Scholars Journal of Applied Medical Sciences (SJAMS) **ISSN 2320-6691 (Online)**
Sch. J. App. Med. Sci., 2015; 3(7B):2522-2526 **ISSN 2347-954X (Print)**

(An International Publisher for Academic and Scientific Resources)
www.saspublishers.com DOI: 10.36347/sjams.2015.v03i07.017

Case Report

Solid Pseudo Papillary Neoplasm of the Pancreas- Rare and Clinically Unexpected: Case Report with Review of Literature

Dr. Vanita Pandey, Dr. V. Indira, Dr. K. Mahesh Kumar, Dr. V. Harishanker
Malla Reddy Institute of Medical Sciences, Jeedimetla, Hyderabad, Telangana, India

***Corresponding author**
Dr. Kandukuri Mahesh Kumar
Email: doctormaheshgoud@gmail.com

Abstract: Solid pseudo papillary tumor (SPN) is a unusual primary neoplasm of the pancreas that customarily affects young women. SPN accounts for less than 1% to 2% of exocrine pancreatic tumors. Most common presenting symptom is abdominal mass sometimes associated with dyspepsia, decreased appetite, and weight loss. Vomiting and nausea occur in few patients but they are less common presenting symptoms. Most patients present with indefinite, non-specific abdominal pain which results in delayed diagnosis. Asymptomatic tumors are seen in up to 25 % of cases and they are incidentally detected on imaging or at operation for some other unrelated pathological condition. Pancreatic SPNs are usually indolent tumors. The exact cell of origin is still disputed. Grossly, size of the tumor may range from 3.0 cm to more than 10 cm in diameter. The microscopic features show alternate variable solid areas with cystic component and pseudo papillary formations. Cells are usually polygonal with bland oval to round nuclei and eosinophilic cytoplasm. Various synonyms were used by various authors around the world, According to the author Kloppel G *et al.;* some of the synonyms used to describe SPPN are 1) solid cystic tumor 2) papillary cystic tumor 3) papillary epithelial neoplasia 4) solid and papillary epithelial neoplasia 5) papillary epithelial tumor 6) Frantz's tumor 7) solid and papillary tumor 8) solid-cystic papillary epithelial neoplasm 9) benign or malignant papillary tumor of the pancreas. In the year 1996, World Health Organization (WHO) came up with the single nomenclature as Solid Pseudo Papillary Neoplasm (SPN). We present a rare case of SPN in a 40 year old female patient who presented to the hospital with complaints of abdominal pain, loss of appetite and fever. Clinically and radiologically diagnosis of pancreatic pseudo cyst was made. Histopathological examination (HPE) gave the definitive diagnosis and proved to be a valuable investigation in this case.
Keywords: Solid Pseudo papillary tumor (SPN), Solid Pseudo papillary Neoplasia, Pseudo cyst of pancreas, Prognosis, IHC, Vimentin, Alpha Antitrypsin, Ki-67.

INTRODUCTION

Pancreatic Solid pseudo papillary neoplasm (SPN) is a very rare neoplasm. It was first described by Frantz in 1959. Usually SPNs are well encapsulated masses, with low malignant potential. It almost always occurs in young females, usually in their third decade of life. Histogenesis or cell of origin of SPN still remains unexplained or unanswerable question. According to world literature, Acinar, endocrine, ductal and progenitor cells could have been the possible cells given rise to this type of tumor. SPNs are frequently asymptomatic or minimally symptomatic tumor. Imaging modalities such as ultrasound imaging (US), computed tomography (CT) and magnetic resonance imaging (MRI) can be used to diagnose and to differentiate SPNs from other pancreatic lesions. On CT scan, SPN presents as a well encapsulated, hypo dense mass lesion with various solid and cystic composition. On Ultrasound imaging, SPNs appear as either homogenous or heterogenous encapsulated mass with both solid echogenic and hypoechogenic components. MRI is most reliable than CT scan in demonstrating the presence of a capsule, hemorrhagic areas, or cystic degeneration [1]. According to the literature, abdominal swelling is the most common complaint by the patient. Nausea and vomiting are less common presenting symptoms. Asymptomatic tumors constitute roughly about 20 % of all the SPNs. Asymptomatic cases are identified either incidentally on imaging or at the time of surgery for unrelated pathology [1-6]. Surgery is the only definitive treatment for SPNs with a excellent cure rate of greater than 95% with total resection.

