A Case Report: Solid Pseudopapillary Tumor of the Head of the Pancreas in a Pregnant Woman

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ABSTRACT

Solid pseudopapillary tumor (SPT) of the pancreas is a rare tumor which account for only 2.5% of all resected pancreatic neoplasm. Though exact pathogenesis of SPT is unknown but it has preponderance among young and middle-aged female population. There is still an enigma regarding optimal management of SPT in pregnant lady. Here we report a case of solid pseudopapillary tumor in the head of the pancreas in a 34 years old pregnant female. Pancreatoduodenectomy (PD) was performed successfully after term delivery.

Introduction

Solid pseudopapillary tumor (SPT) are rare pancreatic tumor that often affect female individual in their twenties and thirties. It was initially described in 1959 by Frantz [1]. Although the SPT is found throughout the pancreas, the most common location is the tail and body and has no racial predilection [2]. Most SPT of pancreas are indolent, but 10-15% of SPT of pancreas shows malignant behavior and metastases [3]. They also can be locally invasive and spontaneously rupture. SPTs are usually asymptomatic but may present with abdominal pain or mass [4]. Jaundice is rare, even in tumors that originate from the head of the pancreas [5]. In most of the instances, these lesions are well-capsulated and typically cured with complete surgical excision. Long-term survival is often observed despite the presence of malignant features [6]. We present a case of SPT in the head of pancreas in a middle-aged pregnant female. SPTs and their relevant aspects of management are discussed.

Case presentation

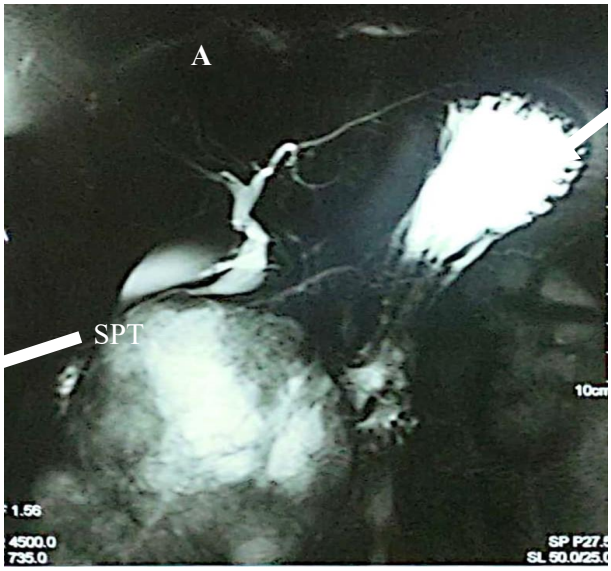
A 34-year-old lady normotensive, non-diabetic, hypothyroid presented to us with the complaint of abdominal mass for last 1 year which was gradually increasing in size which was not associated with pain or any other symptoms. She first noticed this mass in the epigastric region during her 1st trimester of pregnancy. She had no previous medical history of biliary pathology. She was mildly anemic, not icteric. Abdomen was soft, non-tender. There was an ill-defined intra-abdominal lump occupying in the epigastric and right hypochondriac region measuring about 10X10 cm, non-tender and not moving with respiration.

Transabdominal ultrasonogram showed a fairly large soft tissue mass, measuring about 12.4X8.52 cm occupying in the right hypochondriac region which was attached to the inferior surface of liver. As patient was in 1st trimester, CECT was deferred. After consulting with her treating physician, opted for Ultrasonogram guided FNAC which revealed small, uniform epithelial cells arranged in clusters, papillae, groups and acini which have small uniform dark nuclei with scanty cytoplasm suggestive of solid pseudopapillary tumor of pancreas. Routine laboratory test including tumor marker level were within normal range. As patient had no complaint other than lump, after consulting with multidisciplinary team (MDT), definitive treatment was deferred until child delivery and patient was followed up.

