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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.

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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

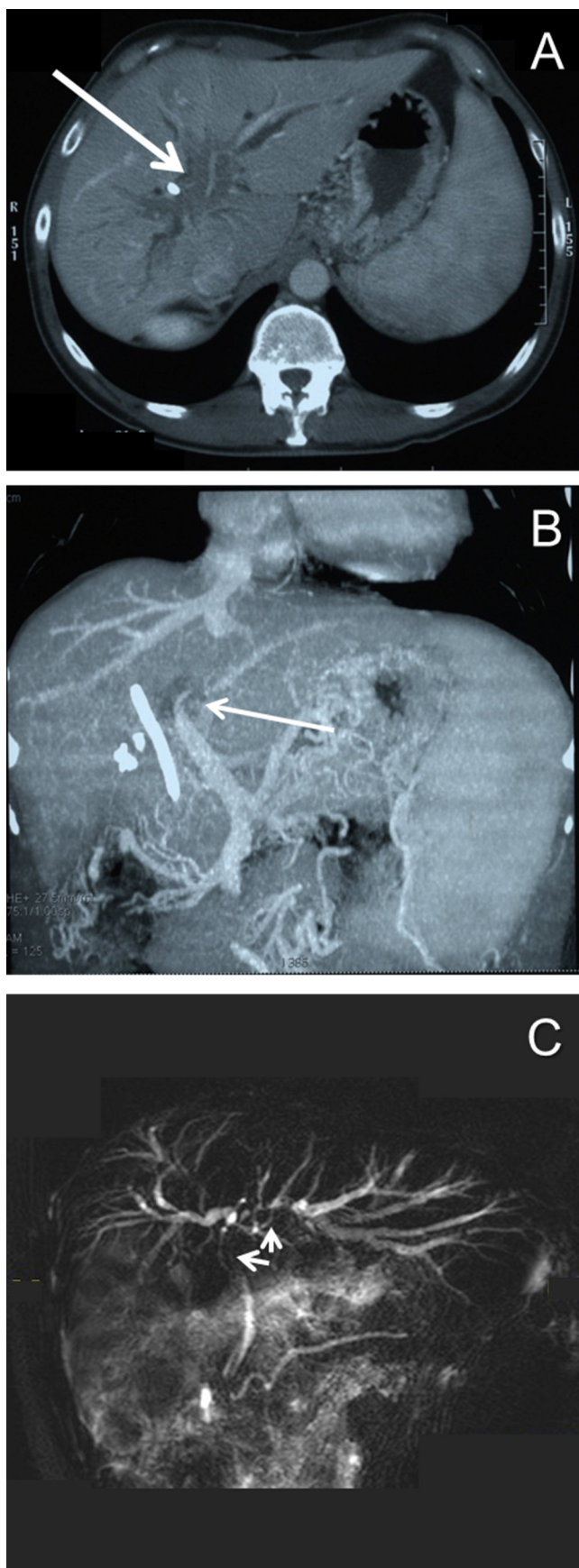
A 36-year-old patient underwent laparoscopic cholecystectomy for Mirizzi syndrome in a town hospital in July 2010. Prior to the

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 derivation³ 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 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).

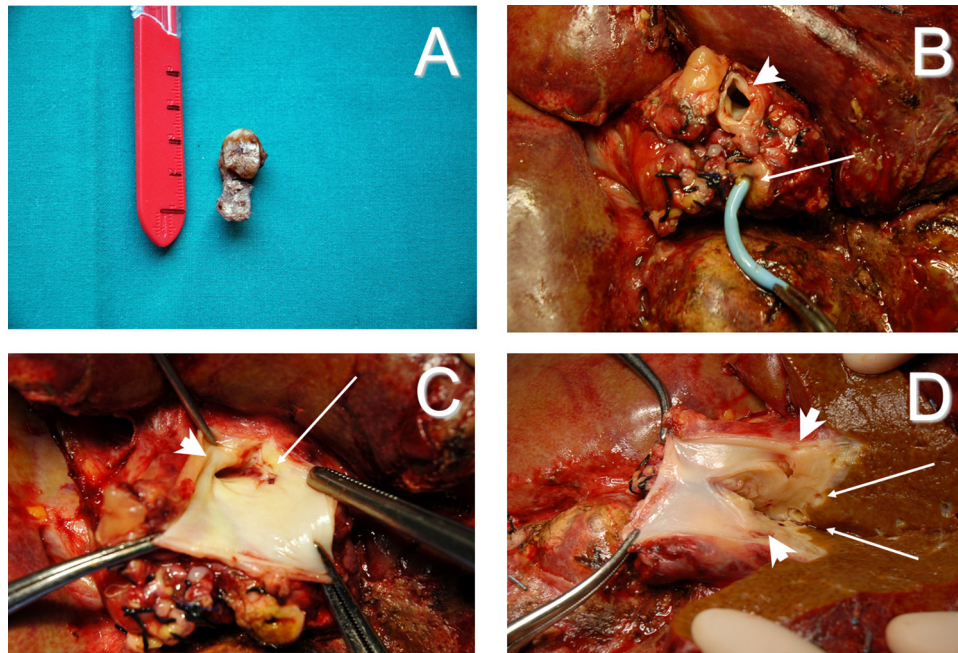


Fig. 2. Explanted liver. (A) Residual 2-cm gallstone; (B) thickened portal vein (arrowhead) and bile duct (arrow); (C) internal aspect of the portal vein after longitudinal section, with the stenotic orifices of the right (arrow) and left (arrowhead) intra-hepatic branches; (D) the portal vein has been severed longitudinally along with the caudate lobe: abnormal thick fibrous tissue surrounding and conglobating the venous branches (arrowheads) and the biliary orifices (arrows).

