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Research Article

Solid Psuedo-Papillary Epithelial Neoplasm of Pancreas: Not So Uncommon Tumor

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ABSTRACT

Introduction: Solid pseudo-papillary epithelial neoplasms (SPEN) of pancreas are rare tumors comprising between 1 and 3% of all pancreatic tumors (1) They are being increasingly diagnosed and treated than before. This is also known as Frantz tumor, owing to its first discovery by Dr Virginia Frantz in 1950 and later by Hamoudi in 1970. Once thought to be rare these low grade malignant potential tumors have shown 7 times increase in incidence over past 2 decades (2). Cystic neoplasms in pancreas other than SPEN account for 2.5% and it increases with age, but the incidence is largely owed to improved cross-sectional imaging. The incidence of pancreatic cysts more than 2cm, supposedly clinically significant, has remained at 0.8% (3). The lesions are predominantly found in children, young females. They are usually discovered because of symptoms of mass or pain or incidentally due to imaging for other reasons. About 10 to 15 % cases are malignant but overall, the prompt recognition and surgical treatment often prompts a long-term survival and cure (4). We present series of four patients that we encountered at the Government Cancer hospital, Aurangabad, Maharashtra, India.

Keywords*: SPEN, Pancreatic tumors*

INTRODUCTION

OBSERVATIONS

Case 1: A 23-year-old female came in with symptoms of dull aching pain in upper abdomen, not localised. There was no relation to food intake and wasn't radiating to back. Clinically she had a barely palpable nontender lump in epigastrium. There was no nausea or vomiting. The Complete blood counts and blood biochemistry was normal. Tumor markers of CA 19-9 and CEA were done. CA 19-9 was 9.2U/L and CEA was 2.4U/L. The CECT showed a 7.4 x 8.1 cm mass replacing head and neck of pancreas. It showed predominantly solid periphery with central cystic areas with necrosis along with discontinuous foci of calcifications at periphery. There was compression to anterior wall of portal vein without abutment or encasement. The main pancreatic duct was 2mm and CBD 6mm in diameter (figure 1). Pre-operative US guided FNAC was tried but showed Pauci-cellular smear and hence was inconclusive. She underwent pylorus sparing pancreato-duodenectomy with pancreatico- jejunostomy. There were no enlarged lymph nodes, pancreas was soft to firm and duct diameters were as indicated in CECT scan. Tumor was not adherent to portal vein. She had an uneventful post-operative period and was discharged on 7th post-operative day. The histopathology report showed the tumor to be well encapsulated SPEN. Solid part of tumor showed numerous pseudo-papillary excrescences. Margins were free, no node of the 12 examined were positive. The Immunohistochemistry showed positivity in B catenin, loss of E cadherin CD 99 and PR positivity. Chromogranin A was not done as radiology and histology were not suggestive of nuero-endocrine tumor.