After child delivery, CECT (Contrast enhanced Computed Tomography) (Figure. 1) abdomen revealed a fairly large well defined lobulated heterogenous soft tissue density mass measuring about CC-11.8 cm X AP-10.1cm X TD-10.8cm is noted involving head and uncinate process of pancreas causing mild compression over the Gallbladder and body and pyloric part of the Stomach. The lesion was also causing compression and displacement of C-loop of duodenum.

Figure 1. CECT Abdomen (A) axial plane (B) coronal plane showing lobulated soft tissue mass involving head and uncinate process of pancreas

Magnetic resonance (MR) (Figure. 2) abdomen showed a fairly large well defined soft tissue mixed signal intensity is noted involving head and uncinate process of the pancreas causing compression and displacement on 2nd part of the duodenum and lower part of CBD. Diameter of main pancreatic duct (MPD) is 2mm. Common bile duct (CBD) is well visualized with normal caliber and homogenous intraluminal signal intensity but distal CBD is not well delineated.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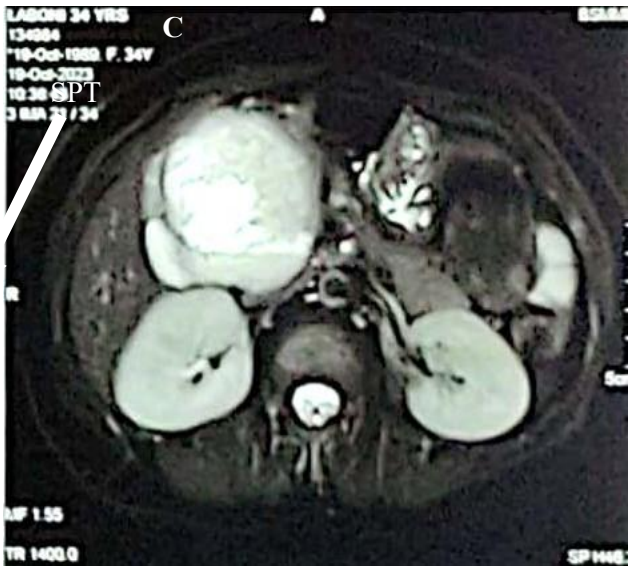
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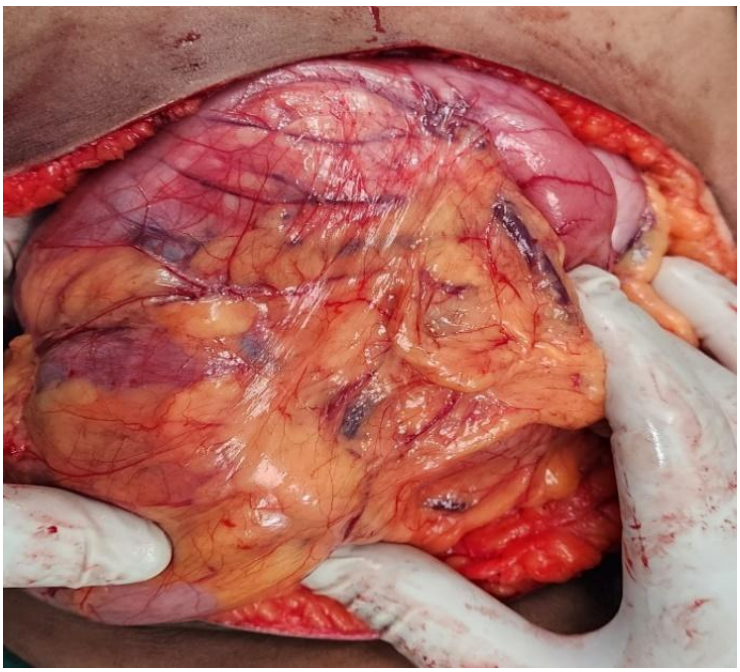
Figure 2: (A) Magnetic resonance cholangiopancreatogram (B) MRI abdomen axial view, T2 (C) MRI abdomen coronal view, T2 showing mixed signal intensity soft tissue mass involving head and uncinate process of the pancreas

The patient was discussed at MDT and considering her clinical, radiographical and tissue diagnosis data, the MDT decided that she is a candidate for surgical treatment for complete excision of the tumor.

