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CASE REPORT | BILIARY

Double Trouble: A Rare Case of Concurrent Biliary Adenofibroma and Hepatobiliary Mucinous Cystic Neoplasm

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ABSTRACT

Biliary adenofibroma and hepatobiliary mucinous cystic neoplasm are exceedingly rare tumors. To our knowledge, no case of co-existent biliary adenofibroma and mucinous cystic neoplasm has been previously reported. We present a patient who was diagnosed with both tumors concurrently. Both can present with abdominal pain, although some are diagnosed incidentally in asymptomatic patients. Imaging and tumor markers can be suggestive, but histologic examination is needed for definitive diagnosis. Both have a propensity for malignant transformation, thus complete surgical resection is the treatment of choice. Although rare, awareness of these tumors leads to earlier diagnosis and treatment.

INTRODUCTION

Biliary adenofibroma (BAF) and hepatobiliary mucinous cystic neoplasm (MCN) are both very rare tumors of the hepatobiliary tract. To date, fewer than 20 cases of BAF have been reported in the medical literature.[1] Hepatobiliary MCN is also very rare, with a reported incidence rate of 1 in 20,000–100,000.[2] Even more rare is finding both tumors in a single patient; to our knowledge, no such case has been published.

CASE REPORT

A 26-year-old previously healthy woman presented with jaundice and pruritus for 1 month. She was initially seen at an outside hospital and was treated with ursodiol with no improvement prior to her transfer. She had no history of oral contraceptive, alcohol, or illicit drug use. The rest of the review of systems was negative. Social history was unremarkable. On physical exam, she had scleral icterus and generalized jaundice. Initial labs were significant for serum total bilirubin 6.6 mg/dL and alkaline phosphatase 277 U/L. Aspartate aminotransferase and alanine aminotransferase were both normal. Hepatitis serologies were negative. T2-Weighted magnetic resonance cholangiopancreatography (MRCP) showed severe intrahepatic biliary ductal dilatation with multiple cystic collections in the left lobe of the liver (Figure 1). No definite mass lesion was noted on T1 sequences, and a presumed diagnosis of Caroli disease with predominant left lobe involvement was made.

On review of imaging at the multidisciplinary meeting, there was concern for common hepatic duct stones. Endoscopic retrograde cholangiography (ERCP) revealed a large filling defect with smooth borders at the bifurcation with marked upstream dilatation of the left intrahepatic ducts and mild dilation of right intrahepatic ducts (Figure 2). The filling defect did not move despite multiple balloon sweeps. Subsequent cholangioscopy revealed a large, soft tissue mass with a long stalk extending into the left main duct (Video 1). Bilateral plastic biliary stents

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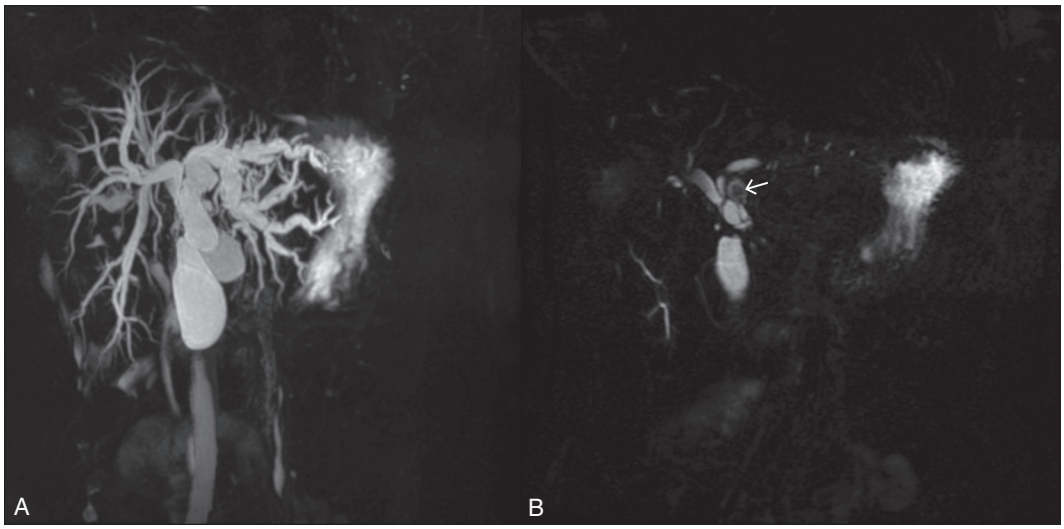

Figure 1. (A) T2-Weighted magnetic resonance cholangiopancreatography (MRCP) showing severe diffuse intrahepatic biliary ductal dilatation with multiple cyst-like dilated areas of the intrahepatic biliary tree. (B) MRCP image concerning for left common hepatic duct stones (arrow).

were placed with improvement in bilirubin. Persistent intrahepatic cystic collections were noted on follow-up MRCP despite ductal decompression after biliary stenting. The patient subsequently underwent left hepatic lobectomy with resection of bile duct mass and hepatico-jejunostomy (Figure 3). Histologic examination revealed a 2.6-cm benign adenofibroma and a 5.5-cm benign multiloculated hepatobiliary mucinous cystic neoplasm with subepithelial ovarian stroma within the left hepatic duct system (Figure 4). No invasive tumor was seen, and resection margins were free of tumor. The patient's postoperative course was unremarkable, and she was eventually discharged home and reported no issues on her 3-month follow-up visit.