CASE HISTORY

A 40 years female patient of Suraram, Hyderabad came to Casualty, Malla Reddy Hospital, with complaints of pain abdomen since 3 months and menorrhagia for last 3 cycles. Pain abdomen was not accompanied with vomiting, constipation and diarrhea. Family history and dietary history were insignificant, except for occasional alcohol intake .General and

systemic examination revealed normotensive, a febrile, average built status. Per abdomen findings were tenderness in Hypo gastric region. CVS, CNS, Respiratory system findings were noncontributory. Investigations showed Hemoglobin 13.4gm/dl, WBC count 8200 cells/cu.mm, Bleeding Time (BT), Clotting Time (CT) within normal units. Random blood sugar (RBS), complete urine examination (CUE), Thyroid function test, Serum urea, creatinine and Serum amylase values within normal limits. Serum Lipase was not significantly raised (81 U/L). Ultrasound abdomen, revealed cystic lesion measuring 6.9 x 6.8 cm over the tail of pancreas suggestive of Pseudo cyst. Mild splenomegaly noted. Bulky uterus with multiple fibroids was also observed. CT scan abdomen (plain and contrast) was reported as acute pancreatitis with Complex per pancreatic Pseudo cyst and bulky uterus with multiple fibroids. Patient was managed, prepared and planned for surgery. Intraoperative a true cystic lesion arising from the tail of the pancreas was observed and cystectomy was done. The patient recovered uneventfully. Gross findings- A grayish brown, circumscribed, bilobed, solid to cystic soft tissue mass measuring 10 x 6x 3 cm was received (FIGURE 1) .Cut surface showed biloculated cyst, with spaces filled with hemorrhagic, necrotic material (FIGURE 2). Cyst wall thickness varied from 0.1 to 1.0 cm. Inner wall showed septations, papillary projections and irregular surface.

Microscopy-Sections showed an encapsulated tumor composed of solid sheets of neoplastic cells showing pseudo papillary configuration with interspersed large thin walled vascular spaces (FIGURE 3 & 4). Tumor cells were seen with bland round to oval nuclei, pale chromatin and inconspicuous nucleoli and scant cytoplasm. Mitotic activity was inconspicuous. These cells are forming pseudo papillae with central hyalinized cores (FIGURE 5). Areas of hemorrhage and hyalinization with presence of hemosiderophages and thick walled blood vessels were seen. Collections of hemosiderophages as an evidence of old hemorrhage in the tumor. In the center of cavity fragments of neoplastic cells, separate from the main fragment due to poorly supported blood vasculature were also found (FIGURE 6). Hence final diagnosis of Solid pseudo papillary neoplasm was made on HPE.

Fig-1: Gross Picture of tumor with partly collapsed cystic area

Fig-2: Cut Surface showing encapsulated mass with cystic and solid areas, blood clot and necrotic debris.

