

Metastatic Solid Pseudopapillary Neoplasm: A Case Series and Scoping Review

J Fulk-Logon, S Rodriguez Franco, R Henkind, F Ho, RA Choudhury, MG Huey, M Patten, YJ Bababekov, M Del Chiaro, RS Schulick

Affiliations: CU Anschutz School of Medicine¹, Department of Surgery²

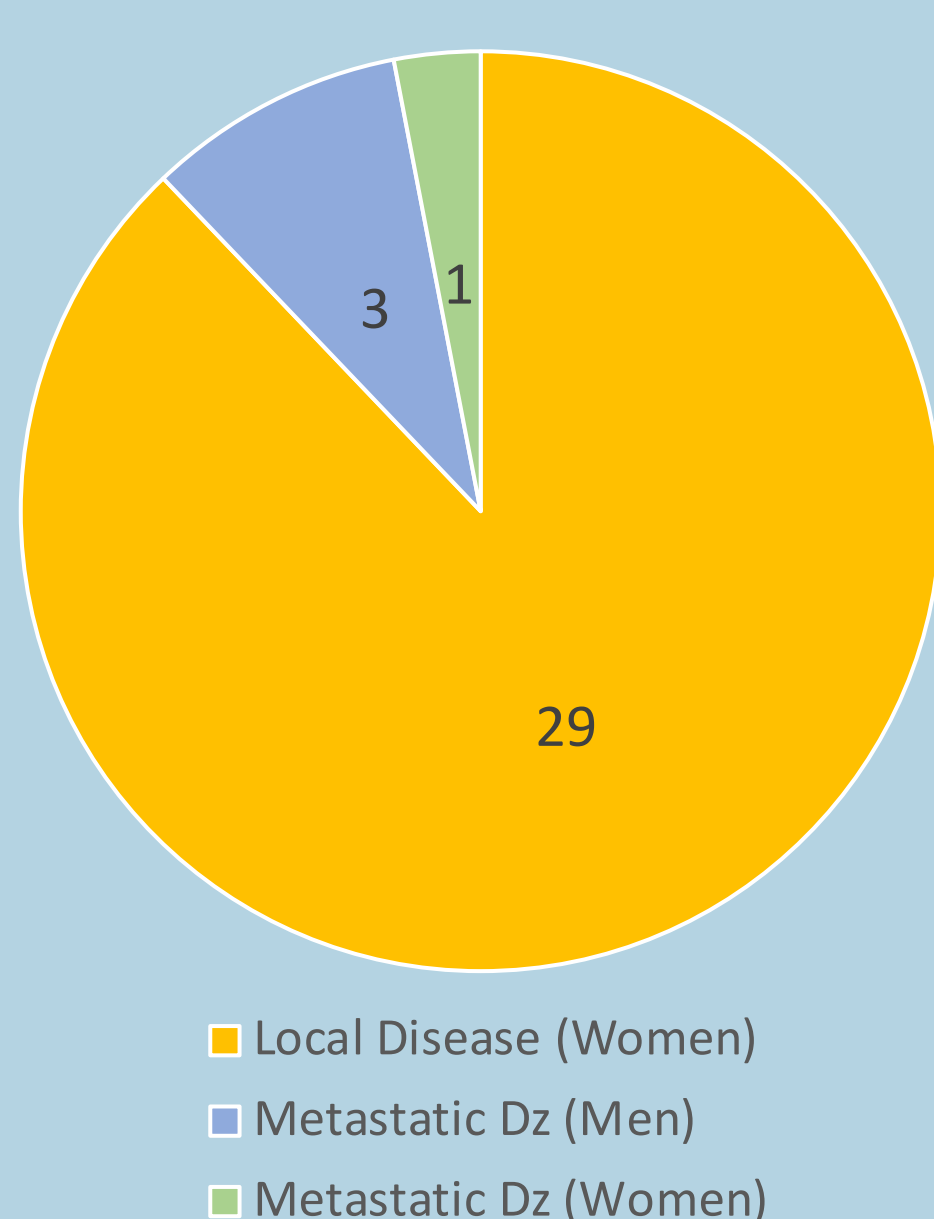


Background

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 predominant in young women, older men appear to be at higher risk for developing metastasis.