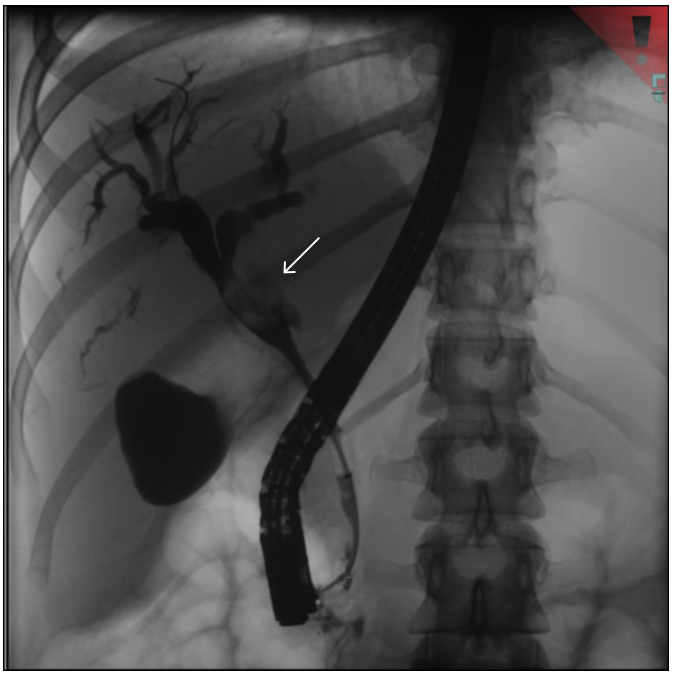

Figure 2. Endoscopic retrograde cholangiography showing soft tissue mass (arrow) at the bile duct bifurcation with upstream biliary dilatation.

Video 1. Cholangioscopy showing soft tissue mass with normal overlying mucosa at the proximal common bile duct. Watch the video: http://s3.gi.org/media/links/ajg/Esteban_Video.mp4.

DISCUSSION

To our knowledge, no other cases with co-existent BAF and hepatobiliary MCN have been reported in literature. BAF was first described in 1993.[3] Since then, fewer than 20 cases have been reported, including 6 patients in one case series.[1] BAF is characterized by cystic and tubular biliary epithelial components surrounded by a bland fibroblastic spindled stroma.[3] Earlier case reports have considered these tumors to be completely benign. However, recent reports suggest that these tumors are more likely slowly progressive neoplasms.[1] There are at least 4 cases of BAF that contained areas considered by investigators to represent frank malignant histology, and pulmonary metastases 2 years after resection of BAF have been reported.[4-8]

Earlier literature about hepatobiliary MCNs included essentially any case with epithelial-lined cystic lesion occurring in the liver and biliary tract. In 2010, the World Health Organization established the presence of ovarian stroma as a requirement for the diagnosis of MCNs.[9] The origin of MCNs is unclear, although the role of hormones in their pathogenesis has been proposed by some authors.[10] After MCNs were defined by the presence of ovarian stroma, the rate of carcinomatous transformation was reported at 6%. Interestingly, this was lower than previously reported in the literature based on cases without the ovarian stroma requirement.[9]

