

7A Case Report on the Solid Pseudopapillary Epithelial Neoplasm (SPEN) of the Pancreas

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I. Introduction

Solid pseudopapillary neoplasm of the pancreas is a rare cystic neoplasm. Its unique characteristics include its occurrence in young females, uncertain cell of origin, unpredictable malignant potential, as well as its excellent long term prognosis, even when it appears to be aggressive at presentation. Accurate diagnosis aids surgical decision making and, where necessary, multimodality management may be required.

Clinical Details- A 16 year old female patient without any significant past medical history presented to our hospital with vague upper abdominal pain since 3 months. There was no other significant history.

On clinical examination patient was moderately built and nourished. Abdomen showed a soft to firm mass felt in right hypochondrium and epigastrium measuring 10*15 cm which moves on respiration. There was no local rise of temperature.

Specific Investigations- Ultrasonography of the abdomen showed a well-defined mixed echogenic lesion in the head of pancreas.

Contrast enhanced computer tomography of abdomen was done, it showed a well circumscribed, heterogeneous pancreatic mass measuring 10 cm in the greatest diameter. (Fig 1)

Treatment- Then surgical resection of the pancreatic mass was done (Fig 2) and then sent for histopathology. Histopathology report showed that the margins were free of the tumor and the features are suggestive of SPEN. (Fig 3) Post surgery patient recovery was uneventful. The patient was not given any adjuvant therapy. She remained asymptomatic and showed no signs of disease after six months of follow-up.

II. Discussion

Solid Pseudopapillary Epithelial Neoplasm (SPEN) is a rare exocrine pancreatic neoplasm first described by Frantz. This tumor is also known by other names such as- solid cystic tumor, papillary cystic tumor and solid pseudopapillary tumor of the pancreas.¹ It accounts for 6% of all exocrine pancreatic tumours in a series published since 2000.² The tumour is thought to arise from ductal or acinar origin and has been mainly described in young women. There are no clear histological features, which establish the clinical behavior of these neoplasms. The tumor cells are usually of uniform size with a solid and pseudopapillary pattern and contain large amounts of eosinophilic cytoplasm and no prominent nucleolus or mitosis.³ The tumours exhibit low-grade malignant potential and metastasize infrequently, malignant degeneration has been reported in $\leq 15\%$ of cases including local invasion or metastasis. Local recurrence has been reported in less than 5% of cases. Metastases typically occur in the liver, lymph nodes and peritoneum.^{4,5} Because these tumours rarely invade adjacent structures, even large tumours have been shown to be resectable. Long-term prognosis is excellent, with 5-year survival rates of 95–97%. The limited data available suggest 10-year survival may be as high as 93%.⁶ Even when tumours are considered “non-resectable”, patients can survive longer than 10 years after surgical debulking. Resection of liver metastases can prolong survival by at least 5 years.²

III. Conclusion

A high index of clinical suspicion is necessary to suspect and diagnose SPEN. This diagnosis should be borne in mind when young female patients present with a pancreatic mass. CT scan and FNAC/biopsy are valuable pointers to the pre-operative diagnosis. Surgical excision offers the best chance for cure and should

always be attempted irrespective of the magnitude of resection involved. Patients with SPEN have an excellent prognosis after surgical excision.

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Fig 1: CT scan showing SPEN in the body and tail of pancreas.

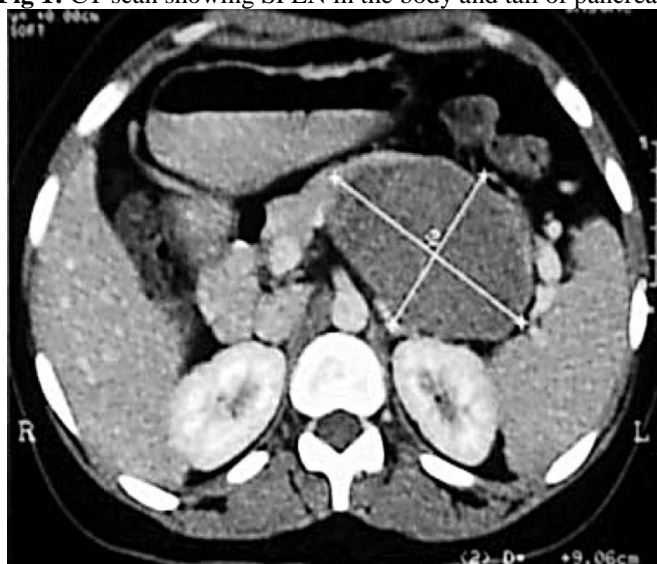
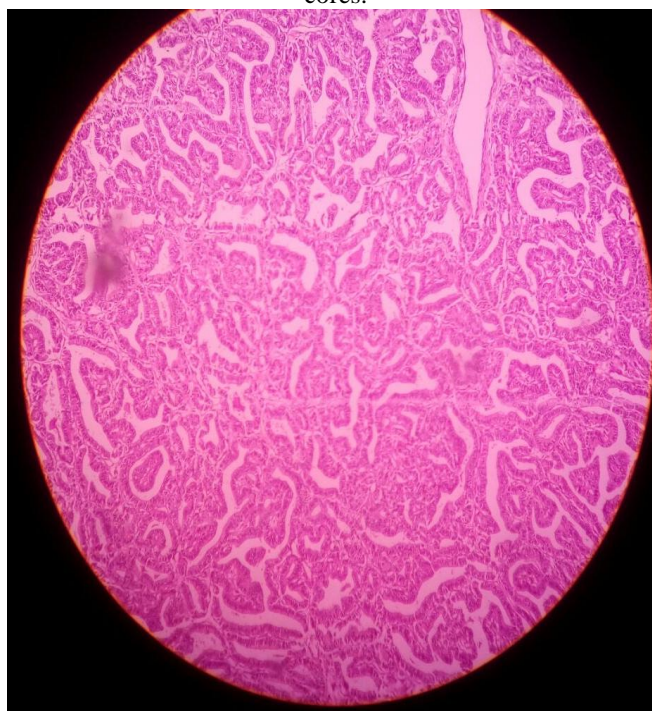


Fig 2: Surgically resected SPEN.



Fig 3: Papillary structures are lined by cytologically bland cells. Marked sclerosis is seen around the central cores.



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