

## Solid Pseudo Papillary Tumor of Pancreas: An institutional experience

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### Abstract

**Introduction:** Solid-pseudo papillary tumor (SPPT) of the pancreas is a very rare tumor and very few case studies has been published from India.

**Aims and Objectives:** To evaluate the clinical presentations SPPT of pancreas and to discuss the treatment, radiological, histopathological and immunohistochemical (IHC) findings with cytogenetic work up and subsequent follow up of the patients.

**Materials and Method:** We retrospectively reviewed the six patients with SPPT managed in our hospital between 2008 to 2016. All the six cases were noted in females and their age ranges from 15 to 25 years. The clinical diagnosis were confirmed with ultrasound and computerized tomography. Distal Pancreatectomy along with splenectomy in two cases and Whipple's procedure was performed in other four cases. Cytogenetic workup was done in two cases.

**Results:** The tumors were large, well encapsulated with mean diameter of 7 cm's, had solid and cystic areas and were distributed in head, body and tail of pancreas. IHC findings revealed tumors, positive for neuron specific enolase, cytokeratin, progesterone, vimentin and focal positivity for chromogranin, and synaptophysin. The follow up of the patient revealed pseudo cyst of pancreas in one case. Cytogenetic work up revealed derivatives of 17<sup>th</sup>, 13<sup>th</sup>, 9<sup>th</sup> chromosomes.

**Conclusion:** SPPT is a rare tumor with good prognosis after surgical resection.

**Keywords:** SPPT, Pancreas, Pseudo papillary, Solid and cystic areas, Immune histochemical, Radiological, cytogenetics.

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### Introduction

Solid-pseudo papillary tumor (SPPT) of the pancreas is a very rare tumor and was first described by Frantz in 1959.<sup>(1)</sup> It accounts for 1-2% of all primary tumors of pancreas. There are many synonyms for solid pseudo papillary tumors which include papillary cystic neoplasm, papillary epithelial neoplasm, solid-cystic papillary tumor or Gruber-Frantz tumor.<sup>(2)</sup> Mostly these tumors are found in young women in the second or third decade.<sup>(3)</sup> In children, these tumors are rarely seen.<sup>(4)</sup> Mostly the patients present with a palpable abdominal mass and a dull abdominal pain. A low malignant potential is suspected in these tumors and their prognosis is extremely good.<sup>(5,6)</sup> WHO in 1996, reclassified this tumor and now solid pseudo papillary tumor of pancreas is a universally recognized entity.<sup>(7)</sup> There are many large series of SPPT from USA, Europe, China, Japan.<sup>(8,9)</sup> In India, Patil et al.,<sup>(10)</sup> Uppin SG et al.,<sup>(11)</sup> studied clinicopathological features and treatment outcome, and various immune histochemical (IHC) patterns of SPPT. In the present study, we evaluated various clinical manifestations, IHC patterns and cytogenetics of SPPT.

### Materials and Method

Prior approval from Hospital Ethical committee was taken. Six consecutive cases of SPPT diagnosed at Department of Pathology at Kamineni Academy of Medical Sciences and Research Center over a period of 9 years from 2008 to 2016 were retrospectively

reviewed. All the six cases were females, age ranging from 15 to 25 years. Symptoms and clinical signs presented were abdominal pain, abdominal mass and back pain. The clinical diagnosis was confirmed with ultrasound (US) and computerized tomography (CT). The tumor localization was body and tail (4 cases), and head of the pancreas (2 cases). The radiological findings were, smooth well-defined pancreatic tumor with solid and cystic areas. The diameter of the tumors varied from 4 to 10 centimeters. Distal pancreatectomy along with splenectomy was done in four cases and Whipple's procedure was performed in other two cases. Routine histopathological and immune-histochemical studies were performed. Follow up of all the patients were done by general surgery department. Chromosomal study using peripheral blood of the patient has been done in two cases using G-banding technique.<sup>(12)</sup>

### Results

There were six consecutive patients diagnosed with SPPT of pancreas during the study period. All the six cases were young females with age ranging from 15 to 25 years (mean 18.6 years). Majority of the patients were in second decade. The duration of the symptoms ranged from 3 days to 6 months. The most common presentation was pain abdomen and mass per abdomen. The location of the tumor in the pancreas included head (2 cases), body and tail (4 cases). (Table 1)

