Metastatic Solid Pseudopapillary Neoplasm of the Pancreas, Rare Presentation of an Uncommon Tumor

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Introduction

- Solid pseudopapillary neoplasms (SPN) make up less than 3% of all diagnosed pancreatic neoplasms.
- Abdominal CT or MRI are the imaging modalities most utilized, and biopsy is necessary to confirm the diagnosis.
- Resection of the mass is standard of care and the prognosis is quite good, with 5-year survival rates of over 95%.
- Both recurrence and metastasis of the tumor are exceedingly rare, occurring in only 1-4% of patients.

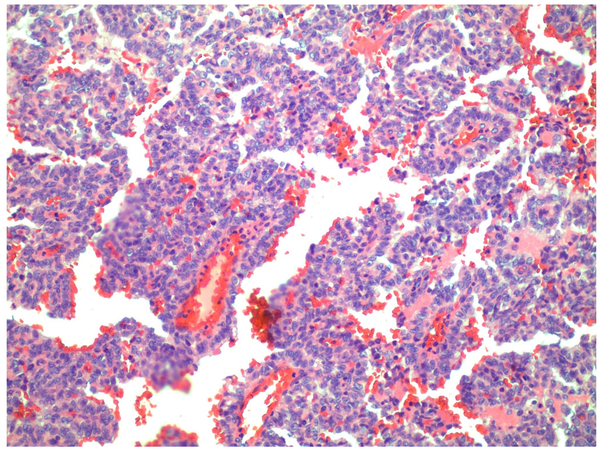

Figure 1: Biopsy of the caudate liver lesion showing clusters of uniform, bland cells with ovoid nuclei and associated fibrillary stroma and hemorrhagic debris, consistent with SPN

Case Report

- A 26-year-old female presented with several years of abdominal pain. CT scan revealed a 10 cm mass in the neck and body of the pancreas and patient underwent a hybrid resection including subtotal pancreatectomy, splenectomy, and cholecystectomy. Pathology showed a neoplasm extending to the inked margins with positive beta-catenin and CD10 nuclear staining with no evidence of metastasis.
- At the age of 32, the patient presented again with abdominal pain. A CT and PET scan showed numerous LUQ, perirenal, pancreatic, and periadrenal soft tissue densities. Laparoscopic resection of a 3.5x3 cm soft tissue mass near the remnant pancreas was performed, showing fibrous stroma with angulated islands of epithelioid cells with cystic degeneration and positive staining for vimentin, CD56, and beta-catenin. Resection of multiple abdominal soft tissue tumors, in bulk measuring more than 10 cm, as well as complete omentectomy, left adrenalectomy, bilateral salpingo-oophorectomy, appendectomy, and hyperthermic intraperitoneal chemotherapy with cisplatin was performed. Pathology showed multiple metastatic tumors all consistent with SPN. The appendix, left adrenal gland, bilateral ovaries, and all lymph nodes were negative for any tumor.
- Two years later, MRI revealed 3.0 x 2.1 cm hyperintense lesion in the caudate lobe of the liver. EUS with biopsy was performed, revealing clusters of uniform, bland cells with ovoid nuclei and associated fibrillary stroma circumscribing vessels with a background of hemorrhagic debris (Figure 1). Immunohistochemistry showed cells positive for vimentin, LEF1, and beta-catenin, again consistent with SPN.

Discussion

- This case represents an extremely rare case of multiple recurrences and metastases of solid pseudopapillary neoplasm after resection.
- Lymph node involvement is present in 2.2% of cases, metastasis in 1-2.9% of cases, and recurrence in 2-4.4% of cases
- Serial imaging studies and close follow-up are needed, even years after resection, and creating a follow up plan on a case-by-case basis while paying special attention to those patients with high-risk features is imperative in cases of SPN

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