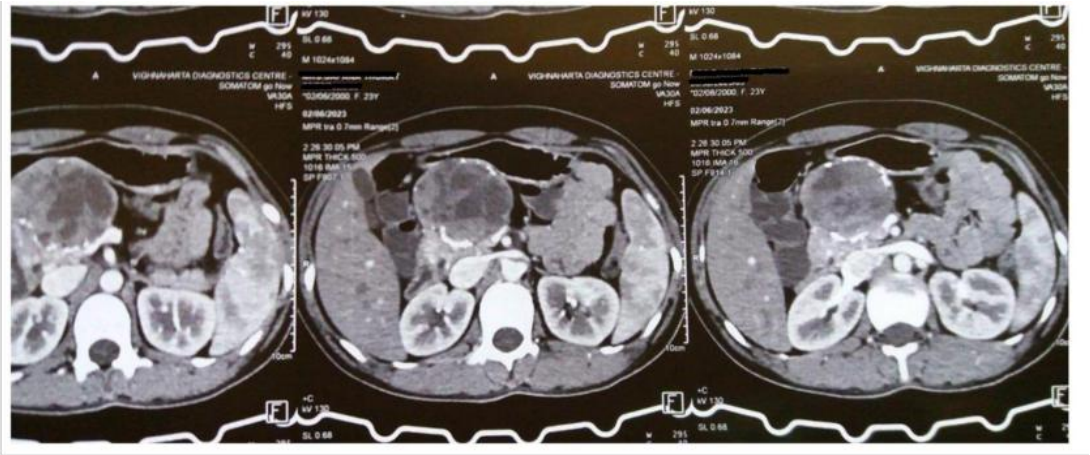

Figure 1: CECT showing a 7.4× 8.1 cm pancreatic head and neck mass with solid peripheral components, central cystic necrosis, and peripheral calcifications, causing compression of the portal vein without encasement.

Case 2: A 21-year-old female came in with symptoms of mass in upper abdomen with occasional pain in it. There were no symptoms of nausea or vomiting and bloating. Clinically she had a palpable nontender firm to hard lump in epigastrium. The Complete blood counts and blood biochemistry was normal. Tumor markers of CA 19-9 was 8.4U/L and CEA was 10.3U/L. The MRI showed a 8.7 x 8.5 x 6.7 cm mass in body and tail of pancreas that was encapsulated except posteriorly over portal vein. It was T1/T2 hyperintense solid cystic lesion with irregular hyperintense areas in between. The plane between tumor and portal vein was not clear. The main pancreatic duct was 3.5mm and CBD 8mm in diameter. US guided FNAC aspirated fluid and showed pauci-cellular smear. At exploration there were no enlarged lymph nodes, pancreas was soft and duct diameters corelated wit MRI finding. Tumor was grossly adherent to portal vein and was free from superior mesenteric artery. A distal pancreato-splenectomy was done along with portal vein sleeve resection. She had a post-operative ancreatic fistula grade B for 19 days and discharged on 20th post-operative day. The histopathology report showed the tumor to be well encapsulated SPEN with foci of high- grade dysplasia and infrequent mitosis. Along with pseudo-papillary excrescences infrequent mitosis were seen. Margins were free, Zero out of 10 examined nodes were positive. The Immunohistochemistry showed positivity in B catenin, CyclinD, CD 99 and PR positivity. Ki-67 was 11% in dysplastic area and 4% in non-dysplastic areas.

Case 3: A 44-year-old female was refererd with symptoms of chronic pain in upper abdomen. There was no other symptom associated with it. Clinically she had unremarkable abdomen She was treated with proton pump inhibitors and underwent ultrasonography for no resolution of pain. Clinically she had unremarkable abdomen. The blood investigations were normal. Tumor markers of CA 19-9 were less than 1U/L and CEA was 1.55ng/ml. The CECT abdomen pelvis showed a well-defined 4 x3.7 x 5 cm mass in head of pancreas. The mass had heterogenous enhancement with a hyper dense soft component and peripheral rim of calcifications (figure 2). The main pancreatic duct was 2mm and CBD 6mm in diameter. US guided FNAC couldn't arrive at diagnosis. At exploration there were no enlarged lymph nodes, pancreas was firm and duct diameters were similar to that mentioned in CECT scan. Tumor was in head pancreas and was free from SMV PV and pylorus preserving pancreato duodenectomy was done with pancreatico-jejunostomy. Post-operative period was uneventful and patient was discharged on 8th post-operative day. The histopathology report showed the tumor to be well encapsulated SPEN with solid and cystic areas with abundant pseudo- papillary projections. Margins were free and no node was positive of 11 nodes. The Immunohistochemistry was positivity in B catenin, CyclinD, Vimentin and PR and negative for chromogranin. Ki-67 was 2%.