Institutional Review

A total of 33 patients with SPN were identified in our institutional review. Among the 33 cases, 4 (12%) had recurrence or metastasis (mSPN) and underwent subsequent resection. 3 of the 4 patients were men, with an average age of 54. Local recurrence occurred in 2 patients and distant liver metastasis in the other 2. Median time to recurrence was 72 months (range 48-120) among our series. All mSPN patients were alive, with no major complications described at the moment of this report. Interestingly, one patient has undergone 6 tumor resections over the last 10 years.



Metastatic Cohort
 Mean age: 54
 Sex: 75% male
 Median time to Recurrence: 72 mo
 Survival rate: 100%

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 pancreatectomy 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 PODX.
4 years post-op, f/u CT PET showed a hypermetabolic soft tissue mass in the splenic fossa (Fig 2). Biopsy verified recurrent SPN.
 Pt underwent cytoreductive surgery including surrounding visceral resection and peritonectomy.
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 capecitabine, 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.
Now 9 years after diagnosis, the patient has undergone one more metastatic resection of recurrent peritoneal 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.



Figure 1.
CT image of 12.6 cm mass in the tail of the pancreas (initial presentation)

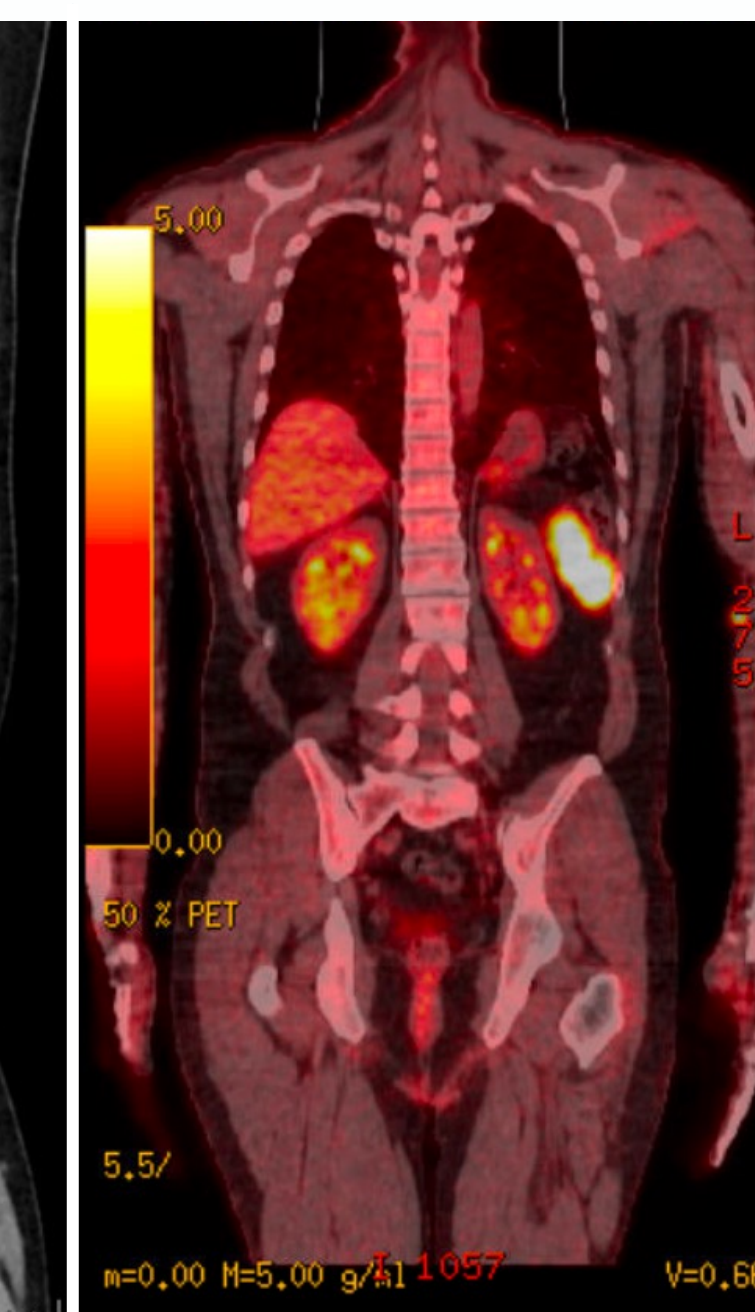


Figure 2.
PET-CT showing hypermetabolic tissue in the splenic fossa, 4 years after initial resection

