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Case Report

Solid Pseudopapillary Carcinoma of the Pancreas Presenting with Acute Kidney Injury and Metastatic Disease in an Elderly Male: A Case Report

Burcin Meryem Atak Tel, Tuba Taslamacioglu Duman, Gulali Aktas*

Department of Internal Medicine, Abant Izzet Baysal University Hospital, Bolu, Turkey

Correspondence author: Gulali Aktas, draliaktas@yahoo.com

Abstract

Background: Solid pseudo-papillary neoplasm (SPN) of the pancreas is a rare malignant neoplasm, typically affecting young females. Occurrence in elderly males is exceedingly uncommon and often associated with aggressive behavior and advanced disease at presentation.

Case Presentation: We report a 75-year-old male with a history of type 2 diabetes mellitus, hypertension, coronary artery disease, and prior coronary bypass surgery, who presented with abdominal distension. Initial evaluation, including laboratory tests and abdominal ultrasonography, was unremarkable. He was discharged with symptomatic treatment following relief in his symptoms. Five months later, the patient presented to the emergency department with nausea, vomiting, and abdominal pain. Laboratory tests revealed severe acute kidney injury (urea 258 mg/dL, creatinine 23 mg/dL, potassium 7 mmol/L). Therefore, emergent hemodialysis was initiated. Abdominal imaging revealed hepatic metastases, a duodenal wall mass, and a large retroperitoneal mass encasing major abdominal vessels (Sonography of the abdomen 5 months ago was normal). Ultrasound-guided biopsy of the retroperitoneal mass demonstrated histological features (variable admixture of solid and pseudopapillary areas) consistent with solid pseudo-papillary carcinoma. Due to poor performance status, palliative care was recommended. The patient died three months after establishing the diagnosis.

Conclusion: This case elaborated the aggressive nature and atypical presentation of SPN in elderly males, including retroperitoneal involvement and severe renal impairment due to obstructive vascular compression.

Keywords

Pancreas cancer, Acute kidney injury, Metastasis

Article History

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1. Introduction

Solid pseudo-papillary neoplasm (SPN) of the pancreas is an uncommon tumor, accounting for 1–3% of all exocrine pancreatic tumors [1]. It predominantly affects young women in their second to fourth decades of life and generally follows an indolent course. Malignant transformation into SPN is rare, with more aggressive behavior, frequent metastases, and poor prognosis [2]. Occurrence in elderly males is exceedingly uncommon and may be associated with atypical and advanced presentations [3]. Since the tumor is relatively large and cause compression symptoms, early detection and management is crucial. Here, we reported an unusual case of SPN in a 75-year-old male presenting with acute kidney injury secondary to tumor encasement of major abdominal vessels, with synchronous hepatic metastases. We report this case to highlight the diagnostic and therapeutic challenges in atypical presentations of SPN.

2. Case Presentation

A 75-year-old male presented to the internal medicine outpatient clinic with abdominal distension. He had a 45-pack-year smoking history and a past medical history of type 2 diabetes mellitus, hypertension, and coronary artery disease. His surgical history included coronary artery bypass grafting performed five years earlier. On physical examination, vital signs were within normal limits and mild abdominal tenderness was noted. The remainder of the examination was unremarkable. Laboratory tests and abdominal ultrasonography were normal. The patient was discharged with treatment for dyspeptic symptoms and scheduled for gastroenterology follow-up.

Persistent symptoms and newly detected iron-deficiency anemia later prompted esophagogastroduodenoscopy, which revealed edematous and hyperemic gastric fundus and corpus mucosa with erosive lesions. Biopsies were obtained, but pathology results were unavailable. Five months afterward, the patient re-presented to the emergency department with nausea, vomiting, and abdominal pain. Laboratory evaluation demonstrated severe acute kidney injury (urea 258 mg/dL, creatinine 23 mg/dL, potassium 7 mmol/L). Baseline laboratory values from 12 months earlier (performed at another institution) were normal. He was admitted to the internal medicine ward, and a temporary femoral dialysis catheter was placed for emergent hemodialysis.

