

Solid pseudopapillary epithelial neoplasm of the Pancreatic Head: A rare case report and literature review

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Abstract

Solid pseudopapillary epithelial neoplasms (SPEN), also known as Frantz tumors, are rare pancreatic exocrine neoplasms that predominantly affect young females. Despite their rarity, the incidence has increased seven-fold over the past two decades. We report a case of a 29-year-old female who presented with vague abdominal pain for one year. Contrast-enhanced computed tomography revealed a 3×3 cm well-circumscribed, encapsulated, heterogeneous lesion with cystic degeneration in the pancreatic head. The patient underwent pancreaticoduodenectomy with uneventful recovery. Histopathological examination confirmed SPEN with pT2N0 staging. SPEN represents a low-grade malignancy with excellent prognosis when diagnosed early and treated appropriately. Accurate diagnosis is crucial as these tumors have low malignant potential and favorable outcomes with surgical resection.

Keywords: Solid pseudopapillary epithelial neoplasm, Frantz tumor, pancreatic neoplasm, pancreaticoduodenectomy

Introduction

Solid pseudopapillary epithelial neoplasms (SPEN), also known as Frantz tumors, represent a rare subset of pancreatic exocrine neoplasms that account for approximately 1.2% of all pancreatic neoplasms. These tumors demonstrate a striking female predominance with a female-to-male ratio of 10:1, typically affecting women in their third to fourth decade of life. Interestingly, epidemiological data suggests a seven-fold increase in SPEN incidence over the past two decades, possibly due to improved imaging techniques and increased awareness among clinicians.

SPEN are characterized as low-grade tumors with low malignant potential, predominantly affecting young and adult females. In contrast, when these tumors occur in males, they tend to present at a later age and exhibit more aggressive behavior. The anatomical distribution shows a predilection for the pancreatic tail, though involvement of the pancreatic head, as in our case, can occur particularly in young females.

The clinical significance of SPEN lies in their generally benign nature, with only 10-15% demonstrating malignant behavior. When metastasis occurs, the liver represents the most common site of secondary involvement. The prolonged asymptomatic period often associated with these tumors can lead to delayed diagnosis, emphasizing the importance of maintaining clinical suspicion in appropriate patient demographics.

Case Summary

A 29-year-old female presented to our surgical outpatient department with a chief complaint of vague abdominal pain persisting for one year. The pain was non-specific in nature without radiation, and the patient denied any associated symptoms such as nausea, vomiting, weight loss, or changes in bowel habits. Her medical history was unremarkable, with no significant past surgical interventions or family history of pancreatic or other gastrointestinal malignancies.

On physical examination, revealed a well-built female in no acute distress. Vital signs were within normal limits, and the patient appeared well-nourished without evidence of weight loss or jaundice. Abdominal examination was largely unremarkable with no palpable masses, organomegaly, or signs of peritoneal irritation.

Initial diagnostic investigations, given the non-specific nature of the patient's symptoms, contrast-enhanced computed tomography (CECT) of the abdomen and pelvis was performed. The imaging revealed a 3×3 cm well-circumscribed, encapsulated, heterogeneous lesion with areas of cystic degeneration located in the head of the pancreas. The radiological characteristics were highly suggestive of SPEN, with no evidence of vascular involvement, lymphadenopathy, or distant metastases.

Fig 2: (Cect Abdomen and Pelvis)



Surgical management

- Based on the imaging findings and clinical presentation, the patient was counseled regarding surgical management. After obtaining informed consent, the patient underwent pancreaticoduodenectomy followed by standard reconstruction consisting of Pancreatico

jejunostomy, Hepatico-jejunostomy, Gastro-jejunostomy (Whipple's procedure) under general anaesthesia.

Intraoperative findings revealed a 3×3 cm well-defined, encapsulated tumor located in the pancreatic head with well-maintained fat planes around the lesion, confirming the preoperative imaging findings. The tumor appeared to have both solid and cystic components, consistent with the radiological appearance. There was no evidence of local invasion or gross metastatic disease. The operation was completed without complications, and hemostasis was achieved.