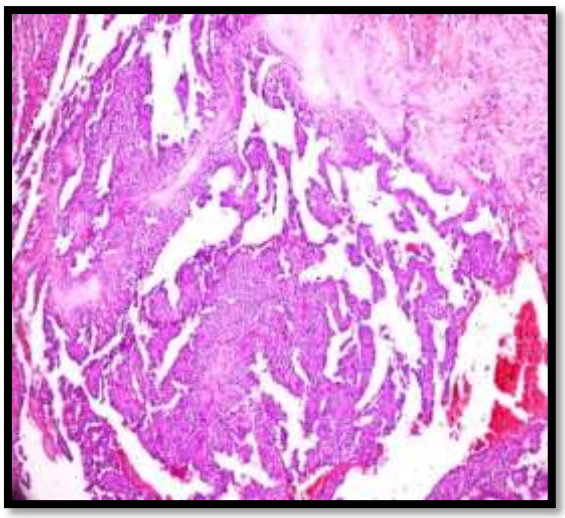

Fig-3: Microphotograph shows capsule and tumor in solid sheets

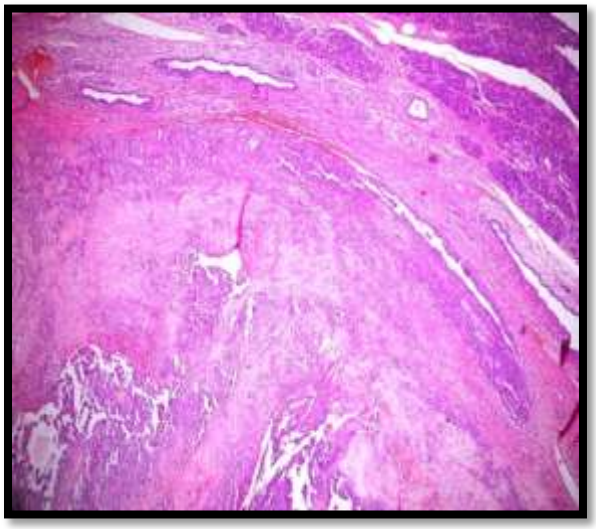

Fig-4: Microphotograph shows cystic areas with pseudo papillary fronds.

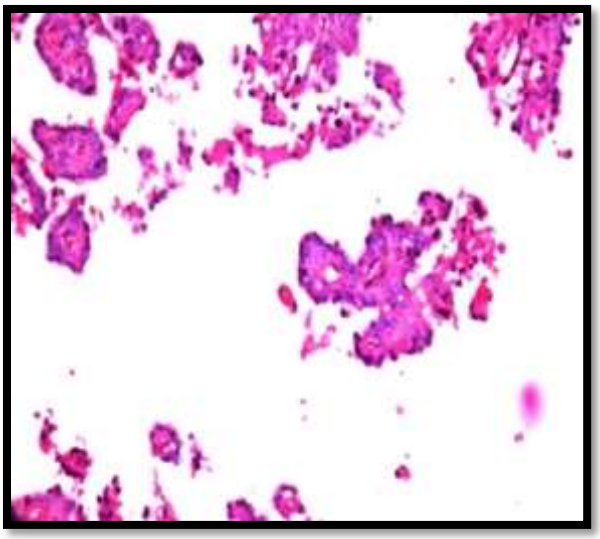

Fig-5: Microphotograph showing pseudo papillary architecture (40 x views)

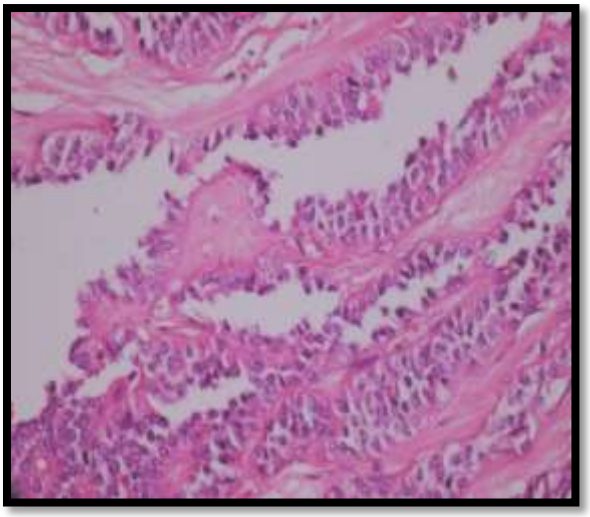

Fig-6: Microphotograph shows Cystic area with free floating tumor cells.

