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Case Report / Olgu Sunumu

Type IV Mirizzi Syndrome at the Hepatic Confluence Mimicking a Hilar Cholangiocarcinoma.

Hepatik Bileşkede Hiler Kolanjiokarsinomayı Taklit Eden Tip IV Mirizzi Sendromu

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Abstract

Mirizzi's syndrome refers to common bile duct obstruction resulting from compression by a gallstone impacted in the cystic duct or neck of the gallbladder. Some cases can not be identified preoperatively, despite modern imaging techniques. Today, treatment of Mirizzi syndrome is surgical. The essential part of the management of patients with Mirizzi syndrome is to determine the best surgical procedure in the preoperative period. In type I patients, simple cholecystectomy is generally enough, but types II-IV require more complex surgical approach, such as cholecystectomy and bilioenteric anastomosis. Here, we presented a 48 year-old man with obstructive jaundice who diagnosed as Mirizzi's syndrome.

Key words: Mirizzi's syndrome, hepatic confluence, hilar cholangiocarcinoma

Özet

Mirizzi sendromu, sistik kanal ya da safra kesesi boynuna impakte taşın, koledok kanalına dıştan basısı sonucu gelişir. Modern görüntüleme tekniklerine rağmen bazı olgular ameliyat öncesi dönemde belirlenememektedir. Mirizzi sendromunun günümüzdeki tedavisi cerrahidir. Mirizzi Sendrom tanılı hastalarda tedavide en önemli noktalardan biri de preoperatif dönemde cerrahi tedavinin belirlenmesidir. Tip I olgularda basit kolesistektomi yeterli olurken, Tip II-IV MS olguları kolesistektomi ve biliyoenterik anastomoz gibi kompleks prosedürler gerektirebilir. Burada tıkanma sarılığı ile müracaat eden, Mirizzi sendromu tespit edilen 48 yaşında bir erkek hasta sunulmuştur.

Anahtar kelimeler: Mirizzi sendromu, hepatik bileşke, kolanjiyokarsinoma

Introduction

MS is a rare cause of jaundice due to extrinsic compression of the CHD and is present in approximately 0.35% of cholecystectomies[1]. Its presentation sometimes varies from obstructive jaundice associated with extrinsic compression or, when the stones migrate through the cystic duct, to the presence of cholecystobiliary fistula. Here, we report the case of a patient with Mirizzi syndrome from ultrasound diagnosis to the surgical operation.

Case Report

We present the case of a 48-year-old male patient, who arrived in the emergency room for recent onset of epigastric and right hypochondrium pain associated with nausea, vomiting, dark urine, grey feces, and scleroskin

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jaundice. He also had history of progressively increasing jaundice for the last two months**.** On examination, he was deeply icteric and dehydrated, with marked tenderness in the upper abdomen. His blood investigations showed leukocytosis and markedly deranged liver functions with a total bilirubin level of 12.7mg/dL (normal range 0.1 to 1.2mg/dL), serum glutamic pyruvic transaminase of 186U/L (normal range 0 to 45IU/L), serum glutamic oxaloacetic transaminase of 64IU/L He was resuscitated and an abdominal ultrasound (US) followed by magnetic resonance cholangiopancreatography (MRCP) (Figure 1), to determine whether there was a gallbladder cancer formation. Endoscopic retrograde cholangiopancreaticography (ERCP) was performed. A plastic stent was placed to drain the biliary system.

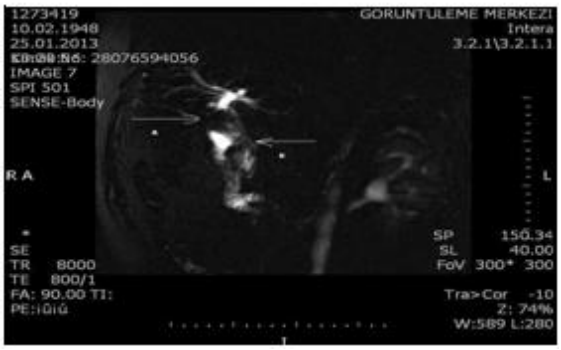

Figure 1. Magnetic resonance cholangio pancreatography demonstrating moderate intrahepatic biliary ducts dilatation due presence of T2 hypointense mass lesion at the junction of right and left hepatic biliary ducts

