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EXTRAPANCREATIC PSEUDOPAPILLARY NEOPLASM ARISING FROM THE MESENTERY, MASQUERADING AS A MESENTERIC CYST – A CASE REPORT

General Surgery

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ABSTRACT

Background: Solid Pseudopapillary Neoplasms of the Pancreas are rare entities, and extrapancreatic origin of SPN are even rarer. (2) **Methods:** We present a case of a 30-year-old female with a slow growing abdominal mass, who underwent exploratory laparotomy owing to the clinical suspicion of a mesenteric cyst. Intraoperatively, there was a nodular growth arising from the mesentery, which on histopathologic exam showed sheets of small round cells arranged in patterns of papillae and pseudopapillae, and on IHC, these cells were positive for β-Catenin, confirming the diagnosis of Solid Pseudopapillary Neoplasms of the Pancreas. (3) **Conclusion:** There are 50 reported cases of Extrapancreatic SPN, with only one other documented case arising from the mesentery, in the English literature. Although Pancreatic SPNs have low malignant potential, Extrapancreatic SPNs have been observed to have higher metastatic potential and recurrence rates, hence excision is recommended.

KEYWORDS

Pseudopapillary Neoplasm of Pancreas; Mesenteric Cyst; Extrapancreatic SPN; Aggressive Neoplasm

INTRODUCTION

Solid Pseudpapillary Neoplasm (SPN) is an unusual primary neoplasm of the pancreas that customarily affects adolescent girls and young women, with a median age of 26 years. SPN accounts for less than 1% to 2% of exocrine pancreatic tumors. It is a low-grade malignancy, with a metastasis rate of 10% to 15% (1). Although patients are often asymptomatic, they may have nonspecific symptoms such as weight loss, dyspepsia, and pain or on rare occasions, they may present with palpable abdominal masses (2). Here we describe an extremely rare case of a 30 year old female with Extrapancreatic Solid Pseudopapillary Neoplasm of the Pancreas arising from the mesentery, initially clinically misdiagnosed as a Mesentery Cyst.

Case Report

A 30 year old female presented to the surgical OPD with complaints of a slow growing mass over the right side of the abdomen since 6 months, associated with vague, nonprogressive pain, of a month's duration. Patient gave no history of abdominal distention, vomiting, bowel disturbances, jaundice, loss of appetite, or significant weight loss. Patient did not give any prior history of abdominal trauma, other comorbidities or previous surgeries, or significant family or dietary history.

Her physical examination revealed a moderate built and nourishment with normal vital signs, with a nontender freely mobile intra-peritoneal mass in the umbilical region, of size 12x 10cm, with a lobulated surface and firm consistency. She had an otherwise unremarkable systemic examination. Laboratory tests demonstrated hypochromic microcytic anemia, with other routine labs being normal. Serum Amylase and Lipase levels were not evaluated.

Abdominal ultrasound revealed a right lumbar and peri-umbilical possible retroperitoneal solid lesion of approximately 117x90mm with root dense calcification, with internal vascularity, and close approximation to, but not infiltrating the Inferior Vena Cava. The mass was seen displacing the bowel anteriorly. Other solid organs were noted to be normal.

Patient underwent a Contrast Enhanced Computed Tomography, which described a large solid mass lesion in the right side of the abdomen – possibly arising from the mesentery, measuring 12.3cm (cranio-caudally), 10.1cm (transverse), and 7.9cm (antero-posterior), with few internal septations, and peripheral dense calcifications, extending upto the uncinate process of the pancreas and displacing the Inferior Vena Cava posteriorly. Pancreas appeared normal in shape and density, with no calcifications, and normal Pancreatic duct diameter. Other solid organs were normal.

Owing to the suspicion of mesenteric cyst, Exploratory Laparotomy was performed through a Midline incision under general anaesthesia. Intra-operatively, clots were noted in the peritoneal cavity, and a nodular mass measuring about 15x10x7 cm was noted arising from the mesentery, related anteriorly to the head and the uncinate process of the pancreas, but with no attachment to it.