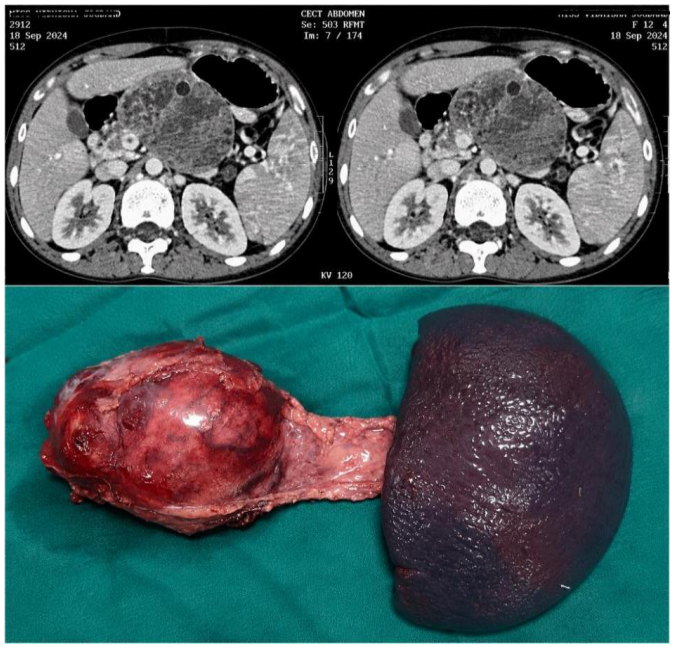

Figure 2: CECT abdomen and pelvis revealed a well-defined 4 × 3.7× 5 cm heterogeneously enhancing mass in the pancreatic head with a hyperdense soft tissue component and peripheral rim calcifications. The MPD and CBD measured 2 mm and 6 mm.

Case 4: A 28-year-old female was referred with incidental finding of tail pancreatic tumor from an ultrasound done for gynecological reason. On asking she could not confirm upper gastrointestinal symptoms. The per-abdominal examination was normal. The blood investigations were normal. Tumor markers of CA 19-9 were 2.4 1U/L and CEA was 1.06ng/ml. The CECT abdomen pelvis showed a well-defined 4 x4.5 x 4cm mass in tail of pancreas. The mass was mostly Solid hyper-enhancing with a very small cystic hypo- enhancing area of 1 cm. There were few specks of calcification in the mass. The main pancreatic duct wasn't measured and CBD 6mm in diameter. US guided FNAC was not done. Intra-operatively tumor was in in tail pancreas 2cm proximal to selenic hilum. No adhesions to splenic artery. There were no nodes along splenic artery or in celiac axis. A Distal pancreatectomy was done. No separate nodal dissection was done. Post-operative period was uneventful; patient was discharged on 7th post-operative day. The histopathology report showed the tumor to be well encapsulated SPEN with solid areas and pseudo-papillary projections. Margins were free and no node was positive of 7 lymph-nodes. The Immunohistochemistry was positivity in B catenin, CD 99, CyclinD, and PR mildly positive for Vimentin.

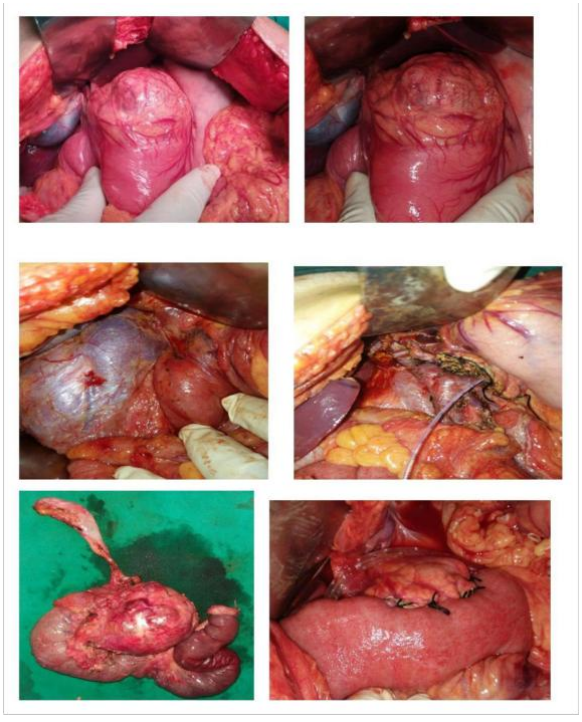

Figure 3: Large well-encapsulated cystic-solid mass arising from the head/body region of the pancreas. Prominent surface vascularity over the tumor capsule. No obvious gross invasion into adjacent major vessels or surrounding organs.

