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Case Report

Reverse Mirizzi Syndrome ☆,☆☆

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ABSTRACT

A man in his 40s presented to our Hospital with abdominal pain, jaundice, and pruritus. He had a history of Alagille Syndrome treated with cholecystojejunostomy in the neonatal period because of initial misdiagnosis of biliary atresia. Laboratory investigations showed hyperbilirubinemia (total bilirubin 1.76 mg/dL [<1.2 mg/dL]; conjugated 1.06 mg/dL [<0.3 mg/dL]) and cholestasis (GGT 78 U/L [<50 U/L]; ALP 200 U/L [<50 U/L]). Transabdominal ultrasound was limited by aerobilia due to the cholecystojejuno-anastomosis. Subsequent basal CT scan revealed an impacted stone within the patient's native common bile duct (CBD). Aerobilia in intrahepatic bile ducts and gallbladder was reported. Magnetic Resonance cholangiopancreatography confirmed the gallstone in the CBD compressing cystic duct and common hepatic duct, with dilation of the upstream bile ducts. Furthermore, the native CBD was obstructed by other gallstones. In Mirizzi syndrome, gallstones impacted in gallbladder's Hartmann's pouch or cystic duct extrinsically compress CBD. We suggest naming the present condition "Reverse Mirizzi Syndrome" (Renzulli Matteo Syndrome, RMS) because it is the exact opposite of Mirizzi syndrome.

Introduction

Mirizzi syndrome is characterized by obstruction of the common bile duct (CBD) with or without a concomitant cholecystobiliary fistula, secondary to mechanical compression caused by gallstones impacted in gallbladder's Hartmann's pouch or cystic duct. Clinical manifestations of Mirizzi syndrome are jaundice, fever, and right upper quadrant pain. Diagnostic evaluation of Mirizzi syndrome comprises abdominal

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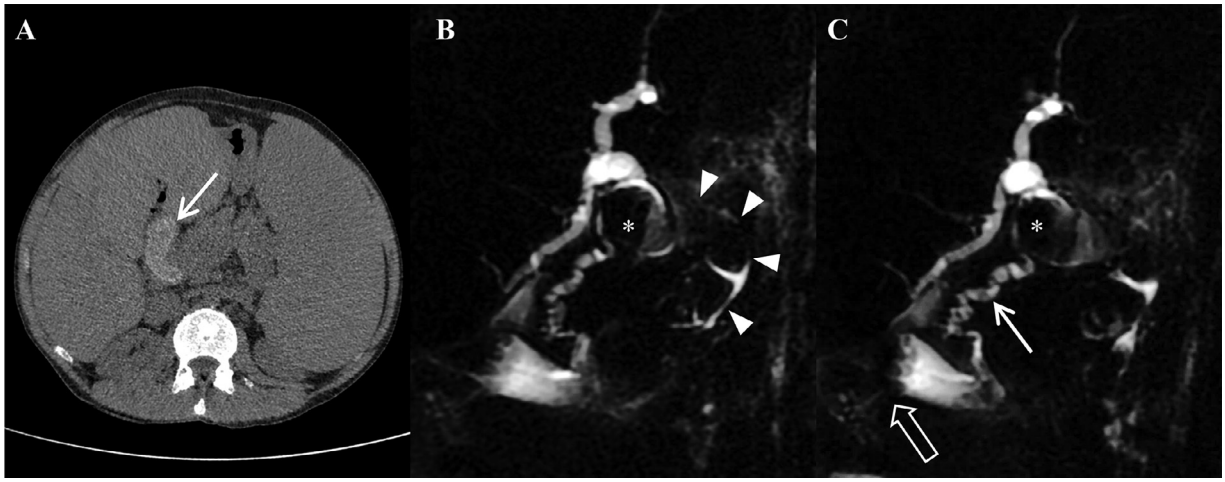

Fig. 1 – (Panel A) Basal CT scan shows a large, partially calcified gallstone (arrow) occupying the patient's native common bile duct. (Panels B and C) Magnetic Resonance cholangiopancreatography (MRCP) shows the gallstone (asterisk) compressing cystic duct (thin arrow, Panel C) and common hepatic duct, with dilation of the upstream bile ducts. The native common bile duct is obstructed by other gallstones (arrowheads, Panel B). Cholecystojejuno-anastomosis is pointed by the empty arrow in Panel C.

ultrasound, computed tomography (CT) and Magnetic Resonance cholangiopancreatography (MRCP). Surgery is the mainstay of therapy [1–3].

