A Rare Cause of Biliary Obstruction

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Abstract

Objective: To discuss a rare cause of obstructive jaundice.

Methods: A case report is presented with emphasis on diagnosis and management. Six month follow up is also presented.

Results: A 42 year old African-American male presented with one week of painless jaundice. He underwent imaging via CT abdomen pancreatic protocol and MRCP demonstrating a massively dilated common bile duct of 12 cm and diffuse intrahepatic ductal dilation. He subsequently developed pruritus, RUQ abdominal pain, and cholangitis thus necessitating an endoscopic retrograde cholangiopancreatography (ERCP). ERCP was performed but not useful in delineating the biliary tree anatomy or relieving biliary obstruction. Later percutaneous transhepatic cholangiography (PTC) was performed to provide drainage and was also not useful in delineating his biliary tree anatomy. Shortly thereafter he underwent laparotomy, choledochal cyst and bile duct resection, Roux en Y pancreaticojejunostomy and hepaticojejunostomy. A review of the pathologic specimen indicates the presence of intrapapillary neoplasm of the bile duct (IPNB) which is a rare variant of a bile duct tumor. The specimen was positive for MUC1 and CEA which indicates a high possibility of recurrence.

Conclusions: Invasive carcinoma has been found to be present in 70-80% of cases of resected IPNB. However, survival has been shown to be better in patients with IPNB compared to those with conventional bile duct tumors. Given the difficulty of preoperative diagnosis of these lesions and their high predisposition for invasion all IPNB should be surgically resected.

Keywords: Bile duct Tumor; Biliary Obstruction; Intrapapillary Bile duct Neoplasm

Introduction

IPNB is a rare variant of bile duct carcinoma and represents 10% of all such cases. This type of bile duct tumor is exceedingly rare and similar to papillary intraductal neoplasm of the pancreas [1]. Typical bile duct carcinoma is characterized initially by flat dysplasia while IPNB is characterized by an epiphic proliferative biliary epithelium on fibro vascular stalks [1]. IPNB is also associated with mucin hyper secretion and cystic dilation of the bile ducts [1-3]. The primary site within the biliary tree does not affect prognosis or course of the disease [1-3]. The most common primary sites within the biliary tree are at the hilum and the left intrahepatic ductal system [1-3]. The presence of MUC1 and CEA positivity in the immunostain indicate a high likelihood of recurrence and poor prognosis [4-6]. Immunohistochemical examination of resection IPNB with invasion demonstrates overexpression of MUC 1 and CEA [4-6]. The degree of invasion is also critical to survival [1]. A significant survival advantage is present when the degree of invasion ranges from 0 mm to < 5 mm [1]. Overall, invasive carcinoma has been found to be present in 70-80% of cases of resected IPNB [1]. However, survival has been shown to be better in patients with IPNB compared to those with conventional bile duct tumors [3]. Given the difficulty of preoperative diagnosis of these lesions and their high predisposition for invasion all IPNB should be surgically resected [7]. The location and extent of IPNB is critical in determining the type of resection [7]. For example, distal IPNB can be resected via a distal bile duct resection and pancreatectoduodenectomy [7]. Tumor in the left and right perihilar bile ducts or in the liver parenchyma necessitates an extended right or left-hemihepatectomy and bile duct resection [7].

