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CASE REPORT | LIVER

Unresectable Metastatic Solid Pseudopapillary Pancreatic Neoplasm Treated With Liver Transplantation

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ABSTRACT

We present a 49-year-old woman requiring living donor liver transplantation after liver metastasis from a pancreatic solid pseudopapillary tumor. After identifying a pancreatic mass and liver lesions, she underwent extensive surgical resection. Pathology revealed a solid pseudopapillary neoplasm of the head and body of the pancreas, extending into the peripancreatic soft tissues and confirmed to have spread to the liver. Subsequently, she underwent adjuvant chemotherapy and radiofrequency ablations of the new liver lesions. Despite immunotherapy and chemotherapy, there was a progression of the lesions. With interval growth of liver lesions, without evidence of extrahepatic disease, she underwent living donor liver transplantation.

KEYWORDS: solid pseudopapillary pancreatic neoplasm; metastasis; liver transplantation; case report

INTRODUCTION

Transplantation has been used to treat primary and secondary liver lesions, including neoplasms and metastases. One scenario that has been scantly described in the literature is liver transplantation because of metastasis from a rare pancreatic neoplasm known as a solid pseudopapillary tumor. Solid pseudopapillary pancreatic tumors were previously considered benign masses but have been reclassified as low-grade malignancy because of potential for spread from the original location in the pancreas.[1] It has been estimated that 10%–15% of individuals will have metastatic lesions or develop tumor recurrence after pancreatectomy.[2] Despite the potential of metastasis, pseudopapillary pancreatic tumors generally follow an indolent course, with studies showing a 5-year survival of up to 97% of patients.[2] These tumors account for 1%–2% of all pancreatic neoplasms and are often discovered incidentally during imaging for unrelated issues because the tumors generally remain asymptomatic until late stages.[3] The most common complaint when symptoms present is abdominal pain.[4] Because of their rarity, their treatment and clinical management guidelines remain unclear. Surgical resection is the mainstay treatment for uncomplicated tumors limited to the pancreas. However, management is further complicated in cases of metastases, particularly for unresectable liver lesions, for which liver transplantation (LT) may be an option.[2,5,6] In this case report, we present a 49-year-old woman requiring liver transplantation after liver metastasis from a pancreatic pseudopapillary tumor.

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CASE REPORT

The patient initially presented in 2013 with severe abdominal pain. Subsequent workup identified a sizable pancreatic mass along with liver lesions. The tumor was tested for genetic changes and found to carry mutations in both CTNNB1 and POLB. The patient also has a germline alteration in MUTYH. Testing further demonstrated elevated tumor markers, including carcinoembryonic antigen: 4.8–10.5 ng/mL and carbohydrate antigen 19-9: 20.6–48 U/mL.

She underwent a total pancreatectomy, central liver resection, distal gastrectomy and splenectomy, partial small bowel resection, and omentectomy. The pancreatectomy was complete while simultaneously resecting liver metastases, classifying it as an R2 resection. The pathology revealed a 15-cm solid pseudopapillary neoplasm of the head and body of the pancreas, with extension into the peripancreatic soft tissues, and the same diagnosis was confirmed in the liver lesions (Figure 1). The liver resections included a 1.5-cm lesion in segment 4, partial resection of a 0.7-cm lesion

