Scholars Journal of Applied Medical Sciences
Abbreviated Key Title: Sch J App Med Sci
ISSN 2347-954X (Print) | ISSN 2320-6691 (Online)
Journal homepage: https://saspublishers.com/sjams/

OPEN ACCESS

Gastrointestinal Surgery

Portal Vein Reconstruction in Solid Pseudopapillary Epithelial Tumor of Pancreas - Case Series and Review of Literature

Dr. Ajay Sharma[1*], Dr. Anjali Sharma[2], Dr. Rudhee Sehgal[3], Dr. Peeyush Varshney[4], Dr. Rajneesh Meena[5], Dr. Priyamvada Gupta[6], Dr. M L Yadav[7]

[1]Associate Professor, Department of Gastrointestinal Surgery, Mahatma Gandhi Medical College Jaipur, Ricco Industrial Area, Sitapura, Jaipur, Rajasthan 302022, India
[2]Head of Department, Department of Pathology, Bhagwan Mahavver Cancer Hospital, Jaipur, Jawahar Lal Nehru Marg, Bajaj Nagar, Jaipur, Rajasthan 302017, India
[3]Final Year Student of Department of General Surgery, Mahatma Gandhi Hospital Jaipur, Rajasthan, India
[4]Assistant Professor, Department of Gastrointestinal Surgery, Mahatma Gandhi Medical College Jaipur, Ricco Industrial Area, Sitapura, Jaipur, Rajasthan 302022, India
[5]Consultant, Department of General Surgery, Apex Ranthambore Sevika Hospital, Sawaimadhopur, Rajasthan, India
[6]Professor Anaesthesiology, Mahatma Gandhi Hospital Jaipur, Rajasthan, India
[7]Professor and Head of the Department, Department of Pathology, Mahatma Gandhi Medical College, Ricco Industrial Area, Sitapura, Jaipur, Rajasthan 302022, India

DOI: 10.36347/sjams.2020.v08i12.043 | **Received:** 12.11.2020 | **Accepted:** 16.12.2020 | **Published:** 30.12.2020

***Corresponding author:** Dr. Ajay Sharma

Abstract **Original Research Article**

Solid Pseudopapillary Epithelial Neoplasm (SPEN) are rare pancreatic tumours, which have generated curiosity of surgeons due to their variable biologic nature. We report three cases of SPEN in young females presenting with hematemesis, nausea, anorexia, abdominal pain and weight loss. Based on computed tomography each of these cases were found to have the neoplasm in pancreas with portal vein (PV) involvement. SPEN is a benign tumour with only 5% displaying malignant features [1]. All three cases were managed by Whipple's pancreaticoduodenectomy with PV resection and reconstruction. Histology and immunohistochemistry confirmed the diagnosis of SPEN in all cases. All cases were followed up and after one year all revealed a patent PV graft with normal flow pattern and no recurrence of disease. Since SPEN has low malignant potential with a favourable prognosis, aggressive management should be pursued. PV invasion by the lesion is neither a contraindication for surgery, nor does it indicate poor prognosis. SPEN with vascular invasion can be managed safely with vascular resection and reconstruction and has a good prognosis.

Keywords: Tumor, Vein Reconstruction, Epithelial Neoplasm.

INTRODUCTION

Virginia Kneeland Frantz in 1959, was the first person to describe solid pseudopapillary epithelial neoplasms (SPEN) or Frantz tumours as benign neoplasms of the pancreas [2]. These pancreatic tumours were known by various names till 1996, when the World Health Organization (WHO) gave the official nomenclature of solid pseudopapillary tumours [3].

SPEN is seen more commonly in females in second to third decade of their life. They are a rare pancreatic tumor with only 1-2% of all pancreatic neoplasms. Usually a benign disease but malignant SPENs of the pancreas are defined by the WHO as those having angio-invasion, perineural invasion, or deep invasion of surrounding pancreatic parenchyma [2]. SPEN tumour is known to have low malignant potential, but 10-15% of these tend to infiltrate into surrounding structures [4].

