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Original Research Article

Management of Solid Papillary Epithelial Neoplasm (SPEN) of Pancreas - Experience of Tertiary Care Centre

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ABSTRACT

Background: Solid Papillary Epithelial Neoplasm (SPEN) Of Pancreas is an uncommon tumor. The present article shares our experience of clinical presentation and management of this rare indolent pancreatic tumor

Materials and Methods: Retrospective analysis of our data of solid pseudopapillary epithelial neoplasm of pancreas was done from period from 2008 to 2015. Parameters taken into consideration were the patient's demographics, clinical presentation, biochemical investigations, tumor markers, radiological imaging, intraoperative findings, pathological report and survival

Results: Six patients of SPEN of pancreas were identified from our data base. All patients were female with mean age of 36.5 years. All patients had vague lump in abdomen with dull aching pain as their clinical presentation. In 4 patients the lesion involved the head, while in two the tail of the pancreas was involved. CECT Scan in all revealed heterogeneous mass in the pancreas. All patients were subjected to curative resection, with four undergoing Whipples procedure and two distal pancreatectomy. All patients are alive with follow up ranging from 7 to 90 months.

Conclusion: SPEN is an indolent neoplasm of pancreas; diagnosis is based mainly on radiological findings and confirmed by pathology. R0 surgical resection is curative for the patient with good 5 year survival.

Keywords: solid pseudopapillary epithelial neoplasm of pancreas.

INTRODUCTION

Solid papillary epithelial neoplasms (SPEN) of the pancreas are very rare accounting for only 0.17 to 2.7 per cent of all non endocrine tumors of the pancreas. [(1,2)]

Frantz was the first to report this neoplasm in 1959, [(3)] and he termed it "papillary tumor of the pancreas-benign or malignant". In 1970 Hamoudi et al [(4)] described the histological characteristics consisting of the presence of solid and papillary patterns, well vascularization and cystic necrosis. This tumor has been described by various terminologies including solid and cystic acinar cell tumor, papillary epithelial neoplasm, solid and papillary neoplasm, papillary-cystic tumor, solid cystic tumor, and papillary tumor of the pancreas (Frantz's tumor).In 1996, the world health organization renamed this tumor as solid-pseudopapillary tumor for the international histologic classification of tumor of the exocrine pancreas.

SPEN differs from other pancreatic neoplasms it being indolent nature with low malignant potential and predilection for young female patients.

Considering the uncommon occurrence of this tumor, the present article presents our experience of management of this rare tumor.

METHODS AND MATERIALS

The present study is an analysis of retrospectively maintained data. A total of 221 patients were identified to have presented with pancreatic neoplasm from year 2008 to 2015 .Of these 6 patients (2.7%) were found to have Solid Pseudo-papillary epithelial neoplasm of Pancreas. We analyzed their data with respect to their demographics, clinical presentation, radiological findings, tumor markers, surgical procedure, intraoperative findings, postoperative course, histopatholgy, follow-up and survival.

Data was analyzed and expressed as median value.

RESULTS

Demographics

A total of 221 patients were identified to have presented with pancreatic neoplasm during the study period year 2008 to 2015 .Of these 6 patients (2.7%) were found to have Solid Pseudo-papillary epithelial neoplasm of Pancreas. All the six patients were female, with their age ranging from 25-46 years (median 36.5 years).

Clinical Presentation

All six patients presented with vague abdominal mass with a dull aching pain (Figure 1). None of the patients had pressure symptoms either in form of gastric outlet obstruction or jaundice. The biochemical investigations and CA 19-9 values were normal in all the patients.