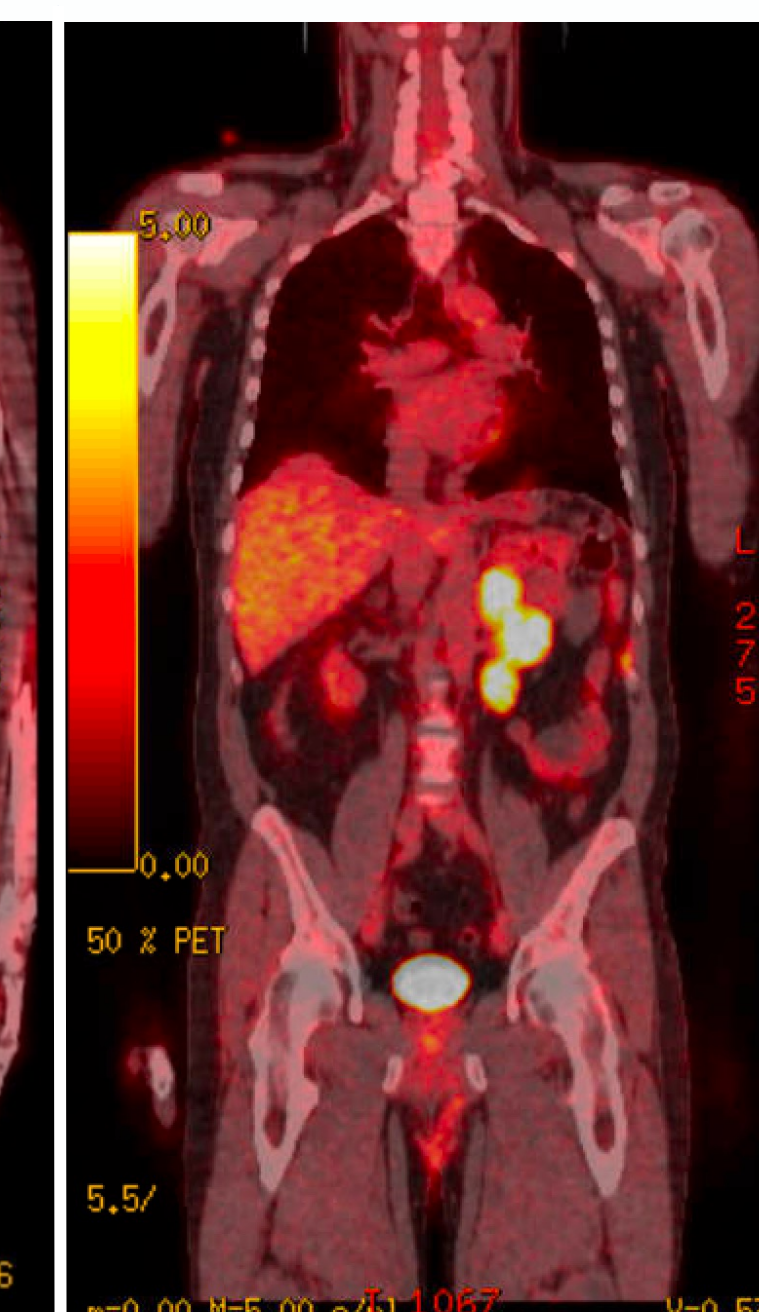


Figure 3.
PET-CT showing hypermetabolic para-aortic nodes, 2 years after second resection