Correct management for infiltrating variant of SPEN still is not yet clearly defined in current literature.

We present three cases of locally aggressive SPEN with PV infiltration managed by Whipple's pancreaticoduodenectomy with en bloc PV resection and reconstruction via three different methods.

Case Series

Case 1

A 27-year-old female presented with complaints of hematemesis, nausea, occasional vomiting, anorexia and weight loss for one month. There was no associated history of fever, jaundice and

an abdominal lump or prior abdominal surgery. Ca 19-9 and carcinoembryonic antigen (CEA) were within normal limits.

Contrast-enhanced computerised tomography (CECT) of the abdomen demonstrated a well-encapsulated mass in the head of the pancreas with both cystic and solid components along with areas of cystic degeneration encasing the extrahepatic PV (Figure 1A & 1B). Endoscopic Ultrasonography (EUS) guided Fine needle aspiration cytology (FNAC) had shown the changes suggestive of SPEN.

At surgery a large hard mass of size 10.2 X 15 cm involving the head of the pancreas infiltrating into the PV, superior mesenteric vein, splenic vein, the root of the mesentery and transverse mesocolon was found. Right hepatic artery and superior mesenteric artery were free from tumour. There were few enlarged lymph nodes at porta hepatis, and along common hepatic artery.

Standard technique for pancreaticoduodenectomy [5] was done until the tumour was attached only at the site of vascular infiltration. The splenic vein was ligated without an accompanying splenectomy. The tumour was resected en bloc along with the involved portion of the PV after taking proximal and distal control with a total clamping time of 32 minutes. An IMPRA®(BARD, UK) ePTFE vascular graft-10 mm x 40 cm was used for reconstruction and was anastomosed proximally to the PV (Figure-2) and distally to the SMV (Figure-3). A colour doppler was done to confirm the patency and adequate flow of PV intraoperatively. Inj Heparin was started intraoperatively and continued postoperatively, till the patient was on normal oral diet when the oral anticoagulant treatment was resumed. The postoperative course was uneventful and the patient was discharged on a postoperative day seven.

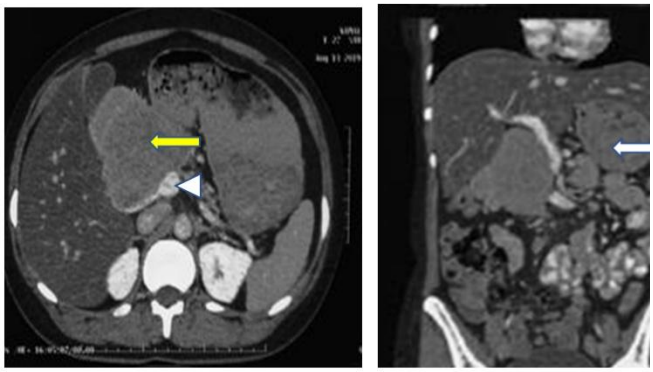

Fig-1 (A & B): Axial and coronal CT scan images large pancreatic mass with a heterogeneous enhancing lesion (yellow arrow) incasing the portal vein (arrow head). Figure 1B shows that portal vein (white arrow) is displaced and tortuous and shows mild irregularity in lumen

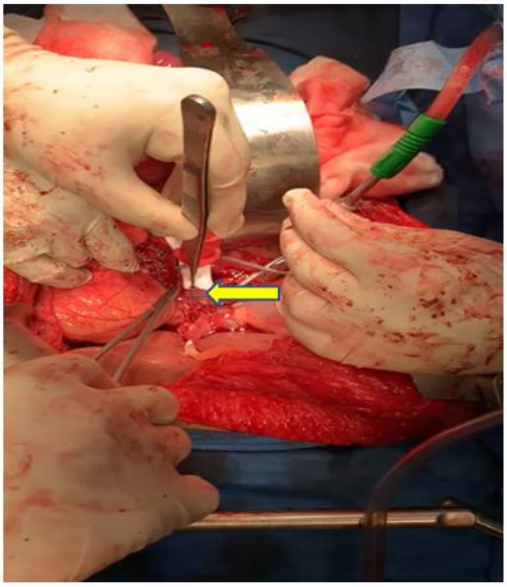

Fig-2: Proximal Reconstruction of the portal vein (yellow arrow)

