylase tested on 2 days postoperatively and subsequent follow-up for 2 more days shows a decline of pancreatic amylase. One week after POD USG of transabdominal on anterior and posterior shows distal pancreatectomy with no significant collection, and the head of the pancreas appears normal on scan.

At the time of discharge, the patient is tolerating a normal diet and not experiencing pain. Discharged with the advice of normal diet, activity, and regular follow-up on OPD.

Case 2 presentation

A 12-year-old female presented with chief complaints of abdominal pain for 1 month and vomiting for 1 day. Abdominal pain was gradually progressive, continuous, and localized to left

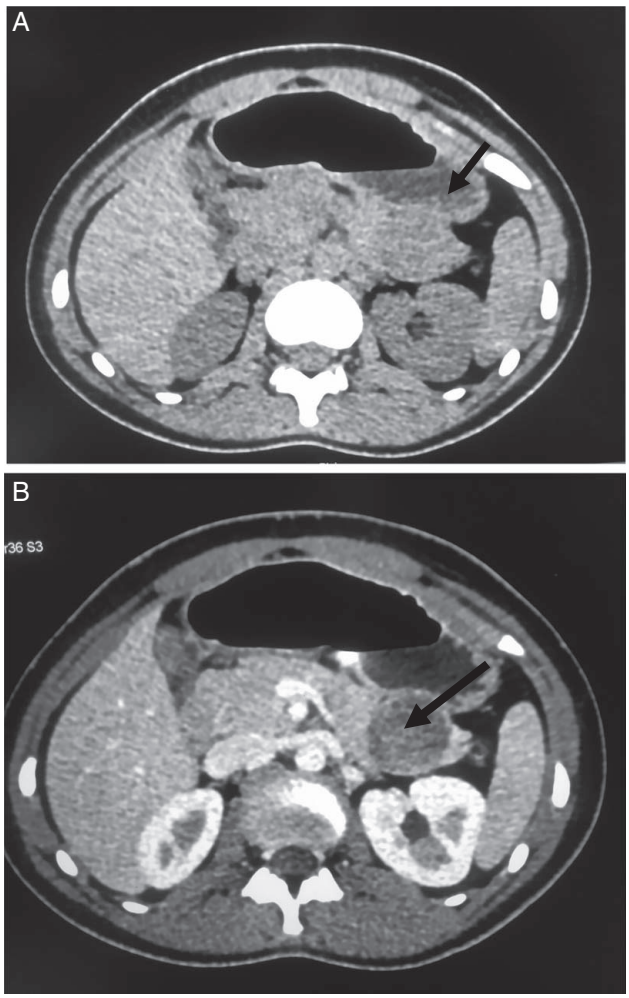

Figure 3. (A) Non-contrast CT abdomen axial image shows an oval heterogenous solid cystic mass in the pancreatic tail (represented by an arrow), no calcification or fat attenuating areas within the lesion. (B) The parenchymal phase of the post-contrast contrast-enhancement CT image of the abdomen shows mild enhancement in the solid areas (represented by an arrowhead) and no enhancement in the cystic areas of the lesion in the pancreatic tail. CT, computed tomography.

lumbar regions, non-radiating, non-referral, with no diurnal variation, and no aggravating and relieving factor. Vomiting was the single episode that was non-bilious, non-blood stained. There is no history of abdominal distension or yellowish discoloration. The bladder and bowel are normal. She didn't complain of any surgery in the past, and there has been no medical illness in the past. On examination General condition is fair and on general examination, there is no pallor, icterus, clubbing, cyanosis, edema, lymphadenopathy, or dehydration. On per abdominal examination, the abdomen is soft, non-tender, and bowel sound is present.

The transabdominal finding of USG shows defined hypoechoic in the regions of the body and tails of the pancreas measuring 43×42×38 mm size with isoechoic septa-like structure with low-level content in the majority part. Figure 4A shows the contrast-enhancement CT abdomen in the sagittal image shows a well-encapsulated oval mass in the pancreatic area (represented by an arrowhead). And Figure 4(B) shows the contrast-enhancement CT shows a well-encapsulated heterogenous mass in the pancreatic tail (represented by an arrowhead).

CECT of the abdomen and pelvis shows well-defined, predominately cystic neoplasm measuring 5×5×4.7 *cm* in size in the body and tail regions of the pancreas. The lesion has caused subtle splaying of the pancreatic parenchyma on either side, hinting at the pancreatic origin of the lesion. On post-contrast study peripheral enhancement of the lesion. Hence the CECT finding shows a cystic neoplasm of the pancreas, which is likely to be a pseudopapillary neoplasm of the pancreas (Table 1).

