Abstract: Background Solid pseudo papillary neoplasia (SPN) of the pancreas is an extremely rare epithelial tumor of low malignant potential. SPN accounts for less than 1 to 2% of exocrine pancreatic tumors. The aim of this Case report is to enumerate our experience with SPN of the pancreas of a young female patient and to give a brief review of literature. Case Report A 21 year young girl with symptom of vague abdominal pain in the epigastric region for 30 days with no other symptoms presented to us. On Abdomen Examination no mass was palpable. USG and CECT scan done showed a well capsulated mass lesion in tail of pancreas of size 7cm x 9cm x 9cm dimension. The lesion was hetero dense with no vascular or adjacent organ involvement. Endoscopic FNAC was done which showed the diagnosis of Solid Pseudo papillary neoplasm. With this diagnosis, we proceeded with distal pancreatectomy and Splenectomy. Post-operative period was uneventful Patient recovered well and discharged on 10th Post op day. The specimen report came as SPN with IHC positive for Vimentin and Neuron Specific Enolase. The margins are clear and no nodes are positive. Hence no adjuvant Chemotherapy given. The patient is on follow up past one year without any recurrence. Conclusion Solid Pseudo papillary tumors must be diagnosed with high index of suspicion because of their rarity and all these patients must be taken up for Surgical removal. Surgery must aim at removing as much as the tumor as possible with negative margins and in cases where there is extensive involvement debulking surgeries also provide definitive improvement in survival. Keywords- SPN, Low malignant potential, Heterodense lesion, Distal Pancreatectomy.
Introduction:
Solid pseudopapillary tumor of the pancreas is a rare but characteristic neoplasm first described by Frantz in 1959 and 1. It afflicts young women, with a 10:1 predominance over men, at an average age of 24 years. Presenting features are usually vague and include abdominal pain, fullness, nausea, and vomiting due to a bulky tumor (mean size 11 cm) compressing local structures in the upper abdomen. The tumor is most commonly, but not always, localized to the tail of the gland. So far only 700 cases are reported in the literature. This paper gives a brief review of the rare case of SPT operated in our Institute with review of literature.

Case report:
21 years young girl presented with Abdominal Pain, which is dull aching and intermittent with no radiation for past 30 days. The patient had no vomiting, jaundice or melena. On examination her Abdomen no mass was palpable. Doppler USG done showed a hetero echoic mass lesion in the tail of pancreas with normal flow in splenic vein and Portal vein. No other lesions are seen on USG abdomen. All routine Blood investigations, Serum Amylase and Tumor Markers- CA 19-9 and CEA are normal. CECT abdomen done showed a single well encapsulated, Isodense heterogenous of size 7* 9* 9 cm in tail of pancreas. Pancreatic Duct is not dilated (Figure 1 a and 1 b). CT arteriography shows the lesion displacing splenic artery Supero- posteriorly (Figure 2).

Figure 1 a. CECT Scan Showing the lesion in AXIAL view
Endo USG guided FNAC done showed clusters of cells with foci of papilloid structures with magenta colored fibrous core to which tumor cells are attached. Tumor cells are oval to polygonal with eccentric nuclei and inconspicuous nucleoli. Occasional nuclear grooves are seen. Final Impression is solid pseudo papillary tumour of pancreas.

Under Epidural/ General Anesthesia, Roof top incision done and on exposure of the abdomen a tumor was found in the body and tail of pancreas with no infiltration to adjacent structures. Spleen along with tail and body of pancreas mobilized and the splenic vessels ligated (Figure 3) The neck of the pancreas stapled and divided using white load stapler (Figure 4) The remnant pancreas stump over sewn with 3-0 mersilk. One drain tube kept in left hypochondrium and removed on 3rd Post op day.

Post op period was uneventful. Patient recovered well and discharged on 10th Post op day. HPE report of the specimen (Figure 5) showed neoplasm composed of solid sheets, pseudo rosettes, Pseudo papillary configuration with Uniform round cells (Figure 6). Focal nuclear grooving, Foam cells and Focal areas of hyalinisation. Final Impression is solid pseudopapillary tumour of pancreas, margins free-2/2 nodes reactive changes only. Immunohistochemical analysis done showed positivity for Vimentin and Neuron specific Enolase.