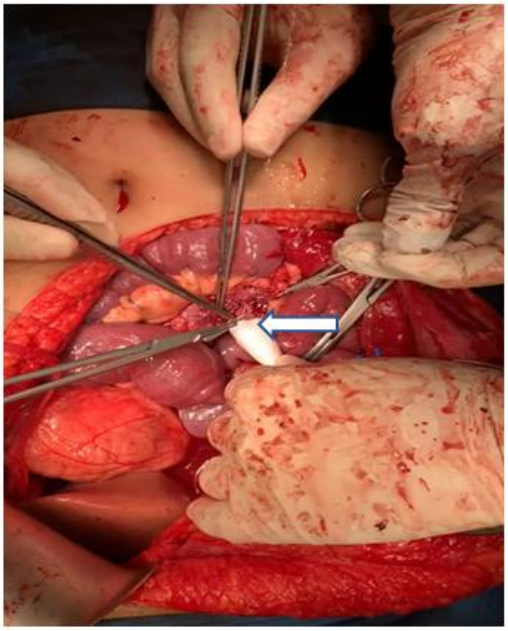

Fig-3: Distal Reconstruction of the portal vein (white arrow)

Case 2

A 26-year-old female presented in our outpatient department with the symptoms of abdominal pain, recurrent vomitings, anorexia, and weight loss for the past three months. Clinical examination did not reveal any specific findings. CECT scan of the abdomen revealed a 4.5 X 5 X 5.7 cm heterogeneous lesion seen in the uncinate process of the pancreas with solid and cystic areas with circumferential involvement of SMV for a length of 2 cm. SMA was closely abutting the mass. Posteriorly it is causing mass effect on left renal vein, IVC and second and third part of duodenum. Inferior pancreato-duodenal artery was involved by tumor and had a loss of plane. Pancreatic duct (PD) was not dilated and Common bile duct (CBD) is of normal

size. EUS guided FNAC was suggestive of SPEN. The operative procedure carried out was Whipple's PD with PV reconstruction using autologous internal jugular vein graft. Intraoperatively done and Post operative Doppler study revealed a good flow in the PV. She was also started on Heparin intraoperatively and oral anticoagulant at discharge.

Case 3

A 30-year-old female presented with nausea, pain in the epigastric region and anorexia for one year. Ultrasonography (USG) of the abdomen revealed a 5 X 4 cm hypoechoic mass arising from the pancreas. CECT scan of the abdomen showed a 5.9 X 4.6 cm mass in the head and uncinate process of the pancreas with a few hypodense non enhancing cystic areas.The mass lesion was closely abutting the PV for around 180 degrees. SMA was free from the mass. It is closely abutting 2nd and 3rd part of duodenum and anterior cortex of right kidney. It is causing mass effect on IVC, Right renal vein and superior mesenteric vein. Adjacent mesenteric and peri-pancreatic nodes were also enlarged. PD and CBD were not dilated. A few thin branches of SMA were seen supplying the lesion. Considering CT findings and EUS guided FNAC report of the patient a provisional diagnosis of SPEN was made. Whipple's PD with PV reconstruction in form of portal sleeve resection and primary repair was done. Intra-operative Doppler showed good flow and retained dimentions of PV at the site of repair. She was discharged on POD 7 after an uneventful postoperative period.