**Table 1: Demographic and clinicopathological features of the six patients with SPPT of the pancreas**

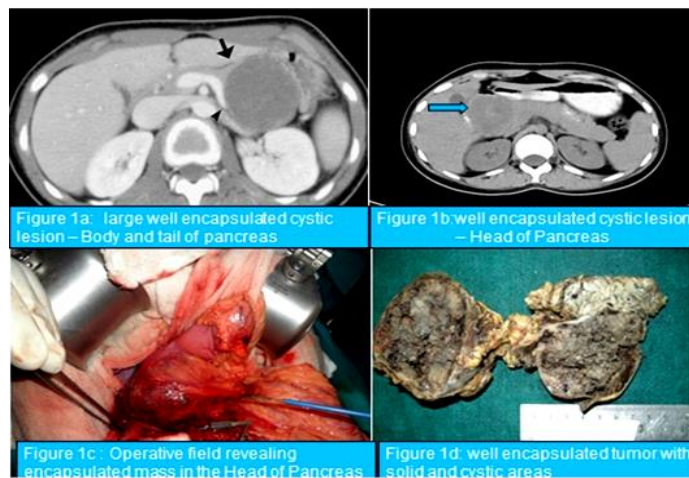
Case	Age/Sex	Clinical presentation	Site	Size	Local invasion	Surgery done	Follow up
1	19/f	Mass per abdomen	Head & body	10 x 10 cm	Nil	Whipple's	5 years
2	15/f	Mass per abdomen	Tail	8 x 8 cm	Nil	DP+S	5 years
3	16/f	Pain abdomen	Tail	10 x 10 cm	Nil	DP+S	4 years
4	25/f	Asymptomatic	Tail	6 x 6 cm	Nil	DP+S	4 years
5	17/f	Mass per abdomen	Tail	5 x 5 cm	Nil	DP+S	3 years
6	20/f	Pain abdomen	Head	4 x 4 cm	Nil	Whipple's	2 years

DP +S: Distal pancreatectomy + Splenectomy

Radiological findings: The patients underwent ultrasound (US) and computerized tomography (CT) examination. The diagnosis of SPPT was considered in 5 cases and cystic neoplasm of the pancreas in 1 case. (Fig. 1a, 1b)

Operative findings: Surgical exploration was done in all cases. (Fig. 1c) For tumors located in head of the pancreas Whipple's procedure (Pancreatico-duodenectomy along with common bile duct and Gall bladder) was performed. And for tumors located in the tail of the pancreas, distal pancreatectomy with splenectomy was done.

Grossly, the tumors were predominantly well encapsulated and globular. The mean diameter of the tumor was 7 centimeters. Cut surface showed variegated appearance, with solid and cystic areas accompanied by hemorrhage (Fig. 1d). Two tumors showed extensive cystic changes.



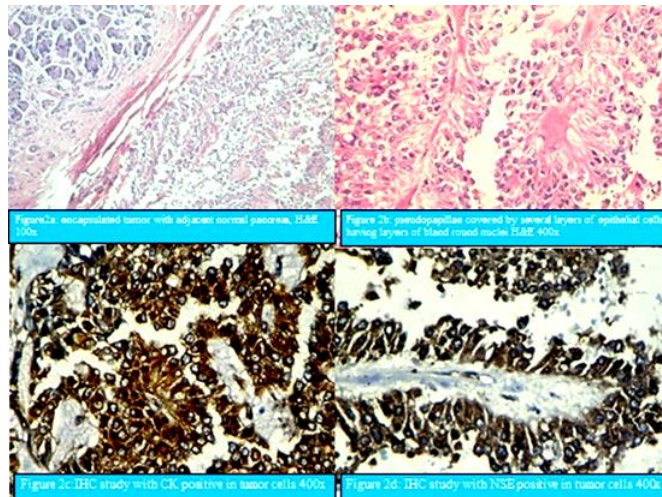
**Fig. 1: Radiological, operative and gross images of SPPT of Pancreas**

Microscopic findings revealed presence of pseudo papillae covered by several layers of epithelial cells, having round to ovoid nucleus with fine chromatin and indistinct nucleoli. The delicate fibro vascular core shows prominent mucinous changes. (Fig. 2a & b) Few cases showed solid component simulating neuroendocrine tumor. Areas of hemorrhage and necrosis were also noted.

