

Two Cases of Type Va Extrahepatic Bile Duct Duplication With Distal Klatskin Tumor Surgically Treated with Whipple Procedure and Hepaticojejunostomy

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Abstract

We describe the diagnostic and therapeutic challenges of a type Va extrahepatic bile duct duplication coexistent with distally located hilar cholangiocarcinoma (Klatskin tumor). We present 2 cases that were diagnosed preoperatively and treated with a modified surgical technique of a combined pylorus-preserving Whipple procedure and hepaticojejunostomy.

Introduction

Biliary tree anomalies are quite common. Approximately 42% of the general population has some anatomical variation in the biliary tree.¹ Biliary tree anomalies have been reported in association with multiple disorders like recurrent pancreatitis, cholelithiasis, cholangitis, and biliary malignancy.² Accurate delineation of biliary tree anatomy is essential for planning a proper surgical treatment and preventing postoperative complications. Duplication of the extrahepatic bile duct is a rare anomaly of the biliary system.^{2,3} A Klatskin tumor is a cholangiocarcinoma involving the biliary hilum at the junction of the right and left main hepatic ducts.

Case Report

Case 1: A 77-year-old woman presented with jaundice and a 9-kg weight loss. Abdominal computed tomography (CT) revealed diffuse bile duct dilatation. No pancreatic mass was seen. Magnetic resonance cholangiopancreatography (MRCP) showed diffuse bile duct dilatation and a stone in the extrahepatic duct with questionable distal common bile duct (CBD) stricture. The pancreatic duct was normal. Endoscopic retrograde cholangiopancreatography (ERCP) was attempted at a community hospital, which reported diffuse bile duct dilatation. The procedure was aborted due to suspected contrast extravasation from the biliary tree, and the patient was transferred to our facility for further evaluation. Liver function tests (LFTs) were consistent with obstructive jaundice. Repeated ERCP revealed extrahepatic bile duct duplication with single biliary drainage to the duodenum (Figure 1). The cystic duct was communicating with the right main extrahepatic duct. These findings were consistent with type Va extrahepatic bile duct duplication (Figure 2). The right and left main extrahepatic bile ducts were markedly dilated (25 mm and 15 mm, respectively) with a stone in the left duct (Figure 1). Brushing cytology of the CBD

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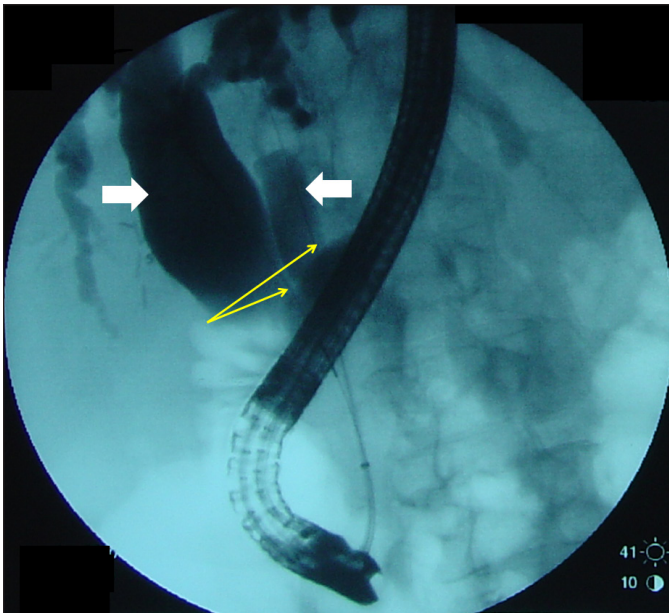


Figure 1. ERCP showing type Va extrahepatic bile duct duplication. Right and left extrahepatic bile ducts (white arrows) are dilated (25 and 15 mm, respectively) with a guidewire in each duct (yellow arrows).

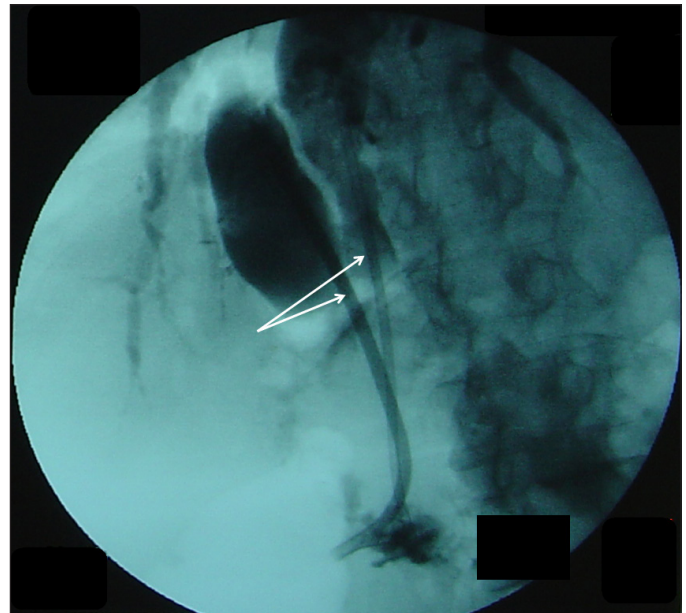


Figure 3. Two plastic stents (arrows) were positioned up the right and left extrahepatic bile ducts.

and main extrahepatic bile ducts revealed adenocarcinoma cells suggestive of a cholangiocarcinoma. Two plastic stents were placed up to the right and left main extrahepatic bile ducts (Figure 3).

