Diagnostic Snapshot

What Is the Cause of This Biliary Stricture?

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Abstract

This article describes the presentation and workup of a 63-year-old male with a bile duct stricture, followed by a discussion on management.

HISTORY

Mr. F is a 63-year-old male with no significant past medical or surgical history. He is physically active, drinks socially, and does not smoke. He presented to his primary care provider with a 2-month history of upper abdominal pain associated with jaundice, pruritis, dark urine, and light stools. He also reported fatigue and unintentional weight loss. Blood work showed elevated liver function tests. A CT scan of the abdomen and pelvis revealed bilateral dilatation of the intrahepatic bile ducts with no evidence of liver or pancreatic lesions. An endoscopic retrograde cholangiopancreatography (ERCP) with endoscopic ultrasound and fine needle aspiration (FNA) was performed next to better evaluate his biliary tract. There was a tight stricture of the proximal common bile duct (CBD) up to the bifurcation of the hepatic ducts (Figure 1). The bile duct was swept and a plastic stent was placed in the CBD up into the right hepatic duct. Stenting relieved Mr. F's symptoms, but cytology from brush biopsy and FNA of an enlarged regional lymph node showed only atypical cells. Mr. F underwent a prereferral open cholecystectomy, with pathology revealing evidence of gallstones and atypical cells at the cystic margin.

CHIEF COMPLAINT

Approximately a month after surgical resection, Mr. F developed recurring obstructive jaundice with pruritis, clay-colored stools, and tea-colored urine, so he came to the surgical oncology clinic. He also complained of worsening fatigue and a 30-pound weight loss but denied experiencing fever, chills, abdominal pain or bloating, nausea, vomiting, or any other symptoms.

PHYSICAL EXAM AND DIAGNOSTIC STUDIES

On physical examination, Mr. F appears to be thin and frail with muscle atrophy. He is afebrile and his vital signs are stable. His sclera are icteric and his skin is jaundiced. The abdominal exam is benign, with no tenderness to palpation or hepatosplenomegaly.

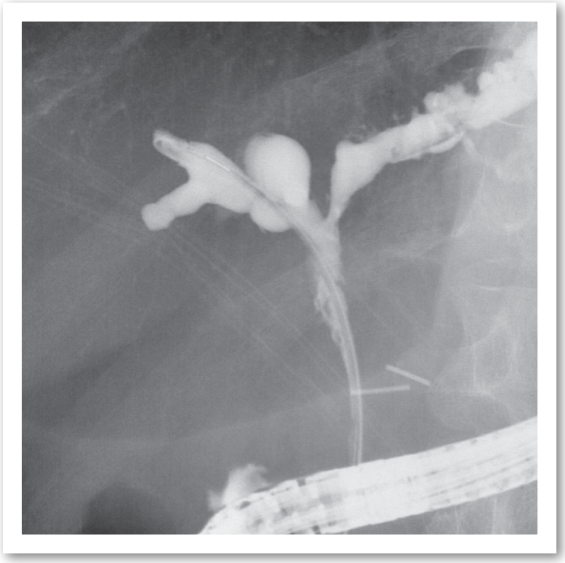

Figure 1

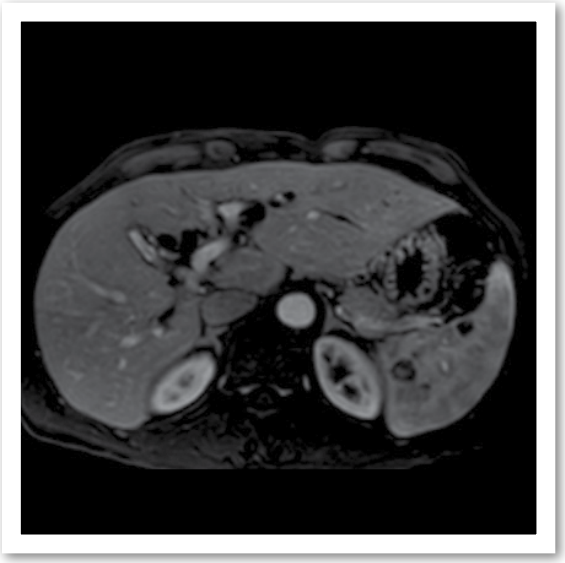

Figure 2

Mr. F underwent laboratory testing. His results were significant for a total bilirubin of 7.4 mg/dL (direct bilirubin of 5.4 mg/dL), aspartate aminotransferase of 93 U/L, alanine transaminase of 110 U/L, alkaline phosphatase of 891 U/L, IgG4 of 9 mg/dL, and CA 19-9 of 261.1 U/mL. The remainder of his labs were within normal limits, including a white blood cell count of 7.5 K/µL.

