Rare solid pseudopapillary neoplasm in a Caucasian male

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Introduction

Solid pseudopapillary neoplasms (SPN) are rare tumors of the pancreas. Epidemiologically they tend to affect women predominantly and occur in patients of Asian or African American ethnic backgrounds. We report a rare case of a solid pseudopapillary neoplasm in a Caucasian man who initially presented with nonspecific acute abdominal pain.

Case Report

A 52-year-old Caucasian man with a past medical history of poor controlled diabetes mellitus and hypertension presented to the emergency room with diffuse abdominal pain and nausea for one day. He described the pain as dull aching, pain score 2 out of 10, and generalized. He has lost 120 Ibs unintentional in one year. Patient denied history of alcohol, tobacco or illicit drug use. His vital signs showed blood pressure 188/117 mm Hg, heart rate 96 beats per minute, respiratory rate 20 breaths/ minute, temperature 97.4°F and oxygen saturation 92% on room air. His body mass index of 27.3 kg/m^2. On examination, he was alert and in no acute distress. He had clear breath sounds on auscultation bilaterally. His abdomen was soft and non-distended, with tenderness at the epigastrium and normal bowel sounds.

Laboratory workup showed white blood cell count of 12.31 k/µl (ref 4.23-9.07), hemoglobin 13.6 g/dL (13.7-17.5 g/dL), and platelet count 219 k/µl (165-400 k/µl). Serum glucose 247 mg/dL, and hemoglobin A1c was 13.2% (4.0-6.0%), alkaline phosphate 115 IU/L (35-129 IU/L), aspartate transaminase 71 IU/L (5-37 IU/L), and alanine transaminase 166 IU/L (5-41 IU/L). Computed tomography (CT) pancreatic protocol showed a 1.7 cm x 1.5 cm solid appearing mass in the body of the pancreas (Figure 1A). Magnetic resonance imaging (MRI) of the abdomen without contrast re-demonstrated a 1.5 cm solid lesion in the body of the pancreas and a 5mm central calcification within the mass. A few mildly prominent lymph nodes are seen in the porta hepatis. An endoscopic ultrasound (EUS) showed a 18.2 mm x 15.4 mm hypoechoic, homogeneous, round mass with central calcification and acoustic shadowing near the genu/proximal body of the pancreas (Figure 1B), and fine needle biopsy (FNB) was obtained. Pathology showed an admixture of solid and pseudopapillary areas forming fibrovascular stalks and rosette-like structures; stroma showed various degrees of hyalinization and evidence of degeneration, foamy macrophages and calcification (Figure 2A and 2B). Tumor cells stained positive for beta-catenin, androgen receptor, CD56, CD10, focally positive for pancytokeratin and synaptophysin and stain negative for E-cadherin and CD45 (Figure 3A and 3B). The morphology and immunophenotype on pathology were most consistent with solid pseudopapillary neoplasm of the pancreas.

Patient was referred to hepato-pancreato-biliary surgery for surgical evaluation. With uncontrolled hypertension and diabetes, patient was considered high risk for surgery at this time. He is currently managed conservatively with annual imaging surveillance.

Discussion

A pancreatic solid pseudopapillary neoplasm (SPN) is a rare exocrine pancreatic tumor and accounts for

1-3% of all pancreatic tumors.[1] It was first described by Frantz in 1959.[2] SPN has a female predominance with a female to male ratio of 10:1. Additionally, it predominantly occurs in Asian and African American women. Women usually present at a younger age compared to men with the mean ages for females and males at the time of presentation being 25 and 35, respectively.[3,4] Typical clinical manifestations include vague abdominal pain (40%) and palpable abdominal mass (33%). Some patients may have poor appetite and nausea which may be secondary to compressive effects of the tumor on the stomach and adjacent organs.[5,6] However, 20% of females and up to 40% of males are asymptomatic[6]. SPN is a rare occurrence in men. The common location of the mass in male patients is body-tail region which account up to 65% of the total cases; the location in females usually involves the pancreatic head.[5]

Solid pseudopapillary neoplasms is detected by imaging incidentally in asymptomatic patients. Multiple imaging modalities for SPN can be utilized. Abdominal ultrasound shows a hypoechoic, clear-bordered cystic or cystic-solid mass. Computed tomography and magnetic resonance imaging of abdomen describe a large well-circumscribed lesion with heterogenous density and solid and cystic components.[6,7] Endoscopic ultrasound (EUS)-guided fine needle aspiration and biopsy (FNA/B) is diagnostic.[8]

The tumor markers for pancreatic cancer are usually negative in patients with pancreatic SPN. These markers include the carcinoembryonic antigen (CEA), serum cancer antigen (CA) 19-9, and serum CA 72. Tumor markers are elevated in less than 10% of patients with SPN. Therefore, no tumor markers or specific laboratory test have been established for diagnosing pancreatic SPN to date. The definite diagnosis is mainly based on imaging studies and pathological tissue analysis from FNA/FNB specimens.

The risk of malignant potential is estimated to be around 15%. The tumor can be locally aggressive and extend into adjacent blood vessels and organs. There is potential for local recurrence and distance metastasis.[8] Distant metastases were reported in about 4% cases at the time of diagnosis. According to a multicenter study by Matos, surgical resection is the treatment of choice for SPN.[9] Patients have an excellent prognosis with 5-year survival rate as high as 97% in patients undergoing surgical resection and 10-year survival rate at 96%.[5,10,11]

Figure legends

Figure 1 (A) Computed tomography (CT) pancreatic protocol showed a mass at the pancreatic body measuring 1.7 cm x 1.5 cm with a central calcification **(B)** Endoscopic ultrasound (EUS) showed an 18.2 mm x 15.4 mm hypoechoic, homogeneous, round mass with central calcification and acoustic shadowing near the genu/proximal body of the pancreas.

Figure 2 (A) Hematoxylin and eosin stain 10x and**(B)** Hematoxylin and eosin stain 20x show admixture of solid and pseudopapillary areas (tumor cells getting detached from blood vessels forming fibrovascular stalks/rosette-like structures). **(A)**Stroma shows various degrees of hyalinization, evidence of degeneration, and hemorrhage.

Figure 3 (A) shows the immunohistochemical stain Beta Catenin with aberrant nuclear expression **(B)** shows the immunohistochemical stain Androgen receptor with strong nuclear positivity.

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