CASE REPORT

An Unusual Case of Obstructive Jaundice: Solid Pseudopapillary Neoplasm of the Pancreatic Head in a 47 Year Old Male

Nina Nandy[1], Joshua Anspach Hanson[2], Dino Beduya[1]

Departments of [1]Medicine and [2]Pathology, Division of Gastroenterology, University of New Mexico School of Medicine, Albuquerque, New Mexico

ABSTRACT

Context Solid pseudopapillary neoplasms of the pancreas are extremely rare, accounting for 1-2% of primary pancreatic exocrine tumors and typically occur in women around the second or third decade of life. Tumors occurring in males are exceptionally uncommon. **Case report** We report herein a case of solid pseudopapillary neoplasm occurring in the pancreatic head in a forty-seven-year-old Hispanic male causing obstructive jaundice. The patient underwent endoscopic ultrasound and fine needle aspiration of the mass followed by endoscopic retrograde cholangiopancreatography with placement of biliary stents. He ultimately had a pancreatoduodenectomy with pathology confirming solid pseudopapillary neoplasm. **Conclusion** We discuss incidence, clinical presentation and overall survival of this rare lesion in male patients along with implications for post-operative surveillance.

INTRODUCTION

Solid pseudopapillary neoplasms of the pancreas are extremely rare, accounting for approximately 1-2% of primary exocrine pancreatic tumors [1]. They are low-grade malignancies that predominantly occur in young females around the second or third decade of life [2]. Tumors occurring in men are exceedingly uncommon. Lesions are typically large, well-circumscribed rounded and encapsulated in appearance and generally do not cause biliary or pancreatic ductal dilatation [3]. Surgery is the mainstay of therapy and 5-year survival approaches 95 % after primary resection. SPNs follow an indolent course but nodal or distant spread may occur especially in larger lesions, but even these have a favorable prognosis [4].

We describe a rather unusual case of a solid pseudopapillary neoplasm of the pancreatic head causing obstructive jaundice in a 47 year-old man treated by biliary stenting and subsequent pancreatoduodenectomy.

CASE REPORT

A forty-seven-year-old Hispanic male electrician was initially admitted after sustaining severe partial and full-thickness high-voltage electrical burns involving 30% of his total body surface area. After multiple escharotomies, debridements and skin grafting procedures he was extubated but developed a ventilator-associated pneumonia. While on vancomycin, cefepime and metronidazole therapy, he developed elevated liver function tests in a cholestatic pattern which was not present on admission, so inpatient gastroenterology was consulted.

Laboratory values were significant for AST 154 u/L (N: 6-58 µ/L), ALT 244 µ/L (N: 14-67 µ/L), AP 614 µ/L (N: 38-150 µ/L), total bilirubin 4.8 mg/dL (N: 0.3-1.2 mg/dL and direct bilirubin of 4.0 mg/dL (N: 0.1-0.4 mg/dL). He had a history of psoriasis but otherwise no significant past medical, surgical or family history. He took no medications or illicit substances, and reported very infrequent alcohol use. At the request of the inpatient gastroenterology service, additional laboratories were obtained for the evaluation of hepatic injury and included negative hepatitis A IgM antibody, hepatitis B surface antigen, hepatitis B core IgM antibody and hepatitis C antibody. Additionally, anti-nuclear antibody was negative as were anti-mitochondrial and anti-smooth muscle antibodies. Ceruloplasmin was 28 mg/dL (N: 17-46 mg/dL) and GGT was 2770 µ/L (N: 6-98 µ/L).

Doppler ultrasound of the abdomen revealed a dilated common bile duct measuring 12 mm and the presence of sludge within the gallbladder. The hepatic parenchyma was noted to be echogenic which was favored to represent hepatic steatosis. For further evaluation of the biliary tree, magnetic resonance cholangiopancreatography (MRCP) was obtained and showed a 3.5 cm x 3.1 cm x 3 cm solid mass in the pancreatic head compressing the distal common bile duct, concerning for pancreatic adenocarcinoma **(Figure 1)**.