The patient underwent surgery one year after the onset of symptoms. After complete preanesthetic checkup, patient was taken for surgery. Patient underwent an exploratory laparotomy and a about 12X11 cm hard consistency mass with engorged vein on its surface arising from head and uncinate process of the pancreas (Figure. 3). The mass was densely adherent with the duodenum, right colic flexure, root of the mesentery, middle colic vessels and partially with portal vein.



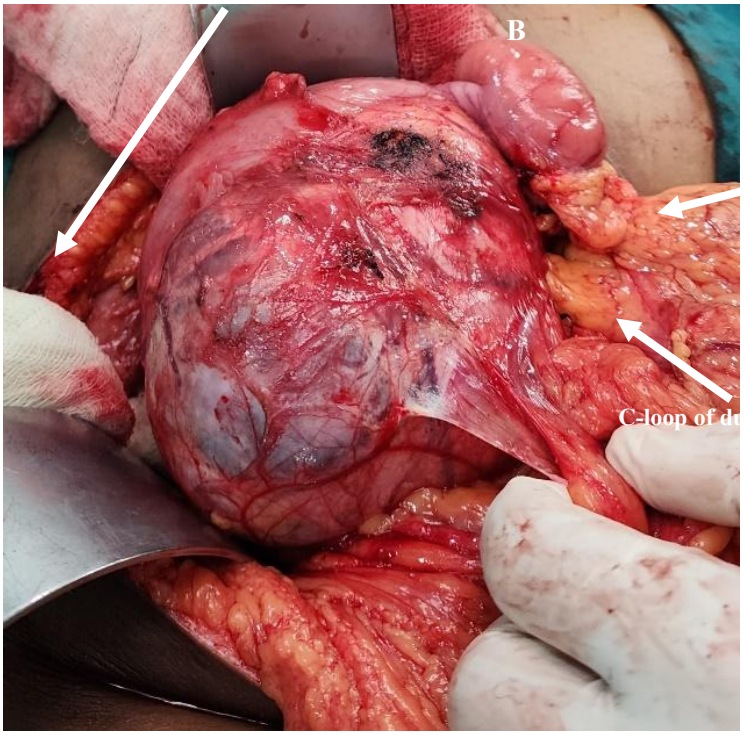


Figure 3: Picture illustrating (A) mass covered with omentum and small bowel mesentery and overlying engorged vein (B) anterior surface of the mass exposed after adhesiolysis.

With meticulous dissection, all the adhesions were taken down. Classical pancreatoduodenectomy performed to achieve complete surgical extirpation (Figure.4). post-operative recovery was uneventful. Oral feeds were started on 5th post-operative day (POD) and patient was discharged on 11th POD.

Histopathology report confirmed SPT. There was no vascular invasion and all resection margins are free of tumor. Excised nine lymph nodes were free of tumor cells.

