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Solid Pseudopapillary Neoplasm of the Pancreas – Frantz Tumor: A Case Report

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1. Abstract

Solid pseudopapillary neoplasm (SPN) of the pancreas, also known as Frantz tumor, is a rare, low-grade malignant tumor that predominantly affects young women. It has a favourable prognosis following complete surgical resection. We report the case of a 24-year-old male patient who presented with nonspecific gastrointestinal symptoms. Imaging revealed a well-defined, rounded lesion in the pancreatic tail, measuring approximately 8.2 × 6.5 × 5.7 cm. After multidisciplinary discussion, the patient underwent partial pancreatectomy (body and tail), splenectomy, omentectomy, and retroperitoneal lymphadenectomy. Histopathology and immunohistochemistry confirmed the diagnosis of SPN, with negative surgical margins and lymph nodes free of neoplasia. Postoperatively, the patient developed a pancreatic fistula and refractory vomiting, requiring abdominal drainage and symptomatic optimization. The patient is currently in good general condition, asymptomatic, and with no evidence of recurrence on follow-up imaging, with resolution of a residual fluid collection. This case illustrates the clinical presentation, diagnostic and therapeutic management, and postoperative course of a patient with SPN, highlighting the role of surgery in treatment and the favourable prognosis of the disease.

2. Introduction

Solid pseudopapillary neoplasm (SPN) of the pancreas, also known as Frantz tumor, is a rare epithelial tumor that accounts for approximately 1–3% of exocrine pancreatic neoplasms and around 5% of cystic pancreatic lesions in adults [1]. It predominantly affects young women in the second or third decade of life, although rare cases have been reported in men and children [2]. Clinically, SPN often remains asymptomatic or presents with nonspecific symptoms such as abdominal pain, nausea, or vomiting, usually related to the tumor's mass effect. The diagnosis is often incidental, based on imaging performed for unrelated reasons [3]. Despite its low malignant potential, SPN can occasionally exhibit locally aggressive behavior or metastasize to the liver or peritoneum in 10–15% of cases [4]. Complete surgical resection remains the mainstay of treatment and offers an excellent prognosis in most patients. This report describes a rare case of SPN in a young male patient, emphasizing the diagnostic process, surgical management, and postoperative outcome.

3. Case Report

The patient, G.H.R.V., a 24-year-old male with a history of depressive-anxiety disorder and no other relevant comorbidities, presented with persistent nausea and vomiting, associated with mesogastric abdominal pain and mild constipation for several weeks. Initial work-up included abdominal imaging. Computed tomography (CT) and magnetic resonance imaging (MRI) of the abdomen revealed a well-defined, rounded lesion in the pancreatic tail, measuring approximately 8.2 × 6.5 × 5.7 cm, with slightly thickened and mildly irregular walls and heterogeneous content. No dilation of the main pancreatic duct was observed [Figures 1 and 2]. The imaging differential diagnosis included complicated pancreatic pseudocyst, hydatid cyst, solid pseudopapillary tumor, and mucinous cystic neoplasm. Staging evaluation showed no regional or distant disease. Tumor

markers (CA19-9, AFP, and CEA) were within normal limits. During hospitalization, the case was thoroughly discussed in a multidisciplinary meeting involving Clinical Oncology, General Surgery, Gastroenterology, Pathology, and Radiology. Given the lesion's characteristics and absence of spread, surgical management was indicated. The patient underwent partial pancreatectomy (body and tail), splenectomy, omentectomy, and retroperitoneal lymphadenectomy. Histopathological examination and subsequent immunohistochemistry confirmed the diagnosis of SPN (Frantz tumor) (Figure 3). Surgical margins were free of neoplasia, and all examined retroperitoneal lymph nodes were negative (0/11), indicating complete resection. Postoperatively, the patient developed a pancreatic fistula and refractory vomiting. Management included abdominal drainage and optimization of supportive care. Gradual clinical improvement ensued. The patient remains in clinical follow-up, in good general condition, asymptomatic, and with no evidence of disease on recent restaging imaging. Follow-up imaging revealed a small, well-defined residual fluid collection in the pancreatic bed adjacent to the surgical margin, which progressively decreased in size over time.

