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CASE REPORT | BILIARY

Granular Cell Tumor of the Common Hepatic Duct as an Unusual Cause of Jaundice in a Hepatitis C Patient

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Abstract

A 33-year-old woman with a history of intravenous cocaine abuse presented with fatigue, nausea, and jaundice. Serologic testing revealed a positive hepatitis C virus (HCV) antibody and HCV RNA. Ultrasound and magnetic resonance imaging/magnetic resonance cholangiopancreatography showed a partially obstructing lesion in the common hepatic duct, which was confirmed by endoscopic retrograde cholangiopancreatography. Surgical excision revealed a granular cell tumor of the common hepatic duct, with immunohistochemical staining of tumor cells positive for S-100.

Introduction

Granular cell tumor (GCT) is a rare, benign neoplasm of Schwann cell origin. Most GCTs have been described in the dermis and oral area, with less than 1% of cases occurring in the biliary tract.[1] Tumors involving the bile duct are difficult to distinguish radiographically from more common etiologies of biliary stricturing, and a tissue diagnosis is difficult to obtain endoscopically. Diagnosis and treatment is most often made via surgical resection.

Case Report

A 33-year-old white woman presented with new onset of jaundice, fatigue, and nausea for 7 days. Her history was significant for intravenous cocaine abuse and frequent alcohol binging. Physical examination was significant for jaundice with no stigmata of chronic liver disease. Significant laboratory results included a total bilirubin of 9.2 mg/dL, aspartate aminotransferase of 1677 IU/L, alanine aminotransferase of 2071 IU/L, and an alkaline phosphatase of 171 IU/L. She had a positive hepatitis C virus (HCV) antibody, with a HCV quantitative viral load of 12 x 10^6 IU/mL (genotype 1A). Her acute jaundice and liver injury was initially presumed to be due to acute HCV infection versus an ischemic hepatitis from cocaine use.

An abdominal ultrasound revealed a normal liver with patent hepatic vessels. A 1.9 x 1-cm isoechoic intraluminal lesion in the mid-common bile duct (CBD) was causing focal CBD expansion with no dilation of the proximal CBD. This finding was suspicious for a neoplasm. Magnetic resonance imaging/magnetic resonance cholangiopancreatography (MRI/MRCP) verified a partially obstructing polypoid lesion (2 x 0.6 cm) in the common hepatic duct (CHD; Figure 1). An endoscopic retrograde cholangiopancreatography (ERCP) with SpyGlass® cholangioscopy (Boston Scientific, Natick, MA) identified a 1.5-cm friable, irregular polypoid lesion in the CHD, which was sampled with CHD brushings and SpyBite™ forceps biopsy (Boston Scientific, Natick, MA; Figure 2). Brushings were negative for malignancy, and the biopsy was insufficient for evaluation. Endoscopic stent placement was not

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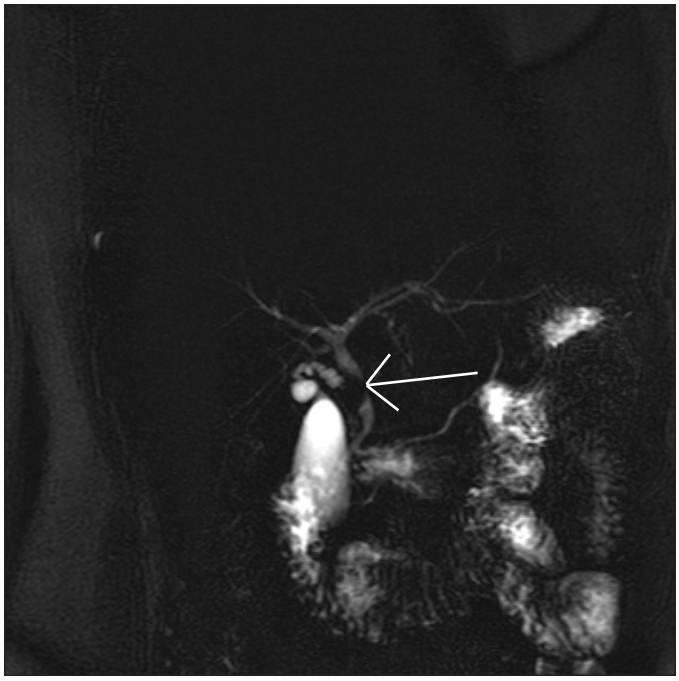

Figure 1. MRCP showing partially obstructing polypoid lesion in proximal extrahepatic bile duct (arrow).

performed. A liver biopsy revealed a biliary obstructive pattern of canalicular and hepatocellular cholestasis with bile ductular proliferation and acute pericholangitis. Trichrome stain revealed stage 1 portal fibrosis. Notably, during this radiographic and endoscopic evaluation, the patient's liver enzymes normalized.

