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CASE REPORT | BILIARY

Metastatic Pancreatic Adenocarcinoma Presenting as a Klatskin Tumor

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ABSTRACT

Cholangiocarcinoma is a rare clinical entity representing approximately 2% of all cancers diagnosed. More than 65% of these tumors can present at the bifurcation of the hepatic duct and are known as Klatskin tumors. Pancreatic cancer is among the top 5 leading causes of cancer death, and it usually presents at an advanced stage with metastasis commonly seen in the liver. We report a patient with established pancreatic adenocarcinoma who presented with an obstructing mass at the hepatic duct bifurcation without any prior history of biliary tract disease. This represents a new diagnosis of either intracholedochal metastatic tumor or a new diagnosis of cholangiocarcinoma found at the hepatic duct bifurcation without liver involvement in the setting of pancreatic cancer, suggesting metachronous cholangiocarcinoma with pancreatic adenocarcinoma.

INTRODUCTION

Approximately 60% of patients with pancreatic cancer present with metastatic disease, with the most common location being the liver.[1] Pancreatic metastasis to the bile duct is exceedingly rare, and this is separated by a long period of time in the very few reported cases.

Cholangiocarcinomas are mucin-producing adenocarcinomas that account for approximately 2% of all diagnosed cancers. They are grouped according to their anatomic site of origin. Perihilar tumors make up more than 65% of cholangiocarcinomas. Tumors arising at the bifurcation of the common hepatic duct were named after Klatskin in 1965.[2]

CASE REPORT

A 59-year-old woman presented to an outside hospital with abdominal pain, nausea, vomiting, and unintentional weight loss. Imaging revealed a pancreatic head-neck mass confirmed to be pancreatic adenocarcinoma. Due to comorbidities and poor functional status, the patient was medically unresectable and given gemcitabine every 3 weeks with good initial response. Between her sixth and seventh cycles, she developed severe abdominal pain associated with nausea and vomiting and presented to our institution. Physical examination revealed jaundice and right upper quadrant tenderness. Laboratory studies were notable for total bilirubin 7.7 mg/dL, alkaline phosphatase 497 U/L, aspartate aminotransferase 160 U/L, alanine aminotransferase 304 U/L, and lipase <50 U/L.

Computed tomography of the abdomen and pelvis revealed irregular intrahepatic biliary ductal dilatation and a hypodensity near the head-neck junction of the pancreas measuring 2.8 × 2.6 cm. Magnetic resonance cholangiopancreatography revealed a defect in the proximal common hepatic duct, dilation of the distal common hepatic duct, and marked irregularity and thread-like narrowing of the proximal common bile duct (Figure 1).

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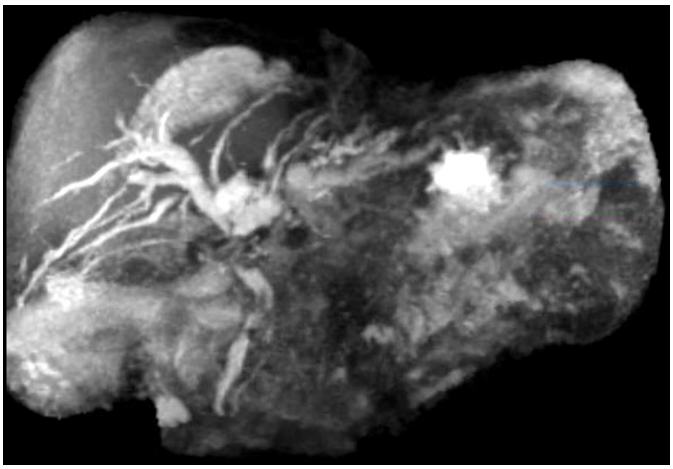

Figure 1. Magnetic resonance cholangiopancreatography showing a filling defect in the proximal common hepatic duct.

An endoscopic ultrasound (EUS) with fine-needle aspiration (FNA) showed an irregular hyperechoic pancreatic head mass measuring 3.2 × 2.6 cm. Endoscopic retrograde cholangiopancreatography showed evidence of a malignant-appearing stricture at the level of hepatic bifurcation with significant proximal dilation (Figure 2). Brushings of the stricture were taken and a 10-Fr 12-cm stent was placed with good bile drainage.

Cytologic analysis of the FNA sample of the pancreatic head mass with Diff-Quik and Papanicolaou stain showed malignant cells with glandular differentiation, significant nuclear pleomorphism, crowding, 3-dimensional clustering, and prominent nucleoli (Figure 3). Biliary tract brushings using Diff-Quik and Papanicolaou stain also showed malignant cells with glandular differentiation, significant pleomorphism, hyperchromasia, nuclear crowding, and 3-dimensional clustering.

Figure 2. Cholangiogram showing a stricture at the level of hepatic bifurcation with significant proximal dilation.

The patient's abdominal pain significantly improved after the stent was placed. She was able to tolerate oral intake and was discharged home in stable condition. She decided to pursue comfort measures and return to her home country to spend her remaining time with her family.

