Background

Solid pseudopapillary neoplasms (SPN) are rare, low-grade, malignant, pancreatic neoplasms that encompass < 3% of all pancreatic tumors. First described by Dr. Frantz in 1959, there have only been 3865 well-documented cases with >1 year follow-up period reported in the literature in the past 50 years; however, their recognition and diagnosis have been increasing due to growing awareness of this disease process and increased incidental findings on imaging. Affecting women compared to men at a 9:1 ratio, SPN are typically identified during the second or third decade of life and are often characterized via CT imaging as large heterogeneous lesions with both solid and cystic components. Local recurrence and metastasis of SPN is very uncommon; however, prognosis in these cases is excellent and surgical resection is potentially curative. There is no clear consensus on role of adjuvant chemotherapy. Although SPN is less aggressive than many other pancreatic neoplasms, with surgical resection providing favorable long-term outcomes and is potentially curative.

Case 1

A healthy 42-year-old man presented with 2 months of bloating and diarrhea. Imaging revealed a 12.6 cm encapsulated mass in the tail of the pancreas (Fig 1). Initial Surgery: distal pancreaticectomy and splenectomy with resection of a 2.5 x 10.5 x 10.0 cm node-negative tumor. Pathology Dx: SPN. Patient was discharged on adjuvant PODIX.

4 years post-op, f/u CT PET showed a hypermetabolic soft tissue mass in the splenic fossa (Fig 2). Biopsy confirmed recurrent SPN.

Pt underwent cytoreductive surgery including surrounding visceral resection and peri toneectomy.

1 year following his second surgery, three enlarging para-aortic nodes were noted on surveillance CT (Fig 3). He was trialed on multiple chemotherapy regimens including capectabine, gemcitabine, FOLFOX, and Keytruda however, nodes continued to enlarge. 6 years after initial diagnosis, the patient underwent an additional cytoreductive surgery, including left nephrectomy (Fig 4) alongside heated intraperitoneal chemotherapy (HIPEC)

1-year post-op, a new small pleural-based left lower lobe nodule was seen on CT. A VATS wedge resection was performed which again revealed metastatic pancreatic SPN.

9 years after diagnosis, the patient has undergone one more metastatic resection of recurrent peri toneal and diaphragmatic metastatic tumors.

The patient has remained active and otherwise healthy despite his extensive metastasectomies comprising 6 major surgeries over ten years. He plans to continue aggressive surgical management for recurrent disease.

Case 2

A 57-year-old male with h/o chronic pancreatitis presented with an incidental pancreatic head mass during a COPD study.

CT showed a 5 cm pancreatic mass containing calcifications and encasing the SMV (Figure 5). EUS-guided biopsy was consistent with SPN. The patient underwent a Whipple procedure with resection and reconstruction of the SMV. The Whipple margins and nodes were negative; however, there were positive margins in the resected mesenteric vein. The patient did not undergo adjunctive chemoradiation and was subsequently lost to follow-up.

6 years after his initial surgery, pt reestablished care. CT A/P showed a 1.1 x 0.3 cm hypodensity in the liver. The lesion continued to grow with f/u imaging. CT showed a hypermetabolic 1.3 cm lesion in the right hepatic dome (Figure 6). Biopsy confirmed metastatic SPN. The patient underwent resection of metastasis including a partial hepatectomy. He had no evidence of recurrence on 3-month follow-up imaging.

Case 3

A 48-year-old female presented with 2 years of epigastric pain radiating to the back. CT showed a 7.6 x 9.7 cm solid-appearing heterogenous mass arising from the body and tail of the pancreas. EUS-guided biopsy was consistent with SPN.

Intraoperatively, a 8 cm mass was found occupying the body and tail of pancreas extending through the left transverse mesocolon. A distal pancreaticectomy, splenectomy, and segmental transverse colectomy were performed with negative margins. The patient did not undergo adjuvant chemotherapy. She discontinued annual surveillance scans 3 years after surgery as imaging continued to show no evidence of disease.

She presented 10 years after her initial surgery with right upper quadrant pain. CT showed a 11.6 x 10.8 x 9.5 cm mass centered in the right hepatic lobe (Figure 8) A biopsy confirmed recurrent SPN. The patient underwent Y-90 therapy to the right lobe to shrink the mass and hypertrophy to the left liver lobe in anticipation of right hepatectomy. 3 months after treatment, CT showed interval decrease in tumor size (Figure 9). A right hepatectomy was performed with negative margins and benign lymph nodes. She has completed 3 years of yearly surveillance imaging (primarily with MRI) with no evidence of recurrent disease.

Discussion / Takeaways

• SPN is less aggressive than many other pancreatic neoplasms, with post-treatment five-year survival rates surpassing 95%.

• Surgical resection provides favorable long-term outcomes and is often curative.

• Recurrence and metastatic disease is rare, between 6-11% of cases.

• The timeline of recurrence is variable, with documentation of metastasis up to 15.8 years after initial surgery.

• Male sex has been identified as a risk factor of both metastases and death, possibly due to the role of progesterone in oncoregulation.

• Additional risk factors include: tumor size >8cm, synchronous metastasis, lymphovascular or parenchymal invasion, and Ki-67 index >4%.

• Our institutional experience and literature review suggest that surgical resection of metastatic disease provides favorable long-term outcomes and is potentially curative.

• Adjuvant chemotherapy has been used with variable success. Common agents include Cisplatin, 5-FU, and Gemcitabine.

• For large liver metastases, Y-90 has been used successfully to shrink the tumor and hypertrophy the FLR. Liver transplantation for unresectable metastases has also been successful in this setting.