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Get to Know the Mirizzi Syndrome

Marco Raharja[1], Marianto[2], Dian Daniella[3]

[1]Kristen Krida Wacana University, Faculty of Medicine, Jakarta, Indonesia (Corresponding author)
marcorahardja120592[at]gmail.com

[2]Sumatera Utara University, Faculty of Medicine, Medan, Indonesia
marianto.lie[at]hotmail.com

[3]Atmajaya Catholic University, Faculty of Medicine, Jakarta, Indonesia
dian.daniella[at]gmail.com

Abstract: ***Mirizzi syndrome describes obstruction of the common hepatic duct by an impacted stone in the cystic duct or gallbladder infundibulum. Mirizzi syndrome is classified based on characteristics such as site of occurance, extent of erosion, cholecystoenteric fistua, with or without stone. Abdominal pain is the most common presenting symptom followed by jaundice and less commonly with the classical triad of cholangitis. Symptoms and laboratory examination of mirizzi syndrome are confused with other abdominal disease especially liver and gallblader due to the many similarities. Therefore, this article is made to get to know more about Mirizzi syndrome.***

Keywords: mirizzi syndrome, classification, abdominal, symptoms

1. Introduction

Mirizzi syndrome is a rare condition caused by the obstruction of common bile duct or common hepatic duct by external compression from multiple impacted gallstones or a single large impacted gallstone in Hartman's pouch. Presenting symptoms are similar to cholecystitis but may be confused with other obstructing conditions such as common bile duct stones and ascending cholangitis due to the presence of jaundice. Preoperative diagnosis is often difficult and usually missed. This syndrome is named after the Argentinean surgeon Pablo Luis Mirizzi. He was born in 1893 in Cordoba Argentina. Mirizzi graduated from the Medical Sciences School at the National University of Cordoba in 1915. His most well-known contribution to surgery is having performed the first intraoperative cholangiogram in 1931. The first published paper describing the syndrome which bears his name today was in 1940.[1]

2. Etiology and Pathophysiology

Gallstones are usually formed from stasis bile. When bile is not fully emptied from the gallbladder, it can precipitate as sludge and subsequently turn into stones. Biliary obstruction may also lead to gallstones including bile duct strictures and cancers, such as pancreatic cancer. The most common cause of cholelithiasis is the precipitation of cholesterol that subsequently forms into cholesterol stones. The second form of gallstones is pigmented gallstones which are the result of increased red blood cell destruction in the intravascular system causing increased concentrations of bilirubin which subsequently get stored in the bile. These stones are typically black. The third type of gallstones is mixed pigmented stones which are a combination of calcium substrates such as calcium carbonate or calcium phosphate, cholesterol and bile. The fourth type is made up primarily of calcium and usually found in patients with hypercalcemia. When multiple gallstones or a singular large gallstone get impacted in Hartman's pouch (the lower outpouching of the gallbladder), external compression of the common bile duct or the common hepatic duct can occur. The exact mechanism as to why this occurs is unknown, but it is suggested to be related to a floppy Hartman's pouch containing a higher mass of stones such as with multiple stones or a single large impacted stone. This causes subsequent inflammation which can also leading to fistula formation.

Gallstones occur when substances in the bile reach their limits of solubility. As bile becomes concentrated in the gallbladder, it becomes supersaturated with these substances, which in time precipitate into small crystals. These crystals, in turn, become stuck in the gallbladder mucus, resulting in gallbladder sludge. Over time, these crystals grow and form large and/or multiple stones. These gallstones may cause symptoms of cholecystitis, but if they become embedded in a floppy Hartman's pouch, they can cause additional findings of jaundice. As this condition progresses, internal fistulas from the gallbladder into the common bile duct, common hepatic duct (CHD) and the duodenum can develop.[2,3]

3. Classification

McSherry described 2 types of Mirizzi Syndrome, type I includes evidence of gallstones impacting upon the Hartmann pouch or cystic duct along with slight external compression on the common bile duct (CBD). While type II show corrosion of the calculus into the CBD, along with cholecystobiliary fistula.[4]

Mcsherry classification of Type II was further redefined by Csendes into 3 furthertypes, based on the extent of erosion in the CBD circumference. Erosion involving less than one-third of the CBD circumference remained known as Type II, erosion up-to two-thirds the circumference of CBD became known as Type III, while type IV involves complete destruction of CBD wall.[5]

In 2008, Beltran described an additional classification. That classification is Type V as the presence of any of the first 4 types plus the formation of a cholecystoenteric fistula. Type

V is further divided into Type Va (without gallstone ileus) and Type Vb (with gallstone ileus).[6]

However, another classification also exist. Staring and Mattalana divide Type I into 2 subtypes, Ia (long cystic duct) and Ib (short cystic duct)[7]