Observations: All the patients were females. Age ranged from 21 to 44 years. The clinical presentation in 2 of 4 patients was that of chronic upper abdominal nonspecific pain. Patients may or may not appreciate lump, one patient had presented as pain and lump while one was incidentally discovered. The tumor markers routinely used for carcinoma pancreas are not elevated. In one case where Se CEA was elevated, the SPEN did show high grade foci along with other features of SPEN. The CECT or MRI findings of solid cystic enhancing masses with peripheral rim of calcification were very suggestive of SPEN and were seen in 3 of 4 patients. Ultrasound guided FNAC did not assist in diagnosis in any of the case. EUS guided FNACs may be more helpful. We do not have EUS in our institute, so they weren't done. Size of the tumor varied from 4.5 to 8.7 cm in maximum diameter. Tumors even if large did not obstruct main pancreatic duct or common bile duct. One tumor did show portal vein adhesion and required sleeve resection of portal vein but in no tumor portal margin was involved. There were 40 nodes examined overall and none was positive. The post-operative recovery of SPEN patients was not different from other pancreatectomies.

DISCUSSION

The solid psuedo-papillary epithelial neoplasms of pancreas are also known as solid papillary tumors of pancreas, Frantz tumor, are classified by the WHO as tumors of low- grade malignant potential that have solid and pseudopapillary areas without a specific epithelial differentiation of the pancreas. They are also considered as uncertain malignant potential with a very rare chance of distant metastasis and even in case of capsular invasion they have a high chance of cure (5). The tumors largely have indolent course and rarely recur after primary surgery. The indicators of carcinoma are metastasis, distant or nodal, orvascular or perinueral invasion (6). Tomioka et al (7) have reported a case of metastatic SPN to liver and noted that half of the primary tumor showed conventional pseudopapillary structures with low Ki-67 index of 6% whie other half showed lobular area with local invasion high mitotic numbers and Ki-67 of 22%. They suggest low grade papillary component might be preceding lesion to the high-grade tumor. They constitute about 1 -2% of all pancreatic neoplasms (8), but the incidence has shown a rise in last two decades (1,8).

Out of all 144 cases in pancreas in 5 years from June 2018 to June 2023. Our incidence therefore is 2.7 %. All patients were females between 21 and 44 years of age. The female preponderance and younger age at presentation is abundantly reported in literature (---) but expression of progesterone receptors as cause of female preponderance is still not confirmed (9) The clinical features of SPEN depend on its location and size presentation but generally are abdominal pain vomiting, discomfort, loss of appetite or early satiety. (10,11). Three of four patients of ours had dull pain in abdomen. Two patients had bloating and were treated for 3 to 4 months with proton pump inhibitors. One patients felt mass in abdomen and no patient had non abdominal symptoms or symptoms like back ache of local invasion and those of metastasis. No patient had jaundice or symptoms of metastasis. One patient incidentally detected to have the tumor. Blood investigations including liver function tests and Se amylase were normal. Tumor markers done were CA19.9 CEA and CA 125. There were no elevations of these in any of patients.We couldn't do CA72-4 due to unavailability of test . The markers of CA19-9, CEA and CA125 were reported normal in most of the studies but one study reported CA72-4 of value in 8.6% of SPEN (12). Fine needle aspiration cytology when done under EUS guidance yields better results CT guided FNAC in diagnosing pancreatic lesion even when they are smaller than 3cm (13). There are conflicting reports on efficacy of EUS guided FNAC in SPEN pancreas but Akira Aso et al (14) and Neelam Mehta et al (15) reported three and four cases of SPEN respectively. EUS guided FNA concurred in all cases with final histo-pathology with both authors. In our case with US guided FNAC, no patient was reported as neoplastic cyst or SPEN, but as hemorrhagic and pauci-cellular smear. It is likely that EUS guided FNAC may help in diagnosis preoperatively. It is not essential to have a preoperative diagnosis of SPEN on tissue diagnosis and surgery can be advised on pre-operative imaging like CT scan. (**16)**