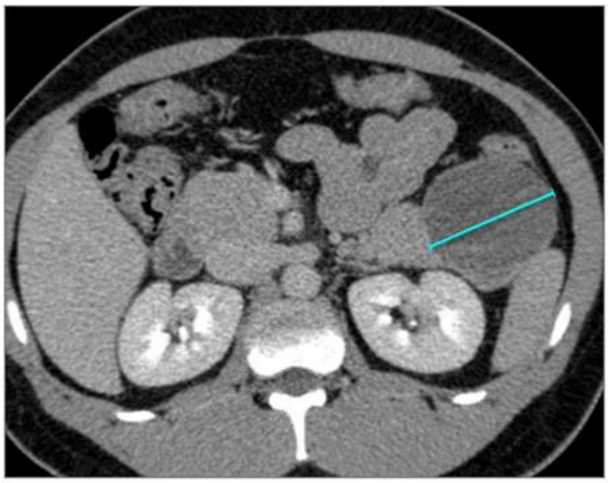

Figure 1: Contrast-enhanced CT of the abdomen, delayed phase: persistent heterogeneous areas within the lesion, suggestive of hemorrhagic components, and enhancement of the capsular margin. The lesion measures approximately 8.5 cm in its largest coronal diameter.

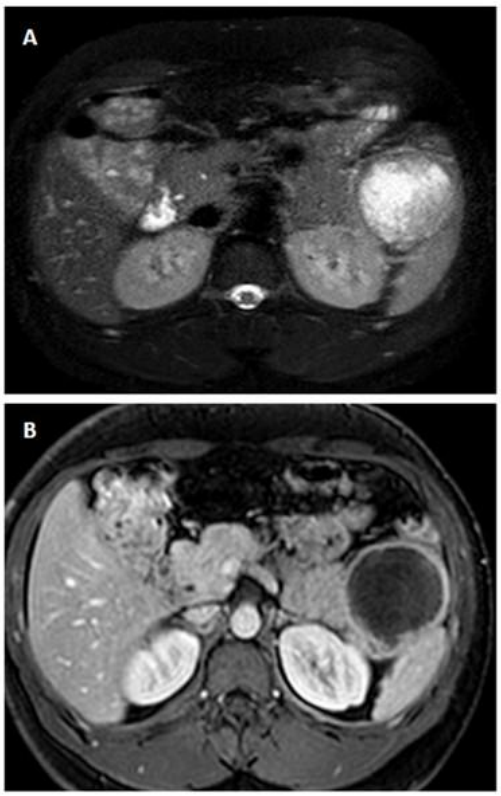

Figure 2. Magnetic resonance imaging (MRI) of the abdomen:
(A) Axial T2-weighted fat-suppressed sequence showing a well-defined round lesion in the pancreatic tail, measuring approximately 8.3 × 6.3 × 6.0 cm (estimated volume of 163 cm^3), with heterogeneous hyperintense signal.
(B) Axial T1-weighted fat-suppressed sequence, venous phase, demonstrating persistent peripheral enhancement, without dilation of the main pancreatic duct or evidence of adjacent tissue infiltration.

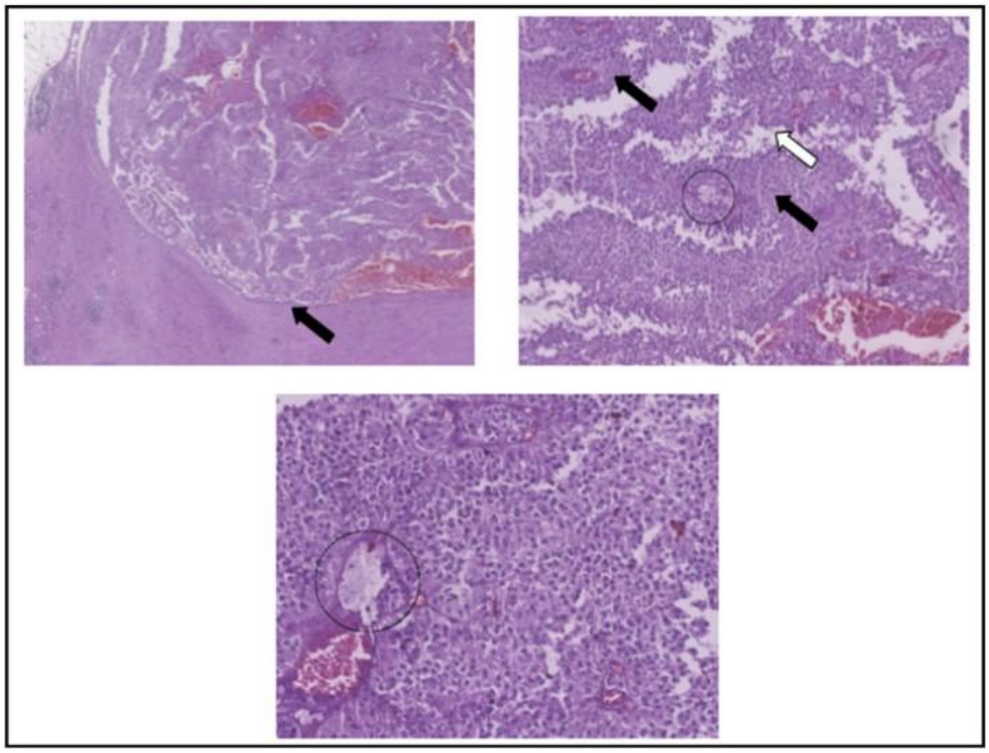

Figure 3: H&E Micrographs of Solid Pseudopapillary Neoplasm of the Pancreas

(A) 2× magnification demonstrating a well-circumscribed lesion with clear demarcation from the adjacent pancreatic parenchyma (arrow).