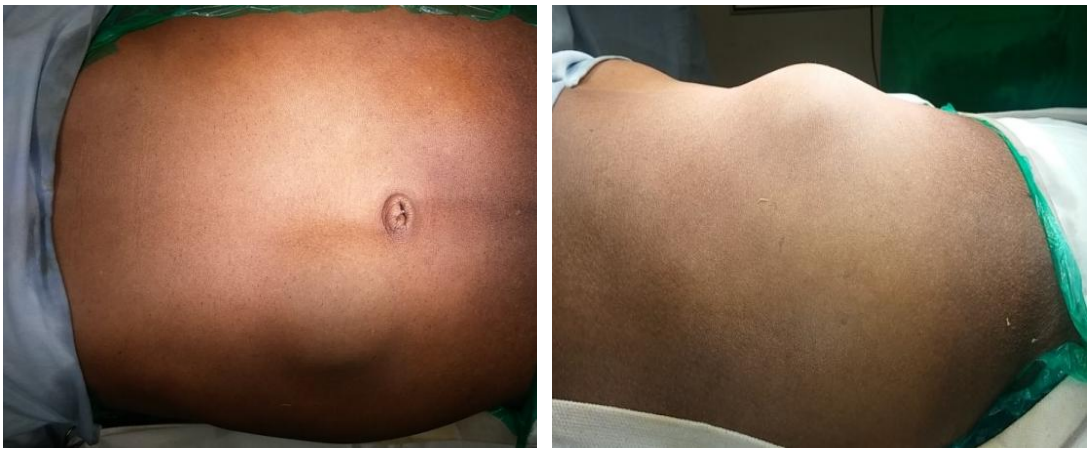

Figure 1: A & B: Lump in abdomen

Tumor characteristics on Imaging:

All the patients were referred to us with USG of abdomen done, which showed a mixed echogenic mass in the abdomen with cystic and solid areas. However the exact organ of origin could not be determined on USG.

On admission with us, these patients were subjected to Contrast enhanced CT scan of abdomen and pelvis (Figure: 2). It revealed heterogenous mass more than 10 cm in size, arising from pancreatic head in 4 patients. The tumor was pushing the superior mesenteric vein and portal vein on left side, without invading them. There were no features suggestive of compression of either biliary system or duodenum. There was no evidence of metastatic disease elsewhere in the abdomen. Two patients were having heterogenous mass arising from tail of the pancreas.

Surgical Intervention

All patients underwent R0 surgical resection with four being subjected to

classical Whipples procedure, and two distal Pancreatectomy with splenectomy. In classical Whipples procedure 2 patients underwent pancreatico-jejunostomy by dunking method as the duct could not be identified and two patients undergoing duct to mucosa pancreatico-jejunal anastomosis (Figure 3). Patients with tumor involving the tail underwent distal pancreatectomy with splenectomy.

Morbidity and Mortality

One of the three patients who underwent Whipples procedure developed Type 1 biochemical pancreatic fistula which was managed successfully with conservative treatment. One patient who underwent distal pancreatectomy with splenectomy developed surgical site infection. There was no mortality in post-operative period. All patients recovered well, with their median hospital stay of 11 days (7-14 days)