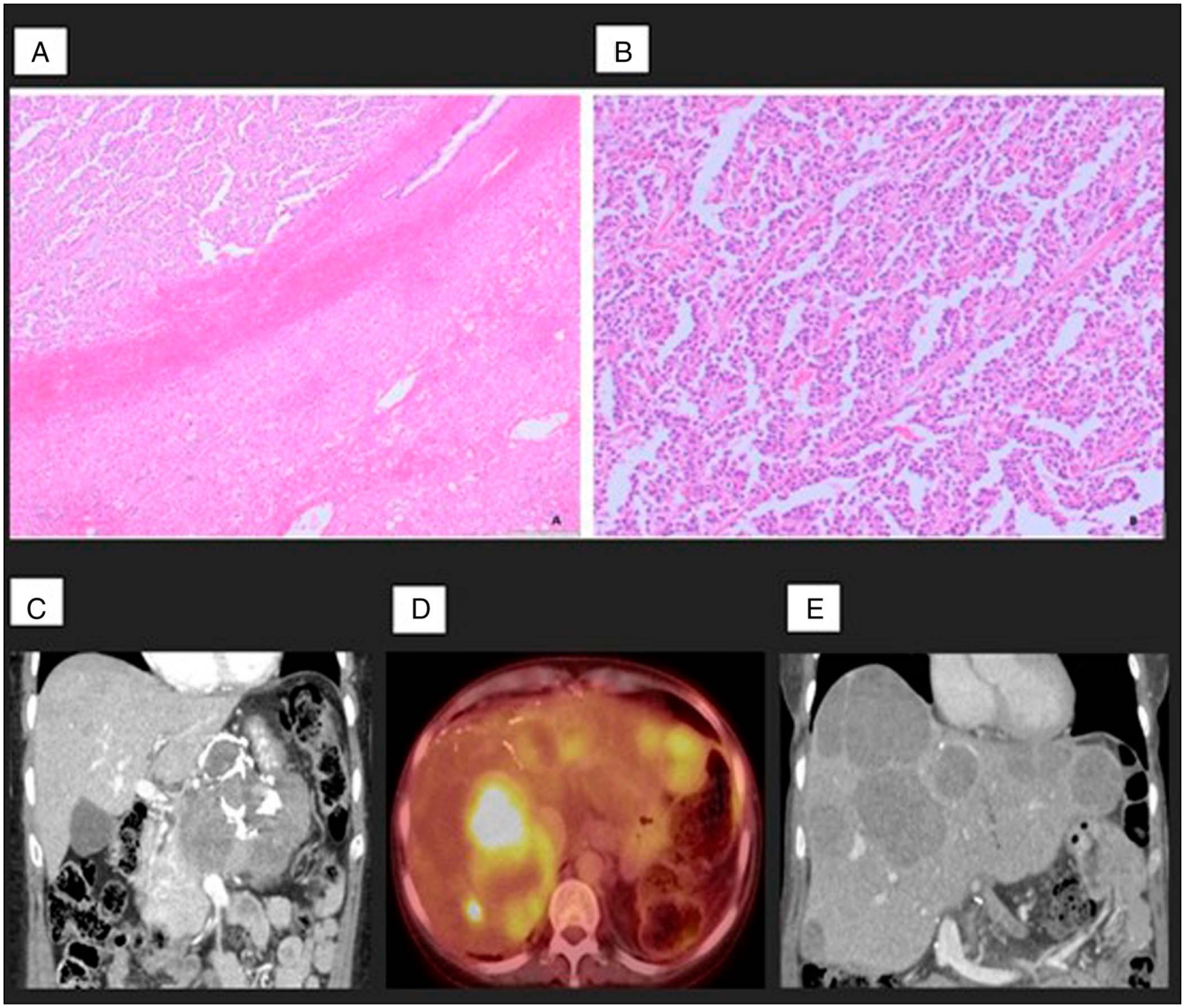

Figure 1. (A) Hematoxylin and eosin (H&E) stain (40× magnification) showing metastatic solid pseudopapillary tumor (upper left corner) involving the liver. (B) H&E stain (100× magnification) showing metastatic tumor forming the characteristic pseudopapillary pattern with thin-walled fibrovascular stalks surrounded by poorly cohesive monomorphic cells. (C) Initial contrast-enhanced computed tomography performed on April 16, 2013. The image shows a large mixed cystic and solid mass with eggshell calcifications occupying the pancreatic body and tail. (D) Axial images of fluorodeoxyglucose positron emission tomography-computed tomography performed on August 20, 2019. The image shows postsurgical changes related to hepatic wedge resection. There are multiple hypermetabolic metastatic focal lesions involving right and left hepatic lobes. (E) Coronal contrast-enhanced computed tomography performed on May 18, 2020, just before living donor liver transplantation. The image shows numerous hepatic metastases throughout the liver.

in segment 5, and a partial resection of a 0.5-cm lesion in segment 8. The tumor showed classic pseudopapillary histomorphology and aberrant nuclear expression for beta-catenin, as well as positivity for synaptophysin, CD10, and progesterone, supportive of the diagnosis. The bile duct, stomach, spleen, and small intestine were free of tumor. Fifty-four examined lymph nodes were negative for malignancy.