Abdominal ultrasonography revealed hepatic cystic lesions, a 9-mm simple cortical cyst in the right kidney, and a 7-cm midline abdominal mass. Contrast-enhanced abdominal and thoracic computed tomography (CT) demonstrated multiple irregular hypodense solid lesions in the right hepatic lobe, the largest measuring 26 × 24 mm, consistent with metastases; duodenal horizontal segment wall thickening up to 28 mm; and a large retroperitoneal mass (76 × 145 × 140 mm) extending to the mesenteric root and encasing the inferior vena cava, abdominal aorta, right renal artery, and left renal vein, contiguous with the posterior wall of the duodenum. Figure 1 shows the CT image of the retroperitoneal mass, and Figure 2 shows the liver lesions.

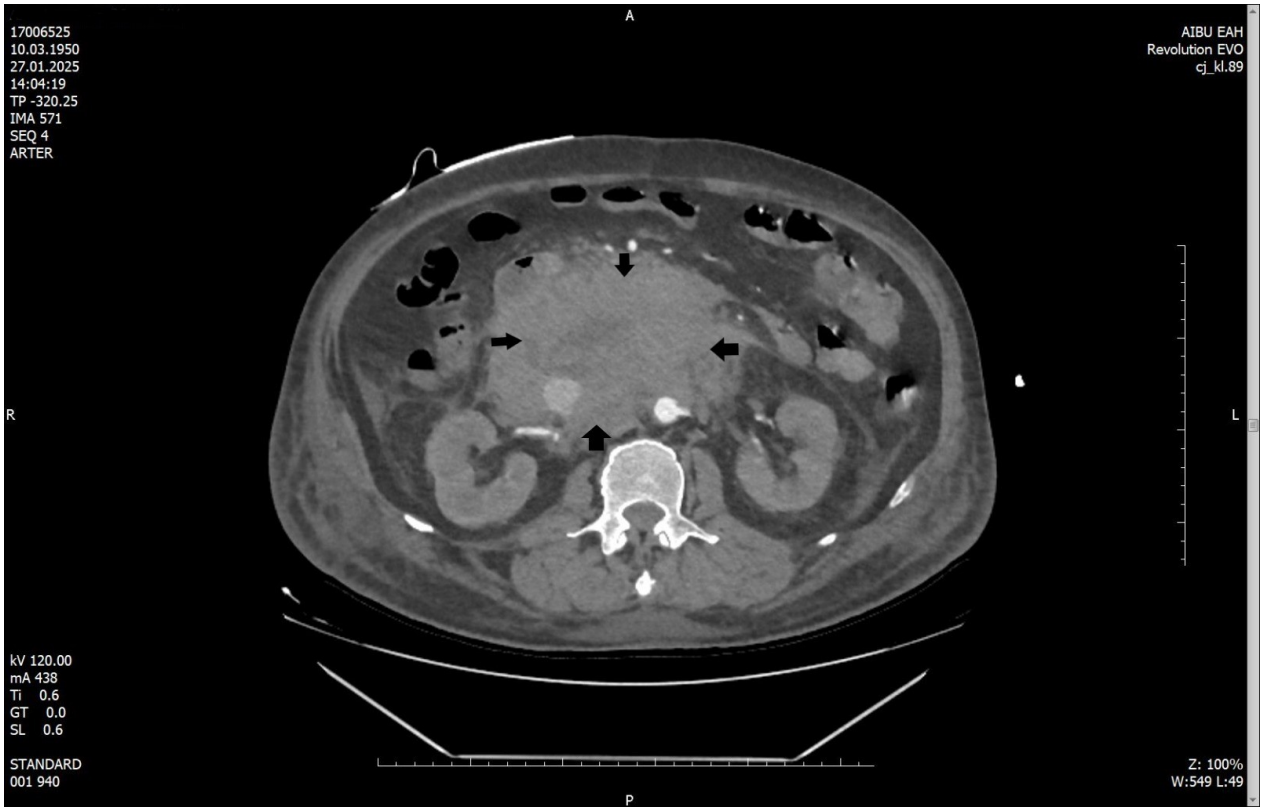

Figure 1. Retroperitoneal mass associated with pancreas and duodenum in CT image. The mass is placed between black arrows.