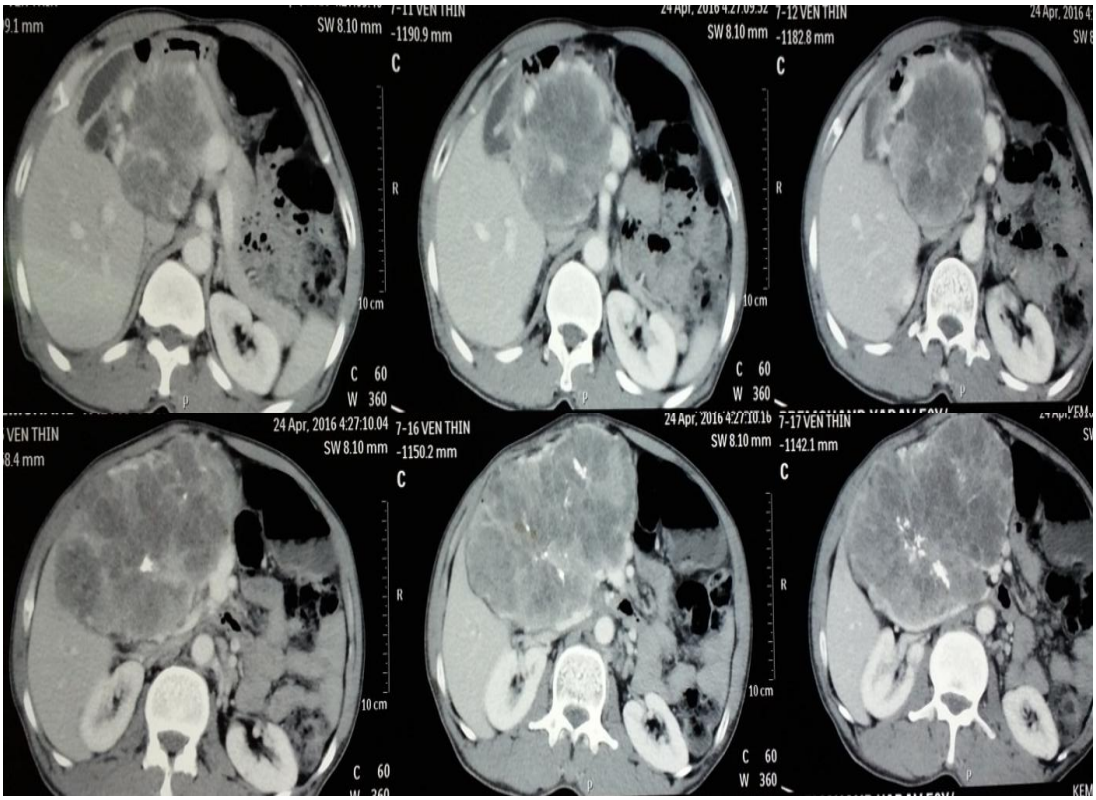

Figure 2: CECT Scan of the abdomen and pelvis

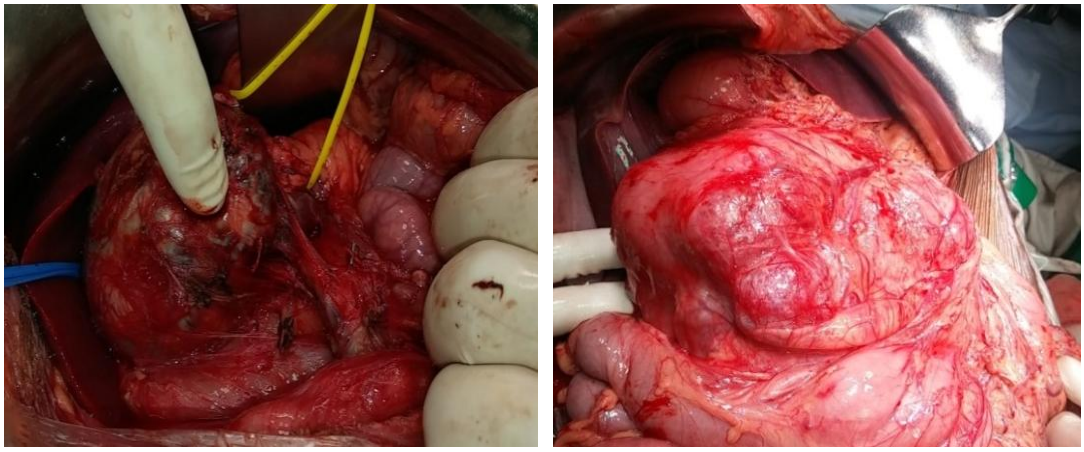

Figure 3: Intraoperative Findings- SPEN arising from head of pancreas

Pathology

On gross examination all tumors were measuring more than 10 cm with solid and cystic areas and hemorrhagic areas within. (Figure 4)