Table 1: Combination of all classification of Mirizzi Syndrome

Type	*Description*
Type I	Gallstones impacting upon the Hartmann pouch or cystic duct along with slight external compression on the common bile duct (CBD)
Type Ia	Long cystic duct
Type Ib	Short cystic duct
Type II	Erosion involving less than one-third of the CBD circumference
Type III	Erosion up-to two-thirds the circumference of CBD
Type IV	Complete destruction of CBD wall.
Type V	Presence of any of the first 4 types plus the formation of a cholecystoenteric fistula.
Type Va	Without gallstone ileus
Type Vb	With gallstone ileus

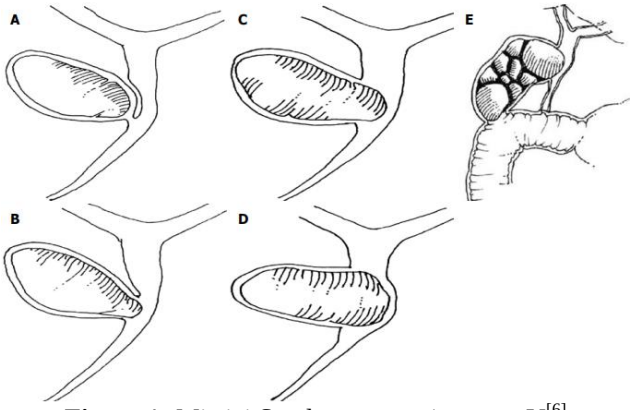

Figure 1: Mirrizi Syndrome type 1 to type V[6]

4. Clinical Sign

Mirrizi syndrome is typically seen in the setting of longstanding biliary symptoms. Obstructive jaundice is the key of feature of Mirizzi syndrome and is frequently accompanied by pain and fever. This presentation often suggests acute cholangitis. Recurent right upper quadrant pain, jaundice, and fever associated with rigors is a typical presentation in up to 80% of the cases due to cholangitis caused by either mechanical obstruction secondary to the stones or inflammation around the common hepatic duct. Patient's presenting with such symptoms should be further evaluated by MRCP (Magnetic Resonance Cholangopancreatography) with endo-biliary stenting if required. Patients can also present with cholecystitis or pancreatitis. Laboratory findings in Mirrizi syndrome, with hyperbilirubinemia as the most encountered laboratory abnormality. Elevated levels of alkaline phosphate and transaminase are also common. Leukocytosis is a frequent presence in concomitant acute cholecystitis, pancreatitis, or cholangitis. These patients are at risk of complications following routine cholecystectomy including ductal injury and bile leaks if the disease is not appreciated pre-or intra-operatively. Once mirizzi syndrome identified, a planned cholecystectomy following a thorough pre-operative evaluation by MRCP with or without pre-op ERCP offers the best chance of a favorable outcome in such difficult disease.[8,9]

5. Diagnostic

Abdominal pain is the most common presenting symptom followed by jaundice and less commonly with the classical triad of cholangitis. And despite advances in imaging, the diagnosis of Mirizzi Syndrome is still difficult and is often made intra-operatively. The diagnosis is usually suggested by biliary ultrasonography and then confirmed by MRCP or ERCP. Ultrasound diagnosis of Mirizzi syndrome demands presence of dilated intrahepatic ducts and common hepatic duct up to the point of external compression by the calculus where the bile duct can be seen to narrow. In post cholecystectomy patients, MRCP or ERCP shows narrowing of the proximal CBD and non-filling of the cystic duct remnant. The criteria used to diagnose Mirizzi Syndrome by other imaging modalities include evidence of compression of the bile duct with a stone impacted in the gallbladder neck or cystic duct with/without the presence of a cholecystocholedochal fistula.

Diagnostic Modalities

Ultrasonography or Computed Tomography (CT) may reveal stones at the junction of the cystic/common hepatic ducts associated with proximal dilation of the biliary tree. However, a dilated cystic duct can be confused with the common hepatic duct of normal diameter, thereby missing the diagnosis of this condition. Ultrasonography is poor at picking up mirizzi syndrome and has reported poor sensitivity (<5%). The use of computed tomography for the diagnosis of mirizzi syndrome is controversial, it caused CT maybe demonstrate the presence of cholecystochoedochal fistula andcases of suspeceted malignancy.