She underwent adjuvant chemotherapy, consisting of a regimen that included pembrolizumab for 11 cycles, followed by folinic acid, fluorouracil, and oxaliplatin (FOLFOX) for 3 cycles, and then gemcitabine every 2 weeks for 16 months. FOLFOX was discontinued after 3 cycles because of progression, with increased size and density of hepatic metastases noted on computed tomography and positron emission tomography. Chemotherapy was followed by several radiofrequency ablations of new lesions in both the left and right liver lobes. There was progression despite immunotherapy and chemotherapy. With interval growth of liver lesions, she underwent living donor liver transplantation (LDLT) in 2020.

Pretransplant imaging revealed a 5.9-cm lesion in segment 2, which had expanded from 4.4 cm, a 3.6-cm lesion in segment 6, formerly 2.2 cm, and an 8.4 × 12.1-cm lesion in segments 7/8, previously measured at 7.7 × 19.9 cm. This growth occurred over a span of 3 months. LDLT was selected because of her ineligibility for a Model for End-Stage Liver Disease (MELD) exception. The absence of known underlying chronic liver disease apart from metastases ultimately rendered her MELD score insufficient to compete for a liver offer. At the time of transplantation, her lesions were progressively growing despite treatment, with no evidence of extrahepatic disease, making the timing for LDLT optimal. On examination after transplant, multiple nodules ranging from 3 to 12 cm were observed in all segments of the liver. In addition, a portal lymph node displayed metastatic disease, measuring 2.4 cm, with involvement of the diaphragm that was ultimately resected. As of 44 months after LDLT, she is doing well on tacrolimus mono-immunosuppressive therapy without evidence of disease recurrence.

DISCUSSION

Our case adds to the literature on the limited use of LT for this rare neoplasm. Reddy et al found 6 cases of LT for metastatic pseudopapillary pancreatic tumors.[5] Of these 6 cases, 2 were LDLT, and 4 were deceased donor transplants. Each case shared a similar approach that included confirming the lack of extrahepatic disease and a period of observation after the identification of liver metastasis.[5-10] The first case was reported in 2007 by Sumida et al, who attributed the success of the transplant to the slow growth of this tumor type, even in cases of metastasis.[6] All 6 cases occurred in females with an average age of 31 years. The average disease-free survival based on the last reported follow-up for the 6 cases was 2 years (2 years, 2 years, 9 months, 5 years, 1 year, and 15 months, respectively).

Solid pseudopapillary neoplasms are uncommon tumors found in the pancreas. Treatment varies based on the presence or absence of metastasis. In cases of metastasis to the liver, liver transplantation may be a viable option if surgical resection of the masses is not possible. Our patient is the seventh documented case of liver transplantation for treating pancreatic pseudopapillary tumors that metastasized to the liver. The MELD score for a patient with metastatic pseudopapillary pancreatic neoplasm would not be expected to be competitive, even in United Network for Organ Sharing regions with low median MELD at transplant. Consideration for MELD exception points should be considered, as should LDLT.

DISCLOSURES

Author contributions: C. Tom: Drafting and proofing of final manuscript. P. Mohamed, Y. Gong, M.D. Hopstone, P.S. Conti, and V. Karne: Interpretation and preparation of the images and proofing of final manuscript. Y. Genyk and N. Kaur: Data acquisition and proofing of final manuscript. J.A. Kahn: Conception and design, data acquisition, analysis, interpretation, drafting, and proofing of the manuscript and is the article guarantor. All authors agree to be accountable for all aspects of the work.

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Informed consent could not be obtained for this case report. All identifying information has been removed.

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