The patient subsequently underwent endoscopic ultrasound (EUS) which again demonstrated a complex

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Correspondence Nina Nandy
Department of Internal Medicine, MSC10-5550,
1 University of New Mexico, Albuquerque, New Mexico, 87131
Phone + 505 272 4756
Fax + 505 272 6819
E-mail nnandy@salud.unm.edu

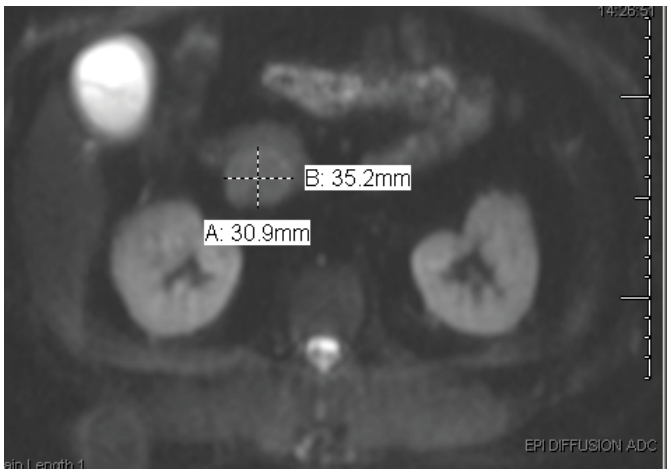

Figure 1. Magnetic resonance cholangiopancreatography (MRCP) showing a 3.5 cm x 3.1 cm x 3 cm solid pancreatic head mass.

pancreatic head mass with calcifications causing compression of the distal common bile duct. Fine needle aspiration (FNA) was obtained and endoscopic retrograde cholangiopancreatography (ERCP) was then performed with successful placement of a plastic biliary stent across the 2 cm distal common bile duct stricture. ERCP brushings showed atypical and reactive ductal cells but were negative for malignancy. FNA revealed dyscohesive neoplastic plasmacytoid cells with finely dispersed chromatin. Immunohistochemical stains performed on the cell block indicated the cells were positive for β-catenin (nuclear and cytoplasmic) and CD10, and were negative for synaptophysin and chromogranin. The absence of synaptophysin and chromogranin excluded a neuroendocrine tumor and nuclear β-catenin positivity combined with CD10 positivity was consistent with a solid pseudopapillary neoplasm **(Figures 2abc)**.

Computerized tomography (CT) of the chest, abdomen and pelvis revealed no evidence of metastatic disease. The patient's biliary obstruction gradually improved and he was discharged. He was re-admitted 2 months later for a planned pancreatoduodenectomy with antrectomy and cholecystectomy, retrocolic end-to-side pancreatojejunostomy and choledochojejunostomy, and anetcolic antiperistaltic side-to-side stapled gastrojejunostomy. The resected pancreas contained a 3.5 cm x 2.6 cm x 2.5 cm solid mass located in the head, with negative margins. The lesion had a largely solid and nested growth pattern interspersed with capillary-sized vascular spaces. Tumor cells were round to oval with nuclear grooves, indistinct nucleoli and pale eosinophilic cytoplasm with some vacuolization and some areas of hyalinization and calcification **(Figures 3ab)**. Immunohistochemical studies demonstrated strong tumor staining with CD10, vimentin, progesterone receptor (PR), nuclear and cytoplasmic β-catenin, CD56 and neuron specific enolase **(Figures 4ab)**. There was no staining noted with cytokeratin AE1/AE3 and chromogranin. Therefore, the staining pattern and morphology confirmed the mass was a solid pseudopapillary neoplasm.

Post-operatively, the patient has been following in the outpatient surgery clinic on a monthly basis mainly to ensure adequate wound healing, and has remained asymptomatic. A repeat CT scan for surveillance of SPN recurrence was planned in one year's time.

DISCUSSION

Solid pseudopapillary neoplasms (SPNs) of the pancreas, also known as Hamoudi or Franz tumors, are extremely rare pancreatic neoplasms of low metastatic potential, accounting for 1-2% of primary exocrine pancreatic tumors [1]. SPNs are unique in that approximately 90% of them occur in young females around the second or third decade of life [2]. A meta-analysis of 718 patients reported in the English literature revealed that approximately 85% of SPNs occurred in females less than 30 years old with a reported mean age of 24 years [4]. There are no known genetic or hormonal factors to explain the strong female predilection but progesterone receptors have been reported as positive in more than 80% of these patients [5]. Lesions are typically large, well- circumscribed, rounded and encapsulated in appearance and generally do not result in biliary or pancreatic ductal dilatation. Our case is unique in that this SPN occurred in the head of the pancreas and caused obstructive jaundice by extrinsic compression of the common bile duct and that it occurred in a 47 year-old male. SPNs arising in males are extremely rare, with very few cases reported in the literature. SPNs are characterized as low-grade malignancies, given expression of abnormal nuclear and cytoplasmic accumulation of β-catenin caused by missense mutations in exon 3 of the CTNNB1 gene that inhibit phosphorylation and degradation of β-catenin [6]. They have potential for local and metastatic spread but despite this have excellent prognosis and low recurrence rates with reported 5-year survival up to 97% even in cases of more aggressive lesions [7]. However, male patients and those with older age, atypical histopathology (i.e. large tumors, diffuse growth, cellular/ nuclear atypia, mitotic activity, necrosis, invasion/metastasis) and incomplete resection may have a higher risk of recurrence and death which deserves particular attention. A recent retrospective analysis of 100 patients with SPNs selected from the National Cancer Institute's Surveillance, Epidemiology and End Results (SEER) database from 2000-2010 suggested that men with SPNs may have compromised survival [8]. A comparison of the 84 female and 16 male patients in the study revealed that men trended toward larger tumor size (7.3 cm *vs.* 6.2 cm p=0.282) with higher rates of extrapancreatic extension (37.5% *vs.* 25.3%, p=0.338), nodal metastasis (25% *vs.* 3.6%, p=0.076) and distant metastasis (18.8% *vs.* 9.5%, p=0.376) but none of the differences were statistically significant. 5-year disease-specific survival in men was also found to be compromised in comparison to women (74.1% *vs.* 91.7%, p=0.026) which did achieve statistical significance. Subset analysis of patients with distant disease showed an even greater reduction in 5-year disease-specific survival in men when compared to women (20.0% *vs.* 71.4%, p=0.072), although not

