

101859_Auto_Edited-check.docx

WORD COUNT

2059

TIME SUBMITTED

16-DEC-2024 07:35PM

PAPER ID

113557616

Name of Journal: *World Journal of Gastrointestinal Oncology*

Manuscript NO: 101859

Manuscript Type: CASE REPORT

Solid pseudopapillary neoplasm in an adolescent: A case report and review of literature

Sapkota A *et al.* SPN in an adolescent

Abstract

BACKGROUND

Solid pseudopapillary neoplasm (SPN) of the pancreas is a rare epithelial tumor that primarily affects young women. Often asymptomatic or presenting with non-specific symptoms, diagnosing the condition can be difficult.

CASE SUMMARY

This report details the case of a 15-year-old girl who presented with a 2-year history of abdominal pain, with no significant findings during physical examination. An abdominal ultrasound revealed a well-defined heterogeneous solid-cystic mass in the epigastric region, likely originating from the tail of the pancreas. A subsequent contrast-enhanced computed tomography scan indicated a well-defined cystic lesion with an enhancing solid component and capsule in the tail of the pancreas, suggestive of a cystic neoplasm. The patient underwent an open distal pancreatectomy with splenectomy, and histopathological analysis confirmed the diagnosis of SPN of the pancreas.

CONCLUSION

This case highlights the risk of SPN in adolescent girls and the necessity of early diagnosis and intervention for better outcomes.

Key Words: Adolescents; Pseudopapillary neoplasm; Pancreas; Pancreatectomy; Splenectomy; Postoperative pancreatic fistula; Case report

Sapkota A, Paudel R, Pandey S, Bhatt N. Solid pseudopapillary neoplasm in an adolescent: A case report and review of literature. *World J Gastrointest Oncol* 2024; In press

Core Tip: Solid pseudopapillary neoplasm (SPN) is an uncommon pancreatic tumor that primarily impacts young women, especially those in their twenties and thirties.

This report highlights the case of a 15-year-old female who experienced a two-year history of abdominal pain and was diagnosed with a solid pseudopapillary neoplasm localized to the pancreatic tail. The patient underwent an open distal pancreatectomy accompanied by splenectomy, and histopathological examination confirmed the diagnosis of SPN. This case underscores the possibility of SPN occurring in adolescent girls and points out the vital importance of early diagnosis and timely intervention, which can significantly enhance clinical outcomes.

INTRODUCTION

Solid pseudopapillary neoplasm (SPN) of the pancreas is a ³ rare epithelial tumor, representing less than 2% of pancreatic exocrine tumors and below 5% of cystic pancreatic tumors[1]. This neoplasm, first explained by Frantz[2], is distinguished for its slow growing behavior and favorable outcome, particularly affecting young women[3]. While often detected incidentally, SPNs may present with nonspecific symptoms such as abdominal pain or distension[4]. The diagnosis is primarily established through abdominal tomography or magnetic resonance imaging[5]. The primary treatment involves the surgical resection of the tumor, which has 95% of 5-year survival rate[6]. Here, we present the condition of a 15-year-old female patient who presented with a 2-year history of abdominal pain and was diagnosed with a SPN localized to the pancreatic tail, for which she underwent an open distal pancreatectomy and splenectomy.

CASE PRESENTATION

Chief complaints

Left upper abdominal pain for 2 years.

History of present illness

A 15-year-old girl presented to the outpatient department with a complaint of upper abdominal pain, primarily in the left hypochondriac region for 2 years. The pain had an

abrupt onset, was dull aching, and ranged from moderate to severe in intensity, significantly impacting her daily activities. The pain was non-radiating and had no known aggravating or relieving factors. She also reported occasional nausea and multiple episodes of vomiting, which were non-projectile, non-foul smelling, and not stained with blood. The patient gives a history of undocumented weight loss, noticeable by the loosening of her previously fitting clothes. She did not have a history of fever, abdominal distension, fat intolerance, shortness of breath, melena, constipation, jaundice, heartburn, or anorexia.

History of past illness

She had no previous medical or surgical history.

Personal and family history

There was no history of alcohol or drug use, and the family history was unremarkable.

Physical examination

On clinical examinations, her vital and cardinal signs were within normal limits. The abdomen was flat, soft, and non-tender with no guarding or rigidity. There was no organomegaly or any palpable mass. Shifting dullness and fluid thrill were absent. Normal bowel sounds were heard on auscultation.

Laboratory examinations

All preoperative investigations were within normal limits, including serum amylase and serum lipase levels.