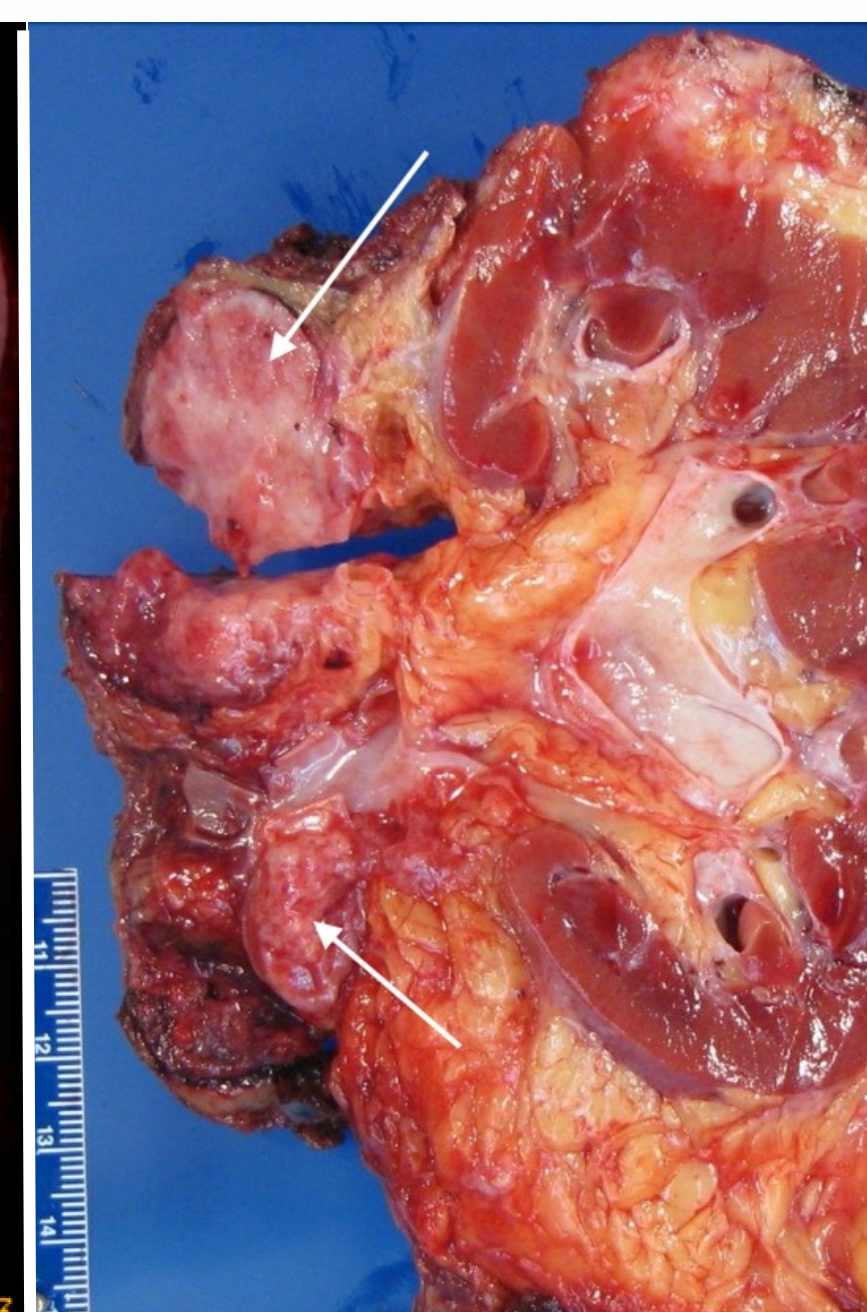


Figure 4.
7.9 x 3.5 x 5.5 cm metastatic lesion in the renal sinus encircling the ureter

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 adjuvant 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 0.9 cm hypodensity in the liver. The lesion continued to grow with f/u imaging. PET 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.