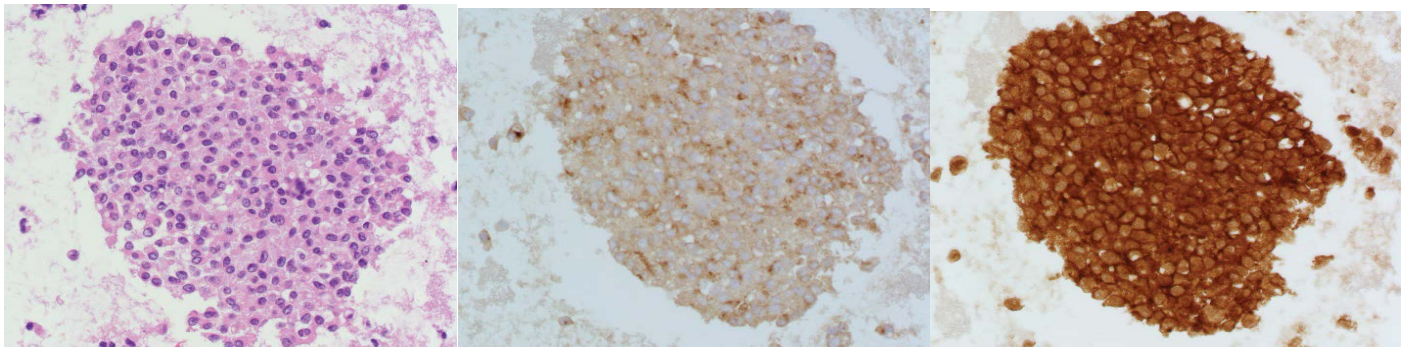

Figure 2. **(a.).** FNA cell block, **(b.).** positive CD10 stain, **(c.).** positive β-catenin stain.