Case 2: A 78-year-old woman presented with pruritus, jaundice, and dark urine for 1 month. LFTs were consistent with obstructive jaundice. Abdominal CT showed diffuse bile duct dilatation. Further evaluation by MRCP showed diffuse bile duct dilatation to the level below the cystic duct take off. No stones were identified. Prior ERCP had shown a distal CBD stricture that was dilated and stented with a 10 French x 5 cm plastic stent. Brush cytology was negative. The patient’s jaundice persisted, and endoscopic ultrasound showed a hypoechoic mass of 21.3 x 17.2 mm at the superior surface of the neck of pancreas (Figure 4). Fine-needle aspiration revealed moderately differentiated adenocarcinoma cells consistent cholangiocarcinoma. The old stent was removed. Repeat ERCP showed type Va extrahepatic bile duct duplication. The cystic duct was communicating with the right main extrahepatic duct. The biliary hilum was obstructed by the mass mimicking a Klatskin tumor (Figure 5). The biliary strictures were dilated and 2 plastic stents were placed up to the right and left main extrahepatic bile ducts (Figure 5).

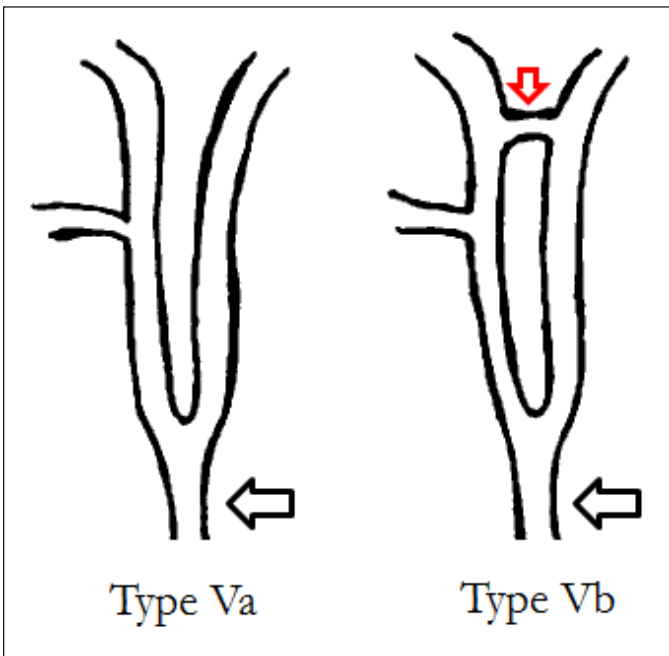


Figure 2. Diagram of type V extrahepatic bile duct duplication. Type Va consists of 2 separate extrahepatic bile ducts with single biliary drainage (black arrow) to the duodenum. Type Vb consists of 2 extrahepatic bile ducts with communicating channel (red arrow) and single biliary drainage (black arrow) to the duodenum.

Both patients were referred for pylorus-preserving Whipple surgery with hepaticojejunostomy. The intra-operative evaluation and surgical pathology confirmed the diagnosis of type Va extrahepatic bile duct duplication with cholangiocarcinoma involving a distally located Klatskin tumor (Figure 6).

Discussion

Extrahepatic bile duct duplication is a rare congenital anomaly.⁴ There are 5 types of extrahepatic bile duct duplication, and type V is the least common.⁴ It represents a single bili-

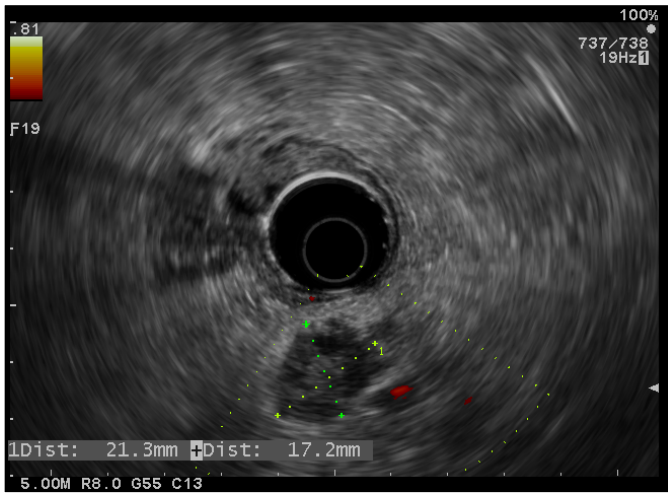


Figure 4. EUS showing a 21.3 x 17.2 mm hypoechoic mass at the superior surface of the neck of pancreas.

any drainage of double extrahepatic bile ducts without (Va) or with (Vb) communicating channels (Figure 2).⁴⁻⁹ Extrahepatic bile duct duplication coexistent with pancreaticobiliary malignancy has been reported. Based on a review of the Japanese medical literature, Yamashita reported gastrointestinal cancers in 25% of 47 cases.² Cancers of the biliary system were common when the pancreaticobiliary maljunction opened into the second portion of the duodenum or the pancreatic duct.² There was no pancreaticobiliary maljunction identified in our cases.

