

## Solid Pseudopapillary Neoplasm: An Unusual Neoplasm of Pancreas

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### Abstract

### Case Report

Solid pseudo papillary tumor of pancreas is a rare neoplasm which has low malignant potential and good prognosis. It constitutes 0.2% to 2.7% of pancreatic tumors. These tumors usually occur in younger females presenting with nausea, vomiting, abdominal pain and abdominal fullness. We report a case of 28 years female presenting with abdominal pain. Ultrasound abdomen and CECT abdomen revealed pancreatic mass of benign nature. Histopathological examination confirmed the diagnosis.

**Keywords:** Pancreatic tumor, Low malignant potential.

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## INTRODUCTION

Solid pseudo papillary tumor of pancreas is an unusual pancreatic tumor representing 0.2 to 2.7% of all the pancreatic tumors [1]. It has been first described in 1959 by Frantz [2]. These tumors mainly occur in Juvenile females and have low malignant potential [3]. Malignant changes have been reported in 10% to 15% of cases as the tumor grows and the most common site of metastasis is liver [4].

Pathogenesis of this tumor is still unclear. Extensive application of imaging structures such as ultrasonography, computed tomography and magnetic resonance imaging helps in detecting lesions. Histopathology remains the gold standard for diagnosis. Treatment of choice is the surgical resection.

## CASE HISTORY

28 years female presented with history of abdominal pain since 25 days which was non radiating. There was no history of fever, jaundice, loss of weight, loss of appetite or altered bowel habits. On physical examination there was no organomegaly.

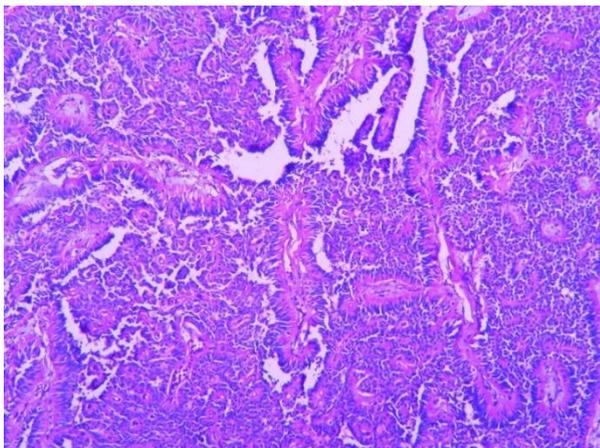
Haematological investigations showed haemoglobin 10.6g/dl, total WBC count – 11,300/cumm, RBC count-3.9millions/cumm, platelet count-3,13,000/cumm, PCV-35Vol%, MCV-88fl. Biochemical investigations revealed serum total

bilirubin - 0.43mg/dl, serum bilirubin- 0.25mg/dl, Serum glutamic oxaloacetic transaminase (SGOT) - 28U/L, total protein 6.06g/dl, serum albumin – 3.08g/dl, globulin-3.0g/dl, serum amylase - 214 v/l, Sodium-143 meq/l, Potassium-3.7 meq/l and Chloride-102mg/l. Ultrasound abdomen showed pancreas with partially distended body and tail and a large heterogeneously hypoechoic solid lesion measuring 5.6x4.2cm with few anechoic areas in the region of head of pancreas. Contrast enhanced computed tomography (CECT) abdomen revealed well defined heterogeneously enhancing mass measuring 6.3x5.2m originating in the region of pancreatic head causing compression on the inferior vena cavae (IVC). Heterogenous nature of the lesion was constituted by scattered areas of fluid density interspersed with irregular areas of soft tissue. There was no obvious calcifications. Benign nature of the lesion was suggested by displacement of adjacent CBD and D2 segment of duodenum laterally, IVC posteriorly, portal confluence medially. Endoscopic ultrasonography revealed a mass of size 5.6x4.7cms seen in the head of pancreas with heterogenous echotexture, internal cystic spaces and sparse vascularity. Portal vein was compressed but not infiltrated. Tumor was enucleated and specimen was sent for histopathological examination. We received soft tissue mass measuring 7x6x1.5cm with hemorrhagic external surface. Cut section showed grey white solid and cystic areas. Cystic areas were filled with papillary excrescences (Figure-1).