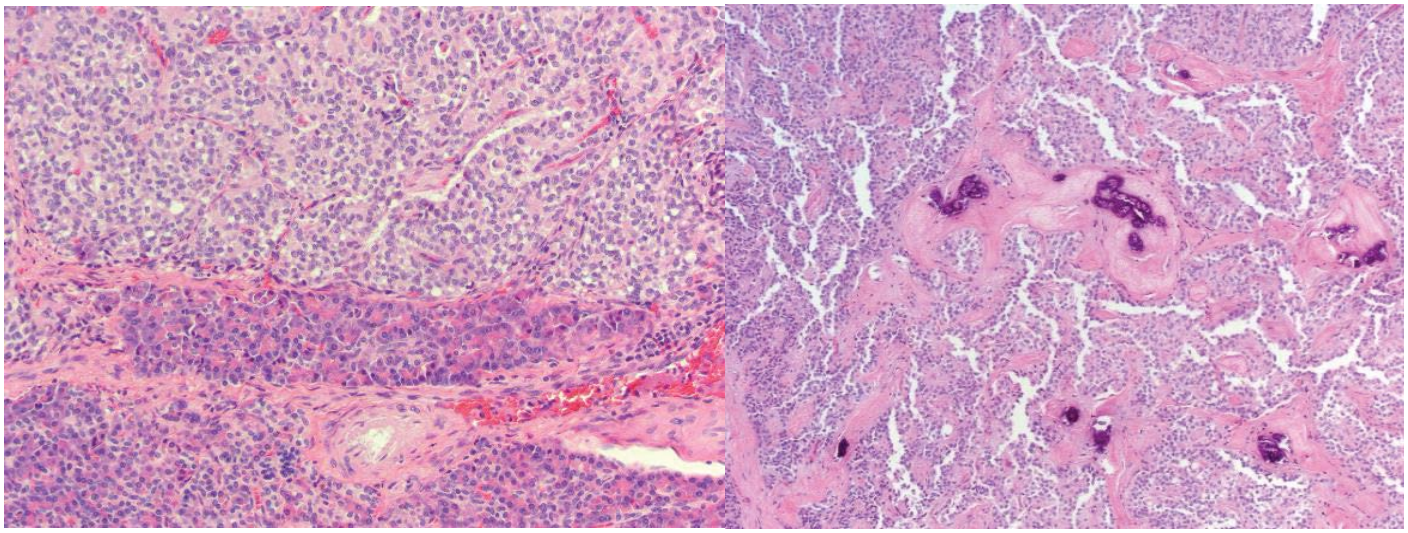

Figure 3. (a.). Tumor seen in upper half of slide of resected pancreas, normal pancreatic tissue seen in the lower half- high power, **(b.).** pseudopapillary architecture of the tumor.

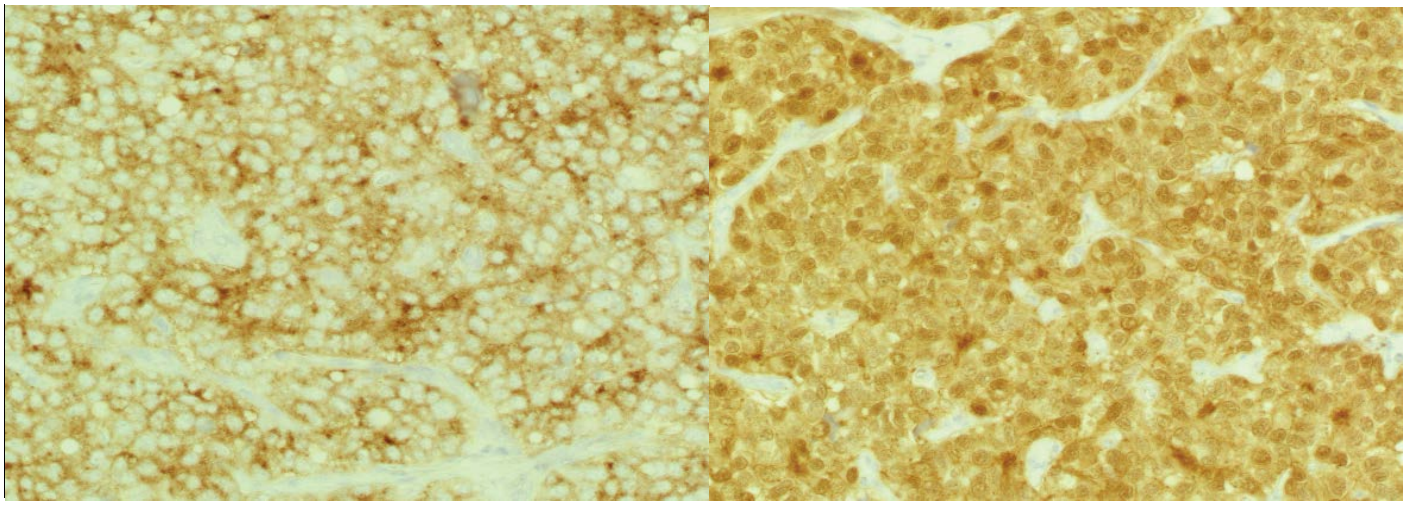

Figure 4. (a.). positive CD10 stain at high power, **(b.).** positive β-catenin stain at high power.

statistically significant [8]. Therefore, even though SPNs have an excellent prognosis in the young women in whom they are typically diagnosed, SPNs occurring in men, although exceedingly rare, may have a more ominous prognosis which could be related to the higher rates of nodal and distant metastasis in this population.

Here we report the occurrence of a solid pseudopapillary pancreatic neoplasm occurring in the head of the pancreas causing obstructive jaundice in a 47 year-old man. SPNs are rare pancreatic exocrine tumors predominantly seen in young women and are extremely uncommon in men. Although the overall prognosis of SPNs is excellent, men with SPNs have been shown to have reduced 5-year survival and may warrant closer post-operative monitoring. Clinical guidelines have not yet been established for post-operative disease monitoring given the rarity of this condition but perhaps as subsequent cases are reported and greater experience is developed, these will need to be revisited especially given the reports of compromised survival in men. Thus, further investigation into recurrence rates after primary resection in men are warranted in order to help create clinical surveillance guidelines.

Conflict of Interest

Authors declare no conflict of interests for this article.

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