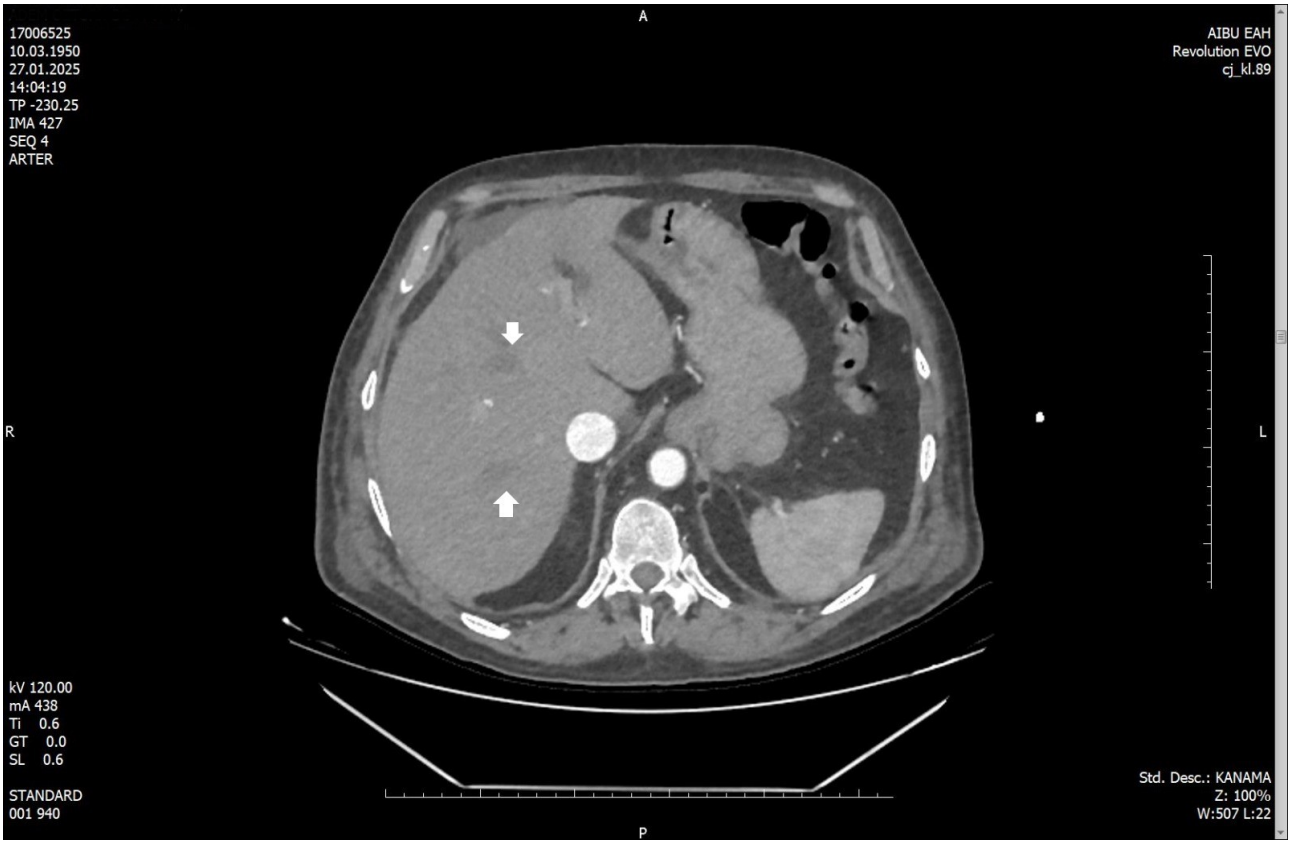

Figure 2. Hepatic metastases in CT image of the patient. Hepatic metastases (White arrows).

Ultrasound-guided biopsy of the retroperitoneal mass demonstrated histopathological features consistent with solid pseudopapillary carcinoma. Tumor cells expressed CD138 and CD38, with positive β-catenin staining and a Ki-67 index of 40%. Chromogranin, synaptophysin, and pancytokeratin were negative. Due to an Eastern Cooperative Oncology Group (ECOG) performance status score of 3, palliative care was recommended, and chemotherapy was not initiated. The patient continued thrice-weekly hemodialysis. His clinical condition declined rapidly, and he died three months after the establishment of the diagnosis.

3. Discussion

Here we presented an interesting SPN case in an elderly patient who eventually died due to primary disease and accompanied conditions.