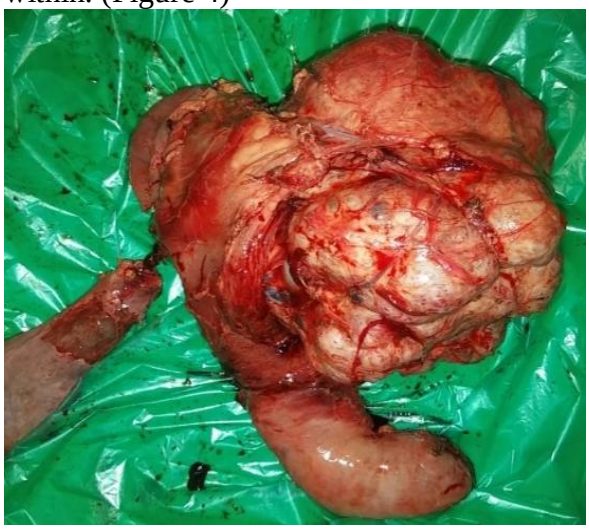

Figure 4: Gross specimen of SPEN arising from head of pancreas

On microscopic examination all patients had papillary pattern with areas of cystic necrosis. The margins were negative in all, and none had lymph node metastasis.

Follow up and Survival

The median follow up period was 57 months (11 to 97 months). All the patients are surviving to date. These patients were followed up routinely initially every two months for 6 months, every three months up to 2 years and there after every 6 months up to 5 years and then annually. Patients were evaluated with clinical examination and CECT Scan and biochemical investigations annually. All patients are recurrence free. The Characteristics of patients is as given in Table 1.

Age	Sex	Presentation	Intraop Findings	Procedure Performed	Resection Margin	Follow Up
25	F	Abdominal mass, dull aching pain	Tumor >10 cm, head region	Whipples procedure	R0 resection	Post operative recovery uneventful
30	F	Abdominal mass, dull aching pain	Tumor >10 cm, head region	Whipples procedure	R0 resection	Post operative pancreatic fistula , recovered conservatively
34	F	Abdominal mass, dull aching pain	Tumor >10 cm, tail region	Distal pancreatectomy with spleenectomy	R0 resection	Post operative superficial surgical site infection, recovered uneventfully
41	F	Abdominal mass, dull aching pain	Tumor >10 cm, head region	Whipples procedure	R0 resection	Post operative recovery uneventful
39	F	Abdominal mass, dull aching pain	Tumor >10 cm, head region	Whipples procedure	R0 resection	Post operative recovery uneventful
46	F	Abdominal mass, dull aching pain	Tumor >10 cm, tail region	Distal pancreatectomy with spleenectomy	R0 resection	Post operative recovery uneventful

DISCUSSION

SPEN is an uncommon exocrine neoplasm of the pancreas accounting for 6% of all exocrine pancreatic neoplasm. It has predilection for young female patients, with female to male ratio of 10:1, and is commonly seen in first three decades. In the present study too it accounts for 2.6% of the pancreatic tumors presenting, all patients were female with their median age being 36.5 years. As in present study, patients present with a slow growing abdominal mass with or without abdominal pain. There is no specific clinical syndrome for SPEN of the pancreas, and the disease tends to be misdiagnosed. Patients often describe their symptoms as an "indescribable" abdominal distension. Besides, there is no evidence of pancreatic insufficiency, abnormal liver function tests, cholestasis, elevated pancreatic enzymes or an endocrine syndrome. Hence diagnosis is frequently delayed as a result of which tumor size at presentation is frequently large. Laboratory tests such as serum amylase or pancreatic tumor markers such as CA19-9, Carcino-embryonic antigen are usually normal. [(5)] A diagnosis of SPEN of the pancreas is therefore considered, especially if the patient is a young woman with no other cause for pancreatic masses. However the diagnosis should not be discounted in male patients. Very often the tumor is diagnosed incidentally on radiological examination.

