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# FRANTZ'S TUMOR IN 13 YEAR CHILD- A CASE REPORT

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**ABSTRACT** Solid pseudo papillary tumor (SPT) or Frantz's tumor is a slow-growing low-grade malignant tumor, commonly seen in young patients with a female predominance, which is commonly located in the body and tail of the pancreas. Though it shows low malignant potential 10% to 15% of the cases show aggressive behavior with metastatic involvement of the liver. The symptoms include abdominal discomfort and abdominal pain. It is very rare in childhood. We report a case of SPT arising from the body of the pancreas in a 13-year-old girl who presented with pain in abdomen and treated successfully by enucleation of the tumor.

**KEYWORDS :** Solid pseudo-papillary tumor of the pancreas ,Frantz's tumor, young female.

## Introduction

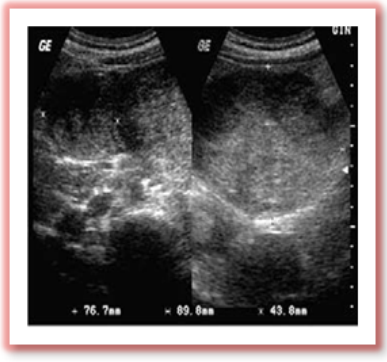
Solid-pseudopapillary tumor of the pancreas was first described by Frantz in 1959.[1] They are considered a rare pathologic entity with minimal malignant potential, affecting mainly juvenile females.[2]The sign and symptom of SPTP is related to mass effect and consists of abdominal pain and abdominal discomfort.

CT scan is an imaging technique of choice for diagnosis and shows a well-defined large solid-cystic mass. Prognosis is excellent and surgical resection can result in complete cure.[3]Although it shows low malignant potential, 10% to 15% of the cases show aggressive behavior with metastatic involvement of the liver. The overall five year survival is 97% even in the presence of disseminated disease [4] We report our clinical experience with a solid pseudopapillary tumor in 13 year female child successfully treated with enucleation of tumor.

## Case report

An 13-year-old girl presented with vague left upper abdominal pain since one month.Patient was haemodynamically stable .Physical examination showed tenderness in the left upper abdomen. Haematological investigation revealed no abnormal findings. Ultrasonography suggested evidence of heterogenous ovoid mass in pancreatic head of pancreas raising possibility of neoplastic origin. (Fig-1)

**Fig-1**




As other routine blood investigations were within normal limits, patient was subjected to CT scan of the abdomen, which revealed a well-demarcated ovoid mass of 4.6 x 4.4 x 4.9 cm sized heterogenously enhancing lesion with central areas of necrosis & contiguous with head of pancreas &compressing CBD. It also showed IHBR dilatation & few sub centimeter mesenteric and para- aortic lymph nodes.(Fig-2&3)

Ultrasound guided FNAC done suggestive of solid pseudopappillary neoplasm of pancreas.During hospital stay child complaint of severe backache & hence bone Scan was done which demonstrated osteoblastic skeletal lesion in left ilium and left femur ,inflammatory in nature. Child responded to NSAID.

After initial stabilization and anesthetic fitness exploratory laparotomy was done.Evidane of solid pancreatic tumor between IVC and portal vein adherent to CBD (Fig-4) .Complete enucleation of pancreatic tumor along with cholecystectomy, Roux-en-Y choledochojejunostomy & hepaticojejunostomy was done. Specimen (Fig-5) sent for histopathological examination suggestive of solid pseudopapillary epithelial neoplasm o pancreas.Tumor margin were negative .Two lymph nodes sent along with tumor show no evidence of malignancy.1year follow-up. Patient doing well after one year of followup.