and correct exposure and dissection of the Calot's triangle is sometimes impossible.⁹ In complex cases, some Authors suggest a wide opening of the gallbladder, extraction of the gallstones and subtotal cholecystectomy, leaving the gallbladder infundibulum in place.¹⁰ According to Antoniou et al.,⁹ laparoscopic cholecystectomy is achieved in 59% of cases, with a 0.8% mortality, 16% complication rate, and 5% reoperation rate. Noteworthy, 25% of complications is represented by residual stones. Preoperative diagnosis of MS is associated with a lower conversion and postoperative complication rate.¹¹ Nevertheless, although several cases of successful management of MS by laparoscopic cholecystectomy have been reported, most Authors remain cautious in recommending the laparoscopic approach, especially for type II MS (i.e. presence of a cholecysto-biliary fistula).⁸ Thus, conversion to an open approach should be

preferred when dissection by laparoscopy becomes hazardous, as it was probably the case for our patient.

This case depicts a peculiar indication for LT, which was not secondary biliary cirrhosis, pruritus or recurrent cholangitis as in most cases of late LT after BDI. The patient did not develop portal hypertension as a consequence of hepatic fibrosis, as demonstrated by the fact that liver parenchyma was histologically normal. A fibrous scar, issue of reiterate episodes of cholangitis and prolonged biliary stenting, surrounded and narrowed the intra-hepatic main branches of the portal vein, causing secondary portal vein thrombosis and pre-sinusoidal portal hypertension. Our first diagnostic hypothesis was a locally advanced Klatskin tumour. This hypothesis was supported by the fact that recurrent cholangitis is a well-known risk factor for cholangiocarcinoma and that bile duct sclerosis-related portal vein encasement has not been reported before. This case highlights the importance of a proper initial management of Mirizzi syndrome and, in general, of any case of biliary obstruction. Even in complex cases, the principles of "critical view of safety"¹² must be respected as much as possible and the threshold for conversion to open cholecystectomy should be kept very low if a safe dissection by laparoscopy is impossible. Intra-operative cholangiography represents a precious aid in defining bile duct anatomy and in diagnosing bile duct injuries.¹³ In this case, if successfully performed, it would have allowed immediate detection of persistent bile duct stenosis and consequently led to a correct timely repair. Finally, it could be argued that a prompt treatment of post-cholecystectomy complications (i.e. removal of the residual gallstone fragment and bilio-enteric derivation) would have avoided the progression towards this unusual form of "mass-forming" sclerosing cholangitis and prevented the need for LT.

4. Conclusion

Chronic cholangitis due to prolonged biliary obstruction may lead to portal vein obstruction and thrombosis. Proper initial management of Mirizzi syndrome and biliary obstruction is of mainstay

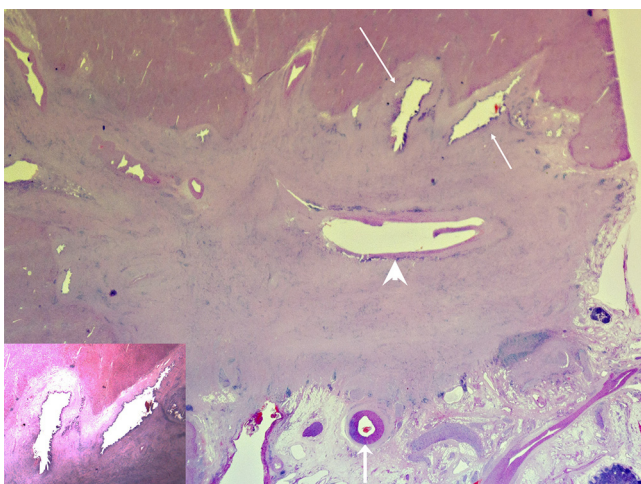


Fig. 3. Explanted liver histology (haematoxylin and eosin staining; original magnification 1x). Section of the hilar plate demonstrating the presence of a dense, star-shaped fibrous tissue conglobating the portal vein (arrowhead), the bile ducts (thin arrows), and the arteries (thick arrow). Inset: biliary ducts surrounded by the fibrous tissue.

importance. The referral to an experienced hepatobiliary unit before the onset of severe complications of chronic cholangitis is strongly advised.

Conflict of interest statement

None.

Funding

None.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

Damiano Patrono wrote the paper, obtained consent, processed digital images and performed literature search; Elena Mazza was the resident in charge of patient care and helped in writing the paper; Gianluca Paraluppi was the second surgeon and jointly performed both operations; Paolo Strignano acquired photos during the operations and contributed to manuscript drafting; Ezio David was the pathologist in charge of pathological examination; Renato Romagnoli performed the transplant operation and critically revised the manuscript; Mauro Salizzoni was the overall responsible for the care of the patient, performed the first operation, supervised and critically revised the manuscript.

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