SPEN has a benign course with a very low-grade malignant potential. [(6)]

Hence if the tumor is recognized early, complete curative resection is possible resulting in complete cure. CECT Scan is the imaging modality of choice for the diagnosis of SPEN. On CECT Scan [7] it is classically seen as a heterogeneous mass with cystic areas in the center and solid components at the periphery. Larger tumors are usually well encapsulated. The enhancement of its capsule and the solid component similar to pancreas in both arterial and venous phase helps to differentiate SPEN from adenocarcinoma and neuroendocrine tumor of pancreas which are the common differential diagnosis of SPEN. Based on enhancement, SPEN tumors can be differentiated from adenocarcinoma and Neuroendocrine tumors. SPEN tumors demonstrate no enhancement of the cystic portions, slight enhancement of the solid portions in the arterial phase and marked enhancement in the portal venous phase. [8] In comparison, adenocarcinoma is hypoattenuating in venous phase whereas neuroendocrine tumor is hyperattenuating in arterial phase. Displacement of adjacent structure rather than its invasion is a feature seen on CECT Scan in patients with large sized SPEN. This was a feature seen in our study too. Differentiation of SPEN from other pancreatic neoplasms especially pancreatic adenocarcinoma is important, as though the appearance of SPEN on CECT looks very aggressive and seems difficult to resect, radical surgery in these patients give a complete cure with 5 year survival more than 90 %. However CECT Scan has limitations in characterizing hemorrhage, cystic degeneration or the integrity of the tumor capsule, features characteristic of SPEN. [9]

MRI owing to its superior contrast resolution, displays capsule and intratumoural hemorrhage better than CT scans. Another indication of use of MRI is presence of smaller tumors (<3.0cm) with atypical imaging characteristics of for SPEN. At such times percutaneous biopsy too may be done for confirmation of diagnosis.

Surgery is the only curative treatment for SPEN of the pancreas, [10] the aim of surgery being complete, and R0 resection. Tumor size is not a predictor of resectability. Large tumors as in our study, depending on their location can be subjected to either pancreatic duodenectomy (if involving the head region) or distal Pancreatectomy (if involving the body and tail). In patients with distant hepatic metastasis or local recurrence, if the lesion is resectable, surgery is not a contraindication. [11,12]

Too large SPENS invading adjacent structures or SPEN with widespread metastasis are deemed as unresectable tumors. Radiotherapy is the treatment in these unresectable tumors as SPEN tumors are radiosensitive.

Local Recurrence is uncommon though distant metastasis can occur in liver. 10 to 15% tumors are found to be malignant as having evidence of metastases or invasion of adjacent structures. Metastasis is known to occur at mean interval of 8.5, warranting long term follow up with periodic imaging. [13] These tumors very rarely recur or metastasize to other organs, so properly planned surgical resection is curative in these patients.

SPEN tumors have a characteristic histologic and cytologic appearance, comprising predominantly of solid, pseudopapillary and cystic regions. The neoplastic cells are uniform and polygonal with abundant cytoplasm and contain oval, regular nuclei. Cellular heteromorphism and mitotic phase may be seen some patients.

Prognosis of patients with SPEN tumors is good, even with local recurrence and metastasis. Patients with unresectable SPEN too have a long survival time. The overall 5-year survival rate of patients with SPEN is about 95%. [14]

CONCLUSION

Solid pseudopapillary epithelial neoplasm of the pancreas is a low-grade

malignant tumor and is prone to occur in young women. Though there are no specific diagnostic laboratories investigations, CECT scan and MRI helps in diagnosis. Comprehensive analysis of clinical features and imaging confirms diagnosis. Surgical resection is a treatment of choice. It has characteristics histopathologic features with papillary pattern with areas of cystic necrosis. Overall prognosis of SPEN tumors is good with 5 year survival rate more than 90%

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