Immunohistochemistry: IHC was done using Cytokeratin, Vimentin, Neuron Specific Enolase, Chromogranin, Synaptophysin and Progesterone in all the six cases. (Fig. 2c & d). Results are shown in Table 2.

**Table 2: Immunohistochemistry results**

IHC Panel	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
Chromogranin	+	+	+	-	-	-
Synaptophysin	+	+	-	-	-	-
Cytokeratin	+	+	+	+	+	+
Vimentin	+	+	+	+	+	+
Progesterone	+	+	+	+	-	-
NSE	+	+	+	+	-	-



**Fig. 2: Microscopic and Immunohistochemical findings of SPPT of Pancreas**

Karyotyping of two patients revealed derivatives of 17<sup>th</sup>, 13<sup>th</sup>, and 9<sup>th</sup> chromosomes. (Fig. 3a & b)



**Fig. 3: Chromosomal studies in two cases of SPPT of Pancreas**

On follow up, one of the patient presented with pseudo cyst of the pancreas after one year. Remaining patients were in good condition.

**Discussion**

SPPT of the pancreas is a very rare tumor, and accounts for 0.13-2.7% of all primary pancreatic tumors,<sup>(8)</sup> first described by Frantz in 1959.<sup>(1)</sup> Majority of the tumors are seen in young patients with a mean age of 28 years and a female to male ratio of 10:1.<sup>(9)</sup> In the present study, mean age of presentation was 18.6 years, and with female predilection/ preponderance. The various clinical presentations include palpable abdominal mass, abdominal discomfort, pain abdomen and asymptomatic. Similar findings were seen in the present study (Table 1). Advances in imaging modalities have led to increase in number of cases diagnosed in recent years. It can be diagnosed preoperatively using Ultrasound, CT scan, and MRI which shows well-circumscribed heterogeneous mass comprising solid and cystic areas. MRI is better than

CT as it can detect marked degenerative changes such as cystic changes, hemorrhage, and integrity of the capsule.<sup>(13)</sup> In our study 5 cases (83%) were diagnosed accurately on imaging, similar to Machado et al.<sup>(14)</sup>

In the present study, tail of the pancreas is the most common site for SPPT, followed by head and body, similar to Vassos N et al.,<sup>(15)</sup> and Uppin SG et al.<sup>(11)</sup> The size of the tumor ranges from 0.5 cm to 34.5 cm, with a mean diameter of 6.8 cm,<sup>(16)</sup> similarly in the present study the size ranges from 4 cm to 10 cm, with a mean diameter of 7 cm. The tumor cut surface showed solid and cystic areas as the name indicates, (Fig. 1a & b) but in one case it showed extensive cystic changes, which lead to radiological misdiagnosis in our study.

The microscopic features of SPPT have been described well in literature. These tumors have solid and cystic areas with pseudo-papillary growth pattern

composed of a thin fibrovascular stalk surrounded by several layers of epithelial cells. Solid areas resemble neuroendocrine tumor. In the present study, the tumor cells showed positivity for Vimentin, Cytokeratin, NSE, Progesterone and focal positivity for Chromogranin, Synaptophysin.<sup>(17,18)</sup> Diffuse positivity of CK and focal positivity of Synaptophysin is in concordance with Pettinato et al.<sup>(18)</sup>

Machado et al.,<sup>(14)</sup> reported PR positivity in 80% of their cases, in the present study we have seen 66.6%, positivity, similar to Uppin SG et al.,<sup>(11)</sup> who reported 64.5% positivity with PR in their case series. The frequent expression of PR indicates hormone dependency of the tumor.

The unbalanced translocation between chromosomes 13 and 17 and the genes flanking the breakpoints may prove to be markers for solid and cystic papillary epithelial neoplasm of the pancreas and provide insight into its histogenesis.<sup>19</sup> In the present study, we attempted to see the chromosomal abnormalities of SPPT in two cases, which revealed derivatives of 17<sup>th</sup>, 13<sup>th</sup>, and 9<sup>th</sup> chromosomes. (Fig. 3a & b)

Surgery is the prime modality of the treatment, which enables cure, and prolonged disease free survival if adequate surgical excision done. In the present study one of the case presented with pseudo cyst of pancreas after one year of follow up, which has been corrected surgically. There is no recurrence or metastasis noted in the present study, which supports the low malignant potential of SPPT.

## Conclusion

The diagnosis of SPPT should be considered in young females with the pancreatic mass comprising solid and cystic components, which is of a rare entity with low malignant potential. Surgical resection is curative.