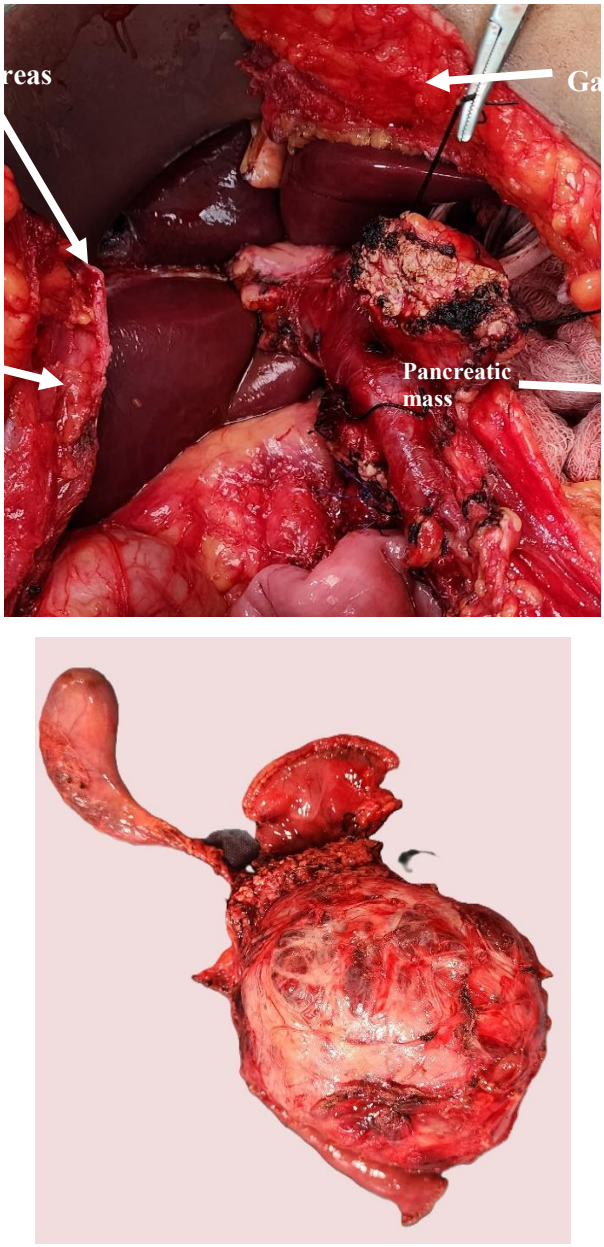


Figure 4: Pictures depicts (A) operating field after complete removal of pancreatic head and uncinate mass (B) resected specimen.

Pancytokeratin (AE1/AE3), Vimentin, E-cadherin and Cyclin D1 expression was observed on immunohistochemistry stains which supports the diagnosis of solid pseudopapillary tumor of pancreas. Later MDT discussion reassured no adjuvant chemotherapy is needed.

Patient was reviewed in outpatient department after 1 month, 3 month and 1 year and till date patient is doing well with no sign of recurrence.

Discussion

SPT is a rare tumor of unknown cellular lineage and exact origin still remains intangible. This tumor was first described by Frantz in 1959 as "papillary tumors of pancreas, benign or malignant [1]. SPT accounts for 0.9 % to 2.7% of all exocrine pancreatic neoplasm in adult and makes up roughly 5% of all cystic pancreatic neoplasm [7]. SPT has striking preponderance among young female population, though no apparent association with endocrine abnormalities has not yet been established. The female to male ratio is 9:1 and the mean age is 29 years [4]. SPT occasionally occurs in 5 to 10 years older men than that of women [8]. Though SPT is an indolent tumor but it tends to be more aggressive in the older male patients.

The SPT is found throughout the pancreas but more frequently in the body or tail in the adult individual [2]. In our case, the lesion was located in the head and uncinate process of the pancreas.

Although the histogenesis of this tumor is not certain, but SPT of the pancreas exhibits a unique pathologic feature. On gross examination, the mass usually as large as 8 to 10 cm in diameter, well-defined and encapsulated. Cystic degeneration and intratumor hemorrhage are common when the tumor attains a large size. On microscopy, it has a heterogenous pattern with various proportions of solid, cystic and pseudopapillary structures. Occasionally calcifications found in the septa and the wall of the tumor. Like other cystic neoplasm of pancreas, SPT rarely cause ductal dilatation due to lack of invasion.

A review of English literature [4], only 38% patients were asymptomatic at presentation. Rest of the patient present with vague abdominal pain or abdominal mass. Jaundice is rare even if the tumor arises from head of the pancreas [5]. Asha Reddy and colleague [8] reported a case of SPT with portal hypertension. In our patient, patient had no jaundice or any other complain except abdominal mass.