Imaging examinations

The patient underwent abdominal ultrasonography, which identified a well-defined heterogeneous solid-cystic mass of size 7.6 cm × 7.2 cm in the epigastric region, seemingly originating from the tail of the pancreas. Notably, the solid component

exhibited demonstrable vascularity raising suspicion for pancreatic solid cystic tumor. A contrast-enhanced computed tomography scan of the abdomen and pelvis was subsequently conducted, revealing a well-defined cystic lesion with an enhancing solid component and capsule, in the region of the tail of the pancreas. The lesion was giving a “claw[7] sign with the pancreas” confirming its intraparenchymal origin. The lesion was observed to be in close proximity to several structures: It abutted the posterior surface of the stomach superiorly, the left kidney, and the left splenic vessel posteriorly, and it displaced the bowel loops anteriorly. These findings were suggestive of a likely cystic neoplasm of the pancreas.

FINAL DIAGNOSIS

The final diagnosis of solid pseudopapillary epithelial neoplasm of the pancreas was confirmed by histopathological reports.

TREATMENT

Surgical resection of the mass was planned, and an open distal pancreatectomy with splenectomy was carried out under general anesthesia. During the procedure, a well-encapsulated solid-cystic mass measuring 7 cm × 7 cm × 8 cm was identified, originating from the tail of the pancreas. An area of whitish patchy tissue was observed anteriorly over the lesion. There were no indications of omental, mesenteric, peritoneal, or hepatic deposits, and there was no regional lymphadenopathy or ascites. The entire tumor was successfully resected and sent for histopathological examination. Postoperatively, the patient exhibited mild leukocytosis with a white blood cell count of 16780/mm³, predominantly composed of neutrophils (86%). Additionally, there was a slight elevation in prothrombin time at 17 seconds and an international normalized ratio of 1.21. On the third postoperative day, a significant increase in serum amylase levels was noted, reaching 1742 U/L, suggestive of a postoperative pancreatic fistula, which gradually resolved on its own.

OUTCOME AND FOLLOW-UP

The patient successfully recovered from the surgery and was discharged from the hospital. She has been kept under regular follow-up to monitor for any potential recurrence of the tumor.

DISCUSSION

The SPN of the pancreas is a very rare condition, primarily affecting young women in their second or third decades of life, nonetheless, it can also occur in children[6]. The significant predominance of females has been suggested to result from the closeness of primitive pancreatic cells to the ovarian ridge during the early stage of development[8]. The clinical features of a SPN of the pancreas are nonspecific and may include unusual abdominal pain, abdominal mass palpable during a clinical examination, or an incidental finding during imaging studies conducted for other purposes[9]. As the tumor is typically found in either the head or tail of the pancreas[6], its growth may lead to symptoms associated with the compression of nearby digestive structures, bile ducts, or blood vessels[10]. In some cases, the tumor may be detected following a spontaneous bleeding episode or as a result of bleeding due to abdominal trauma[11].

Despite the advancements in diagnostic techniques, accurately diagnosing SPNs preoperatively remains a clinical challenge[12]. This is due to their radiological and clinical similarities with variety of differential diagnoses, such as pseudocysts, pancreatic mucinous neoplasms, well-differentiated ductal adenocarcinoma, pancreatic endocrine neoplasms, and acinic cell carcinoma[13]. In children, pancreatic tumors that arise from secondary origins, such as neuroblastoma, leukemia, lymphoma, and lymphoproliferative disorders, are more commonly observed[6].

A contrast-enhanced computed tomography scan of the abdomen is considered the most effective imaging modality for SPNs as it offers comprehensive information about tumors including their origin, size, and configuration, as well as details regarding the local invasion and the presence of metastasis[6]. The encapsulated lesions, characterized by a mixture of solid and cystic components display both enhancing and non-enhancing

areas and often contain intra-tumoral calcifications[6]. In addition, hemorrhage can arise from the growth of the tumor, that results into internal degeneration[11]. Thus, a mass covered with a capsule, containing both cystic and solid components along with intra-tumoral hemorrhage distinguish SPNs from their other malignant tumors[6,14,11]. Macroscopically, SPNs are typically large, well-circumscribed tumors with a diverse appearance, often exhibiting areas of necrosis, hemorrhage, and cystic degeneration, which can be attributed to vascular ischemia[15]. Pseudo-papillae are created when tumor cells detach from blood vessels and develop fibrovascular stalks or rosette-like structures[16].

Microscopically, SPNs are typically characterized by solid sheets of neoplastic cells, which are often interspersed with regions where the tumor cells are organized around the fibrovascular cores[17]. A distinctive feature of these tumors is the presence of Periodic Acid-Schiff-positive hyaline globules[18]. Other histological characteristics include the presence of pseudo-papillary architecture, microcystic changes, clear cells, nuclear grooves, eosinophilic cytoplasm, myxoid stroma, and hyaline globules. Additional features such as atypical cells, tumor giant cells, mitotic activity, calcification, cholesterol clefts, fibrosis, hemorrhage, infarction, and tumor necrosis might also be noted during histological evaluation[18].