DISCUSSION

We report a rare case of obstructive jaundice in the setting of pancreatic cancer and a new diagnosis of either intracholedochal metastatic lesion or cholangiocarcinoma found at the proximal hepatic duct bifurcation without metasynchronous liver involvement. Computed tomography, magnetic resonance cholangiopancreatography, and EUS imaging all confirmed a defect in the proximal common hepatic duct that was separate from the pancreatic head mass, ruling out a tumor growth extension.

Distinguishing between cholangiocarcinoma and a metastatic pancreatic cancer lesion poses a great challenge. Both present histologically as adenocarcinoma.[3] A number of malignancies can mimic Klatskin tumor, but pancreatic cancer is seldom described.[4] In the literature, we found 3 cases of pancreatic cancer with simultaneous involvement of the biliary tract, but none involving the bifurcation of the hepatic duct.[5,6,7]

All of the bile duct tumors in the literature were in the distal duct near the pancreas.[5,6,7] Our patient had 2 malignancies that were discovered less than 1 year apart and were at considerable anatomical distance from each other without observable extension. Curiously, our patient presented with symptoms of the pancreatic head lesion first without any obstructive symptoms, whereas other cases were diagnosed on the basis of bile duct obstruction and were incidentally noted to have a pancreatic tumor.

Extrahepatic cholangiocarcinoma and pancreatic cancer typically present with abdominal pain, jaundice, and weight loss.[8,9] Schmuck et al.[10] propose that both tumors hold a common developmental origin, tumor genesis, and molecular pattern. Both are difficult to distinguish on diagnostic imaging in late stages, respond poorly to conventional chemotherapy, and require surgical resection.

Cholangiocarcinoma with extrahepatic adjacent organ involvement can be resected with a 5-year survival rate of 38% for a node-negative diagnosis versus 10% for a node-positive diagnosis.[11,12] Survival rates and long-term outcomes for potentially curative resection for cholangiocarcinoma are based on several factors, with the 2 main prognostic factors

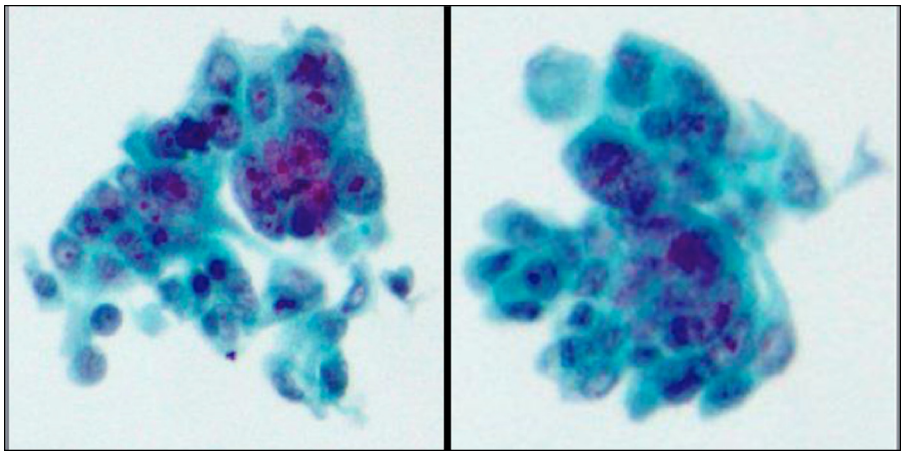

Figure 3. Papanicolaou stain of a fine-needle aspiration sample of the pancreatic head mass. Note the significant nuclear pleomorphism, nuclear crowding, prominent nucleoli, and 3-dimensional cellular arrangement.

being lymph node involvement and histologic margin involvement.[11] Schmuck et al.[10] recommend surgery when a suspected malignant lesion is located in the pancreatic head, regardless of whether it arises from the pancreatic or the bile duct.

An EUS-guided FNA biopsy provides the pathologist with more tissue than cytological brushings for histologic assessment of lesion morphology.[13] In our case, the finding of signet ring features in the FNA from the pancreatic mass raises the possibility of 2 primary tumors because this morphology was not identified in the preparation of the bile duct brushing. Because both cholangiocarcinoma and pancreatic adenocarcinoma demonstrate similar patterns of differentiation as well as immunohistochemical expression, further immunohistochemistry or molecular testing cannot reliably distinguish between these 2 malignancies. Currently, distinction between the 2 still remains a clinical judgment based largely upon clinical history, impression, and anatomic location.

In conclusion, our case is a very rare presentation of pancreatic cancer with either bile duct metastasis at the bifurcation or with an accompanying primary cholangiocarcinoma. Neither situation has been described before in the literature. This highlights the need for further studies exploring the carcinogenesis in these patients and for further advancements in histopathological analysis, such as those of biliary tree brushings, to clearly distinguish between metastatic pancreatic adenocarcinoma and primary cholangiocarcinoma.

DISCLOSURES

Author contributions: All authors contributed equally to reviewing the literature, acquiring the figures, and writing the manuscript. H. Omar is the article guarantor.

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Informed consent was obtained for this case report.

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