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Single Case

Successful Endoscopic Management of Late Biliary Cast Syndrome in a Liver Transplant Recipient: A Case Report

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Keywords

Liver transplant · Endoscopic retrograde cholangiopancreatography · Biliary cast syndrome · Sphincterotomy

Abstract

Biliary cast syndrome (BCS) is an unusual complication of liver transplantation (LT). The pathophysiology is not known, and it is thought to develop because of mucosal damage in the bile duct related to obstruction, ischemia, or bacterial infection. It occurs in 2.5–18% of LT patients and is associated with increased graft failure, need for retransplantation, and mortality. Here we report on a case of BCS of late appearance after LT who was successfully treated by endoscopic means.

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Introduction

Biliary cast syndrome (BCS) was first described in 1975 and refers to the presence of multiple hard pigmented dark materials that mold the bile ducts. BCS is reported to occur in 2.5–18% of patients with liver transplantation (LT) [1, 2] and much less frequently in non-LT settings, including postoperative status, fasting associated with total parenteral nutrition,

and gallbladder hypomotility [3–6]. BCS in an LT setting is associated with ischemic injury to the biliary endothelium and strictures. Other mechanisms, such as acute cellular rejection and infection, have been proposed to play an etiological role in the development of BCS. With respect to the timing of cast formation, it has mostly been reported within the first year following LT, and rarely up to 5 years after LT [7].

Case Report

A 67-year-old male with a history of LT 6 years earlier for hepatitis B-induced cirrhosis presented at the emergency room with epigastric pain irradiating into the right upper quadrant. Physical examination revealed tenderness in the upper abdomen and no other abnormality. Routine laboratory evaluation only revealed a slight elevation in gamma-glutamyl transferase at 71 IU/L (reference range, 9–51 IU/L), with normal alkaline phosphatases, aminotransferases, bilirubin, and prothrombin time. Abdominal ultrasound disclosed a normal common bile duct diameter with normal portal and hepatic artery Doppler flows. Liver biopsy showed bile duct proliferation, biliary clots, and neutrophilic cholangitis, suggesting cholestasis and no acute rejection. Magnetic resonance cholangiopancreatography revealed a filling defect in the common hepatic and cystic ducts, extending down from the main hepatic confluence to the ampullary level. These findings were consistent with BCS (Fig. 1).

Endoscopic retrograde cholangiopancreatography confirmed the magnetic resonance cholangiopancreatography findings (Fig. 2), and endoscopic therapy was performed. After biliary sphincterotomy, a stone retrieval balloon was passed into the bile duct and positioned below the main hepatic confluence, inflated, and withdrawn distally to extract the cast (Fig. 3). Subsequently, regular saline flushes were employed to facilitate drainage of remainder cast from the cystic duct, and an occlusion cholangiogram allowed to exclude the presence of any anastomotic stricture. The patient's course was uneventful, pain disappeared, and he was discharged 24 h after admission. He was followed with clinical and imaging controls on a bimonthly basis during the first year and then annually up to 2 years, without any sign or symptom of recurrence.

Discussion

BCS is an uncommon condition occurring mostly after LT, defined by the presence of casts within the intra- or extrahepatic biliary system. It morphologically appears as dark, hardened material molded to the bile ducts, different from the usual round shape of biliary stones. Although bile duct stones and biliary casts have a similar microstructure [8, 9], their mechanism of formation differs significantly. The risk factors for BCS following LT include ischemia-reperfusion injury, prolonged cold preservation, prolonged warm ischemia, and hepatic artery thrombosis [10].

Biochemically, biliary casts consist of bilirubin as the primary component (approximately 10–50%), followed by bile acid synthesis products and cholesterol (10–15%) and protein or collagen from necrotic biliary epithelial cells (5–10%) [8]. Using choledochoscopy, patients with BCS have been classified into 6 categories according to the level of injury to the biliary tract epithelium [10]. From type 1 to type 6, clinical symptoms and biliary tract strictures are more frequent and the size of biliary casts increases. In type 1 (intact bile duct epithelium), clinical symptoms or laboratory index changes are absent, while type 6 (necrotic

biliary duct epithelium in the common hepatic duct plus left and right hepatic ducts) usually develops early in the postoperative period and frequently requires retransplantation. We believe that the case reported here belongs to type 1 or 2 based on the indolent patient course, including the long delay since LT at presentation, the paucity of symptoms at the time of diagnosis, and the unremarkable patient follow-up for 2 years following endoscopic extraction of the biliary cast. In particular, to our knowledge the delay of 6 years between LT and BCS diagnosis is one of the longest reported in the literature.

Statement of Ethics

Written informed consent for the procedure was signed by the patient.

Disclosure Statement

All authors disclose no financial relationships relevant to this publication.

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Fig. 1. Magnetic resonance cholangiopancreatography showing a long filling defect (arrow) in the extrahepatic ducts suspicious for biliary cast.

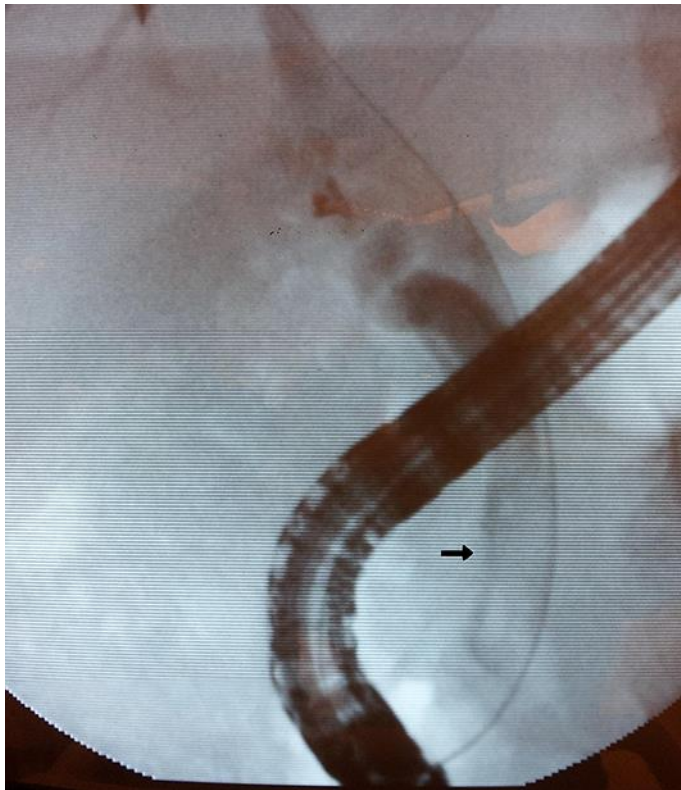


Fig. 2. Endoscopic retrograde cholangiopancreatography confirming the presence of a filling defect (arrow) in the slightly dilated extrahepatic ducts.



Fig. 3. Endoscopic view of the biliary cast in the duodenum after sphincterotomy and balloon-assisted extraction.