The coexistence of extrahepatic bile duct duplication with cholangiocarcinoma contributed to a challenging diagnosis and management strategy in our cases. The accurate preoperative delineation of the biliary tree anatomy was an essential step for further management. Preoperative diagnosis of type V extrahepatic bile duct duplication is essential for proper surgical planning, especially when coexistent with distal hilar cholangiocarcinoma (Klatskin tumor). In our

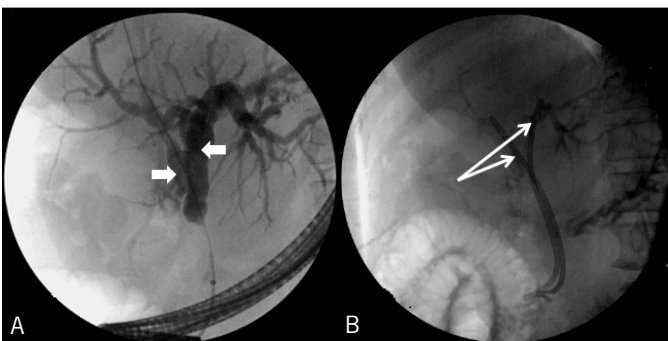


Figure 5. (A) ERCP showing dilated right and left extrahepatic bile ducts (arrows). The biliary hilum is invaded by the mass-like Klatskin tumor. (B) Two plastic stents (arrows) positioned up to the right and left extrahepatic bile ducts.

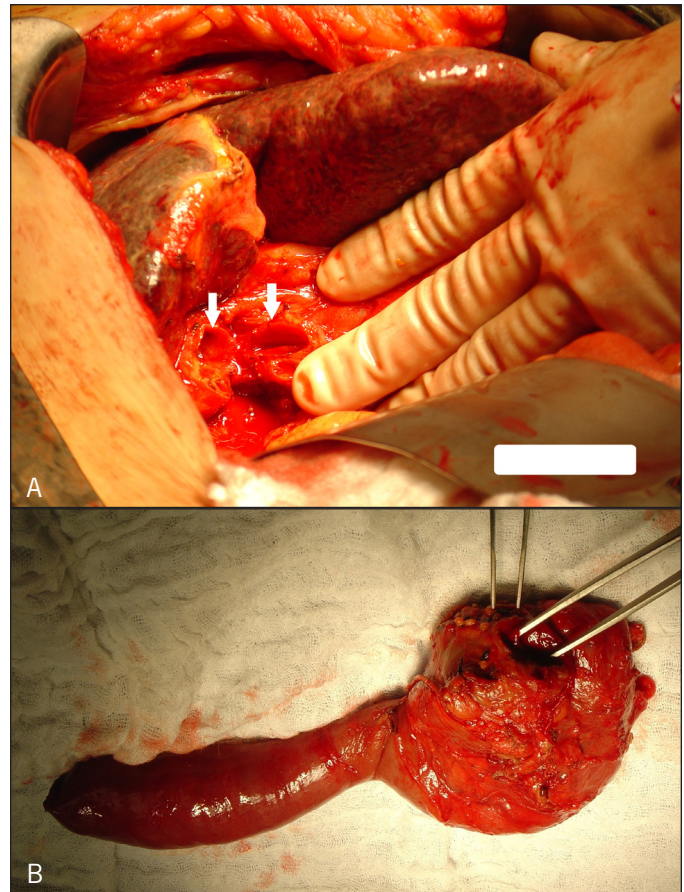


Figure 6. (A) Proximal ends (arrows) of the right and left extrahepatic bile ducts after surgical transection. (B) The head of pancreas with the duodenum after pancreateoduodenectomy. The surgical clamps point to the distal ends of the right and left extrahepatic bile ducts after transection.

cases, biliary drainage was achieved by pylorus-preserving Whipple surgery and hepaticojejunostomy, an approach that has only been described once in the literature.¹⁰⁻¹² Both of our patients had an uneventful post-operative course.

Disclosures

Author contributions: TA Hammad and Y. Alastal wrote the manuscript and reviewed the literature. MA Khan, O. Alaradi, A. Nigam, and TC Sodeman reviewed the article for important intellectual content. M. Hammad revised the article. A. Nawras supervised the process, critically revised the article, and approved the final draft. TA Hammad is the article guarantor.

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