Routine laboratory tests including tumor markers are usually within normal range. CECT is the choice of investigation with an additional MRI if further anatomical delineation is required [9]. CECT will show the location of the tumor, involvement of vessels, relation with the surrounding structures and tumor calcification. On MRI, SPT presents as an area of heterogenous high or low signal intensity on T1-weighted images and heterogenous high signal intensity on T2- weighted images. MRI has some certain advantage over CECT in showing certain tissue characteristics such as hemorrhage, cystic degeneration or the presence of capsule [10]. Sometimes ultrasonogram (USG) and endoscopic ultrasonogram (EUS) may also be useful. Though use of a biopsy is

controversial, CT guided biopsy of the tumor is required for the establishment of diagnosis and to exclude malignancy. Albeit, the accuracy of the biopsy is limited [3]. In our case, as patient was presented to us at 1st trimester, we opted to perform USG guided biopsy to confirm diagnosis.

Serous microcystic adenoma, mucinous cystic neoplasm, cystic islet cell tumor, pancreaticoblastoma and calcified hemorrhagic pseudocyst should be considered as differential diagnosis when a pancreatic mass consists of cystic and solid component [11]. Nevertheless, SPT should be considered as a top differential diagnosis if a young female is encountered with a well-encapsulated pancreatic mass with solid and cystic components. In some instances, immunology and molecular markers can be helpful in reaching the diagnosis. Immunohistochemically the tumor cells are positive for β-catenin, vimentin, α1-antitrypsin, α1chymotrypsin, cyclin D1, CD10, and CD 56. There is a loss of membranous expression of E-cadherin. In some cases, there is immunoreactivity for neuron-specific enolase, synaptophysin, and cytokeratin [12].

SPT is regarded as a tumor of low-grade malignant potential and complete surgical excision is the preferred course of treatment. The form of surgical resection varies on location and size of the tumor such as pancreatoduodenectomy (Whipple's procedure) in case of head lesion, central pancreatectomy for the lesion located in the neck region, while lesion located in body and tail are best treated with distal pancreatectomy with or without splenectomy. Many series have showed long term survival after resection. Even long-term survival is often achievable despite the presence of malignant factors such as metastatic disease involving the liver and/or peritoneum and should not withhold resection in selected cases [6]. Local invasion, recurrence or limited metastases are not contraindications to resection.

There is currently limited knowledge regarding chemotherapy and radiotherapy with or without the presence of metastatic disease. Radiotherapy has been suggested in cases of unresectable SPT, as these tumors appear to be radiosensitive [13].

SPT is extremely rare during pregnancy and poses a great challenge to reach a diagnosis. Before confirming the diagnosis as SPT, other more common disorder related to pregnancy should be ruled out. Safe monitoring of the rate of tumor growth and possibility of rupture is suggested by some authors [14]. Some authors support surgical excision of the SPT, since this tumor is responsive to progesterone blood levels [10,15,16]. USG and MRI are the choice of investigation during pregnancy to assess and monitor the lesion. But there is still no consensus regarding optimal management approach of SPT during pregnancy.

Conclusion

Albeit SPT is a rare tumor, but advent of modern imaging technique has increased the pick-up rate of this lesion early. Till date, there is no specific guideline to manage SPT during pregnancy. In most instances, the SPTs are indolent and remain static, monitoring should be sufficed and active management could be deferred till delivery at term. With meticulous dissection, large SPT involving head and uncinate process of pancreas complete resection can be possible.