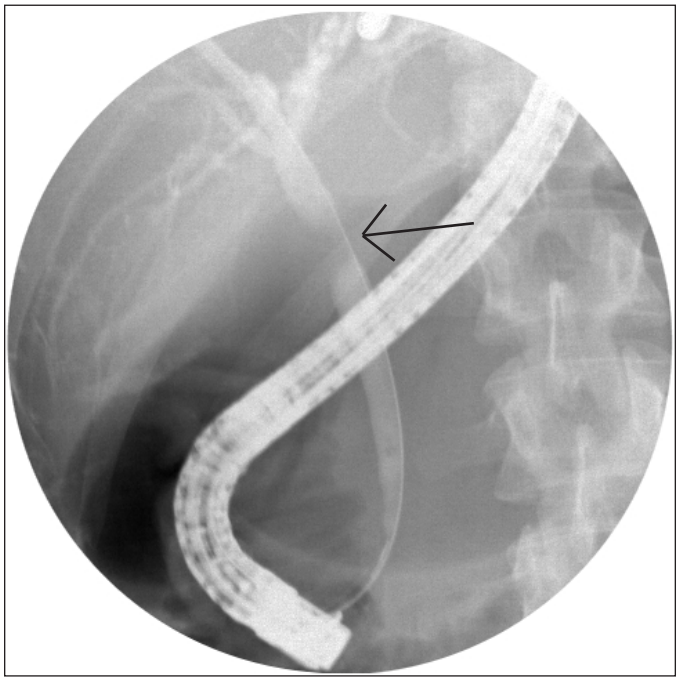

Figure 2. ERCP revealing a 15-mm irregular fixed filling defect in common hepatic duct just proximal to the cystic duct (arrow).

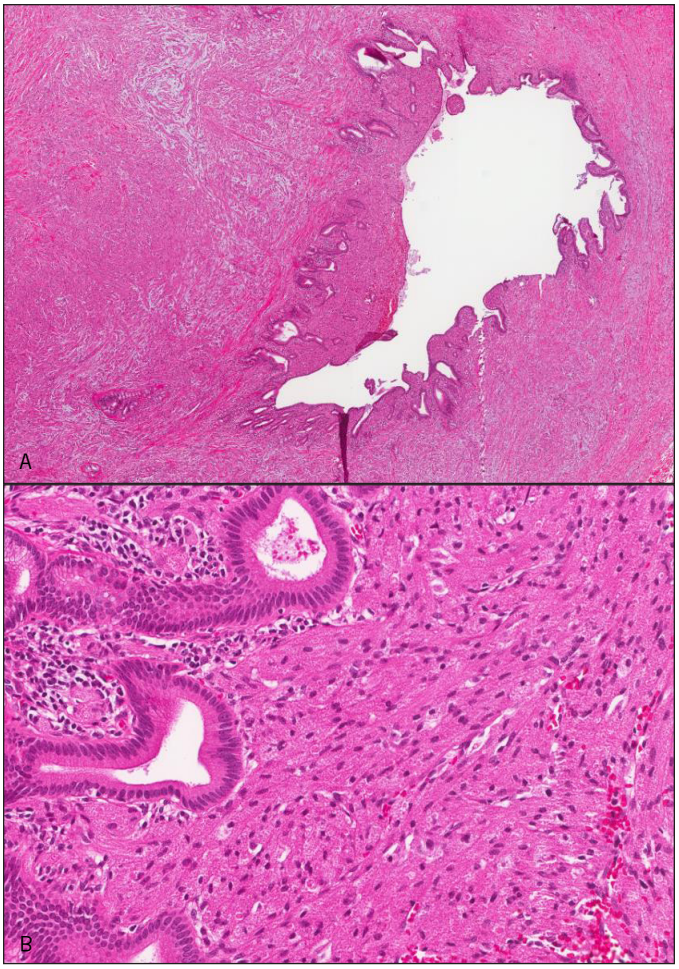

Figure 3. Photomicrographs showing a proliferation of tumor cells within the bile duct mucosa and submucosa. The cells have eosinophilic and granular cytoplasm, with no significant cytologic atypia. No mitosis is identified. H&E stain at (A) x20 magnification and (B) x200 magnification.