As the margins were free and there was no evidence of malignancy no adjuvant chemotherapy was given. The patient is still on follow up for the past 1 year and there is no recurrence.

Discussion:
SPN is very rare; in fact, they only constitute about 5% of cystic pancreatic tumors and about 1 to 2% of exocrine pancreatic neoplasms. They present mainly in the second and third decades of life. The origin of solid pseudo papillary tumors still remains unclear. These neoplasms have been suggested to have a ductal epithelial, neuroendocrine, multipotent primordial cell, or even an extra-pancreatic genital ridge angle-related cell origin.

The most common symptom is non-specific abdominal pain and a few patients present with gradual abdominal mass with compressive symptoms. Usually there is no evidence of pancreatic insufficiency, abnormal liver function tests, cholestasis, elevated pancreatic enzymes or an endocrine syndrome. Tumor markers are also generally unremarkable. In one series by yagci et al, seven patients (70%) presented with abdominal pain or abdominal dullness.
Ki-67 and sarcomatoid areas may be associated with aggressive high mitotic rate, immunohistochemistry findings of expression of pancreatic parenchyma. A recent study showed that some perineural invasion; and 3) deep invasion of the surrounding which are classified as ‘SP carcinoma’ 1) angioinvasion; 2) Adenomatoid tumors have already metastasized at the time of presentation13. The most common sites for metastasis are the liver, regional lymph nodes, mesentery, omentum and peritoneum. Once the diagnosis of SPN is made, surgery is the first choice of treatment. SPN is usually surrounded by a pseudocapsule and exhibits benign or low-grade malignancy. Conservative resection with preservation of as much pancreatic tissue as possible is the treatment of choice. Surgery differs based on location of tumor. It may extend from Simple Enucleation, Distal Pancreatectomy + Splenectomy, Pancreaticoduodenectomy to even total pancreatectomy. Extensive lymphatic dissection or more radical approaches are not indicated when the disease is localized. Local invasion and metastases are not contraindications for resection. Portal vein resection is advocated when there is evidence of tumor invasion. For the metastases, surgical debulking should be performed, in contrast to other pancreatic malignancies. Metastases can be removed with enucleations or lobectomies and some patients with unresectable SPN may also have a long term survival13. The overall five-year survival rate of patients with SPN is about 95%6. Though most of these tumors are benign, some may be considered low grade malignancies with local invasion into contiguous structures and occasional distant metastasis (roughly 15–20% of cases). Malignant SPT is more common in old age and ames (Lam et al 1999). Criteria which distinguish potentially malignant tumors and which are classified as ‘SP carcinoma’ 1) angioinvasion; 2) perineural invasion; and 3) deep invasion of the surrounding pancreatic parenchyma. A recent study showed that some histological features, such as extensive necrosis, nuclear atypia, high mitotic rate, immunohistochemistry findings of expression of KI-67 and sarcomatoid areas may be associated with aggressive behavior14.

Adjuvant therapy is used only in a small number of patients because of the high resectability of SPN. The role of chemotherapy or chemoradiotherapy in the treatment of SPN is also unclear. In some studies, adjuvant chemotherapy and radiotherapy are reported in some unresectable cases with good results.15,16. Neoadjuvant chemotherapy or chemoradiotherapy is also reported to have been successful in a few cases.17. We didn’t advice adjuvant chemotherapy for our case because of the complete R0 resection of the lesion and absence of malignant features in the pathology report.

Conclusion:
Solid Pseudopapillary tumors must be diagnosed with high index of suspicion because of their rarity and all these patients must be taken up for surgical removal. Surgery must aim at removing as much as the tumor as possible with negative margins and in cases where there is extensive involvement debulking surgeries also provide definitive improvement in survival.

Conflict of Interest:
There is no Conflict of interest

References