**Fig -2 CT scan image**

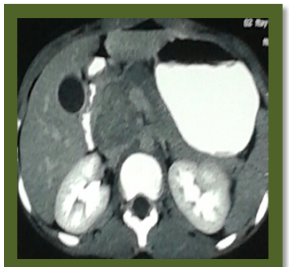


**Fig-3 CT scan image**

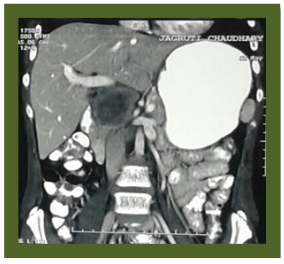


**Fig-4 Intraop pic**

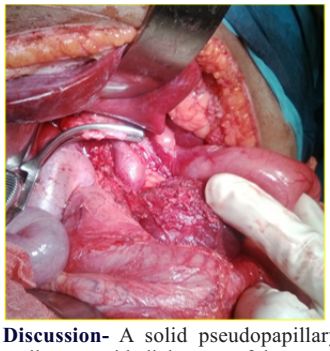
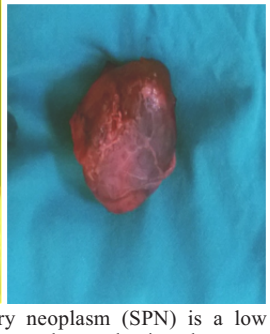


**Fig-5 Specimen**



**Discussion-** A solid pseudopapillary neoplasm (SPN) is a low malignant epithelial tumor of the pancreas that predominantly occurs in adolescent & young female.[5,6]

SPN usually affects young women at an average age of 28 years with a female: male ratio of 10:1[7]. About 20-25% of the cases are seen in pediatric patients.[8]

SPT was classified according to the WHO criteria as either an SPT with an uncertain potential for malignancy or as a solid pseudopapillary carcinoma (SPC)[6].Criteria that could distinguish potentially malignant tumors, classified as SPC, included the following: 1) perineural

invasion, 2) angioinvasion, 3) deep invasion into the surrounding tissue, and 4) distant metastases.The origin of this tumor remains an enigma. Kosmahl [9] favors an endocrine origin. The female predominance along with the presence of progesterone receptors in some reported cases suggests a neuroendocrine origin.The clinical presentation of SPN is nonspecific. Most of patients present with non-specific symptoms including abdominal discomfort, mild abdominal pain or palpable abdominal mass [1o].

A large encapsulated pancreatic mass with well-defined borders that contains areas of calcifications as well as intratumoral hemorrhage on CT scan in a young female is virtually diagnostic of an SPPN.[11-13]

SPN often remains asymptomatic, until the tumor has enlarged considerably. Accordingly, many are detected incidentally on diagnostic imaging for unrelated diseases or after a blunt abdominal trauma [14] The most common localization of SPN is the tail of the pancreas, followed by the head and the body. Unusual presentations include multicentric tumors in the pancreas and extrapancreatic sites, such as the mesocolon, retroperitoneum, omentum, liver and duodenum, possibly representing synchronous tumor spread [15]

There are no pathognomonic features on blood investigations and tumor markers are usually unremarkable.The diagnosis is usually made on cross-sectional imaging when pathognomonic features are present [16]: encapsulated, well defined mass with central areas of calcification, necrosis,haemorrhage, and/or cystic degeneration. In both the arterial and venous phases, there is usually peripheral enhancement with similar hounsfield unit density as the nearby pancreatic parenchyma[16]. The diagnosis can usually be made onmultiphase contrast enhanced CT with an estimated 60% overall accuracy[17]

Complete surgical resection (R0) is the most effective therapy for PPT [18]. Pancreatoduodenectomy, distal pancreatectomy (with or without splenectomy), middle pancreatectomy, or enucleation can be performed based on the location, size, angioinvasion, and adjacent organ compromise.

The prognosis of SPT patients even with local recurrence and metastasis or invasion is good. It has been reported that the overall 5-year survival rate of SPT patients is about 95%[8]

Park et al.[19] found that primary tumor resection resulted in a 100% cure rate, regardless of tumor size, even in patients positive for tumor at the resection margin. Their findings indicate that, despite the large tumor size and its ability to extend locally, complete excision can be beneficial in most patients. Metastases are uncommon in SPT (10-15% of patients with advanced disease)[20]. Although complete resection was more frequently performed on patients with SPT, Rojas et al.[20] reported that positive margins did not affect the outcome of patients with SPT, suggesting that enucleation when feasible, rather than radical resection, is likely sufficient to achieve long-term survival. The role of chemotherapy and radiation is controversial, but it has been used in some cases with aggressive disease[20].

To conclude considering low malignant potential and the excellent overall prognosis, surgical resection has been the standard of care in the management of SPN .

Conclusion, SPN is a rare pancreatic neoplasm of unclear histogenesis that typically affects young females without significant symptoms. Appearance on imaging is fairly characteristic and may suggest diagnosis Complete surgical resection of the tumor is the only effective treatment option.

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