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Liver transplantation for "mass-forming" sclerosing cholangitis after laparoscopic cholecystectomy☆

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ABSTRACT

INTRODUCTION: Chronic biliary obstruction consequence of a bile duct injury may require liver transplantation (LT) in case of secondary biliary cirrhosis, intractable pruritus or reiterate episodes of cholangitis. "Mass-forming" sclerosing cholangitis leading to secondary portal vein thrombosis and pre-sinusoidal portal hypertension has not been reported so far.

PRESENTATION OF CASE: We present the case of a patient who underwent laparoscopic cholecystectomy for Mirizzi syndrome. The persistent bile duct obstruction due to a residual gallstone fragment was treated by a prolonged biliary stenting. Following repeated bouts of cholangitis, a fibrous centrohepatic scar developed, conglobating and obstructing the main branches of the portal vein and of the biliary tree. The patient developed secondary portal vein thrombosis and portal hypertension. After an extensive diagnostic work-up, including surgical exploration to rule out malignancy, the case was successfully managed by liver transplantation.

DISCUSSION: Mass-forming sclerosis of the bile duct and biliary bifurcation may develop as a consequence of chronic biliary obstruction and prolonged stenting. Secondary portal vein thrombosis and pre-sinusoidal portal hypertension represents an unusual complication, mimicking Klatskin tumor.

CONCLUSION: A timely and proper management of post-cholecystectomy complications is of mainstay importance. Early referral to a specialized hepato-biliary center is strongly advised.

1. Introduction

We present the case of a patient who developed a sclerosis of the bile duct and of the biliary bifurcation as a consequence of a residual gallstone fragment after laparoscopic cholecystectomy for Mirizzi syndrome (MS) and prolonged biliary stenting. The thickening of the biliary ducts at the hilar plate conglobated the portal vein branches leading to portal vein thrombosis and portal hypertension and was finally treated by liver transplantation (LT).

2. Presentation of case

A 36-year-old patient underwent laparoscopic cholecystectomy for Mirizzi syndrome in a town hospital in July 2010. Prior to the operation, a biliary stent had been placed by endoscopic retrograde cholangio-pancreatography to relieve biliary obstruction. During the operation, due to the presence of important adhesions precluding a safe dissection of the Calot's triangle, the gallbladder was opened in the attempt to remove a huge gallstone impacted in the infundibulum. As the gallstone was firmly adhering to the gallbladder wall and impossible to dislodge, it was fragmented using a cadmium laser. Cholecystectomy was achieved by laparoscopy leaving in place a part of the gallbladder infundibulum. Intraoperative cholangiography was not even attempted. The early postoperative course was uneventful and the first biliary stent was removed, but the patient suffered two episodes of cholangitis due to recurrent bile duct obstruction in the following three months, which were treated again by endoscopic stenting of the bile duct. A first plastic stent was replaced by a fully covered metallic stent, which was left in place until March 2012, when it was substituted after the patient presented a further episode of cholangitis. The total duration of the biliary stenting was 21 months. In April 2012, Doppler ultrasonography showed a previously unrecognized splenomegaly and partial thrombosis of intrahepatic portal vein branches, thus low molecular weight heparin therapy was started. Only in June 2012, when he presented an episode of upper gastrointestinal bleeding from large gastric fundus varices, the patient was referred to our Institution.

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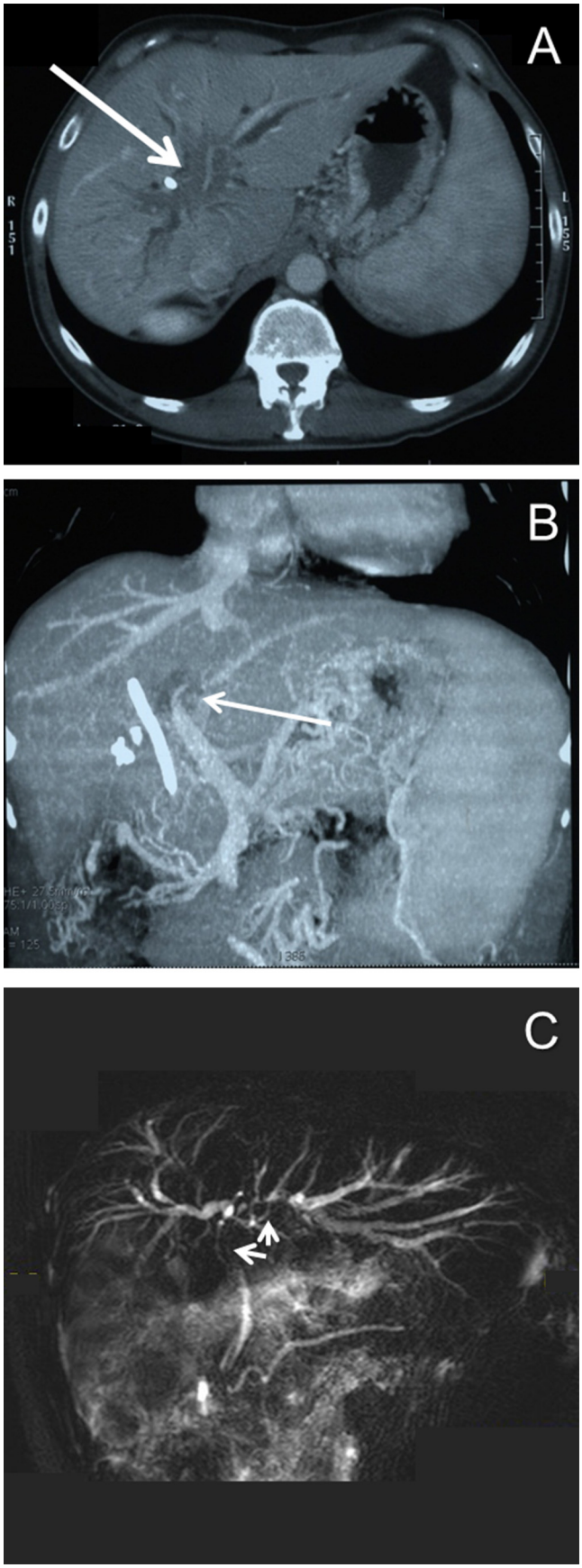