Fig 2: (A well defined mass with solid and cystic consistency in the pancreatic head region)

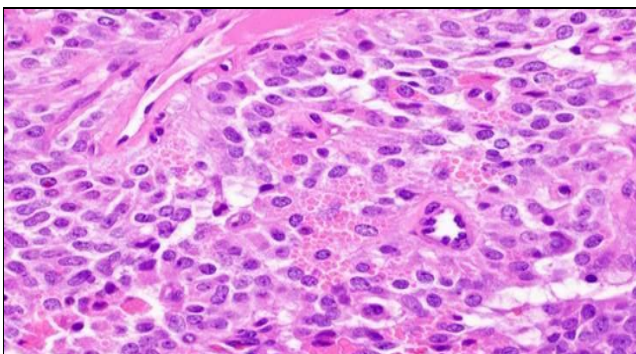


Postoperative course

The patient's postoperative recovery was uneventful. She tolerated oral intake well, demonstrated appropriate wound healing, and showed no signs of pancreatic fistula, delayed gastric emptying, or other common complications associated with pancreaticoduodenectomy. The patient was discharged on postoperative day 14 in stable condition with appropriate follow-up arrangements.

Histopathological Findings

Histopathological examination of the resected specimen confirmed the diagnosis of solid pseudopapillary epithelial neoplasm. The tumor was staged as pT2N0, indicating invasion beyond the pancreas but without lymph node involvement. The tumor showed characteristic morphological features of SPEN with areas of solid growth pattern interspersed with pseudopapillary structures and cystic degeneration (fig-



Follow-up and management

Given the low malignant potential of SPEN and the complete surgical resection achieved, the patient was advised regular follow-up without adjuvant therapy. The

multidisciplinary team consensus supported surveillance as the appropriate management strategy given the favorable histological features and complete resection margins.

Discussion

SPEN represents a unique entity among pancreatic neoplasms, characterized by its distinct demographic predilection and generally favorable prognosis. The seven-fold increase in incidence over the past two decades likely reflects improved diagnostic capabilities rather than a true increase in disease occurrence. Advanced imaging modalities, particularly high-resolution CT and MRI, have enhanced our ability to detect and characterize these lesions. The striking female predominance (10:1 ratio) and typical age of presentation in the third to fourth decade of life are consistent with our case presentation. The patient's age of 29 years falls within the expected demographic range for SPEN occurrence in females. The location in the pancreatic head, while less common than tail involvement, is not unusual in young females, as demonstrated in our case.

The prolonged asymptomatic period observed in our patient, with vague abdominal pain as the only symptom, is characteristic of SPEN. This non-specific presentation often leads to delayed diagnosis, emphasizing the importance of appropriate imaging in patients with persistent abdominal symptoms, particularly in the demographic group at risk for SPEN.

The radiological characteristics observed in our case, including the well-circumscribed, encapsulated appearance with heterogeneous enhancement and cystic degeneration, are typical features of SPEN on contrast-enhanced CT. These imaging findings, combined with the patient's demographic profile, strongly suggested the diagnosis preoperatively.

Surgical management remains the treatment of choice for SPEN, with the extent of resection depending on tumor location and size. In our case, pancreaticoduodenectomy was performed due to the tumor's location in the pancreatic head. The excellent postoperative recovery and absence of complications highlight the generally favorable surgical outcomes associated with SPEN resection.

The histopathological staging of pT2N0 indicates local invasion beyond the pancreas but without lymph node involvement, which is consistent with the low malignant potential of these tumors. The absence of lymph node metastases supports the favorable prognosis associated with SPEN, even when local invasion is present.

The decision to forgo adjuvant therapy in our patient reflects the current understanding of SPEN biology and treatment recommendations. Given the low malignant potential and excellent prognosis following complete surgical resection, surveillance rather than adjuvant treatment is the standard approach for most patients with SPEN.

Conclusion

SPEN represents a rare but increasingly recognized pancreatic neoplasm with distinct clinical and pathological characteristics. Accurate preoperative diagnosis is crucial given the tumor's low malignant potential and excellent prognosis following appropriate surgical management. The case presented demonstrates the typical presentation, diagnostic approach, and favorable outcomes associated with SPEN in young females.

Healthcare providers should maintain awareness of SPEN in the differential diagnosis of pancreatic lesions, particularly

in young females presenting with non-specific abdominal symptoms. Early diagnosis and appropriate surgical intervention can result in excellent long-term outcomes, as demonstrated in our case. The favorable prognosis associated with SPEN, even in cases with local invasion, supports the importance of prompt surgical management when feasible.

Continued surveillance and long-term follow-up remain important components of SPEN management, though the overall prognosis remains excellent with appropriate treatment. This case contributes to the growing body of literature documenting successful outcomes in SPEN management and reinforces the importance of maintaining clinical suspicion for this rare but treatable pancreatic neoplasm.

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