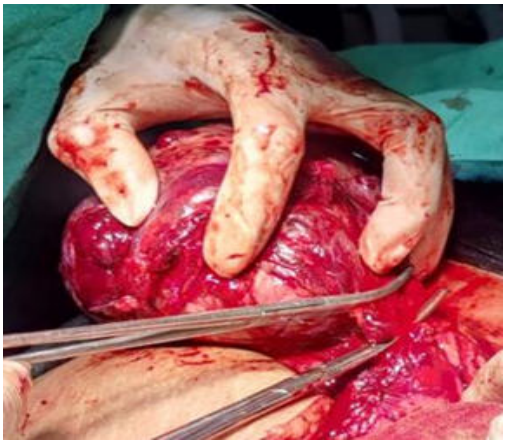

Figure 1. Nodular growth with a pedicle arising from the mesentery

The tumor was excised intact and intoto, with placement of an intra-abdominal drain, and surgery was completed within 3 hours, with minimal blood loss. Patient shifted to the ICU and was managed on mechanical ventilation, but developed ventilator-associated pneumonia, deteriorated, and succumbed on post-operative day 7, due to respiratory causes.

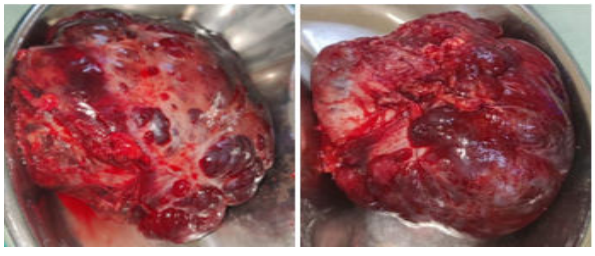

Figure 2. Mass excised intact

The cut surface of the tumor had friable nodules, measuring 0.5x2cm, with the cut surface of the nodules showing brown colored fluid and cystic spaces.

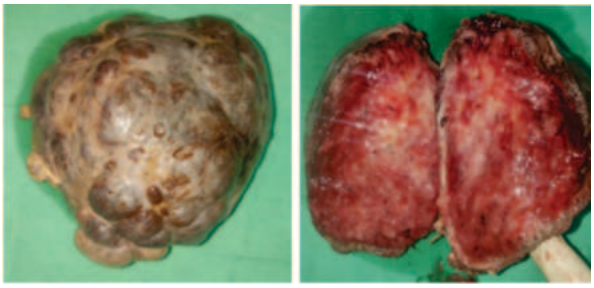

Figure 3. (a) Gross specimen (b) Cut surface of the specimen

Histologically, the solid portions contained cells arranged in various patterns like papillae, pseudopapillae, solid sheets and trabeculae. The papillae were lined by multiple layers of rather small, and fairly round cells with small round oval nuclei with moderate eosinophilic cytoplasm arranged around fibrovascular core with few areas showing eosinophilic cytoplasmic deposits. The tumor cells farthest from the central vessel showed degenerative changes. On immuno histochemical analysis, the cells stained positive for β-Catenin. All the above features were suggestive of Solid Pseudopapillary Neoplasm of the Pancreas.

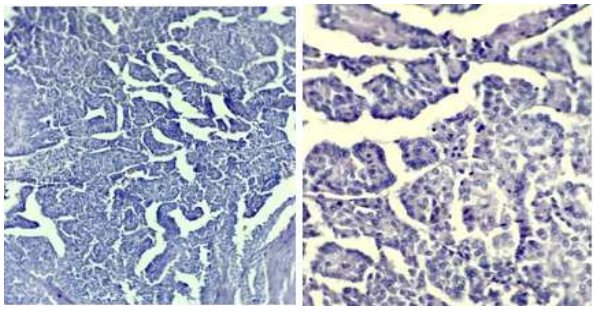

Figure 4. Histopathological examination showing sheets of small round cells arranged in patterns of papillae and pseudopapillae.

DISCUSSION

Solid Pseudopapillary Neoplasm was first described by V.K. Frantz in 1959 and incorporated into the WHO classification in 1996 (3). WHO describes it as low malignant potential tumors with unclear cell of origin and pathogenesis. It has a female preponderance of 10:1 and accounts for 30% tumors in < 40 year old women (4). Occasional cases have been reported in patients with familial adenomatous polyposis (5,6).