Histopathology shows circumscribed but unencapsulated tumors with central cystic, cavitary degeneration. The residual tumor is a wall of cavitary lesions that shows tumor cells arranged in sheets, lobules, and pseudopaillae. The cells are uniform with oval nuclei, fine chromatin, occasional prominent nucleoli and nuclear grooves, and a moderate amount of eosinophilic cytoplasm. Clear cells are also seen necrosis is also seen. Calcification, hemorrhage, cholesterol clefts, and foamy macrophage are also seen. Mitosis is infrequent, scanty pancreatic tissue is seen at the periphery of the tumor.

Postoperatively, the patient is stable. Pancreatic amylase was tested on 2 days.

At discharge, the patient tolerates a normal diet and does not experience pain. Discharged with the advice of normal diet, activity, and regular follow-up on an OPD basis.

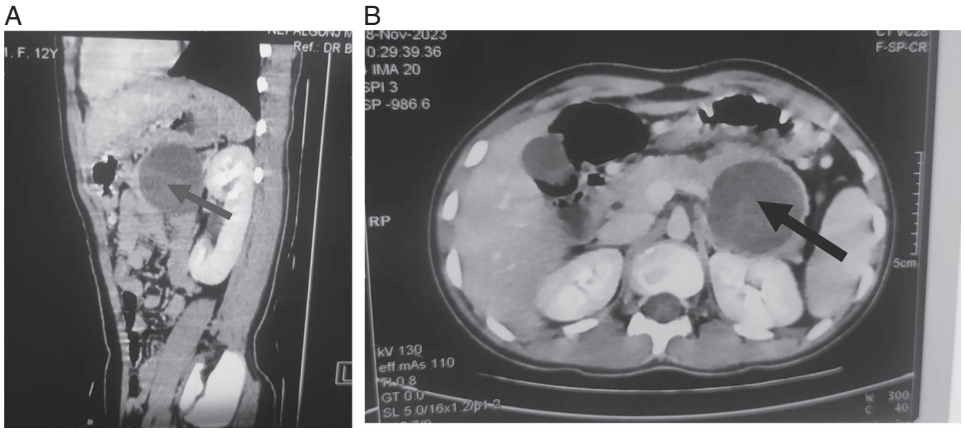

Figure 4. (A) Contrast-enhancement CT abdomen in the sagittal image shows a well-encapsulated oval mass in the pancreatic area (represented by an arrowhead). (B) Contrast-enhancement CT shows a well-encapsulated heterogenous mass in the pancreatic tail (represented by an arrowhead). CT, computed tomography.

Table 1

Comparison of two cases of SPN.

Case	Sex	Age	Clinical presentation	Location in pancreas	Size of tumor	Treatment	Follow-up	Recurrence
1	Female	12 years	Abdominal pain- 3 months	Tail	7 cm	Distal pancreatectomy	No complaints about illness.	No
2	Female	12 years	Abdominal pain -1 months	Body and tail	5 cm	Enucleation	No complaints about illness	No

Discussions

SPN is a rare pancreatic tumor that constitutes 1–3% of all pancreatic neoplasm. It predominately affects young female patients in the third or fourth decades of life. Although SPN is defined as a low-grade malignant pancreatic tumor, it has an excellent prognosis after curative resection[9]. It is composed of monomorphic epithelial cells that form solid and pseudopapillary structures. It is one of the four major pancreatic cystic neoplasms besides intraductal papillary mucinous neoplasm, mucinous cystic neoplasm, and serous cystic neoplasm[1]. Patients with SPN usually lack specific manifestations. Common chief presentations include Nonspecific abdominal pain, discomfort, and incidental detection[4].

Here, we present 2 cases of SPN in children with a median age of presentation of 12 years of age. Our findings are summarized in the table as follows;

We have reported two cases of SPN in pediatrics at 12 years of age. Both cases are in females with different treatment modalities. The first case is treatment with distal pancreatectomy, and the second case is with enucleation of the tumor.

Conclusion

We discussed two cases of solid-pseudopapillary neoplasm of the pancreas both in a 12-year-old female—both the Patients presented with abdominal pain. The patient underwent distal pancreatectomy in the first case and enucleation of mass from the distal pancreas in the second case because the size of the tumor is 5 cm and does not involve the bile duct and major pancreatic duct. The diagnosis was confirmed by radiological investigation through Non-contrast and contrast CT and supported by histopathology findings.

Ethical approval

NA.

Consent

Parental consent for minors; Written informed consent was obtained from the patient's parents for publication and any 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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Author contribution

K.K.Y. is the main principle investing authors and had taken the case in detail and written introduction and about patient presenting parts. K.Y. and P.Y. helped in the methodology parts. B.A. and P.Y. helped in writing discussion.

Conflicts of interest disclosure

The authors declare no conflicts of interest.

Research registration unique identifying number (UIN)

1. Name of registry; None.
2. Unique Identification number or registration ID; None.
3. Hyperlink to your specific registration (must be publicly accessible and will be checked):

Guarantor

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All data are available in manuscript itself.

Provenance and peer review

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