## References

1. Frantz VK. Tumors of the pancreas. In: Blumberg CW, editors. Atlas of tumor pathology. VII. Fascicles 27 and 28. Washington, DC: Armed Forces Institute of Pathology; 1959:32-3.
2. Casadei R, Santini D, Calculli L, Pezzilli R, Zanini N, Minni F. Pancreatic solid-cystic papillary tumor: clinical features, image findings and operative management. JOP.2006;7(1):137-44.
3. Ulasan S, Bal N, Kizilkilic O, Bolat F, Yildirim S, Yildirim T, et al. Solid pseudo papillary tumor of the pancreas associated with dorsal agenesis. Br J Radiol 2005;78:441-3.
4. Rebhandl W, Felberbauer FX, Puig S, Paya K, Hochschoner S, Barlan M, et al. Solid pseudo papillary tumor of the pancreas (Frantz tumor) in children: Report of four cases and review of literature. J Surg Oncol 2001;76:289-96.
5. Vincent PJ, Nagpal BM, Singh P, Harjan MM, Nagi GS, Satyanarayan S. Gruber-Frantz tumor (Medical J Armed Forces India) MJAFI 2003;59:355-7.
6. Gupta ML, Ohri A, Singh J, Singla S, Singh D. Solid pseudo papillary tumor of the pancreas: A case report. Indian J Med Pediatr Oncol 2004;25:42-3.
7. Martin RCG, Klimtra DS, Brennan MF, Conlon KC. Solid-pseudo papillary tumor of the pancreas: a surgical enigma? Ann Surg Oncol. 2002;9(1):35-40.
8. Papavramidis T, Papavramidis S. Solid pseudo papillary tumor of the pancreas: Review of 718 patients reported in English literature. J Am Coll Surg 2005;200:965-72.
9. Yu PF, Hu ZH, Wang XB, Guo JM, Cheng XD, Zhang YL., et al. Solid pseudo papillary tumor of the pancreas: A review of 553 cases in Chinese literature. World J Gastroenterol 2010;16:1209-14.
10. Patil TB, Shrikhande SV, Kanhere HA, Saoji RR, Ramadwar MR, Shukla PJ. Solid pseudo papillary neoplasm of the pancreas: A single institution experience of 14 cases. HPB (Oxford) 2006;8:148-50.
11. Uppin SG, Hui M, Thumma V, Challa S, Uppin MS, Bheerappa N., et al. Solid-pseudo papillary 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.
12. Veerabhadrapa SK, Chandrapa PR, Roodmal SY, Shetty SJ, Madhushankari GS, Mohan kumar KP. Karyotyping: current perspectives in diagnosis of chromosomal disorders. Sifa Med J 2016;3:35-40.
13. Zhang H, Liang TB, Wang WL, Shen Y, Ren GP, Zheng SS. Diagnosis and treatment of solid-pseudo papillary tumor of the pancreas. Hepatobiliary Pancreat Dis Int 2006;5:454-8.
14. Machado MC, Machado MA, Bacchella T, Jukemura J, Almeida JL, Cunha JE. Solid pseudo papillary neoplasm of pancreas: Distinct patterns of onset, diagnosis, and prognosis of male versus female patients. Surgery 2008;143:29-34.
15. Vassos N, Agaimy A, Klein P, Hohenberger W, Croner RS. Solid-pseudo papillary neoplasm (SPN) of the pancreas: Case series and literature review on an enigmatic entity. Int J Clin Exp Pathol 2013;6:1051-9.
16. Chakhachiro ZI, Zaatari G, tari G. Solid-pseudo papillary neoplasm: A pancreatic enigma. Arch Pathol Lab Med 2009; 133:1989-93.
17. Santini D, Poli F, Lega S. Solid-pseudo papillary tumors of the pancreas: histopathology. JOP 2006;7:131-36.
18. Pattinato G, Di Vizio, Manivel JC, et al., Solid-pseudo papillary neoplasm of distinct and highly characteristic cytologic features. Diagn cytopathol 2002;27:325-334.
19. Grant LD, Lauwers GY, Meloni AM, Stone JF, Betz JL, Vogel S, Sandberg AA. Unbalanced chromosomal translocation, der(17)t(13;17)(q14;p11) in a solid and cystic papillary epithelial neoplasm of the pancreas. Am J Surg Pathol. 1996; 20(3):339-45.