Magnetic Resonance Imaging: All patients with deranged of liver function tes should undergo magnetic resonance cholangopancreaticography (MRCP) as this would confirm the diagnosis and guide therapy. MRI is the best non-invasive imaging modality for diagnosis of mirizzi syndrome. It can demonstrate with precision the presence of biliary dilation, degree of obstruction, intraluminal or external location of the biliary stones, identifies and evaluates the degree of inflammation around the gallbladder. It can also demonstrate any extrinsic narrowing of the common bile duct, reveal complications such as fistulae and helps to identify and anatomical

Endoscopic Ultrasonography (EUS): Mirizzi Syndrome is usually encountered intra-operatively in cases of long standing gall bladder diseases. EUS is an important diagnostic modality in ultrasound negative cases of suspected biliary pancreatitis i.e. for microlithiasis ,and EUS is used in some units for confirmation of choledocholithiasis prior to therapeutic ERCP's, it does not seem to play any major role in the diagnostic or management planning protocols of mirizzi syndrome. On the other hand, intra-operative laparascopic ultrasound if available could diagnose Mirizzi Syndrome and aid management planning.

Endoscopic Retrograde Cholangiopancreatography (ERCP): may reveal narrowing or compression of the common hepatic duct. The diagnosis is traditionally made by ERCP during evaluation of the biilary tree in jaundiced patients. Lateral filling gap of the common hepatic duct and central dilation of the biliary tree on ERCP provide a strong suspicion of the syndrome. ERCP may also assist in differentiating benign from malignant strictures by smooth outline of the stricture and delineation of a cholecystocholedochal fistulae.[10]

Management

Mirizzi syndrome is important to surgeons because a preoperative diagnosis is not always possible, and because surgical management is associated with a significantly increased risk of bile duct injury. In addition, it can also cause inflammation with thick, hard, dense adhesions, and is associated with tissue swelling. The presence of a cholecystobiliary fistula increases the risk of injury to the biliary duct. During surgery, Calot's Triangle surgery can cause bile duct injury or excessive bleeding, as well as the appearance of other morbidities such as sepsis, bile duct strictures, and secondary biliary cirrhosis. The surgical management of mirizzi syndrome does not take a strictly standardized approach and must be individualized, depending on the ease of the case and the expertise of the surgical team.[11,12,13]

Mirizzi Type I syndrome can be resolved only with classic cholecystectomy. Cases of extensive chronic or acute inflammation of the Calot's Triangle can safely be performed subtotal cholecystectomy with excellent results.[12,13,14]

Mirizzi Type II syndrome is treated with the initial approach of subtotal cholecystectomy in a patient with cholecystobiliary fistula, a dissection from the gallbladder fundus to the Hartmann's Pouch. Most cholecystobiliary fistulas are diagnosed during surgery, and are not found on preoperative examination.[11,13,15]

Most cases of Mirizzi Type III Syndrome can be treated with subtotal cholecystectomy leaving a gallbladder wall flap at least 1 cm in size to repair the bile duct. However, some cases with significant inflammation of the gallbladder wall will require other procedures such as a bilioenteric anastomosis to the duodenum or Roux-en-Y hepaticojejunostomy

Management of Type IV Mirizzi Syndrome with extensive damage to the bile duct wall is by bilioenteric anastomosis. Roux-en-Y hepaticojejunostomy is usually also considered.
Mirizzi syndrome Type V can be associated with an acute or chronic serious condition with active or inactive bilioenteric fistula, as a result of which treatment differs according to type. Mirizzi Type Va syndrome is treated by simple suturing with absorbable sutures, the biliary fistula above the involved viscera (duodenum, stomach, large intestine, or small intestine) and cholecystectomy, either total or subtotal according to the presence of a cholecystobiliary fistula or simple external compression of the bile ducts. Mirizzi Type Vb syndrome remains controversial, but it seems advisable to treat prior acute condition first (gallstone ileus) and after the patient has recovered from surgery (3 months or more later), gallbladder management is appropriate for the presence or absence of external compression of the bile ducts. or cholecystobiliary fistula.[11,13,15]

Laparoscopic cholecystectomy can be performed with caution in selected patients with Type I Mirizzi Syndrome. However, it is not recommended for patients with Type II Mirizzi Syndrome or higher. In addition, the success of laparoscopic cholecystectomy in patients can be predicted based on visualization of the cystic duct at the time of initial dissection over the Calot's Triangle. A series of reports on laparoscopic cholecystectomy in Mirizzi syndrome had complication rates from 0% to 60%, bile duct injury from 0% to 22%, and mortality ranging from 0% to 25%.

Endoscopic Retrograde Cholangiopancreatography (ERCP) apart from being diagnostic, also allows sphincterotomy for stone extraction and facilitates other interventions such as placement of a stent, or nasobiliary tube, or other procedures. Patients with cholangitis will benefit from preoperative biliary drainage as a temporary measure before definitive surgery. In general, endoscopic management includes, bile drainage and stone removal, and finally stent insertion.[11]

6. Conclusion

Approach the suspect Mirizzi Syndrome patient must be carefully and thorough. Every effort should be made to establish a diagnosis correct before surgery, and if the case encountered during the operation, every effort should be made to perform an accurate operation and carefully tried to identify the type of Mirizzi, and undertake the most adequate management for every case.

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