Solid pseudo-papillary neoplasms are rare pancreatic tumors, comprising approximately 1-3% of all pancreatic neoplasms [4,5]. Most commonly, they affect young women in their second or third decades of life, with a strong female-to-male predominance (about 10:1) [3,5,6]. SPNs generally exhibit low-grade malignant potential, often presenting as encapsulated masses with cystic and solid components [7].

The current case involved a 75-year-old male, a demographic rarely described in SPN literature. Among 1320 pancreatic resections in one series, only 3 male cases (9.4%) were identified, with a mean age of 28.4 years [4]. Another study reconfirmed the strong prevalence in young females [6]. Thus, this case represents an extremely unusual age and sex profile, complicating clinical suspicion early on.

SPN is a rare pancreatic malignancy, with a marked female predominance (female-to-male ratio ≈ 9:1) and mean age of diagnosis around 22 years [8]. The occurrence in elderly males is rare and generally portends a poorer prognosis. Common presentations include abdominal pain or mass [9,10], however, our patient's initial presentation with dyspeptic symptoms and subsequent acute kidney injury due to vascular compression is unusual. Standard SPNs originate in the pancreas (body and tail in two-thirds of cases) [4,5]. SPN may mimic other diseases such as adrenal malignancy [11]. It also may cause portal hypertension [12]. Primary extra-pancreatic SPNs are exceedingly rare, with fewer than 50 cases reported, including retroperitoneal occurrences [6,13]. These tumors share characteristic histopathology and immuno-histochemical profiles: β-catenin nuclear accumulation, CD99 punctate staining, and loss of E-cadherin [6,13]. In this patient, the retroperitoneal mass conforming to SPN histology, with surrounding vessel encasement and hepatic metastases, is consistent with reports of aggressive extra-pancreatic SPNs [6,13]. Such cases underscore the diagnostic dilemma posed by atypical location and imaging.

Metastases of SPN most often occur in the liver, in approximately 10–15% of the cases, but extensive retroperitoneal involvement with major vessel encasement is rarely reported [14]. In our case, the aggressive local invasion likely contributed to the rapid deterioration in renal function. Despite the generally indolent behavior of SPNs, up to 10–15% exhibit metastatic spread, most commonly to the liver or peritoneum [6,7,13]. A comprehensive review of 6,651 SPN cases reported a metastasis rate of around 1.9% at presentation, with an overall recurrence rate of 3%, mortality of 0.2%, and a mean size of 5.8 cm [5]. Yet, more aggressive phenotypes, especially in extra-pancreatic tumors, demonstrate metastatic disease with poor outcomes [6,13]. Consistent with that, this patient's tumor was sizable (76 × 145 × 140 mm), encasing vital structures and already metastatic to the liver at presentation, correlating with rapid clinical decline.

Treatment of SPN involves surgical resection, which offers excellent long-term survival in localized disease [15]. However, in unresectable or metastatic cases, there is limited evidence for the efficacy of chemotherapy or radiotherapy. Our patient’s poor performance status precluded systemic therapy, and palliative care was prioritized. Standard of care favors margin-negative surgical resection, which is often curative given the low malignant potential of SPNs [4,5]. In younger, fit patients, surgery provides excellent long-term survival. Emerging small lesion management via EUS-guided radiofrequency ablation (RFA) holds promise, particularly for tumors under 2 cm [5]. In contrast, this patient’s advanced age, comorbidities (e.g., coronary artery disease, diabetes, renal failure requiring dialysis), poor performance status (ECOG 3), and unresectable tumor precluded surgical or ablative therapy, necessitating palliative care. This aligns with current guidelines that recommend conservative management for patients with limited functional reserve or widespread disease [4,5].

The delay in diagnosis, initial nonspecific gastrointestinal symptoms, normal labs and ultrasound, and lack of early endoscopic biopsy results, demonstrates the difficulty of detecting SPNs in atypical patients. The presentation in an elderly male with extra-pancreatic location and aggressive spread is among the rarest described, with few analogous cases reported [4,6,13]. Once liver metastases and retroperitoneal mass encasing major vessels manifested, the prognosis was poor, consistent with metastatic SPN data [5,7]. This case emphasized the importance of considering SPN in the differential, even in atypical demographics, when encountering retroperitoneal tumors with pseudo-papillary histology. Early tissue diagnosis (e.g., EUS-FNA) in patients with unusual but persistent symptoms might offer better diagnostic yield and allow timely multidisciplinary decision-making [4,6,13].