Complete surgical excision with cholecystectomy and hepaticojejunostomy was performed followed by histopathological examination, confirming the diagnosis of GCT of the CBD/CHD (Figure 3). Immunohistochemical staining of tumor cells was S-100 positive and neurofilament, smooth muscle actin and c-kit negative (Figure 4). There was no significant cytologic atypia or mitosis identified, and the lesion was felt to be benign. The postoperative course was uneventful. The patient was asymptomatic at 1-month follow-up and was to receive outpatient management of her HCV infection.

Discussion

Originally described by Abrikossoff in 1926, GCTs are mesenchymal tumors of Schwannian origin.[1] The Schwannian origin is supported by the immunohistochemical findings of S-100 positivity and presence of neuron-specific enolase in the cysoplasmic granules. Most cases arise in the dermis or subcutaneous tissue of the chest and arms, with only 5–9% of cases reported involving the GI tract.[2] First described by

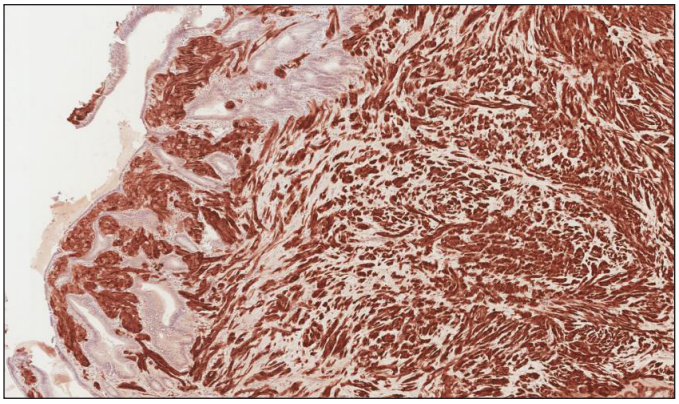

Figure 4. Photomicrograph showing tumor cells that are diffusely and strongly positive for S-100 on immunohistochemical stain (x100 magnification).

Coggins in 1952, GCT of the bile duct is extremely rare, with fewer than 80 cases reported in the literature.[1] Malignant GCT is quite unusual, and no cases of malignant biliary GCT have been reported.[2] These benign lesions are more common in young women, especially in African Americans; clinicians should have a high index of suspicion when evaluating a biliary mass in this population.[2]

Within the biliary tree, the most common location is the common bile duct (49%) followed by the cystic duct (19%), common hepatic duct (9%), intrahepatic duct (1%), and gallbladder (1%).[2] Imaging may not help distinguish GCT from other lesions causing biliary obstruction, such as a choledochal cyst, cholangiocarcinoma, or a benign stricture. As in our case, obtaining tissue endoscopically can be difficult, and surgical excision is both diagnostic and curative.[2-5] GCTs involving the common hepatic duct or common bile duct require wide local excision and bilio-enteric anastomosis. Temporary decompression can be achieved with percutaneous or endoscopic stents.[6] If left untreated, these lesions can cause luminal obstruction leading to secondary biliary cirrhosis and hepatic failure necessitating liver transplant.[2] A thorough excision is required to prevent local recurrences secondary to incomplete removal.[7] Given the rarity of this tumor, there are no consensus guidelines for surveillance imaging after resection.

This case emphasizes the difficulty in diagnosing GCT of the biliary tree with radiographic and endoscopic techniques. Furthermore, the patient's acute hepatitis, possibly due to a HCV infection or ischemic injury from cocaine, further confounded the diagnosis. To our knowledge, there are no other cases of concomitant hepatitis C and granular cell tumors of the biliary tract. Timely investigation of any bile duct lesion is important to prevent complications of biliary obstruction and exclude malignancy. Long-term follow-up after removal is essential to survey for any recurrence and in order to prevent the complication of secondary biliary cirrhosis.

Disclosures

Author contributions: All editors wrote and edited the manuscript. CL Smith is the article guarantor.

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Informed consent was not obtained for this case report, as the patient relocated to another state before we had an opportunity to consent her. We were unable to locate her at the address or phone number listed in her medical record.

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