Figure 5.
CT image of 5cm pancreatic head mass



Figure 7.
Right hepatic lobe metastatic lesion

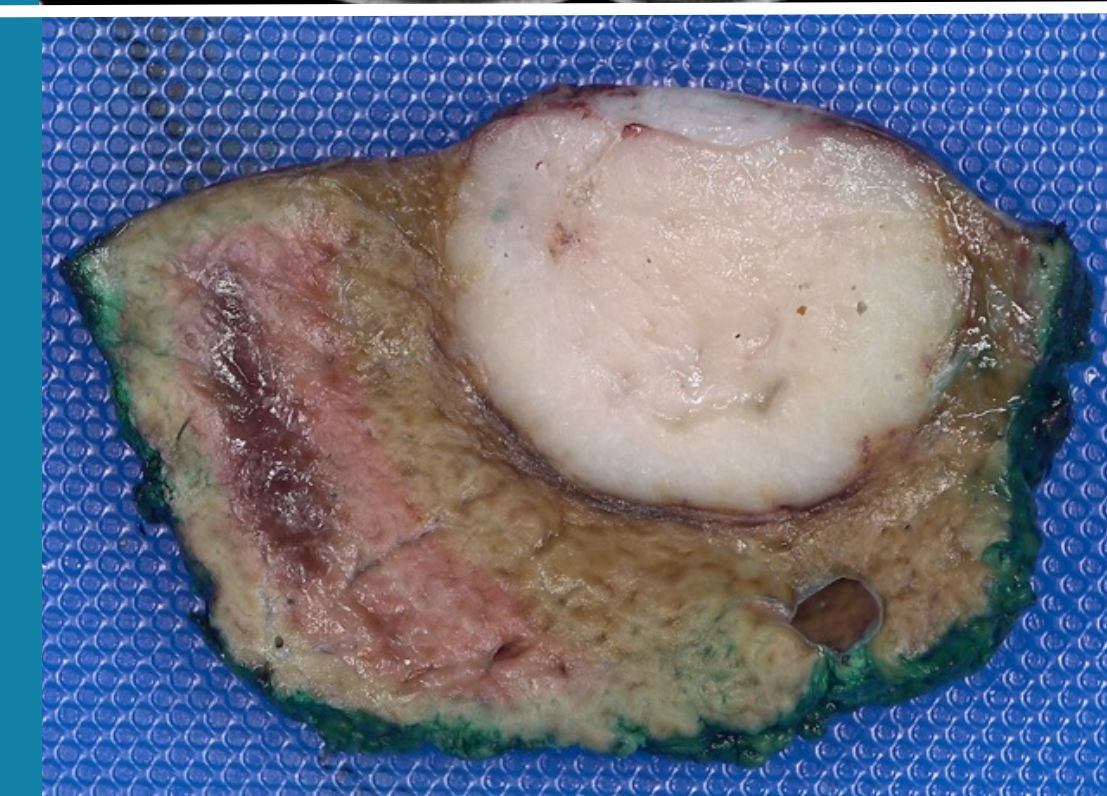
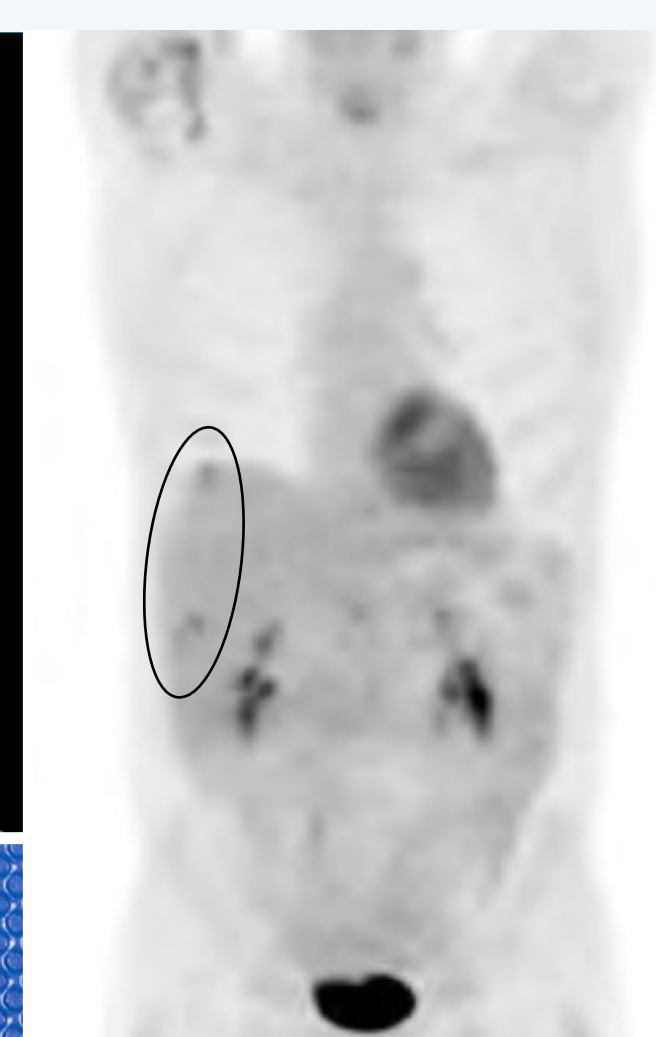