In literature, Lubezky et al reported 31 SPN cases with a mean age of 29 years and most of which were females (91%) [4]. However, we presented SPN case in an elderly male.

In Yagci et al’s study, there were 10 SPN cases (90% of those cases were female) with the most common presenting symptom of abdominal pain [16]. Only one patient was over 70 years of age in that report and only three of them died during follow up of 24 months [16]. Abdominal distension and pain were the presenting symptom in the present case and his survival was poor. The present case had aggressive course in contrast of the usual slow progression of the disease. The deteriorated kidney functions were likely due to compression of the retroperitoneal tumor mass.

Cummins et al reported a SPN case in a 59 year old woman who presented to general practitioner for wheeze [17]. Tumor showed positive β catenin in histological examination [17]. Presenting symptoms of the present case do not include respiratory symptoms and similarly, the tumor cells were positive for β catenin. Table 1 summarizes the cases presented in previous reports in the literature.

A retroperitoneal mass with concurrent hepatic lesions necessitates consideration of a broad differential diagnosis, including pancreatic adenocarcinoma, neuroendocrine tumor, retroperitoneal sarcoma, adrenal neoplasm, lymphoma, metastatic disease, and extramedullary plasmacytoma. While imaging characteristics may provide suggestive features, such as hypervascularity in neuroendocrine tumors, infiltrative retroperitoneal patterns in lymphoma, or fat-containing elements in sarcoma, radiological findings alone are not definitive. Therefore, tissue biopsy was essential to distinguish among these entities. The biopsy specifically aimed to exclude epithelial malignancies such as pancreatic adenocarcinoma, hematologic malignancies such as lymphoma or plasmacytoma, and mesenchymal tumors including retroperitoneal sarcoma. Histopathology combined with immunohistochemistry (IHC) ultimately supported the diagnosis of SPN, demonstrated by characteristic nuclear β-catenin positivity, CD10 and CD 138 and CD 38 expression. Moreover, markers typical for alternative diagnoses such as, synaptophysin/chromogranin for neuroendocrine tumors, cytokeratins for adenocarcinoma, CD45 for lymphoma were negative.

Table 1. Characteristics of some of the SPN cases in the literature and present case.

Case	Author	Age	Gender	Survival	Reference
1	Yagci et al.	30	female	healthy	[16]
2	Yagci et al.	18	female	healthy	[16]
3	Yagci et al.	21	female	healthy	[16]
4	Yagci et al.	18	female	healthy	[16]
5	Yagci et al.	62	female	healthy	[16]
6	Yagci et al.	50	female	deceased	[16]
7	Yagci et al.	40	female	deceased	[16]
8	Yagci et al.	33	female	healthy	[16]
9	Yagci et al.	71	female	deceased	[16]
10	Yagci et al.	45	male	healthy	[16]
11	Cummins et al.	59	female	healthy	[17]
12	Present case	75	male	deceased	-

4. Conclusion

This unfortunate case of a solid pseudo-papillary neoplasm arising extra-pancreatic in an elderly male with an aggressive clinical course and metastases provided several key insights; SPNs, while usually benign and affecting young women, can present atypically, and thus may be overlooked. Extra-pancreatic presentations, especially in unusual age/sex groups, remain diagnostically challenging. Aggressive variants, although rare, can rapidly progress, emphasizing the importance of early recognition and intervention where feasible. In patients with limited physiological reserve or advanced disease, the focus may shift to palliative care, recognizing the limits of curative therapy. Further documentation of such rare presentations in the literature is crucial to enriching clinical awareness and informing future diagnostic and management strategies. This case highlights the importance of considering SPN in the differential diagnosis of retroperitoneal masses in elderly males. Early biopsy and multidisciplinary evaluation are essential to avoid diagnostic delay, even in atypical demographics.

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Informed Consent

Informed consent was obtained from the patient.

Data Availability Statement

Anonymized data and imaging are available upon reasonable request

Conflict of Interest

There is none.

Generative AI Statement

The authors declare that no Gen AI was used in the creation of this manuscript.

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