(B) 10× magnification highlighting pseudopapillae (black arrows) and intervening degenerative areas (white arrow), histiocytes (circle), as well as foci of hemorrhage in the lower right corner.

(C) 20× magnification revealing monotonous, isomorphic cells without atypia, characterized by round, vesicular nuclei. Aggregates of foamy histiocytes are also observed (circle).

4. Discussion

Solid pseudopapillary neoplasm (SPN) of the pancreas-also known as Frantz tumor-is a rare oncologic entity with low malignant potential and excellent prognosis after surgical resection [5]. This case illustrates the typical disease course, from nonspecific clinical presentation to diagnosis and surgical management. The patient's presentation with nausea, vomiting, and abdominal pain is common in pancreatic tumors, particularly those of considerable size producing mass effect. Normal tumor markers (as CA19-9 and CEA) are often observed in SPN, distinguishing it from more aggressive pancreatic malignancies such as adenocarcinoma, where these markers are usually elevated [6]. The young age and male sex of this patient-despite the higher prevalence in young women-underscore the importance of considering SPN in all patients with cystic or solid pancreatic lesions. In men, SPNs tend to be more solid and less cystic; in children, SPN accounts for 6%–17% of all pancreatic neoplasms [7]. Histopathologically, SPNs are typically well-encapsulated, partially solid and cystic tumors with areas of hemorrhage and necrosis. Microscopically, they exhibit solid and pseudopapillary structures composed of monomorphic cells loosely arranged around delicate vessels. Immunohistochemical profiling is essential for diagnosis, with characteristic nuclear and cytoplasmic β-catenin positivity, CD99 dot-like pattern, and expression of CD10, vimentin, CD56, and progesterone receptor, along with absence of chromogranin A and loss of membranous E-cadherin [8]. These features were consistent with the findings in the present case. Imaging studies, particularly CT and MRI, are key for lesion detection and characterization. The description of a well-circumscribed, rounded mass with thickened walls and heterogeneous content is consistent with SPN, which often presents with peripheral solid components and central cystic areas due to hemorrhage and degeneration. On contrast-enhanced CT, the solid component typically shows mild arterial enhancement with progressive portal-venous phase uptake. On MRI, hemorrhagic areas appear hyperintense on T1-weighted images, while T2 signal intensity varies, reflecting tumor heterogeneity. The absence of main pancreatic duct dilation is also a common finding. The imaging differential is broad, and preoperative confirmation via endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) is considered the gold standard [9,10,11]. Multidisciplinary decision-making is essential. Distal pancreatectomy, often including splenectomy due to vascular proximity, remains the standard approach for tumors located in the pancreatic body/tail. Splenectomy is often required due to anatomical proximity and shared vascular supply. Routine lymphadenectomy is not performed, as lymph node metastasis is rare (~1.6%) [12,13]. Histopathology and immunohistochemistry remain the gold standard for definitive diagnosis. Monomorphic cells arranged in solid and pseudopapillary patterns, the presence of eosinophilic hyaline globules, and positivity for specific markers are essential. Aberrant nuclear β-catenin expression-a hallmark of SPN-results from

CTNNB1 mutations present in over 90% of cases [14]. Negative margins and lymph node involvement (0/11) in this case are indicative of R0 resection, correlating with an excellent prognosis. Postoperative complications, such as pancreatic fistula, are relatively common after pancreatectomy. Conservative management with adequate drainage and supportive care generally leads to resolution, as in this patient. The absence of recurrence on follow-up imaging supports the favorable outcome after complete resection. This case reinforces the importance of maintaining diagnostic suspicion, appropriate imaging work-up, radical surgical approach, and close postoperative surveillance of this rare but curable pancreatic neoplasm.

5. Conclusion

Solid pseudopapillary neoplasm of the pancreas is a rare tumor with a favorable prognosis after complete surgical resection. This report describes a young patient with nonspecific gastrointestinal symptoms whose diagnosis of Frantz tumor was established through imaging and confirmed by histopathology and immunohistochemistry following distal pancreatectomy with splenectomy. Despite postoperative complications, which were successfully managed, the patient remains in good general condition and free of disease recurrence. This case underscores the importance of early recognition, a multidisciplinary approach, and radical surgery as the cornerstone of successful management and excellent prognosis in SPN.

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