DISCUSSION

Pancreatic Solid Pseudo papillary Neoplasm (SPN) is a very rare neoplasm. It was first described by Frantz in 1959. SPN constitute approximately 5% of cystic pancreatic lesions and about 1 - 2% of exocrine pancreatic neoplasms [7]. They are mainly encountered in the second and third decades of life. According to the study conducted by Chen X *et al.;* the male to female ratio is 1:10 and the mean age at presentation is 22 years. It usually presents as a gradually enlarging abdominal mass but can be asymptomatic in few cases. Jaundice is a late and infrequent presentation [2]. Different titles were used for this tumor till 1996, later they were defined by the World Health Organization (WHO) as a ‘‘solid pseudo papillary tumor’’ (SPN) of the pancreas in 1996 [8]. The cell of origin of solid pseudo papillary tumors still remains unclear. According to Eder F et al, SPNs probably may originate from ductal epithelial cells, neuroendocrine cells, multi potent primordial cell, or even from an extra-pancreatic genital ridge angle-related cell [9]. Head and tail of the pancreas are most common sites; however they can arise from any part of the pancreas [3, 10]. SPNs are mostly confined to pancreatic tissue but sometimes adjacent anatomical tissues like the mesenteric vessels, stomach, and first and second part of the duodenum may also be involved [4]. Depending on the site involved, the contrasting diagnosis include adrenal tumors, pancreatic endocrine neoplasm, cysts or tumor of the liver, or a pancreatic pseudo cyst when tail is involved. In our case, tail of the pancreas was involved mimicking pseudo cyst of the pancreas. In a study conducted by Cameron D Adkisson *et al.;* involving 718 patients with SPN, involvement of tail of the pancreas was seen in 35.9% of SPN patients, 34% of tumors were detected in head of the pancreas, and 10.3% were seen at the junction of body and tail [3]. In approximately 1% of patients SPNs are seen in areas other than pancreatic tissue [3]. Usually there are no derangements of pancreatic functioning, deviated liver function tests, cholestasis, and abnormal elevation of pancreatic enzymes or an endocrine manifestation. Tumor markers are also generally unreliable and unremarkable [3, 11]. These tumors have low-malignant potential and metastasize infrequently. Histologic markers of poor prognosis include a high mitotic index, spindling of tumor cells; anaplastic giant cells, capsular invasion, and lympho-vascular involvement .None of the features are seen in our case. Immuno histo chemistry (IHC) can be additionally helpful as SPNs express strong Vimentin, Progesterone Receptor, and Alpha anti trypsin, CD 10 and Beta Catenin positivity. Patchy Cytokeratin (CK) and Neuron specific enolase (NSE) reactivity helps to distinguish it from Adenocarcinoma and Neuroendocrine neoplasms involving pancreas. So the tumor is considered hormone dependent, and different from epithelial and endocrine neoplasms. High Ki-67 reactivity seen in malignant counterparts. According to Sclafani LM *et al.;* local recurrence rate is less than 5% [12]. Among local recurrence sites liver, lymph nodes and peritoneum are more commonly involved. Rarely, superior mesenteric artery or portal vein involvement can be seen and forms a major limitation in resecting the tumor. Complete resection of lesion is curative. According to Lam KY and Fried et al, even patients with unresectable, left over disease or metastases have been reported to have a good survival rate following surgical treatment. Very few reports in the literature quoted the use of chemotherapy and radiotherapy for these tumors but only with limited response [13,14]. According to Zang H and Liang T, the characteristic features of malignancy include vascular invasion, peri neural invasion and deep invasion of pancreatic tissue [1]. Incidence of malignant transformation is 15% of adults and in children it is 13%. Greater risk of malignant transformation is noted in males and the elderly [1, 15]. According to the authors, Papavramidis T *et al.;* and Martin R *et al.;* recurrence was seen in 5–7% of patients after surgical resection [3, 16]. Metastatic spread is more frequent to liver, lymph nodes, and peritoneum and its incidence is 10–15%. Poor

prognostic factors are presence of vascular invasion on microscopy, tumor size greater than five cm and low nuclear grade [16, 17, 18]. Surgery is the only definitive treatment; with complete resection of the tumor the cure rate of greater than 95% can be achieved. Radical resection of tumor with removal of enlarged lymph nodes can be undertaken in addition to resection of synchronous or metachronous metastases [19]. Unlike other pancreatic tumors, invasion of the portal vein or superior mesenteric artery does not contraindicate tumor respectability [20]. In approximately 7% of SPN patients pancreatic fistula is the most common complication after complete resection [5]. The role of adjuvant therapy in treatment of SPNs is not clear and not well documented so far, with few studies demonstrated role for gemcitabine and radiotherapy to decrease the size of large tumor(s) or treat the rare case of unresectable disease [15]. Five year survival after complete resection is 94–100% [3, 16, 21].

CONCLUSION

Pancreatic Solid Pseudo papillary Neoplasm (SPN) is a very rare neoplasm which presents with vague and non-specific clinical features. SPN can arise from any part of the pancreatic tissue. Grossly, they are characterized by solid and cystic areas with papillary excrescences .Surgical excision of the lesion is the definitive treatment of choice even when there is local metastasis. Prognosis is favorable even in the presence of distant metastasis with 5 year survival rate up to 100 %.