Histopathology and Immunohistochemistry

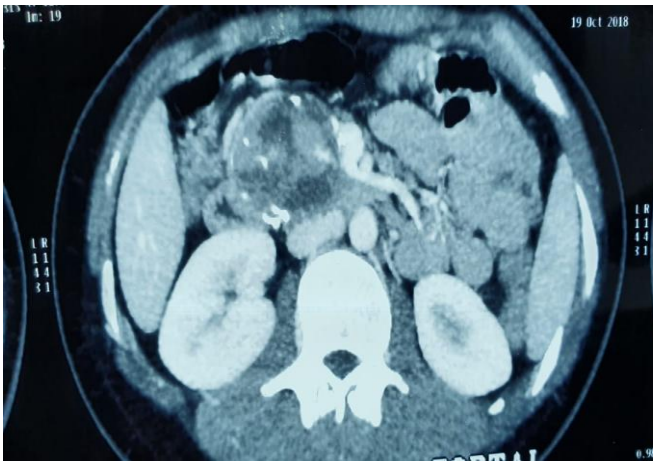

Fig-4: Axial CT scan image shows large pancreatic mass with a heterogeneous 4.5 cm X 5.5 X 5.7 cm heterogeneous lesion seen in the uncinate process of the pancreas with solid and cystic areas

Leeds protocol [6] was used for all histopathology reporting. The biopsy of all three cases on gross pathology showed a pancreatic tumour partially demarcated by a fibrous capsule (Figure-5 image of case one). The cut surface showed a grey-white soft friable mass. Discontinuous tumour deposits were seen in pancreaticoduodenal fat in case one. There was an infiltration of the PV but no lymphatic, vascular or perineural invasion was present in all three cases. The four lymph node examined were free of metastatic deposits. Resected margins were free from the tumor in all three patients.

Microscopic examination in all cases had tumour cells arranged in solid sheets, nests (low power) Figure 6A. Small round cells with round to oval nucleus and granular chromatin with eosinophilic cytoplasm (high power) in Figure 6B**.**

Immunohistochemistry of all three cases showed that the tumour cells had immunopositivity for synaptophysin, PR, vimentin, dim positive for CD10 & CD 56 and dim focal positive for chromogranin. Beta-catenin shows cytoplasmic positivity and Pan CK shows dim cytoplasmic positivity. Ki index was less than 5% in all three. The tumour cells were immune-negative for CA19.9, CK19, E-Cadherin.

Based on these pathological and immunohistochemical findings, the diagnosis of a solid pseudopapillary epithelial tumour of the pancreas was confirmed in all three cases.

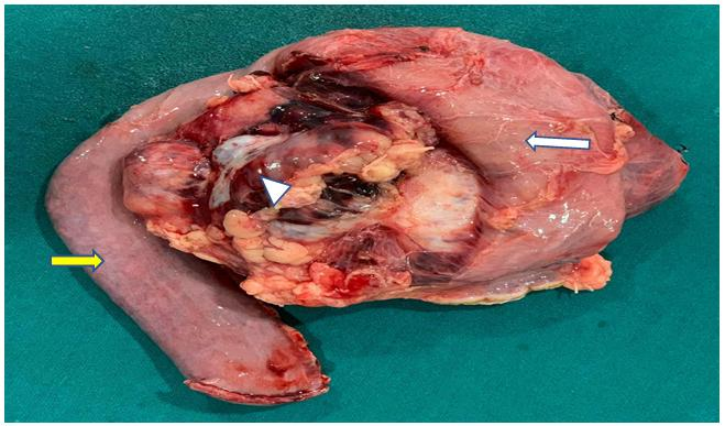

Fig-5: Whipple's specimen demonstrates duodenum (yellow arrow), pancreas (white arrow) and the tumour present at the head of the pancreas (arrow head)