As is the case with our patient, they most commonly present with abdominal pain and a palpable, nontender, upper abdominal mass. They may also present with symptoms related to mass effect, such as discomfort, nausea, vomiting and early satiety, all of which, our patient did not complain of. However, most cases are incidentally discovered on imaging (7). Although it is most commonly seen arising from the pancreas, there are a few reported cases in extra-pancreatic sites, such as mesocolon, omentum, ovary and testis (8,9,10).

Although SPNs characteristically appear as solid or solid-cystic masses in the tail of pancreas on imaging studies, in our case, there was suspicion of the mass arising from the mesentery or the retroperitoneum. Wu H et al. have described a case of a 40 year old man with SPN arising from the mesentery, who developed recurrent disease after 4 years, while another case report by Chen L et al. describes a 13-year-old male patient with retroperitoneal SPN and multiple hepatic metastases (11,12). Depending on the site involved, the differentials include adrenal tumors, pancreatic endocrine neoplasm, cysts or tumor of the liver, or a pancreatic pseudo cyst when tail is involved. Biochemical analysis of the cyst fluid shows low CEA and amylase (13).

Since SPNs are typically clearly distinguished from adjacent non-neoplastic pancreatic tissue, they can be curatively treated with surgery with a 5-year survival rate of 95–98%. Procedures such as Distal Pancreatectomy, Pancreaticoduodenectomy or Enucleation can be done, depending on the size and location of the tumor (14,15). Due to the clinical suspicion of a mesenteric cyst, Exploratory Laparotomy and Excision of the tumor was warranted.

Poor prognostic factors in a case of SPN have been described as - size > 5 cm, male gender, presence of necrosis, cellular atypia, vascular invasion, perineural invasion and invasion into adjacent structures (16). Although SPNs have a good prognosis after surgery, there is no consensus on whether postoperative chemotherapy and other treatments are needed for patients with high recurrence risk. Rupture of the tumor or laparoscopic biopsy may seed the tumor cells into the peritoneal cavity, and could be a possible factor responsible for peritoneal recurrence (17).

CONCLUSIONS

Extrapancreatic SPNs are extremely rare entities, with about only 50 cases identified upto 2022 (18). There has been only one reported case of SPN arising from the mesentery, and 5 reported to be arising from the mesocolon in the English literature (11,18). Although Pancreatic SPNs have low malignant potential, Extrapancreatic SPNs have been observed to have higher metastatic potential and recurrence rates. Further studies are warranted to increase our understanding about the biological behavior of Extrapancreatic SPN to improve our diagnostic and treatment modalities.

REFERENCES:

[1] Kang CM, Choi SH, Kim SC, et al. Predicting recurrence of pancreatic solid pseudopapillary tumors after surgical resection: a multicenter analysis in Korea. Ann Surg. 2014;260:348e355.

[2] Tasar, Pinar MDa; Kilicturgay, Sadik Ayhan MDa,*. Solid pseudopapillary neoplasms of the pancreas: Is there a factor determining the prognosis? Experience of a single institution. Medicine 101(34):p e30101, August 26, 2022. | DOI: 10.1097/MD.0000000000030101

[3] Atlas of tumor pathology, Section VII, Fascicles 27 and 28. Tumors of the Pancreas. By Virginia Kneeland Frantz, M.D., Professor of Surgery, College of Physicians and Surgeons, Columbia University. 10⅝ × 7¼ in. Pp. 149, with 92 illustrations. 1959. Washington, D.C.: Armed Forces Institute of Pathology. $1.50. (1959). *British Journal of Surgery, 47(203), 334–334.* doi:10.1002/bjs.18004720344

[4] La Rosa S, Bongiovanni M. Pancreatic Solid Pseudopapillary Neoplasm: Key Pathologic and Genetic Features. Arch Pathol Lab Med. 2020 Jul 1;144(7):829-837. doi: 10.5858/arpa.2019-0473-RA. PMID: 31958381.

[5] Naoi D, Koinuma K, Sasanuma H, Sakuma Y, Horie H, Lefor AK, Sugano K, Ushiama M, Yoshida T, Sata N. Solid-pseudopapillary neoplasm of the pancreas in a patient with familial adenomatous polyposis: a case report. Surg Case Rep. 2021 Jan 28;7(1):35. doi: 10.1186/s40792-021-01121-x. PMID: 33511474; PMCID: PMC7843828.