Case Report

A 42 year old African-American male with no past medical history presented with one week of painless jaundice. He denied a history of liver disease, alcohol use, smoking, viral hepatitis, blood transfusions, tattoos and needle sharing. Laboratory testing indicated a total bilirubin of 28 and a direct bilirubin of 16 (Table 1). Hepatology labs were negative for viral hepatitis, autoimmune hepatitis, hemochromatosis, Wilson’s disease, alpha 1 antitrypsin deficiency and primary biliary cirrhosis. He underwent radiologic imaging via CT abdomen pancreatic protocol and MRCP demonstrating a massively dilated common bile duct of 12 cm with a mural nodule and diffuse intrahepatic ductal dilation (Figure 1a/b and 2). There was no evidence of a pancreatic mass or cirrhosis. He subsequently developed pruritus, RUQ abdominal pain, and cholangitis thus necessitating an endoscopic retrograde cholangiopancreatography (ERCP) to attempt biliary drainage. ERCP was performed but not useful in delineating the
biliary tree anatomy or relieving the biliary obstruction. Later percutaneous transhepatic cholangiography (PTC) was performed to provide drainage but was also not useful in delineating his biliary tree anatomy. After placement of a PTC drain his bilirubin decreased to 5 and his pruritus improved. Shortly thereafter he underwent laparotomy, choledochal cyst and bile duct resection, Roux en Y pancreaticojejunostomy and hepaticojejunostomy. An endoscopic ultrasound performed 6 months after his surgery demonstrates a normal pancreas and no evidence of recurrence.

Discussion

IPNB is a very rare variant of a bile duct tumor and difficult to manage [1-3]. All cases necessitate surgical resection [7]. In our patient, the pathology specimen contained a small area of intrapapillary neoplasm of the bile duct (IPNB) (Figure 3 and 4). The specimen contained an area of papillary growth with high grade dysplasia but did not demonstrate any definite evidence of an invasive carcinoma. An immunostain for MUC1 was strongly positive and stains with polyclonal and monoclonal antibodies to CEA were focally positive in this area. Thus, despite the fact that the pathology specimen did not show any evidence of an invasive carcinoma expression of MUC1 and CEA portends a poor prognosis and high risk of recurrence [4-6]. Currently, there are not established surveillance guidelines for a high risk IPNB after resection.

In our patient, on the gross specimen this large type IV choledochal cyst invaded the pancreas medially necessitating some pancreatic dissection. The distal intra-ampullary common bile duct was occluded and the gallbladder was mostly intrahepatic at the fundus. The mural nodule which was later found to be an IPNB was located near the hilum. In our patient, it’s not entirely clear if the tumor arose in the pancreas and then extended into the bile duct or vice versa. Complete resection of the tumor in the pancreas is essential to reduce the chance of a future recurrence [7].

Intrapapillary mucinous neoplasm of the bile duct is difficult to diagnose and often diagnosed when cancerous. ERCP and PTC were not very useful in making a diagnosis given the enormous size of this tumor. ERCP and PTC can be useful if the entire tumor can be filled with contrast to delineate it’s anatomy. However, both ERCP and PTC can be useful as a drainage procedure to relieve the biliary obstruction. In this case, both CT pancreatic protocol and MRCP were useful in delineating the anatomy of the type IV choledochal cyst but did not outline the intrapancreatic portion clearly. In retrospect, an endoscopic ultrasound with FNA may have aided in better delineation of the tumor in the pancreas.

In conclusion, IPNB remains a challenging diagnosis often necessitating multiple imaging modalities for complete characterization. IPNB always requires surgical resection and close follow up [7]. Endoscopic ultrasound may aid in the diagnosis of these tumors especially given that IPNB may also arise in the pancreas. Surveillance guidelines are currently lacking and will be critical in future management.

<table>
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<tr>
<th>Laboratory Values</th>
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<tbody>
<tr>
<td>WBC count</td>
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<td></td>
<td>4-11 K/uL</td>
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<td>Hemoglobin</td>
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<td>13.2-17.3 g/dL</td>
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<td>Hematocrit</td>
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<td>36.6-51.9%</td>
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Table 1: Laboratory values for our patient.

Figure 1a: CT pancreatic protocol axial images of a large type IV choledochal cyst (arrow) with mural nodule extending into the liver.

Figure 1b: CT pancreatic protocol coronal image of a severely dilated common bile duct (arrow) and intrahepatic ducts.

Figure 2: MRCP showing a massively dilated common bile duct (arrow) and intrahepatic ducts.

Figure 3: H and E stain showing papillary projections and an exophytic growth (arrow).

Figure 4: Magnified view of IPNB highlighting abnormal cells (nuclei) in the midst of papillary projections (arrow).

References