Computed tomography and magnetic resonance imaging showed the obstruction of the intrahepatic portal vein branches related to the presence of a thick, dense tissue in the hilar plate region, determining also the stenosis of the proximal portion of the common bile duct and of the bifurcation (Fig. 1). This presentation was deemed consistent with a locally advanced Klatskin tumour involving the intra-hepatic portal vein branches with secondary thrombosis. Nevertheless, CA19.9 level was normal despite the increased bilirubin and positron emission tomography was consistent with an inflammatory reaction at the hepatic hilum. Furthermore, several trans-luminal and percutaneous ultrasound-guided biopsies were negative for malignancy. In order to formally rule out cholangiocarcinoma we proceeded to open surgical exploration. At operation, the liver was cholestatic but with no sign of cirrhosis, the hepatic pedicle presented a cavernomatous transformation and a dense scar-like tissue occupied and retracted the hilar plate. After dissection of the hepatic pedicle cavernoma and removal of a residual gallstone fragment (Fig. 2A), we carried out an extensive sampling of the extremely stiff hilar tissue. Pathologic examination was again negative for cancer. The dissection of the biliary bifurcation and of the hilar plate was abandoned when it became evident that the intra-hepatic involvement of the biliary ducts precluded any attempt at a bilio-enteric derivation. Thus, considering the portal hypertension status, the complex biliary lesion not amenable to a standard repair by hepatico-jejunostomy and the absence of any argument in favour of malignant disease, we enlisted the patient for liver transplantation (LT). Due to the complicated surgical history and the reiterate episodes of cholangitis, the patient was granted an upgrade on the waiting list and was transplanted in August 2012 with a biochemical Model for End-stage Liver Disease score of 16. No major post-operative complication occurred, and he is currently alive with good liver function ten months after LT. Macroscopic (Fig. 2B and C) and pathological (Fig. 3) examination of the explanted liver confirmed the absence of malignancy and the presence of a "mass-forming" sclerosis of the hilar bile ducts encasing the portal vein branches.

3. Discussion

LT represents the last resource in the treatment of bile duct injuries (BDI) in case of secondary biliary cirrhosis, repeated episodes of cholangitis, intractable pruritus and poor quality of life.[1] Several cases of LT after a BDI occurring during laparoscopic cholecystectomy have been reported. The timing of LT varies according to the type of injury: patients with associated vascular injuries may develop a fulminant hepatic failure consequent to the massive hepatic necrosis, thus requiring urgent LT.[2] On the other hand, patients with complex biliary injuries not amenable to bilio-enteric derivation[3] or hepatic resection,[4] or who develop secondary biliary cirrhosis, usually undergo LT months to years after the BDI.[1,5,6]

Mirizzi syndrome is the obstruction of the common bile duct due to the extrinsic compression by a gallstone impacted in the gallbladder neck. Surgical treatment depends on the local anatomy and spaces from laparoscopic cholecystectomy to hepatico-jejunostomy.[7,8] The role of laparoscopic cholecystectomy in the treatment of MS is still debated. Due to the presence of tenacious inflammatory adhesions, grasping of the Hartmann's pouch

Fig. 1. Pre-operative work-up. (A) Contrast-enhanced computed tomography showing the portal vein occluded by dense tissue at the porta hepatis (thick arrow). Note the presence of visceral varices, the splenomegaly and the biliary stent; (B) the hypodense mass conglobates the hepatic vessels (thin arrow) and the bile ducts at the hilar plate; (C) magnetic resonance cholangiopancreatography showing the stenosis of the proximal common bile duct extended to the junction of the main biliary branches (arrowheads).