[6] Inoue T, Nishi Y, Okumura F, Mizushima T, Nishie H, Iwasaki H, Anbe K, Ozeki T, Kachi K, Fukusada S, Suzuki Y, Mizuno A, Kajikawa M, Watanabe K, Sano H. Solid pseudopapillary neoplasm of the pancreas associated with familial adenomatous polyposis. Intern Med. 2015;54(11):1349-55. doi: 10.2169/internalmedicine.54.4061. Epub 2015 Jun 1. PMID: 26027985.

[7] Dinarvand P, Lai J. Solid Pseudopapillary Neoplasm of the Pancreas: A Rare Entity With Unique Features. Arch Pathol Lab Med. 2017 Jul;141(7):990-995. doi: 10.5858/arpa.2016- 0322-RS. PMID: 28661210.

[8] Klöppel G, Maurer R, Hofmann E, Lüthold K, Oscarson J, Forsby N, Ihse I, Ljungberg O, Heitz PU. Solid-cystic (papillary-cystic) tumours within and outside the pancreas in men: report of two patients. Virchows Arch A Pathol Anat Histopathol. 1991;418(2):179-83. doi: 10.1007/BF01600295. PMID: 1705067.

[9] Deshpande V, Oliva E, Young RH. Solid pseudopapillary neoplasm of the ovary: a report of 3 primary ovarian tumors resembling those of the pancreas. Am J Surg Pathol. 2010 Oct;34(10):1514-20. doi: 10.1097/PAS.0b013e3181f133e9. PMID: 20871224.

[10] Cheuk W, Beavon I, Chui DT, Chan JK. Extrapancreatic solid pseudopapillary neoplasm: report of a case of primary ovarian origin and review of the literature. Int J Gynecol Pathol. 2011 Nov;30(6):539-43. doi: 10.1097/PGP.0b013e31821724fb. PMID: 21979589.

[11] Wu H, Huang YF, Liu XH, Xu MH. Extrapancreatic solid pseudopapillary neoplasm followed by multiple metastases: Case report. World J Gastrointest Oncol. 2017 Dec 15;9(12):497-501. doi: 10.4251/wjgo.v9.i12.497. PMID: 29290920; PMCID: PMC 5740090.

[12] Chen L, Yuan M, Wang M, Luo C, Gao M, Huang Q, Li Z, Zhou Z. Case report: Retroperitoneal solid pseudopapillary neoplasm associated with multiple hepatic metastases. Front Oncol. 2024 Jan 30;14:1335930. doi: 10.3389/fonc.2024.1335930. PMID: 38352895; PMCID: PMC10861742.

[13] Rockacy M, Khalid A. Update on pancreatic cyst fluid analysis. Ann Gastroenterol. 2013;26(2):122-127. PMID: 24714589; PMCID: PMC3959935.

[14] Zhang C, Liu F, Chang H, et al. Less aggressive surgical procedure for treatment of solid pseudopapillary tumor: limited experience from a single institute. Plus One. 2015;10:e0143452.

[15] Yağci 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 Oncol. 2013;11:308.

[16] Del Chiaro M, Verbeke C, Salvia R, Klöppel G, Werner J, McKay C, Friess H, Manfredi R, Van Cutsem E, Löhr M, Segersvärd R; European Study Group on Cystic Tumours of the Pancreas. European experts consensus statement on cystic tumours of the pancreas. Dig Liver Dis. 2013 Sep;45(9):703-11. doi: 10.1016/j.dld.2013.01.010. Epub 2013 Feb 14. PMID: 23415799.

[17] Tajima, Y., Kohara, N., Maeda, J., Inoue, K., Kitasato, A., Natsuda, K., … Kanematsu, T. (2012). *Peritoneal and nodal recurrence 7 years after the excision of a ruptured solid pseudopapillary neoplasm of the pancreas: report of a case. Surgery Today, 42(8), 776–780.* doi:10.1007/s00595-012-0208-7

[18] Liu, H. *et al.* (2022) 'Case report: A case report and literature review of extrapancreatic solid pseudopapillary neoplasm', *Frontiers in Surgery*, 9. doi:10.3389/fsurg. 2022. 1020044.