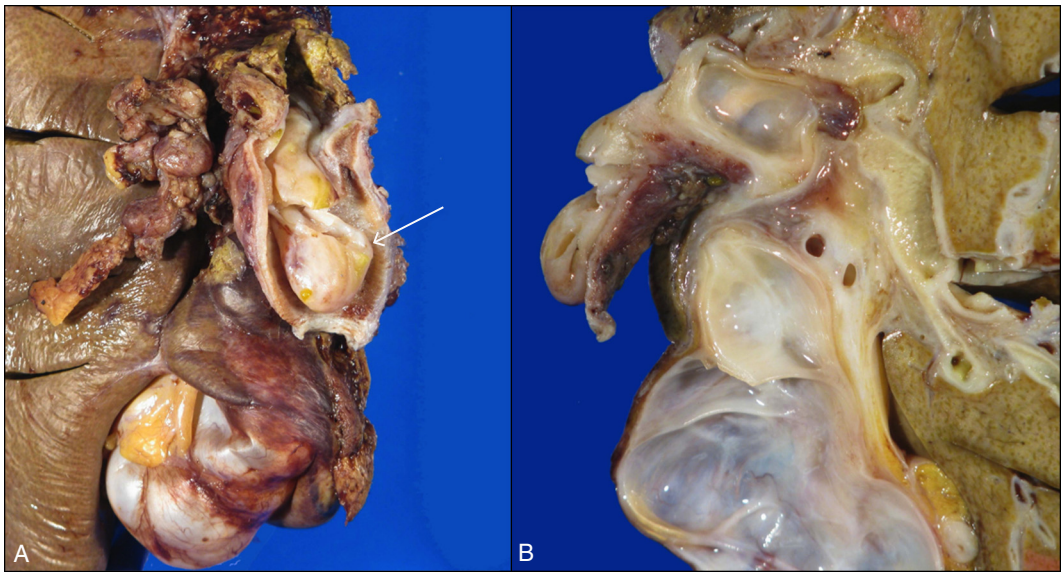

Figure 3. Gross specimen showing (A) a polyp within the bile duct (arrow) and (B) a multiloculated smooth-walled cyst.

Both BAF and MCNs usually present with abdominal pain, although some lesions have been found incidentally in patients who were asymptomatic.[1,4,10] Measurement of CA 19-9, CEA, and AFP levels should be performed.[10,11] Imaging such as ultrasound, computed tomography, magnetic resonance imaging (MRI), or MRCP could be suggestive. MRI findings suggestive of BAF include lesions that are T1 hypointense, are heterogeneously T2 hyperintense, and display varying peripheral enhancement on post-contrast sequences with washout.[12] For MCNs, T1-weighted MRI can show a fluid-containing multilocular mass with homogeneous low signal intensity, and wall enhancement can be seen after gadolinium administration. On T2-weighted images, the fluid collection within the tumor demonstrate variable, homogeneous high signal intensity, while the wall of the mass is represented by a rim with low signal intensity.[10] Differential diagnoses for both conditions include simple liver cysts, parasitic cysts, hematomas or post-traumatic cysts, liver abscesses, Caroli disease, and von Meyenburg complex. Of note, our patient was initially diagnosed with Caroli disease based on imaging. One important point that could be learned from our case is not to solely rely on imaging. Cholangiography and cholangioscopy can be helpful as was demonstrated in our case. Another important point that could be derived from our case is that although stones are the most common cause of biliary tract-filling defects, cysts should also be considered in the differential. Histologic examination will ultimately distinguish BAF and MCN from other choledochal cystic diseases.

Both BAF and MCNs have the possibility of malignant transformation.[1,4-9] Given this possibility, complete surgical resection with negative margins is the treatment of choice for both conditions.[1,2,4,9-11,13] The importance of complete resection is emphasized by finding that, in 2 patients who had recurrence of BAF, both had positive surgical margin at resection.[1] Close clinical follow-up and observation is suggested for all patients

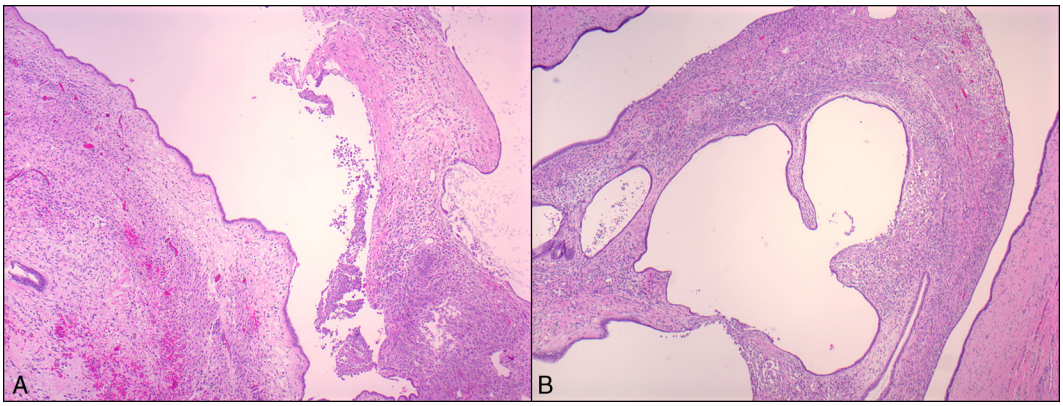

Figure 4. Histologic image showing (A) left hepatic duct adenofibroma and (B) hepatobiliary mucinous cystic neoplasm with underlying ovarian stroma.

after surgery.[4,12] Some authors even suggest imaging follow-up for potential recurrence.[12] Although rare, clinicians should be aware of these disease conditions. A high index of suspicion results in earlier diagnosis and treatment and leads to better outcomes for patients.

DISCLOSURES

Author contributions: M. Esteban and J. Amin wrote the manuscript and reviewed the literature. S. Jakate provided the pathology images. M. Hertl provided the gross surgical images. A. Singh edited the manuscript. M. Esteban is the article guarantor.

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