Abstract:
The solid-pseudopapillary neoplasm is a rare solid-cystic neoplasm of pancreas which has characters of benign tumors predominantly. There has been very low incidence of malignant transformation and metastasis. Hence surgical excision becomes mandatory and it is curative. This report brings two cases of this rare tumor with indolent nature necessitating Whipple’s Pancreatico-duodenectomy.

Keyword: SOLID-PSEUDOPAPILLARY NEOPLASM OF PANCREAS, SOLID-CYSTIC NEOPLASM OF PANCREAS, GRUBER TUMOUR

INTRODUCTION
The neoplasms of pancreas are classified based on the tumor content by computed tomography imaging as solid and cystic neoplasms. The cystic neoplasms of pancreas are considered malignant unless proven otherwise. These neoplasms are indolent in presentation and rarely do they present early without any spread. The solid-pseudopapillary neoplasm is a rare solid-cystic neoplasm of pancreas which has characters of benign tumors predominantly. There has been very low incidence of malignant transformation(< 5%) and metastasis(5 – 10%) in these patients[6,7]. Hence surgical excision becomes mandatory and the pathologist only can decide the rare incidence of malignancy in these cases.

CASE REPORT
Case 1: A 38 years old Hindu female agricultural laborer from Madurai presented with complaints of abdominal pain for 6 months which has insidious onset and progressively disturbing her daily activity. Pain is confined to the upper abdomen and radiates to the back. No complaints of fever, hematemesis, malena, jaundice, bowel disturbances. No past history of chronic medical illness.
No previous surgeries. She attained menarche at 14 yrs. and has regular menstrual cycles. She has two children by full term normal vaginal delivery. Last child birth was 15 years earlier. Her general physical examination was unremarkable. On examination of abdomen, well defined mass of size 6x6 cm is palpable in the epigastrium, which is not moving with respiration. Percussion over the mass was impaired. No organomegaly/free fluid. Her hemoglobin was 9 g/dl and blood counts and blood chemistry were non-contributory. Chest skiagram was normal. The markers for pancreas - amylase, lipase and CA 19-9 were within normal limits. There is no history of pancreatic tumours/cancer running in the family. Ultrasonogram of abdomen showed an 8x8 cm cystic lesion involving head of pancreas with altered echoes internally. CECT abdomen detected a hypo intense, 8x8 cm, cystic lesion with areas of hemorrhage involving the head of the pancreas. OGD scope study was normal. Patient was prepared for surgery with the pre op diagnosis of cystic lesion of pancreas for wide excision. Intra-op finding was a cystic lesion of size 8x8 cm involving the head of the pancreas, no lymph nodes, no ascites, and no sign of metastasis. Since the mass was very close to c-loop of duodenum, the resection of which will involve injury to the duodenum, it was decided and proceeded with pylorus preserving pancreatecoduodenectomy (modified whipple’s procedure). Post-operative period was uneventful and patient was discharged on post op-day 15. HPE was suggestive of Solid pseudo-papillary tumor of pancreas with no evidence of malignancy. IHC was positive Keratin, desmoplakin, trypsin, chymotrypsin, amylase. Patient is on regular 6 monthly follow up.
Case 2: A 22 years old Hindu female home maker from Salem presented with complaints of abdominal pain for 3 months which has insidious onset and progressively increasing with radiation to back, disturbing her daily activity. Pain is confined to the upper abdomen. No complaints of fever, hematemesis, melena, jaundice, altered bowel habits. No past history of chronic medical illness. No previous surgeries. She attained menarche at 14 yrs. and has regular menstrual cycles. She has one child by full term normal vaginal delivery 2 years earlier. Her general physical examination was unremarkable. On examination of abdomen, well defined mass of size 8x8 cm is palpable in the epigastrium, which is not moving with respiration. Percussion over the mass was impaired. No organomegaly, no free fluid. Her hemoglobin was 12 g/dl and blood counts and blood chemistry were non-contributory. Chest skiagram was normal. The markers for pancreas - amylase, lipase and CA 19-9 were within normal limits. Ultrasonogram of abdomen showed a 10x8 cm cystic lesion probably arising from inferior surface of liver with altered echoes internally – provisionally diagnosed as hydatid cyst. CECT abdomen detected an hypo intense, 12x10 cm, cystic lesion with areas of solid structures involving - ? Inferior surface of liver/ ? Lesser sac with features suspicious of hydatid cyst.

Figure 4: CECT Abdomen of patient 2 – AXIAL SECTION
MRCP showed features suggestive of cystic neoplasm of head of the pancreas. OGD scopy study was normal. Patient was prepared for surgery with the pre op diagnosis of cystic lesion of pancreas for wide excision. Precautions were taken as there was suspicion of hydatid disease. Intra-op finding was a solid-cystic lesion of size 15x10 cm involving the head of the pancreas very close to portal vein, Liver was normal, no lymph nodes, no ascites, and no sign of metastasis. Since the mass was very close to portal vein, proceeded with classical pancreatoduodenectomy(whipple’s procedure) with portal venorrhaphy for injury at the junction of SMV and PV. Post operatively patient had reactionary hemorrhage and re-exploration and corrective surgery was done. Postoperative course was uneventful otherwise and patient was discharged on post op-day 25. HPE was suggestive of Benign Solid pseudo-papillary tumor of pancreas. IHC was positive Keratin, desmoplakin, trypsin, chymotrypsin ,amylase and progesterone receptor. Patient is on regular follow up.
Figure 5: Resected specimen of Head of Pancreas of Patient 2

DISCUSSION:
The presentation of cystic lesions of pancreas should be considered malignant unless proven otherwise. In 1959 Frantz described a third type termed papillary tumour of the pancreas [3]. Our case belongs to the category of cystic papillary tumour of the pancreas or Gruber-Frantz tumour of which only 150 cases have been reported in the literature. It has a low grade malignant potential manifested by invasion of the capsule and contiguous structures. The cell of origin of these rare tumours is unknown; investigators have suggested an origin from primordial pancreatic cells and it has been classified under neoplasms of indeterminate type [4]. It begins as a solid mass in which there are many poorly supported tiny vessels. Cells farthest from the vessels degenerate where as the cells near the vessels remain intact resulting in a pseudopapillary pattern and cystic spaces. Groups of foamy macrophages accumulate along with giant cells [4,5]. Gruber-Frantz tumours characteristically occur in young females as it was in our case [6]. The most frequent symptom of these tumours is upper abdominal pain seen in nearly half the patients; this was the only presenting symptom in our case. Symptoms may be present for years before correct diagnosis is made. The most important distinction to be made is the differentiation of cystic neoplasms from other cystic lesions of the pancreas, particularly pancreatic pseudocyst. This distinction is often very difficult without an operative biopsy. All these tumours should be viewed as malignant tumours and treated with anatomic pancreatic resection [1]. The diagnosis of a “benign” cystic neoplasm can only be made confidently with pathologist’s assistance because a small focus of malignant epithelium may be found in an otherwise benign lesion [2]. The Immunohistochemistry of these tumours show Positivity for Keratin, Desmoplakin, Trypsin, Chymotrypsin, Amylase, Vimentin; Focal positivity for Neuron specific enolase, Glucagon, Insulin; Progesterone receptor positive and hence also considered a hormone dependent tumor[1,3,6] A wide local excision would suffice treatment for this lesion if possible and no compromise should be made for an R(0) resection as this surgical exercise would be the prime and sole management option, with 10 year survival approaching 100% [6,7].

References