The CECT is first imaging investigation of choice in pancreatic tumors. SPEN is seen as shows a large tumor with heterogenous enhancement due to variable solid and cystic components and often with peripheral calcification. The lesion is well encapsulated in most of the cases. It is commonly seen in tail and head of pancreas but can be seen in any part of pancreas (17) The lesion doesn't appear to cast effect on pancreas like duct dilatations and firmness secondary to duct obstruction. Duct dilatation or strictures and tapering or abrupt cut off of ducts are present in mass forming pancreatitis and carcinomas (18) are conspicuously absent in SPEN. We had 3 of four cases in head of pancreas and one case in neck of pancreas anterior to superior mesenteric artery. The lesions were between 4.5 to 8.7 cm in maximum diameters. They were all solid cystic with discernible capsule except one patient had a break in capsule near portal vein.

The choice of treatment is surgery. Since it is low malignant potential organ preservation surgeries are done when possible. But they can range from enucleation for a small tumor less than 3 cm and away from duct to head pancreas resection with or without duodenal resection, distal pancreatectomy, central pancreatectomy depending on size location and proximity to main duct. Involvement of vein Superior mesenteric or portal may necessitate respective vein resections (19). Two of our cases underwent Pylorus preserving pancreatoduodenectomy and two patients underwent distal pancreatectomy one of which had sleeve resection of portal vein. Regional lymphadenectomy was performed in

one cases which showed doubtful capsular breach, considering it probably malignant. Of 24 patients with SPEN pancreas Ning Guo et al (20) reported only one patient with jaundice, this means CBD and pancreatic duct compression are rare with headpancreatic lesion. The lesion doesn't appear to cast effect on pancreas like duct dilatations and firmness secondary to duct obstruction. In the four cases of ours no patient had dilatation of common bile duct. The CBD diameters ranged from 6 mm to 9mm. The main pancreatic duct also didn't show effects of obstruction. The diameters were 2 to 3.5mm. The pancreas was soft to firm indicating no subclinical inflammation causing firm to hard pancreas seen in ca head of pancreas. A complete Surgical excision, if necessary, with metastasectomy is suggested by Martin RC et al (21) in a series presented by MSKCC.

SPEN on histology sometimes may have similarities with neuroendocrine tumors. Sometimes neuroendocrine tumors may show cystic and necrotic areas composed of discohesive cells appearing like papillary structure and some SPEN may not show pseudopapillary structures (22). It was reported in a series of 93 cases by Tiemann K et al et al (23) Cyclin D1 (92.3%), Beta-Catenin (93.7%),p27 (100%), CD56(97%), p16 (86%) and PR(100%) are useful positive stains to arrive at diagnosis. Vimentin, CK, NSE, Synptophysin E cadherin have been suggested by others (24).

Chromogranin A is used to differentiate nuero-endocrine tumors from SPEN in some cases. The other differential diagnosis should be mucinous cystic carcinomas and acinar cell carcinoma. (25)

CONCLUSION

We are more likely to encounter solid pseudo-papillary neoplasms of pancreas than before owing either to increase in incidence or detection. A SPEN should be considered in diagnosis in paediatric and early adulthood females in pancreatic tumors. CECT is suggestive in diagnosing SPEN. A excision in form of suitable pancreatectomy without lymphadenectomy suffices in most of the cases. Malignant transformations are rare. Differential diagnosis of Cystic neuroendocrine tumors and mucinous cystadenocarcinoma should be considered along.

