Granular cell tumor of the common hepatic duct as an unusual cause of jaundice in a hepatitis C patient: looking beyond the tip of the iceberg!

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Introduction

Granular cell tumors (GCTs) are rare, benign mesenchymal tumors of Schwann cell origin. Since they were originally described by Abrikosoff in 1926, most GCTs have been described in the dermis, oral area, or subcutaneous tissue of the chest and arms. Only 5%-9% of reported cases have involved the gastrointestinal tract and fewer than 1% of cases have occurred in the biliary tract. 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 32 year old Caucasian woman presented with new onset of jaundice, and fatigue and nausea for seven days. Her social history was significant for intravenous cocaine abuse for fifteen years with recent use three weeks prior to onset of the symptoms, as well as frequent alcohol binging.

Pertinent physical exam findings: Bilateral icteric sclera with no stigmata of chronic liver disease.

Laboratory data: Total bilirubin 9.2 mg/dL, aspartate aminotransferase of 1677 IU/L, alanine aminotransferase of 2071 IU/L, and an alkaline phosphatase of 171 IU/L, positive hepatitis C virus (HCV) antibody, with a HCV quantitative viral load of 12x10^4 IU/mL (genotype 1A).

She was presumed to have a HCV infection versus an ischemic hepatitis from cocaine.

Imaging: An abdominal ultrasound revealed a normal liver with patent hepatic vessels. In the mid common bile duct (CBD) there was a 1.9 x 1 cm isoechoic intrahepatic lesion causing focal CBD expansion with no dilation of the proximal CBD. This finding was suspicious for a neoplasm.

Magnetic Resonance Cholangiopancreatoscopy (MRCP) verified a partially obstructing polypoid lesion (2 x 0.6 cm) in the common hepatic duct (CHD) (Figure 1).

Endoscopy: An endoscopic retrograde cholangiopancreatography (ERCP) with SpyGlass cholangioscopy identified a 1.5 cm friable, irregular polypoid lesion in the CHD (Figure 2A, B), which was sampled with CBD brushes and SpyBite forceps biopsy. Brushings were negative for malignancy, and the biopsy was insufficient for evaluation.

A liver biopsy revealed a biliary obstructive pattern of canaliculal 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.

Surgical Evaluation: Complete surgical excision with cholecystectomy and hepatectomy was performed followed by histopathological examination, confirming the diagnosis of GCT of the CBD (Figure 3). Immunohistochemical staining of tumor cells was S-100 positive (Figure 4) and neurofilament, smooth muscle actin and c-kit negative.

Postoperative course: The patient had no postoperative complications. She was asymptomatic at one-month follow-up and was to receive outpatient management of her HCV infection.

Discussion

- GCTs of the bile ducts are extremely rare, with fewer than eighty cases reported in the literature.
- These benign lesions are more common in young women, especially in African Americans; a high index of suspicion is needed when evaluating a biliary mass in this population.
- Timely investigation of any bile duct lesion is important to prevent complications of biliary obstruction and exclude malignancy.
- Obtaining tissue endoscopically can be difficult, and surgical excision is both diagnostic and curative.
- Thorough excision is required to prevent local recurrences secondary to incomplete removal.
- If left untreated, GCT can cause luminal obstruction leading to secondary biliary cirrhosis and hepatic failure necessitating liver transplant.
- Long-term follow-up after removal is essential to monitor for any recurrence, in order to prevent the complication of secondary biliary cirrhosis.
- 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 hepatitis C infection or an ischemic injury from cocaine, further confounded the diagnosis.

References


Photomicrographs credit: Dr. John Farber, MD; Dr. Wei Jiang, MD
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NONE OF THE AUTHORS HAVE ANY RELEVANT DISCLOSURES TO REPORT