Intraductal Papillary Neoplasm of the Bile Duct Diagnosed Post Liver Transplant in Patient with Recurrent Hepatocellular Carcinoma

Leanna Rucker, MD[1]; Ahmad Al-Dwairy, MD[2]; Dalal Alhaqqan, MD[1]; Amol S. Rangnekar, MD[1]

[1]MedStar Georgetown University Hospital, [2]MedStar Health/Georgetown-Washington Hospital Center

Introduction

- Intraductal papillary neoplasm of the bile duct (IPNB) is a rare tumor originating from the biliary epithelium
- Shares features with intraductal papillary mucinous neoplasm (IPMN) of the pancreas
- May involve extra- and intrahepatic bile ducts and defined histologically by mucin-secreting papillary and/or cystic proliferation[1]
- Recognized as precancerous lesion with more favorable postoperative prognosis compared to infiltrating biliary tumors (i.e., cholangiocarcinoma)[2]
- Diagnosis can be challenging and requires direct cholangiography or cholangioscopy
- Only 11% of IPNB cases arise from United States, with majority described in countries such as Japan, China, Korea, and Taiwan[3]

Our case describes a patient diagnosed with IPNB during his post liver transplant course, with evidence of local metastasis. The case further highlights the challenge in diagnosing IPNB and its malignant potential.

Description of Case

A 67-year-old Middle Eastern male with recurrent hepatocellular carcinoma (HCC) and decompensated hepatitis C cirrhosis was referred to our center for liver transplant evaluation. His prior HCC treatments included hepatic resection as well as transarterial chemoembolization and radioembolization (TACE, TARE) which were complicated by the development of biliary strictures requiring percutaneous drainage. This was further complicated by multiple episodes of cholangitis with drug-resistant organisms. At that time, endoscopic retrograde cholangiopancreatography (ERCP) was performed revealing biliary strictures with pathology negative for dysplasia or malignancy.

The patient ultimately underwent orthotopic liver transplant with Roux-en-Y biliary reconstruction; MELD score of 33 at time of transplant. Intraoperative findings were notable for chronic inflammation and intra-abdominal abscesses.

Case continued...

Notably, surgical pathology from his native liver and bile ducts revealed IPNB with mucin production, not detected on prior specimens from cholangiography. Three months later, he underwent Whipple procedure for locally advanced IPNB. Chemotherapy with gemcitabine and cisplatin was initiated.

Histopathology Findings

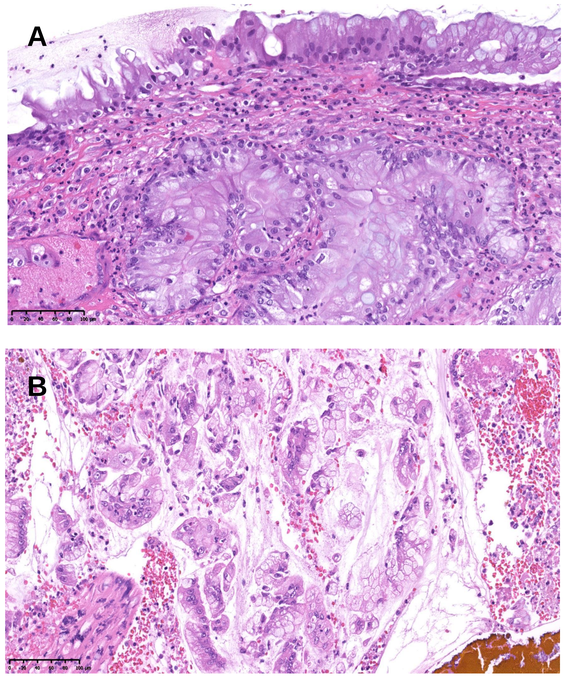

Figure 1
Hematoxylin and eosin tissue staining obtained from patient's native bile ducts (A) and hilar margin of explanted liver (B). Magnification x20.
A) Bile duct collection demonstrates multiple fragments of intraductal papillary neoplasm of bile duct with mucin production and high-grade dysplasia.
B) Microscopic foci of moderately differentiated invasive mucinous adenocarcinoma in hilar soft tissue.

Preoperative Imaging

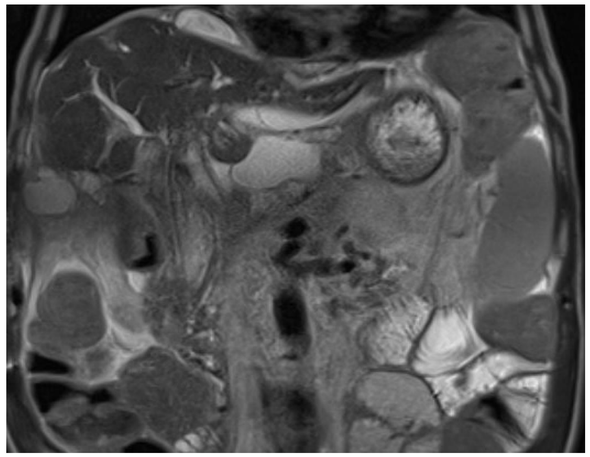

Figure 2
Magnetic resonance cholangiopancreatography (MRCP) imaging with liver/pancreas protocol. Coronal (top) and axial (bottom) T2 weighted images showing moderate to severe intrahepatic biliary ductal dilation. There is also a short segment narrowing of the proximal extrahepatic bile duct with abnormal wall thickening and enhancement, raising concern for malignant stricture.

Discussion

Though rare, IPNB has been increasingly reported in medical literature and in 2010 was recognized by the World Health Organization as a precancerous lesion of biliary carcinoma.[2] Excessive mucin production and friability of the tumor are thought to cause intermittent biliary obstruction and are responsible for its symptoms, including jaundice, abdominal pain, and cholangitis. Epidemiologically, IPNB has been mainly described in patients of Far East Asian descent, where hepatolithiasis and clonorchiasis are endemic.[3] Although a specific etiology and pathogenesis are unclear, these two scenarios appear to be risk factors for the development of IPNB.

In the case of our patient, locally invasive IPNB was only found post liver transplant requiring the patient to undergo an additional major surgery with a Whipple procedure. It is questionable whether his recurrent HCC or prior TACE and TARE procedures were linked to such tumor development. This further highlights the need to have a high index of suspicion and awareness for the elusive presentation of IPNB. It should also be considered as a potential diagnosis in patients presenting with recurrent cholangitis.

Conclusion

This case demonstrates the challenge in diagnosing IPNB given its rarity and elusive presentation. It also highlights the potential for malignant transformation and metastasis. There remains more to be discovered about its pathogenesis and clinical course.

References

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