REFERENCES

1. Zalatnai A, Kis-Orha V. Solid-pseudopapillary neoplasms of the pancreas is still an enigma: a Clinicopathological review. Pathol Oncol Res. 2020;26:641–9.
2. Law JK, AhemadA, Singh VK, AkshintalaVS, OslonMT et al: A systemic review of Solid pseudo papillary neoplasms: are these rare lesions? Pancreas :2014Apr;43(3):331-7.
3. Matsubara S, Tada M, Akahane M et al. Incidental pancreatic cysts found by magnetic resonance imagingand their relationship with pancreatic cancer. Pancreas2012;41:1241-1246. Doi 10.1097/MPA.0b013e31824f5970.
4. Pahlavan PS, Khiyami A, Ganesan S. 2021 Jul: Solid Psuedopapillary Neoplasm of the pancreas: cytology, pitfalls, and literature review; Ann Pancreat Cancer 2021;4:5 doi:10.21037/apc-20-42.
5. Goh B. K. P., Tan Y.-M., Cheow P.-C., et al. Solid pseudopapillary neoplasms of the pancreas: an updated experience. *Journal of Surgical Oncology*. 2007;95(8):640–644. doi: 10.1002/jso.20735).
6. Kodai Tomioka,1Nobuyuki Ohike,2Takeshi Aoki,1Yuta Enami,1Akira Fujimori,1Tomotake Koizumi,1Tomokazu Kusano,1Koji Nogaki,1Yoshihiko Tashiro: 2020 Jan: Solid pseudopapillary neoplasm of pancreas with high grade malignant transformation involving p16-RB pathway alterations: Vol 2020 doi: 10.1155/2020/5980382.
7. Julio C, Marco AR da Costa, Eduardo JB Ramos, Andre Ritzmann Torres Maiane Christina Savio Christiano MP Claus: 2018 Dec. 2018;22(4):e2018.00032 doi:10.4293/JSLS.2018.00032).
8. Feiyang Wang, Zibo Meng, Shoukang Li, Yushun Zhang, Heshui Wu,BMC Gastroenterol.2018 Dec;18:187, doi 10.1186/s12876-018-0914-8).
9. Syed Saad Mujtahedi, Sunil Kumar Shetty, Flora Dorothy Lobo. Solid pseudopapillary epithelial neoplasm of the pancreas involving the distal body and proximal tail: A case report 2021 Mar; Int J Surg Case Rep 80:105519. Doi 10.1016/j.ijscr.2021.01.013).
10. Yagcı, A., Yakan, S., Coskun, A. *et al.* Diagnosis and treatment of solid pseudopapillary tumor of the pancreas: experience of one single institution from Turkey. *World J Surg Onc* **11**, 308 (2013). https://doi.org/10.1186/1477-7819-11- 308.
11. Liu Mengqi, Liu Jiang, Hu Qiangsheng, Xu Wenyan, Liu Wensheng, Zhang Zheng, Sun Qiqing. Management of solid pseudopapillary neoplasms of pancreas: a. a single center experience of 243 consecutive patients. Pancreatology. 2019;19(5):681–685. doi: 10.1016/j.pan.2019.07.001.
12. Horwhat JD, Paulson EK, McGrath K, Branch MS, Baillie J, Tyler D, Pappas T, Enns R, Robuck G, Stiffler H, Jowell P. A randomized comparison of EUS-guided FNA versus CT or US-guided FNA for the evaluation of pancreatic mass lesions. Gastrointest Endosc. 2006 Jun;63(7):966-75. doi: 10.1016/j.gie.2005.09.028.
13. Akira Aso, Eikichi Ihara, Kazuhiko Nakamura, Irina Sudovykh, Tetsuhide Ito, Masafumi Nakamura, Tetsuo Ikeda, Nobuyoshi Takizawa, Yoshinao Oda, Shuji Shimizu, "Solid Pseudopapillary Neoplasm of the Pancreas in Young Male Patients: Three Case Reports", *Case Reports in Gastrointestinal Medicine*, vol. 2017, Article ID 9071678, 4 pages, 2017. https://doi.org/10.1155/2017/9071678
14. Neelam Mehta, Lopa Modi, Trupti Patel, Manoj Shah. Study of cytomorphology of solid pseudopapillary tumors of pancreas and its differential diagnosis; J Cytol.2010 Oct;27(4):118- 122. Doi 10.4103/0970-9371.73293.
15. **T** B Patil, S V Shrikhande, H A Kanhere, R R Saoji, M R Ramadwar, P J Shukla. Solid papillaryneoplasm of pancreas: a single institution experience of 14 cases. HPB (Oxford) 2006 apr;8(2):148-150.
16. Jelena Djokic Kovac, Aleksandra Djikic-Rom, Aleksander Bogdanovic, Aleksandra Jankovic, Nikica Grubor,