Figure 6.
PET image of 1.3 cm hypermetabolic lesion in the right hepatic dome



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 pancreatectomy, 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.

Figure 8.
CT image of 7.6 x 9.7 cm mass in the body and tail of the pancreas

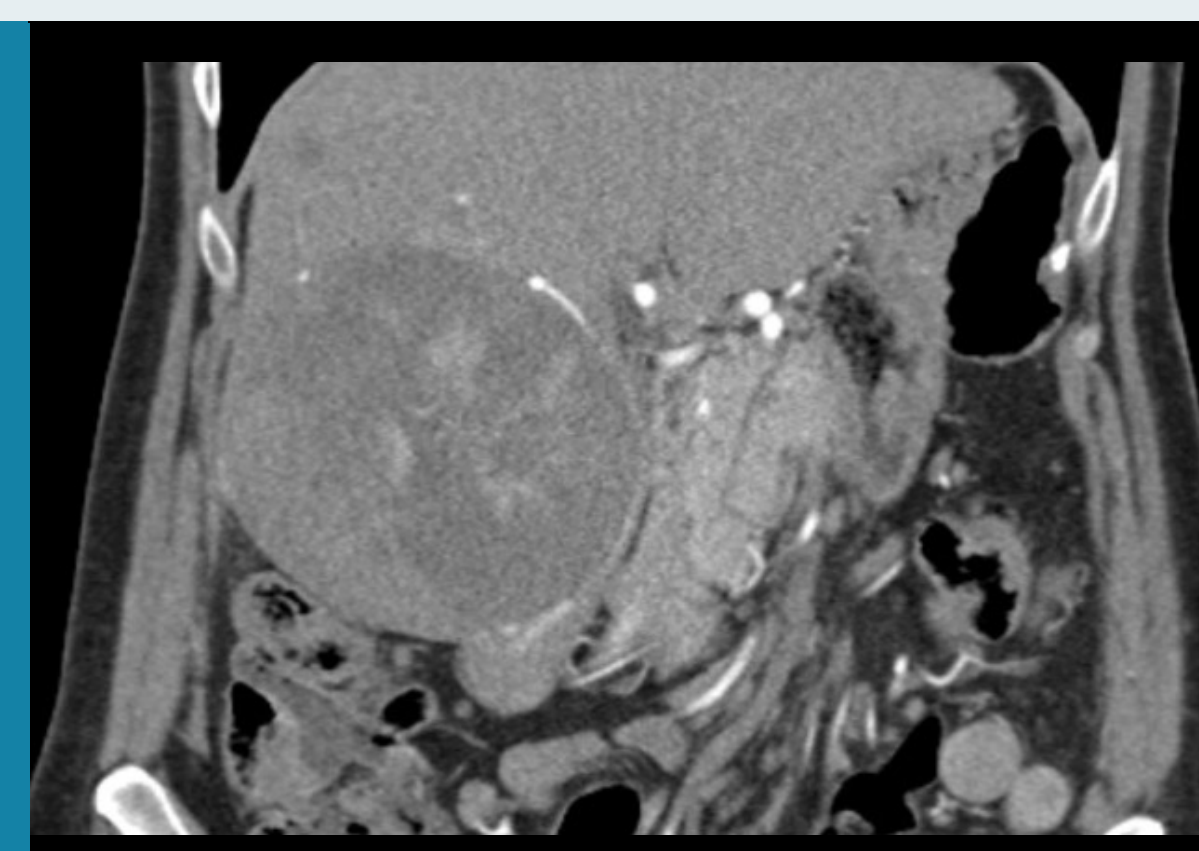
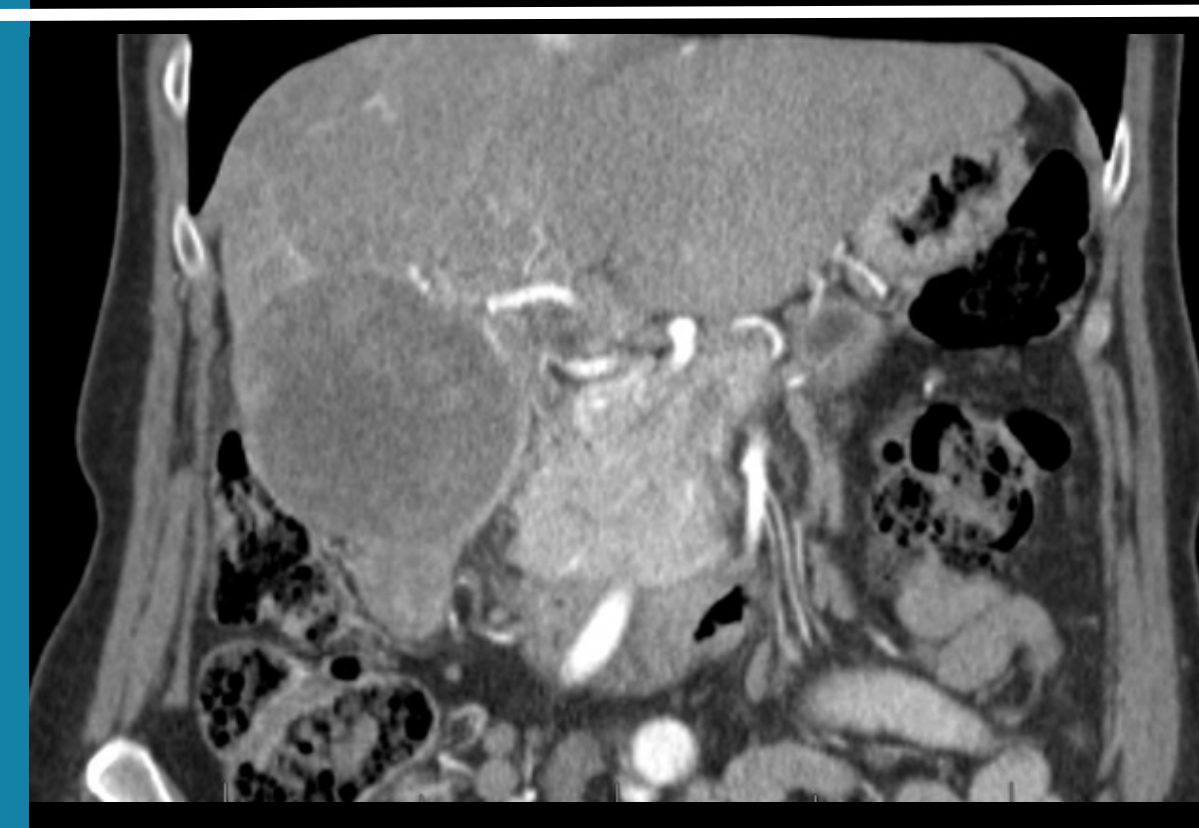


Figure 9.
CT image of hepatic mass status post Y-90 radio-embolization



Case Series Data Table

Case #	Patient Gender	Age	Primary Tumor Location	Primary Tumor Size (cm)	Initial Stage	Surgery	Recurrence/ Metastasis Location	Time to recurrence
1	Male	42	Pancreatic tail	10.5	T3N0, stage IIA	DP	Local, para-aortic nodes, lung	4 years
2	Male	57	Pancreatic head	5	T3N0, stage IIA	PD	Liver	6 years
3	Female	48	Pancreatic body/tail	9.7	T3N0, stage IIA	DP	Liver	10 years

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.