In order to delineate the underlying pathology, further investigation was performed. His CA19-9 levels were found to be elevated at 58.10U/mL (normal range 0 to 33U/mL, median 5.0U/mL). A computed tomography (CT) scan of his abdomen showed moderately dilated intra-hepatic ducts up to the porta hepatis, soft tissue thickening involving the gall bladder neck and proximal bile duct and a few enlarged upper abdominal nodes. The findings were suggestive of neoplastic lesion involving the gall bladder neck and proximal common bile duct causing bile duct stricture. Taking into consideration the findings from the ERCP, CA19-9 level, and CT scan findings, a provisional diagnosis of cholangiocarcinoma involving the gall bladder neck was made. Two weeks after his initial presentation, our patient was scheduled for resection of the tumor.

On surgical exploration, the gallbladder was detached from the liver, then it was opened, and after removing the bigger stone, the smaller one was seen in the cystic duct with a fistula involving up to complete destruction of the common bile duct (Figure 2), hereby the confirmation of a type IV Mirizzi syndrome.

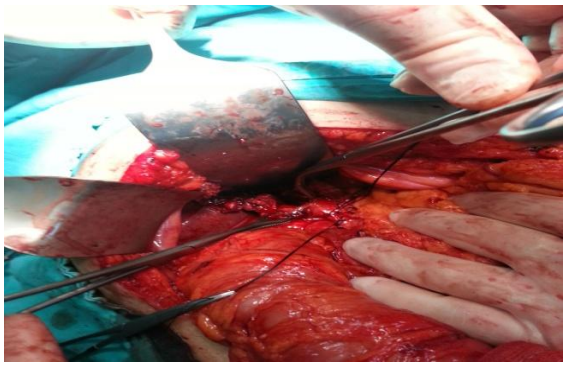

Figure 2. A fistula involving up to complete destruction of the common bile duct.

In view of the high clinical suspicion, a biopsy of the lateral wall of the bile duct wall was performed and sent for frozen section, which was reported as benign. After removing the smaller stone an operative cholangiogram was performed to confirrm the diagnosis and exclude the presence of other stones in the choledocus. After inflamed segment of bile duct and gallbladder remnant were resected, we also performed Roux-en-Y hepatico-jejunostomy. The postoperative course was

uneventful with progressive normalization of the hepatic stasis parameters and of transaminases. The patient was discharged on the eighteenth postoperative day in good health conditions.

Discussion

The syndrome was initially described in 1948 by Pablo Luis Mirizzi[2], Mirizzi syndrome (MS) is a rare cause of jaundice due to extrinsic compression of the CHD and is present in approximately 0.35% of cholecystectomies[1]. McSherry[3] classified the syndrome into 2 groups: type I where extrinsic compression of the bile duct occurs and type II where erosion occurs of the wall of the CHD by the stone with formation of a cholecysto-choledochal fistula.

The Csendes classification of Mirizzi syndrome is broken down as follows: Type 1: external compression of the common bile duct Type 2: a cholecystobiliary fistula is present involving less than one thirdvthe circumference of the bile ductv Type 3: a fistula is present involving up to two thirds the circumference of the bile duct Type 4: a fistula is present with complete destruction of the wall of the bile duct[4]. (Figure 3) In a large study (219 patients), Csendes et al. reported that 11% of their patients with MS had type I lesions, 41% had type II, 44% had type III, and 4% had type IV.