To complement the histological findings and to differentiate SPNs from histological mimics, immunohistochemical staining is crucial. A panel of commonly used immunohistochemical markers includes CD10, CD56, beta-catenin, cyclin D1, CD99, cytokeratins, chromogranin A, synaptophysin, and progesterone receptor[17]. All patients with SPN possess activating somatic mutations in the β -catenin gene (catenin beta1, located on chromosome 3p)[19]. Increased expression of certain proteins involved in Wnt signaling, such as Dickkopf-related protein 4, along with other proteins like non-POU domain-containing octamer-binding and Fused in sarcoma, that interact directly with β -catenin, are upregulated in solid pseudopapillary tumor[20]. Moreover, nine metabolic proteins such as SLC25A13, glycosylphosphatidylinositol, phosphoglycerate kinase 1, hexokinase 1, enzyme enolase 2, pyruvate dehydrogenase E1 component

subunit beta, ALDH7A1, pyruvate kinase M2, and DLD are overexpressed[21]. But in our case, due to the limited resources available, the diagnosis was largely made based on the typical clinical presentation, imaging results, and histopathological features, which are often sufficient to confirm the diagnosis in such situations.

The mainstay of treatment of SPNs remains the surgical removal of the tumor[6]. Depending on the location, procedures such as distal pancreatectomy with or without splenectomy, pylorus-preserving pancreatoduodenectomy, Whipple procedure, or enucleation may be carried out[22]. Small tumors located away from the main pancreatic duct are enucleated, those located on the head or uncinate process of the pancreas are treated with pancreatoduodenectomy, while central pancreatectomy is carried out for the tumors of the neck or body of the pancreas, without vessel involvement[23]. The surgical resection must be performed carefully, as the spillage of the tumor contents can lead to the implantation of the tumor cells into the peritoneum[13,24].

Studies show that the ² patients who had limited resection with microscopically positive margins showed outcomes similar to those who underwent extensive surgery with R0 resection[25]. Thus, longevity can be attained with minimally invasive procedures[26], even in patients with advanced or metastatic disease[27]. Moreover, chemotherapy (primarily 5-fluorouracil and gemcitabine) and radiotherapy have been reported to be effective treatment modalities in a limited number of patients[28]. Other modalities such as radiofrequency ablation[29], transcatheter arterial chemoembolization with pharmorubicin and iodized oil[30], and selective internal radiotherapy[31] have also been found to be suitable for inducing long-term remissions of the strongly vascularized liver metastases. In a study done by Dovigo *et al*[32], liver transplantation was done and found to have survival free of recurrence at 1 year later.

The reported recurrence rate after surgery lies between 3%-9%[14]. According to a study done by Serrano *et al*[33], recurrence commonly occurred 5 to 7 years after surgery, indicating that a follow-up period of more than 5 years is necessary with routine imaging, especially for patients with lymphatic and vascular invasion,

metastases, and probable tumor capsule invasion. Another meta-analysis reported a recurrence rate of 2% following resection, highlighting that male patients, those with positive lymph nodes, and individuals with lymphovascular invasion were at an increased risk for recurrence[34]. The patient in our case underwent surgery on April 24, 2024, and is under regular follow-up since then.

CONCLUSION

In conclusion, this case highlights the significant possibility of SPN occurring in adolescent girls, which is often overlooked in clinical practice. It underscores the importance of vigilance and thorough assessment in adolescent patients, ensuring that conditions like SPN are not missed and that appropriate care is provided promptly. Early intervention can significantly alter the course of the disease, leading to more favorable results and ultimately improving the quality of life for young patients.

3%

SIMILARITY INDEX

PRIMARY SOURCES

- 1** www.ncbi.nlm.nih.gov 32 words — 1%
Internet
 - 2** Li You, Feng Yang, De-Liang Fu. "Prediction of malignancy and adverse outcome of solid pseudopapillary tumor of the pancreas", *World Journal of Gastrointestinal Oncology*, 2018 17 words — 1%
Crossref
 - 3** Taoufik El Abbassi, Abdessamad Elazhary, Mohamed Ouchane, Anas El Wassi, M. Rachid Lefriyekh. "Solid pseudopapillary tumors of the pancreas in young women: Case report", *International Journal of Surgery Case Reports*, 2021 17 words — 1%
Crossref
-

EXCLUDE QUOTES ON

EXCLUDE BIBLIOGRAPHY ON

EXCLUDE SOURCES < 15 WORDS

EXCLUDE MATCHES < 15 WORDS