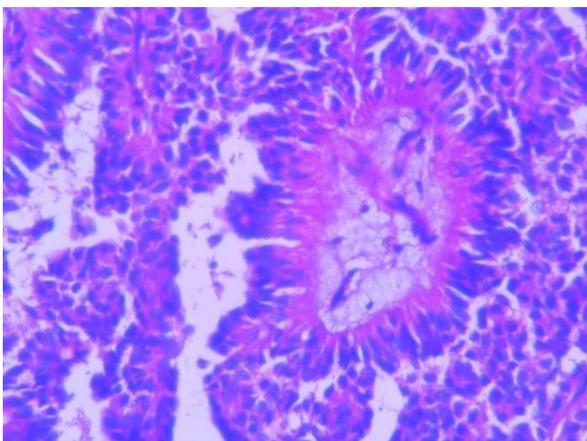


**Fig-1: Cut section showed grey white solid and cystic areas filled with papillary excrescences**

Histopathological examination revealed circumscribed tumor composed of uniform population of tumor cells admixed with delicate capillaries. Tumor cells were arranged in sheets, pseudopapillary pattern and occasional of glandular pattern (Figure-2). Few foci shows perivascular pattern of arrangement. Tumor cells have round to oval nuclei and eosinophilic cytoplasm (Figure-3).

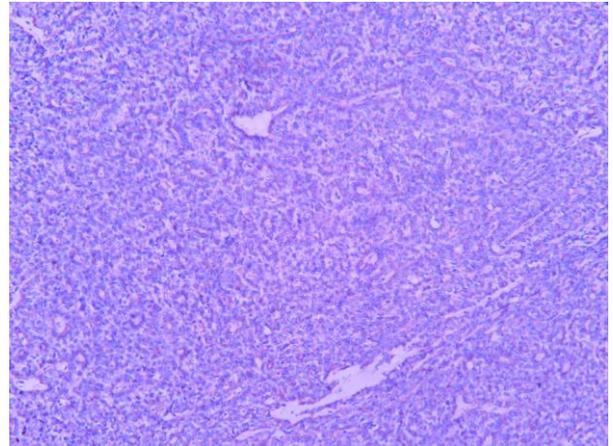


**Fig-2: Tumor cells arranged in pseudo papillary pattern (H&E, X100)**

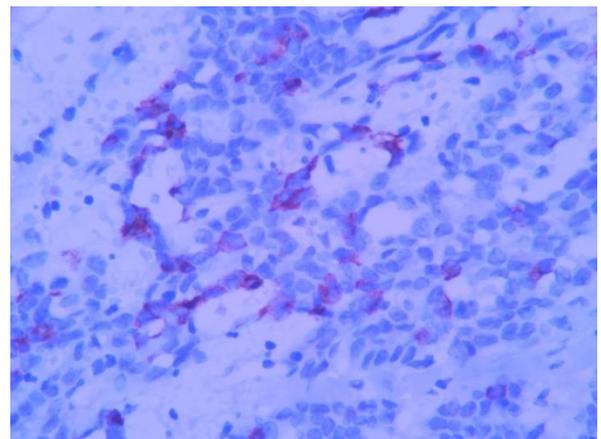


**Fig-3: Tumor cells arranged in pseudo papillary pattern with nuclei of tumor cells oriented away from vessel (H&E, X400)**

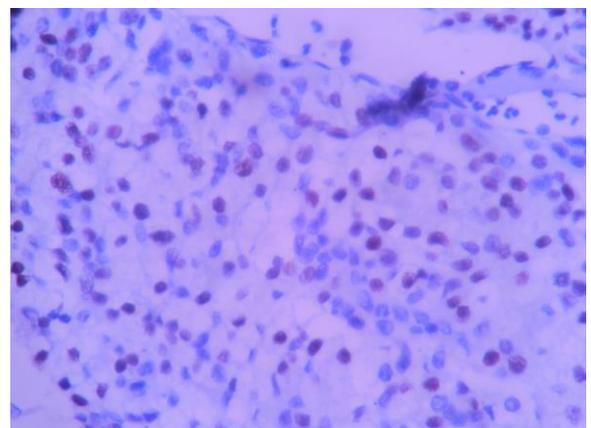
Few foci shows collection of foamy macrophages and hyaline globules. Immunohistochemistry shows positive for Vimentin (Figure-4), focal positivity with synaptophysin (Figure-5) and progesterone receptors (Figure-6), negative for chromogranin. Depending on the above histopathological and immunohistochemical findings, diagnosis of solid pseudopapillary tumor was made.



**Fig-4: Tumor cells showing positivity with vimentin (Vimentin, X100)**



**Fig-5: Tumor cells showing focal positivity for synaptophysin (Synaptophysin, X400)**



**Fig-6: Tumor cells show positivity with Progesterone receptors (PR, X400)**

## DISCUSSION

Solid pseudo papillary tumor, also termed as Frantz tumour is a rare pancreatic neoplasm which has low malignant potential. It shows female predilection and occurs at younger age group [5] Histogenesis of this tumor is not clear due to its rarity [1].