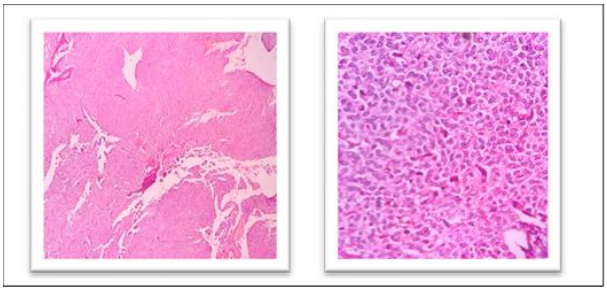

Fig 6(A & B): Microscopy showed 4X image of tumor cells in solid sheet and nest of poorly cohesive cells around blood vessels) in Figure 5A. Figure 5B shows 40x image of small round cells with round to oval nucleus and granular chromatin with eosinophilic cytoplasm

Follow Up

Follow-up included clinical examination, routine laboratory tests, Doppler sonography and CECT abdomen every 6 months. During the follow up two patients had fatty food intolerance, gastritis, and frequency of defecation. One patient was asymptomatic during the post operative follow up. All had normal laboratory routine investigations reported. Doppler and CECT showed a patent PTFE graft with the normal flow in PV in all three cases (Figure-7). There was no evidence of recurrence of disease seen in follow up to 12 months post surgery.

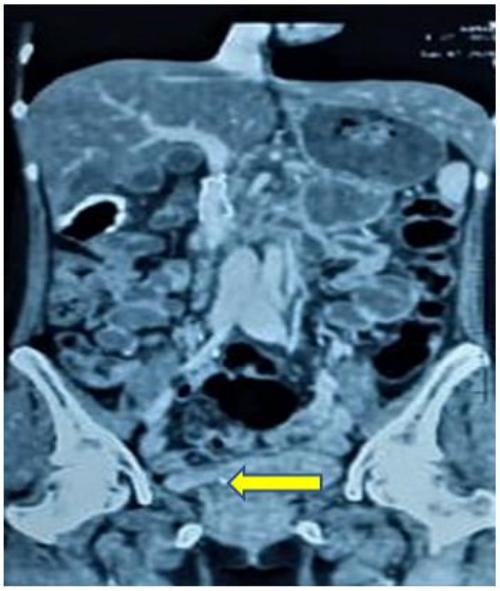

Fig-7: Coronal CT scan images showing patent PTFE graft with normal flow (yellow arrow)

DISCUSSION

Solid-pseudopapillary is an indolent tumour of the pancreas with a distinct (>90%) female preponderance [4]. Usually the tumor has a benign course but it can point towards malignancy if there is lymphovascular or perineural invasion or spread to the surrounding organs [7].

Multiple theories have been suggested for the pathogenesis of the tumour but the cellular derivation is still unclear. One of the theories suggested by Abraham was mutation of Beta-catenin played a role in tumour genesis of SPEN [8]. Another theory by Tanaka had diffuse cytoplasmatic and nuclear positivity for BETA-catenin as the cause [9].

The clinical presentation varies with each patient. Mostly the patients are asymptomatic and the diagnosis is made incidentally on imaging studies [7]. Although some of them can present with abdominal pain, abdominal distension, palpable abdominal mass and nausea [10]. A review of 34 patients with SPEN showed that 58.8% (20 patients) were asymptomatic and 41.2% displayed symptoms like abdominal pain, nausea, abdominal distension and abdominal lump [10].

Kato *et al*., [9] using Schwartz [11] formula showed the tumor doubling time of the SPEN tumour to be 765 days. This can be once of the cause the tumor being asymptomatic for a long time. More over such long tumor doubling time with low risk of metastasis may support long term behaviour after aggressive surgical management of the tumor.

SPEN can occur in different sites of the pancreas. A large review of 718 cases was conducted which displayed the common sites of tumour to be localized to the tail(35.9%) and head 34 % [12]. Less common sites included the body(14.8%), 1% in the neck and 0.43% in the uncinate process of the pancreas. The extrapancreatic localization consisted of 1.01% [12].

Diagnosis of SPEN is generally made by CT or MRI imaging. Typically, characteristics like a large, well-circumscribed, heterogeneous mass with varying solid and cystic components are seen. It may also demonstrate a peripheral capsule and occasional intralesional calcifications [13].