Alagille syndrome (ALGS) is an autosomal dominant, multisystem disorder caused by mutations in genes of the Notch signaling pathway. Hepatic manifestations are the most consistent feature of ALGS and include intrahepatic bile duct paucity and chronic cholestasis. They can lead to cirrhosis and hepatic failure, requiring liver transplantation (LT) [4].

ALGS enters differential diagnosis with biliary atresia (BA), a progressive, idiopathic, fibro-obliterative disease of the extrahepatic biliary tree. BA is the most common cause of obstructive jaundice in the first 3 months of life. The therapeutic management of BA varies depending on the location of the most proximal obstruction in the extrahepatic biliary tree. In the majority of cases, the biliary obstruction is at the hepatic hilum, therefore the surgical procedure is the Kasai portoenterostomy. In cases where the most proximal level of obstruction is within the CBD, a cholecystojejuno-anastomosis can be performed. This procedure connects the gallbladder fundus to a jejunal loop and establishes a new biliary outflow tract through the cystic duct and gallbladder [5,6].

Case report

A man in his 40s presented to our hospital with abdominal pain, worsening jaundice, and pruritus. He had a history of ALGS treated with cholecystojejunostomy in the neonatal period because of initial misdiagnosis of BA. On examination, the patient was jaundiced, apyrexial, and reported right flank pain. Laboratory investigations showed hyperbilirubinemia (total bilirubin 1.76 mg/dL [<1.2 mg/dL]; conjugated 1.06 mg/dL [<0.3 mg/dL]) and cholestasis (GGT 78 U/L [<50 U/L]; ALP 200 U/L [<50 U/L]). Serum C-reactive protein and fibrinogen concentrations were only slightly elevated, and white cell count was normal. A transabdominal ultrasound was performed but it was limited by aerobilia due to the cholecystojejunoanastomosis. Therefore, the patient underwent a basal CT scan that showed a large, partially calcified gallstone (arrow, Panel A Fig. 1) completely occupying the patient's native CBD. Aerobilia in intrahepatic bile ducts and gallbladder was reported. Magnetic Resonance cholangiopancreatography confirmed the gallstone (asterisk, Panel B-C Fig. 1) compressing cystic duct (thin arrow, Panel C Fig. 1) and common hepatic duct, with dilation of the upstream bile ducts. Furthermore, the native CBD was obstructed by other gallstones (arrowheads, Panel B Fig. 1). The patient was scheduled for endoscopic procedures but in the meanwhile underwent LT. Thereafter, the patient demonstrated good clinical conditions.

Discussion

We propose the term "Reverse Mirizzi Syndrome" (Renzulli Matteo Syndrome, RMS) to describe the condition presented in this paper, as it represents the exact opposite of Mirizzi Syndrome. In Mirizzi syndrome, gallstones impacted in gallbladder's Hartmann's pouch or cystic duct extrinsically compress CBD. Conversely, in our patient with a cholecystojejunoanastomosis (empty arrow, Panel C Fig. 1), gallstones impacted in CBD (asterisk, Panel C Fig. 1) extrinsically compress the cystic duct (thin arrow, Panel C Fig. 1). In fact, in our patient the role of CBD and gallbladder is reversed: CBD assumes to some extent the role of bile reservoir, physiologically accomplished by the gallbladder, favoring the formation of stones within the CBD itself; the stones piled in the CBD extrinsically compress the cystic duct and Hartmann's pouch which, being connected to the jejunum through the gallbladder fundus, serve as biliary outflow. To our knowledge, this is the first reported case of Reverse Mirizzi Syndrome in the medical literature.

Patient consent

Obtained.

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