Goran Djuricic, Vladimir Dugalic. The Role a. of MRI in diagnosis of Solid Psuedopapillary neoplasm of the Pancreas and its Mimickers: A case Based review with Emphasis on Differential diagnosis:Diagnostics;2023,13(6),1074. Doi: 10.3390/diagnostics13061074.
17. Wolfgang Schima, Gernot Bohm, Christine S. Rosch , Alexander Klaus, Reinhold Fugger Helmut Kopf. Mass-forming Pancreatitis versus pancreatic ductal adenocarcinoma: CT and MR imaging for differentiation. Cancer Imaging.2020;20:52. Doi 10.1186/s40644-022-00324-z.
18. He Song, Ming Dong, Jianping Zhou, Weiwei Sheng, Banghua Zhong, Wei Gao. Solid Psuedopapillary Neoplasm of the Pancreas:Clinicopathologic Feature, Risk Factors of Malignancy , and Survival Analysis of 53 Cases from a single Center: Biomed Research International;2017, Article ID 5465261, 1-7 a.. doi: 10.1155/2017/545261.
19. Ning Guo, Quan B. Zhou, Ru F. Chen, Sheng Q. Zhou, Zhi H.Li, Qing Lin, Jie Wang, Ji S. Chen. Diagnosis and surgical treatment of solid pseudopapillary neoplasm a. of the pancreas: analysis of 24 cases: Can J Surg; 2011 Dec; 54(6);368-374. Doi 10.1503/cjs.011810.
20. Martin RC, Klimstra DS, Bennan MF, Conlon KC, Solid-psuedopapillary tumor of pancreas; a surgical enigma? Ann Surg Oncol.2002; 9:35-40.
21. Yusuke Ohara, Tatsuya Oda, Shinji Hashimoto, Yoshimasa Akashi, Ryuichi Miyamoto, Tsuyoshi Enomoto, Kaishi Satomi, Yukio Morishita, Nobuhiro Ohkohchi. Pancreatic neuroendocrine tumor and solid-psuedopapillary neoplasm: Key immunohistochemical profiles fordifferential diagnosis. World J Gastroenterol.2016 Oct 14;22(38):8596-8604 , a. doi: 10.3748/wjg.v22.i38.8596
22. TiemannK, Heitling U, Kosmahl M, Kloppel G, Solid pseudopapillary neoplasms of the pancreas show an interruption of Wnt- signaling pathway and express gene products of 11q.Mod pathol.2007 Sep 20 (9): 955-60. Doi: 10.1038/ modpathol.3800902.
23. Ayse Yagci, Savas Yakan, Ali Coskun, Nazif Erkan,Mehmet Yildirim, Evrim Yalcin, Hakan Postaci. Diagnosisand treatment of solid pseudopapillary tumor of the pancreas: experience of one single institution from Turkey. World J Surg Onc 11,308(2013) doi: 10.1186/1477-7819-11-308
24. Ka Ram Kang. Ok Ran Shin, SU Lim Lee, Young Mi Ku. Imaging Findings of Pancreatic Solid Psuedopapillary Neoplasm with High Grade Malignant transformation: Focussing on Diffusion- Weighted Imaging and NormlisedApparent Diffusion Coefficient Values. J Korean Soc Radiol. 2018;78(3) 163-169. Doi:10.3348/jksr.201878.3.163.