The Cloaked Intruder: An Unexpected Case of Eosinophilic Cholangitis

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ABSTRACT

Often, when biliary strictures are evaluated, malignancy is high on the differential. However, it is important to consider benign and malignant causes because approximately 10% of biliary strictures are benign. Eosinophilic cholangitis is an extremely rare benign disorder of the biliary tract caused by fibrosis and stricture from eosinophilic infiltration. The etiology of the disease remains unclear, and often, patients present with obstructive jaundice. We present a young man who presented with obstructive jaundice and abdominal pain, with a biliary stricture and peripheral eosinophilia. The final surgical pathology revealed eosinophilic cholangitis.

INTRODUCTION

When evaluating a patient with obstructive jaundice, one must always rule out malignant causes. However, benign conditions, such as eosinophilic cholangitis (EC), can sometimes be the cause of obstructive jaundice. EC is a very rare benign disorder of the biliary tract caused by fibrosis and stricture from eosinophilic infiltration.

CASE REPORT

A previously healthy 23-year-old man presented with worsening abdominal pain over several weeks, vomiting, 17-pound weight loss, and jaundice. He reported having clay-colored stools, dark urine, and pruritus. Before his admission, the patient had been evaluated at multiple local emergency departments for abdominal pain but left against medical advice on multiple occasions. Physical examination showed mild epigastric tenderness with a negative Murphy sign. Laboratory work up upon admission revealed the following: white blood cell count of 10.56 × 10³ cells/L with 22.9% eosinophils, total bilirubin of 18.7 mg/dL, direct bilirubin >10.0 mg/dL, aspartate aminotransferase of 247 U/L, alanine aminotransferase of 606 U/L, and alkaline phosphatase of 588 U/L.

Liver ultrasound showed mild intrahepatic and extrahepatic biliary dilation. Autoimmune antibodies and viral hepatitis serology were negative. Magnetic resonance cholangiopancreatography depicted moderate to severe intrahepatic biliary ductal dilation at the level of the porta hepatis with consideration for acute obstruction vs postinflammatory changes (Figure 1). Endoscopic retrograde cholangiopancreatography demonstrated no identifiable filling defects (Figure 2). Although a 25-mm localized stenosis was appreciated, biliary balloon sweep was negative. A 10 Fr × 9 mm plastic stent was deployed at the biliary stenosis site. Brush biopsy of the common bile duct was negative for a malignant cytopathology. Histopathology of common bile duct biopsy showed the benign ductal mucosa with subepithelial fibrosis and mixed eosinophils and lymphocytic inflammation (Figure 3). The patient was started on high-dose steroids for 3 months. Follow-up complete blood count revealed a white blood cell count of 9.3 × 10³ cells/L with eosinophils downtrending to 4.1%. Aspartate aminotransferase, alanine aminotransferase, total bilirubin, and direct bilirubin were downtrending as well. A repeat endoscopic retrograde cholangiopancreatography with cholangioscopy was performed, which revealed incomplete resolution of the stricture. The patient was referred to hepatobiliary surgery for a Roux-en-Y hepaticojejunostomy.
DISCUSSION

EC is an extremely rare, benign cause of biliary obstruction that can mimic malignancy. The exact origin of EC is unknown, but it has some similarity to hypereosinophilic syndromes. Both disease processes are associated with eosinophilic involvement of the gastrointestinal viscera; however, EC does not require a definitive peripheral eosinophil threshold because it is neither a sensitive nor a specific finding for the condition. Furthermore, peripheral eosinophilia is only reported in half of the cases of EC. Diagnosis is best determined by histopathology. It presents with a characteristic dense transmural eosinophilic infiltration in association with other inflammatory cell infiltration in the bile ducts, as well as with a thickened fibromuscular layer and fibrosis of the subserosal layer. This appearance is very similar to the case we have presented here. The pathophysiology of the disease is ambiguous, and it is thought that eosinophil granulocytes release radicals and tissue-damaging proteins (such as growth factor-b) that induce tissue fibrosis.

Furthermore, similar to our case, radiographic imaging of EC is rather nonspecific and can present another challenge toward achieving a preoperative diagnosis. Typical findings include intrahepatic and extrahepatic biliary dilation. In the setting of bile duct wall thickening involving the cystic duct with peripheral eosinophilia, EC should always be considered.

Rodgers et al labeled EC “a malignant masquerade.” Not only has eosinophilia been seen to occur in malignancy, but the obstruction from strictures also causes subsequent weight loss and malabsorption. Fortunately, the strictures caused by EC are benign and can be proven either with contrast-enhanced ultrasonography, as was used by Matusumoto et al, or with histopathology, as was the case with our patient.

The treatment for EC can vary. Butler et al described their case in which eosinophilia and obstructive jaundice resolved on its own, with no evidence of allergy, parasitic infection, or hypereosinophilic syndromes. Other cases have shown resolution of symptoms completely after treatment with steroids. The only definitive treatment for EC is surgical removal. Roux-en-Y hepaticojejunostomy has been the surgical procedure of choice.
for EC. It has only been documented in the literature 6 times before this case. Furthermore, surgical intervention prevents the recurrence of disease and the medical complications associated with prolonged steroid treatment.

EC can cause fibrosis and stricture of the biliary tract from eosinophilic and lymphocytic infiltration. Although this condition is benign, it can present with certain red flag symptoms such as weight loss and obstructive jaundice that make exclusion of malignancy crucial to the diagnosis. Diagnosis can be difficult, but with more documented cases of this rare condition, we can become adept at excluding malignancy.

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