References

1. Frantz VK: Atlas of tumor pathology. Section VII-Fascicles 27 and 28. Tumors of the Pancreas. Bumberg CW (ed): Armed Forced Institute of Pathology, Washington, DC; 1959. 32-3.
2. Kuo TT, Su IJ, Chien CH. Solid and papillary neoplasm of the pancreas. Report of three cases from Taiwan. Cancer. 1984 Oct 1;54(7):1469-74. doi: 10.1002/1097-0142(19841001)54:7<1469: aid-cncr2820540742>3.0.co;2-e.
3. Papavramidis T, Papavramidis S. Solid pseudopapillary tumors of the pancreas: review of 718 patients reported in English literature. J Am Coll Surg. 2005 Jun;200(6):965-72. doi: 10.1016/j.jamcollsurg.2005.02.011.
4. Law JK, Ahmed A, Singh VK, Akshintala VS, Olson MT, Raman SP, et al. A systematic review of solid-pseudopapillary neoplasms: are these rare lesions? Pancreas. 2014 Apr;43(3):331-7. doi: 10.1097/MPA.0000000000000061.
5. Orlando CA, Bowman RL, Loose JH. Multicentric papillary-cystic neoplasm of the pancreas. Arch Pathol Lab Med. 1991; 115:958–960.
6. Reddy S, Cameron JL, Scudiere J, Hruban RH, Fishman EK, Ahuja N, et al. Surgical management of solid-pseudopapillary neoplasms of the pancreas (Franz or Hamoudi tumors): a large single-institutional series. J Am Coll Surg. 2009 May;208(5):950-7; discussion 957-9. doi: 10.1016/j.jamcollsurg.2009.01.044.
7. WHO. WHO Classification of Tumors: Digestive System Tumors. 5th ed. International Agency for Research on Cancer (IARC); 2019
8. Reddy A, Sanniyasi S, George DJ, Narayanan CD. A rare case report of Solid Pseudopapillary Tumor of the pancreas with portal hypertension. Int J Surg Case Rep. 2016;22:35-8. doi: 10.1016/j.ijscr.2016.03.030.
9. B. Edwin, T. Mala, O. Mathisen, et al., Laparoscopic resection of the pancreas: a feasibility study of the short-term outcome, Surg. Endosc. 18 (3) (2004) 407–411
10. Antoniou EA, Damaskos C, Garmpis N, Salakos C, Margonis GA, Kontzoglou K, et

al.Solid Pseudopapillary Tumor of the Pancreas: A Single-center Experience and Review of the Literature. In Vivo. 2017 Jul-Aug;31(4):501-510. doi: 10.21873/invivo.11089.

11. Cantisani V, Mortele KJ, Levy A, Glickman JN, Ricci P, Passariello R, Ros PR, Silverman SG. MR imaging features of solid pseudopapillary tumor of the pancreas in adult and pediatric patients. AJR Am J Roentgenol. 2003 Aug;181(2):395-401. doi: 10.2214/ajr.181.2.1810395.
12. Uppin SG, Hui M, Thumma V, et al.: Solid-pseudopapillary neoplasm of the pancreas: a clinicopathological and immunohistochemical study of 33 cases from a single institution in Southern India. Indian J Pathol Microbiol. 2015, 58:163-9. 10.4103/0377-4929.155305
13. Zauls JA, Dragun AE, Sharma AK. Intensity-modulated radiation therapy for unresectable solid pseudopapillary tumor of the pancreas. Am J Clin Oncol. 2006 Dec;29(6):639-40. doi: 10.1097/01.coc.0000190457.43060.fd.
14. Yee AM, Kelly BG, Gonzalez-Velez JM, Nakakura EK. Solid pseudopapillary neoplasm of the pancreas head in a pregnant woman: safe pancreaticoduodenectomy postpartum. J Surg Case Rep. 2015 Aug 20;2015(8):rjv108. doi: 10.1093/jscr/rjv108.
15. Levy C, Pereira L, Dardarian T, Cardonick E. Solid pseudopapillary pancreatic tumor in pregnancy. A case report. J Reprod Med. 2004 Jan;49(1):61-4.
16. Huang SC, Wu TH, Chen CC, Chen TC. Spontaneous rupture of solid pseudopapillary neoplasm of the pancreas during pregnancy. Obstet Gynecol. 2013 Feb;121(2 Pt 2 Suppl 1):486-8. doi: 10.1097/aog.0b013e31826d292f.