Solid pseudo papillary tumors occur in the pancreas and also in the extra pancreatic sites like mesentery, retroperitoneum, greater omentum, duodenum, stomach, liver, ovary or lung [6] WHO has categorised this tumour as borderline tumour of the pancreas in 1996 [7]. These tumors present often with non-specific symptoms like abdominal discomfort or pain or abdominal fullness associated with nausea. Nearly 15% of the patients will be asymptomatic. [1].

Imaging techniques like ultrasound, computerised tomography and magnetic resonance imaging are helpful in diagnosis. On imaging the tumour appears as well circumscribed neoplasm with solid and cystic components. On ultrasound this tumor appears as well circumscribed mass, which is surrounded by a pseudo capsule formed by reactive fibrosis and compressed pancreatic tissue [8].

Contrast enhanced CT shows mild enhancement of both the solid component of the tumour and tumour pseudo capsule [9]. It also identifies areas of haemorrhages and calcification. In children MRI is considered as best technique for detecting the tumor as there is no radiation in this technique. MRI is preferred due its improved ability to visualise tumor components such as intratumoral hemorrhage. Intratumoral hemorrhage and fibrous capsule are clues for diagnosis as these findings are rarely present in other pancreatic neoplasm [10].

Preoperative ultrasound guided FNAC may be helpful in diagnosis. The preoperative cytological diagnosis of the tumor helps to plan the surgical treatment with preservation of uninvolved pancreas. Cytology smears will be highly cellular with tumor cells arranged as papillary structure with fibrovascular core. Pseudo rosette pattern can also be present [11].

Grossly cut section of the tumor has variegated appearance with solid, cystic components, areas of hemorrhages and necrosis. On histopathological examination, tumour has a pseudo capsule and is composed of solid and cystic components. Solid areas consists of discohesive polygonal tumor cells surrounding the delicate vascular channels, forming the pseudo papillae. In this region nuclei will be arranged away from vessels, which results in cytoplasmic zone separating the nuclei from capillaries. Mucopolysaccharide rich ground substance can be present. Collection of foamy macrophages, coagulation necrosis, cholesterol clefts, peripheral blood pools, intracytoplasmic hyaline globules can also be present in

this tumor [7]. Immunohistochemically, tumor cells are positive for Vimentin, Alpha-1 antitrypsin, Neuron specific enolase, Progesterone receptors. They variably express Synaptophysin, Cytokeratin and Cyclin D1. Tumor cells are negative for Chromogranin, Somatostatin, Insulin, Glucagon, Lipase, Calretinin, Estrogen receptors, S-100, EMA and Placental alkaline phosphatase.

Solid pseudo papillary tumor should be differentiated from other tumors like pseudocyst of pancreas due its cystic component and well differentiated pancreatic endocrine neoplasm as this tumor also have uniform population of tumor cells.

Pseudocyst of pancreas resemble solid pseudo papillary neoplasm on imaging. Pseudocyst are more common in men and patient has history of recurrent pancreatitis and raised serum amylase. In contrast solid pseudo papillary neoplasm of pancreas is common in women, do not have associated pancreatitis and serum amylase levels will be normal. Histologically, pseudocysts has no lining where as solid pseudo papillary neoplasm has tumor cells at periphery [12].

Well differentiated endocrine neoplasms resemble solid pseudo papillary neoplasm on microscopy due to the uniform population of tumor cells. However presence of pseudo papillae, foamy macrophages, cholesterol clefts and eosinophilic hyaline globules favours the diagnosis of solid pseudo papillary neoplasm. Immunohistochemically endocrine neoplasms of pancreas are positive for Synaptophysin, Chromogranin, and pancreatic hormones like insulin, glucagon and somatostatin whereas solid pseudo papillary neoplasms are negative for these markers and only show variable positivity with Synaptophysin. Well differentiated endocrine neoplasms of pancreas have tumor cells with coarse speckled "salt and pepper" chromatin pattern which is absent in solid pseudo papillary neoplasm [13].

Treatment of choice is surgical resection. If the tumor is not large to produce compression or if there is no involvement of adjacent organs, enucleation or local resection of tumor is preferred [14]. If the tumor is large pancreateo duodenectomy or distal pancreatectomy can be performed. This prevents loss of normal pancreatic parenchyma which may affect the exocrine and endocrine function of pancreas

In few cases where the tumor is unresectable and has metastasis, radiotherapy and chemotherapy can be applied to increase the survival of patient [15].

## CONCLUSION

Solid pseudo papillary tumor is a rare pancreatic tumor having female predilection and presents with vague symptoms like pain in abdomen and abdominal mass. This tumor has low malignant

potential. Imaging techniques and cytology gives clue for preoperative diagnosis which helps surgeon for planning proper treatment, thus preventing the unnecessary loss of normal pancreatic tissue.

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