EUS with FNA has recently been advocated as a useful diagnostic tool in SPEN. The diagnostic accuracy of EUS with FNA was found to be 75% in a multicentric experience conducted by Jani in 2008 [14]. More recently, a research study done by Law [15] showed that addition of EUS-FNA to a preoperative workup of SPEN significantly increased the diagnostic yield to 82.4%.

Papavramidis T demonstrated the involvement of PV in 26 patients (5%) of 497 total patients operated for SPEN. The percentage of involvement of other organs like spleen, colon and duodenum was seen to be 9% [12].

Cheng found that a favourable outcome can be achieved if a case of SPEN having vascular invasion was managed appropriately. His study had 8 patients

with SPEN infiltrating the portal mesenteric vein, who underwent pancreatectomy associated with vascular resection. Out of 8 patients, 7 were alive and free of disease after a median follow up 67.5 months and one patient died of liver metastasis [16].

Baltreme published a case report of a SPEN with vascular invasion requiring pancreaticoduodenectomy and PV reconstruction by internal jugular vein graft. This patient had a survival of 77 months [17].

In 2008 Cosimo sperti diagnosed SPEN tumour that invaded a long segment of the portal-mesenteric vein confluence. Total pancreatectomy with resection of the PV and reconstruction with the internal jugular vein was carried out. The patient developed liver metastasis 32 months after the surgery [4].

A research study of SPEN management done by M. Frost [18] had 1 out of the total 21 patients having vascular invasion. This patient underwent a total pancreatectomy, splenectomy, PV resection with interposition Dacron graft. Post-operatively patient developed graft thrombosis, portal hypertension and oesophageal varices for which band ligation was done [18]. The follow up of two years showed no signs of recurrence.

In summary, our case series describes three extremely rare cases of SPEN with vascular invasion which was successfully managed by PD with PV reconstruction. All three patients after a mean follow up of 24 months had a patent PV graft with no evidence of recurrence of disease with a good general condition of the patients. Therefore, we conclude that SPEN tumour having vascular invasion should not act as an obstacle for the surgeon as it pertains a good resectability with a long-term prognosis.

Funding: Self-funded.

Availability of Data and Material

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Competing Interests: There are no competing interest in our research.

Consent for Publication

All the participants were given prior knowledge about publishing the results after the end of the research.