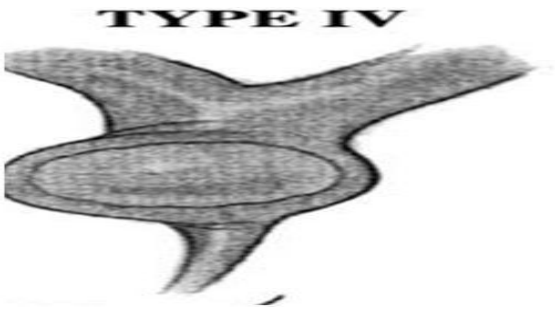

Figure 3. Presence of cholecystobiliary fistula which involves the entire circumference of the common hepatic duct wall.

The mechanism of the pathology includes a possible explanations:externalcompressing of the common bile duct and followed complications: empyema of gallbladder, abscess, cholecystocholedochal fistula, CHD structure and bile ducts obstructions[2]**.** Impaction results in the Mirizzi syndrome in two ways: chronic or acute inflammatory changes lead to gallbladder shrunken and secondary structure of CBD, or large impacted stones lead to compression, ischemia and necrosis, and cholecystocholedochal communication[5].

Symptoms of MS are essentially those of cholecystitis or choledocholithiasis. Most patients present with epigastric or right upper quadrant pain, jaundice, and elevated liver function tests[6]. the correct diagnosis was made in 8% to 62% of patients until ERCP was used regularly[7]. Most patients who have suspected biliary tract disease undergo ultrasound as a first test, with MRI or CT often following. Cholangiography, either per-cutaneous or endoscopic, is performed when liver function tests are sufficiently abnormal[8].

There is purportedly a 5 times higher rate of gallbladder malignancy in Mirizzi sydrome compared with that in uncomplicated gallstone disease[9]. Prasad et al[9] found 5.3% of patients with MS had gallbladder cancer compared with 1% in non-MS cases, and most were diagnosed on histology after cholecystectomy. CA19-9 is synthesized from normal human pancreatic and biliary ductal cells. the actual mechanism for elevated serum CA19-9 concentration is uncertain. It seems that extremely high and continuously increasing CA19-9 levels, together with well assessed clinical information, may point towards neoplasia[10] The diagnosis should always take

into account medical history, clinical examination, qualitative radiology studies, and careful follow-up. If all of the above features are suggestive, a high serum CA19-9 value may be of great help in favor of a diagnosis of biliary malignancy.

Treatment of Mirizzi syndrome is challenging for a surgeon. Operative methods depend on type of pathology. In Type I partial cholecystectomy is a method of choice: open or laparoscopic. If inflammation may permit total, usually antegrade with tube drain is the best. In Type II Mirizzi syndrome opportunities are dependent of biliary communication and sorrowing inflamed tissues: Choledochoplasty with neighborhoods tissues and cholecystoduodenostomy has been described, but not have introduced as good results[11].

Type II defects can usually be treated successfully with either complete or partial cholecystectomy followed by closure of the fistula with T-tube placement in CBD[5]. Baer and colleagues[12] suggested placement of a T-tube through a separate choledochotomy in the distal CBD in order to prevent excessive leakage and stricture at the fistula site. These authors also suggested biliaryenteric bypass via Roux-en-Y choledochojejunostomy or a choledochoduodenostomy to reduce the mortality and morbidity risk of CBD stricture[11]. The remaining cases with type II and all of the type III MS cases underwent cholecystectomy with excision of the external bile ducts and reconstruction with Roux-en-Y hepatico-jejunostomy. Type II-IV patients require complex management. Type IV Bilio-enteric anastomosis is preferred since the entire wall of the common bile duct has been destroyed.

The prognosis of MS is very good for type 1 lesions, as simple cholecystectomy is all that is necessary for cure. In treating more serious types with fistulous destruction of the common duct, postoperative morbidity rises, with 10% or more biliary fistulae, biliary stricturing requiring dilation or reoperation, or hepatic abscesses requiring drainage[6].

In conclusion, Surgery is the mainstay of therapy of Mirizzi syndrome. The aberrant anatomy intrinsic to this syndrome presents a difficult challenge to surgeons.Type IV patients require complex management. Total isolation of inflamed segment with Roux-en-Y hepaticojejunostomy may have the best long-term outcome.

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