The abdominal MRI/magnetic resonance cholangiopancreatography for Mr. F showed multiple biliary strictures, including a predominate stricture involving the CBD up to the biliary bifurcation consistent with primary sclerosing cholangitis (PSC) and bilobar intrahepatic ductal dilatation, despite the presence of endobiliary stent (Figure 2).

DIFFERENTIAL DIAGNOSIS

WHAT IS THE CORRECT DIAGNOSIS?

A Autoimmune pancreatitis

B Extrahepatic cholangiocarcinoma

C Choledocholithiasis

D Bile duct injury

E Mirizzi syndrome

SEE BACK FOR ANSWER ▶

WHAT IS THE CORRECT DIAGNOSIS?

A Autoimmune pancreatitis

B Extrahepatic cholangiocarcinoma (correct answer)

C Choledocholithiasis

D Bile duct injury

E Mirizzi syndrome

A **Autoimmune pancreatitis (AIP).** While this is a disease of chronic inflammation of the pancreas, it also affects extrapancreatic organs, including the bile ducts up to 88% of the time (Nishino et al., 2005). Characteristics supporting this diagnosis include elevated IgG4 levels in both the serum and histology, CT findings of pancreatitis, and response to steroid therapy (Majumder et al., 2017).

B **Extrahepatic Cholangiocarcinoma.** Cholangiocarcinoma is a primary tumor of the biliary tract, which can be classified as intrahepatic or extrahepatic (further broken down into distal or hilar, also referred to as a Klatskin tumor). Cholangiocarcinoma is rare, with an incidence of 2.1 cases per 100,000 in the United States (Khan & Dageford, 2019). Many patients with extrahepatic cholangiocarcinoma present with a late stage at diagnosis with symptoms of obstructive jaundice as the bile duct becomes occluded by tumor, and with an elevation of CA 19-9. While many patients do not possess any known risk factors, cholangiocarcinoma has been associated with primary sclerosing cholangitis, parasitic infection, hepatolithiasis, viral hepatitis, and cirrhosis (Esnaola et al., 2016).

C **Choledocholithiasis.** Gallstones can cause biliary strictures by creating inflammation and scarring as they migrate down the biliary tract into the CBD and are still a possibility after cholecystectomy. This case is more likely a picture of tumor growing through the flimsy stent material rather than chronic inflammation.

D **Bile Duct Injury.** Bile duct injury following a cholecystectomy, especially laparoscopic, is a primary cause of benign bile duct strictures. Strictures are caused by misplacement or clips, overuse of cautery, or other missteps causing ischemic injury to the ducts (Dadhwal & Kumar, 2012). However, with Mr. F's elevated CA 19-9, risk factors of PSC, and failure of plastic stent, this is more suggestive of a malignant stricture.

E **Mirizzi Syndrome.** This condition is a result of an impacted gallstone in the infundibulum of the gallbladder or cystic duct causing extrinsic compression on the CBD or common hepatic duct that can lead to fistula formation or destruction of the bile duct wall (Witte, 1984). However, this is usually relieved by cholecystectomy.

MANAGEMENT

Cholangiocarcinoma is a rare disease that is difficult to work up and diagnose. When considering a bile duct stricture, it is wise to consider malignancy until proven otherwise, and to consult a gastrointestinal medical or surgical oncologist. This presentation is not uncommon, as imaging can be nonspecific and ERCP with bile duct brushings often provide inadequate tissue to establish a diagnosis. Delay to diagnosis can have devastating consequences.

The prognosis rates for cholangiocarcinoma are dismal, with a 5-year survival rate of 2% to 16% based on stage (Howlander et al., 2020). However, if disease is detected while still localized (stages I–IIIB), it can be treated surgically with curative intent (Ho & Curley, 2016). Surgical options for nonmetastatic tumors are driven based on location of tumor along the biliary tree and include a Whipple procedure, biliary reconstruction, lymph node dissection, wedge or anatomical hepatectomy, or even liver transplant. Otherwise, treatments are limited to palliative radiation, systemic chemotherapy, or clinical trials.

FOLLOW-UP

Following another ERCP, brushings of the proximal bile duct stricture confirmed cholangiocarcinoma, and multiphasic CT unfortunately revealed a hilar lesion with extension into the second-order bile ducts on the right, unclear extension on the left, with abutment of the common hepatic artery. Mr. F was taken to the operating room for a diagnostic laparoscopy, which did not reveal evidence of metastatic disease (stage IIIB).

Mr. F was started on systemic chemotherapy with gemcitabine and cisplatin with the hopes of downstaging the tumor to possible surgical resection. However, after three cycles of chemotherapy, no treatment response was identified on restaging imaging, and the disease was deemed unresectable. He was placed on palliative chemotherapy, the plastic stent was exchanged for a permanent metal stent, and referral was placed to radiation oncology for definitive chemoradiation. ●

Disclosure

The author has no conflicts of interest to disclose.

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