REFERENCES

1. Mirminachi B, Farrokhzad S, Sharifi AH, Nikfam S, Nikmanesh A, Malekzadeh R, Pourshams A. Solid pseudopapillary neoplasm of pancreas; a case series and review literature. Middle East journal of digestive diseases. 2016 Apr;8(2):102-108.
2. Go VL. Tumors of the Pancreas (Atlas of Tumor Pathology Series 4). Pancreas, 2007; 35:388.
3. Klöppel G, Solcia E, Longnecker DS, Capella C. Histological Typing of Tumours of the Exocrine Pancreas. Google Books. https://books.google.mw/books?id=WJ0OswEACAAJ&source=gbs_book_other_versions_r&cad=4.
4. Sperti C, Berselli M, Pasquali C, Pastorelli D, Pedrazzoli S. Aggressive behaviour of solid-pseudopapillary tumor of the pancreas in adults: a case report and review of the literature. World journal of gastroenterology: WJG. 2008 Feb 14;14(6):960-965.
5. Warshaw AL, Thayer SP. Pancreaticoduodenectomy. Journal of Gastrointestinal Surgery: official journal of the Society for Surgery of the Alimentary Tract. 2004 Sep;8(6):733-741.
6. Carlin PS, Aguirre EP, del Cid EA, Botella AG, Valladares LD, Garcia AT. Implementation of Leeds Protocol in Duodenopancreatectomies for Pancreatic Adenocarcinoma and Impact of Resection Margins in the Survival. HPB. 2016 Apr 1;18:e778-9.
7. Paluri R, Babiker HM. Cancer, Solid and Papillary Epithelial Neoplasm (SPEN). StatPearls (StatPearls Publishing, 2019).
8. Abraham SC, Klimstra DS, Wilentz RE, Yeo CJ, Conlon K, Brennan M, Cameron JL, Wu TT, Hruban RH. Solid-pseudopapillary tumors of the pancreas are genetically distinct from pancreatic ductal adenocarcinomas and almost always harbor β-catenin mutations. The American journal of pathology. 2002 Apr 1;160(4):1361-9.
9. Volkan A, Olca B. Surgical Pathology of the Pancreas, Surgical Pathology Clinics. Google Books. https://books.google.co.in/books?id=_Pox2riPhvoC&pg=PA588&lpg=PA588&dq=Tanaka+Y,+Kato+K,+Notohara+K,+et+al.+Frequent+-catenin+mutation+and+cytoplasmic/nuclear+accumulation+in+pancreatic+solid+pseudopapillary+neoplasm&source=bl&ots=QlKUr0PZ3J&sig=ACfU3U1G2yT2D6ZmXte27Vmx2eDxV_v_lw&hl=en&sa=X&ved=2ahUKEwjR9vqglvLrAhXkzTgGHX9UBtgQ6AEwAHoECAEQAQ#v=onepage&q=Tanaka Y%2C Kato K%2C Notohara K%2C et al. Frequent -catenin mutation and cytoplasmic%2Fnuclear accumulation in pancreatic solid pseudopapillary neoplasm&f=false.
10. Suzuki S, Hatori T, Furukawa T, Shiratori K, Yamamoto M. Clinical and pathological features of solid pseudopapillary neoplasms of the pancreas at a single institution. Digestive surgery. 2014;31(2):143-50.
11. Schwartz M. A biomathematical approach to clinical tumor growth. Cancer. 1961 Nov;14(6):1272-94.
12. Papavramidis T, Papavramidis S. Solid

pseudopapillary tumors of the pancreas: review of 718 patients reported in English literature. Journal of the American College of Surgeons. 2005 Jun 1;200(6):965-72.

13. Vassos N, Agaimy A, Klein P, Hohenberger W, Croner RS. Solid-pseudopapillary neoplasm (SPN) of the pancreas: case series and literature review on an enigmatic entity. International Journal of Clinical and Experimental Pathology. 2013;6(6):1051-1059.
14. Jani N, Dewitt J, Eloubeidi M, Varadarajulu S, Appalaneni V, Hoffman B, Brugge W, Lee K, Khalid A, McGrath K. Endoscopic ultrasound-guided fine-needle aspiration for diagnosis of solid pseudopapillary tumors of the pancreas: a multicenter experience. Endoscopy. 2008 Mar;40(03):200-3.
15. Law JK, Stoita A, Weaver W, Gleeson FC, Dries AM, Blackford A, Kiswani V, Shin EJ, Khashab MA, Canto MI, Singh VK. Endoscopic ultrasound-guided fine needle aspiration improves the pre-operative diagnostic yield of solid-pseudopapillary neoplasm of the pancreas: an international multicenter case series (with video). Surgical endoscopy. 2014 Sep;28(9):2592-8.
16. Cheng K, Shen B, Peng C, Yuan F, Yin Q. Synchronous portal-superior mesenteric vein or adjacent organ resection for solid pseudopapillary neoplasms of the pancreas: a single-institution experience. The American Surgeon. 2013 May;79(5):534-9.
17. Beltrame V, Pozza G, Dalla Bona E, Fantin A, Valmasoni M, Sperti C. Solid-pseudopapillary tumor of the pancreas: a single center experience. Gastroenterology research and practice. 2016 Dec 29;2016.
18. Frost M, Krige JE, Bornman PC, Panieri E, Beningfield SJ, Wainwright H. solid pseudopapillary epithelial neoplasm–a rare but curable pancreatic tumour in young women. South African Journal of Surgery. 2011;49(2):75-81.