REFERENCES

1. Zhang H, Liang T, Wang W, Shen Y, Ren GP, Zheng SS; Diagnosis and treatment of solid pseudo papillary tumor of the pancreas. Pancreas 2006;5 (3):454–8.
2. Chen Z, Zhou GW, Zhou HJ, Peng CH, Li HW; Diagnosis and treatment of solid pseudo papillary tumors of the pancreas. Hepatobiliary and Pancreatic Dis Int 2005;4:456–9.
3. Papavramidis T, Papavramidis S; Solid pseudo papillary tumors of the pancreas: review of 718 patients reported in English literature. J Am Coll Surg 2005;200:965–72.
4. Dong D, Zhang, S; Solid pseudo papillary tumor of the pancreas: CT and MRI features of 3 cases. Hepatobiliary and Pancreatic Diseases International 2006;5 (2):300–4.
5. Chen S, Zou S, Dai Q, Li H; Clinical analysis of solid pseudo papillary tumor of the pancreas: report of 15 cases. Hepatobiliary Panc Dis Int 2008;7 (2):196–200.
6. de Castro S, Singhal D, Aronson D, Busch O, Gulik T, *et al.;* Management of solid pseudo papillary neoplasms of the pancreas: a comparison with standard pancreatic neoplasms. World J Surgery 2007;31:1129–34.
7. Klimstra DS, Wenig BM, Heffess CS; Solid pseudo papillary tumor of the pancreas: a typically cystic carcinoma of low malignant potential. Semin Diagn Pathol 2000, 17:66–80.
8. Coleman KM, Doherty MC, Bigler SA; Solid pseudo papillary tumor of the pancreas. Radiographics 2003; 23:1644–48.
9. Eder F, Schulz HU, Röcken C, Lippert H; Solid-pseudo papillary tumor of the pancreatic tail. World J Gastroenterol 2005, 11:4117–4119.
10. Pettinato G, Manivel JC, Ravetto C; Papillary cystic tumor of the pancreas: a clinico pathologic study of 20 cases with cytologic, immuno histo chemical, ultra structural, and flow cytometric observations, and a review of literature. Am J Clin Pathol 1992;5:478–88.
11. Yu PF, Hu ZH, Wang XB, Guo JM, Cheng XD, Zhang YL, Xu Q; Solid pseudo papillary tumor of the pancreas: a review of 553 cases in Chinese literature. World J Gastroenterol 2010, 16:1209–1214.
12. Sclafani LM, Reuter VE, Coit DG, Brennan MF; The malignant nature of papillary and cystic neoplasm of the pancreas. Cancer 1991, 68:153-8.
13. Lam KY, Lo CY, Fan ST; Pancreatic solid-cystic-papillary tumor: clinico pathologic features in eight patients from Hong Kong and review of the literature. World J Surg 1999, 23:1045-50.
14. Fried P, Cooper J, Balthazar E, Fazzini E, Newall J; A role for radiotherapy in the treatment of solid and papillary neoplasms of the pancreas. Cancer 1985, 56:2783-5.
15. Machado CM, Machado AM, Bacchella T, Jukemura J, *et al.;* Solid pseudo papillary neoplasm of the pancreas: distinct patterns of onset, diagnosis, and prognosis for male versus female patients. Surgery 2008;29–34.
16. Martin R., Klimstra D, Brennan M, Conlon K. Solid pseudo papillary tumor of the pancreas: a surgical enigma? Annals of Surgical Oncology 2002;9 (1):35–40.
17. Kang C, Kim K, Choi J, Kim H; Solid pseudo papillary tumor of the pancreas suggesting malignant potential. Pancreas 2006;32 (3):276–80.
18. Nishihara K, Nagoshi M, Tsuneyoshi M, Yamaguchi K., Hayashi I; Papillary cystic tumors of the pancreas: assessment of their malignant potential. Cancer 1993;71:8292.
19. Tipton S, Smyrk T, Sarr MG, Thompson GB; Malignant potential of solid pseudo papillary neoplasm of the pancreas. British Journal of Surgery 2006;93:733–7.
20. Jeng L, Chen M, Tang R; Solid and papillary neoplasm of the pancreas: emphasis on surgical treatment. Arch Surg 1993;128:433–6.
21. Reddy S, Cameron J, Scudiere J, Hruban R, Fishman EK, Ahuja N, *et al.;* Surgical management of solid pseudo papillary neoplasms of the pancreas (Franz or Hamoudi Tumors): a large single institutional series. Journal Am Coll Surg 2009;208 (5):950–7.