Mirizzi's syndrome -An update on diagnosis and management: Narrative review article

Abstract

Mirizzi's syndrome is one of the rare complications of acute cholecystitis, but it is associated with significant morbidity. The diagnosis is difficult, and it is obtainedpreoperatively in 50% of cases, as the clinical symptoms are non-specific. The diagnosis is usually obtained from endoscopic retrograde cholangiopancreatography (ERCP) or by a Magnetic resonance cholangiopancreatography (MRCP). Ultrasound and computerized tomography have a low sensitivity to diagnose this condition. The treatment of Mirizzi's syndrome depends on its grade. Grades 1 and 2 involve performing a cholecystectomy, grade 3 a subtotal cholecystectomy and grade 4 may require performing a hepatic-enterostomy or choledochal-enterostomy. We have conducted this review article to look at the diagnosis and management of Mirizzi's syndrome.

Keywords- "Mirizzi's syndrome", "Obstructive jaundice"," Cholecystectomy"." Subtotal cholecystectomy", "Endoscopic retrograde cholangiopancreatography", "Choledochal-enterostomy" and "Gallbladder fistula".

Introduction

Mirizzi's syndrome is a condition that is seen in patients with longstanding gallstone disease which is characterized by extra-hepatic bile duct obstruction by a stone in the Hartmann's pouch or cystic duct. It is seen in 0.05% to 4% of all patients who undergo a cholecystectomy for gallstone disease. Mirizzi's syndrome is characterized by the cystic duct that is parallel to the common hepatic duct, the impacted stone in the cystic duct that causes partial obstruction of the common hepatic duct and the clinical presentation of obstructive jaundice or acute cholangitis. The clinical presentation includes the presence of jaundice and abdominal pain, but this is seen in 40% of patients(1,2).

Mirizzi's syndrome is classified by McSherry into Type 1 where there is external compression of the common hepatic duct by an impacted calculus in the cystic duct and Type 2 where the calculus has eroded into the common hepatic duct to cause a cholecystic-choledochal fistula. Csendes further subdivided the McSherry type 2 into three categories, types 2 to 4 depending on the size of the cholecystic-choledochal fistula. The diagnosis is usually obtained during the investigation for obstructive jaundice, with ultrasound of the abdomen and computerized tomography are associated with a low sensitivity. Magnetic resonance cholangiopancreatography

(MRCP) and Endoscopic retrograde cholangiopancreatography (ERCP) have a higher sensitivity of diagnosing this condition. Up to 50% of cases of Mirizzi's syndrome are also detected intra-operatively during cholecystectomy(3,4).

The management of Mirizzi's syndrome can be divided into surgical and endoscopic management. Surgical therapy involves performing a cholecystectomy, removing the obstructing stone and repairing the underlying fistula. Performing a cholecystectomy is difficult due to the position of the gallbladder and a fundus first approach may be required. In some cases, a subtotal cholecystectomy may be performed which opens the fundus of the gallbladder and removes the stone and the infundibulum with the cystic duct is closed with sutures. Patients with a cholecystic-choledochal fistula will require performing a choledochal-enterostomy or closure of the fistula with an underlying T-tube. Endoscopic therapy is often limited to performing an endoscopic retrograde cholangiopancreatography (ERCP) to relive the obstruction by placement of a stent and this is usually reserved for patients who are not fit for surgery(5–8).

We have conducted this review article to investigate the diagnosis and management options in Mirizzi's syndrome. The role of computerized tomography, magnetic resonance cholangiopancreatography and endoscopic retrograde cholangiopancreatography in the diagnosis of Mirizzi's syndrome is reviewed. We have reviewed the various surgical options that are available for the management of Mirizzi's syndrome. We conducted a literature review using PUBMED, Cochrane database of clinical reviews and Google scholar looking for clinical trial, observational studies, cohort studies systemic reviews, and meta-analysis from 1980 to 2024.We used the following keywords, "Mirizzi's syndrome", "obstructive jaundice", "cholecystectomy", "subtotal cholecystectomy "" Endoscopic retrograde cholangiopancreatography "," choledochal-enterostomy" and "gallbladder fistula". All articles were in English language only. Further articles were obtained by manual cross referencing of the literature. Commentaries and letters to the editor and editorials were excluded. Adult male and female patients were included in this study. Pregnant patients and pediatric patients were excluded.

Discussion

Classification of Mirizzi's syndrome

McSherry was the first to classify Mirizzi's syndrome into a type 1 which involves external compression of the common hepatic duct by a stone that is impacted at the cystic duct or

Hartmann's pouch. Type 2 is characterized by a stone that has partially or completely eroded into the common hepatic duct to form a cholecystic- choledochal fistula(9).Csendes further modified the classification into type 1 which was external compression of the common bile duct, type 2 is a cholecystic biliary fistula involving less than one third of the circumference of the bile duct, type 3 is a fistula involving more than two thirds of the bile duct circumference and type 4 is complete destruction of the common bile duct by the fistula(10).Csendes and Beltran validated this classification and further subdivided type 4 into a type 4a which is a cholecystic- enteric fistula without gallstone ileus and type 4b which refers to a cholecystic-enteric fistula with gallstone ileus(11).

Nagakawa also classified Mirizzi's syndrome into a type 1 which was external compression of the bile duct, type 2 which was a cholecystic biliary fistula, type 3 which was gallstone in the cystic duct and common hepatic duct confluence and type 4 which was stricture without stones. Paya Lorente proposed a modified classification which was divided into type 1 which was external bile duct compression, type 2 which was a cholecystic biliary- fistula involving less than 50% of the bile duct diameter and type 3 which was a cholecystic- biliary fistula involving more than 50% of the bile duct diameter .The classification of Mirizzi's syndrome is important when deciding on what management options that may be required and if the patient will need to be referred to a specialized unit(12–14).

Table □ : Table showing the various classifications for Mirizzi's syndrome

Classification	McSherry	Csendes	Beltran	Paya-Llorente	Nagakawa

Type	□-External compression of the bile duct	□-external compression of the bile duct	□-external compression of the bile duct	□-external compression of the bile duct	□-external compression of the bile duct
	□-Cholecystic-biliary fistula	□-cholecystic-biliary fistula up to 1/3 of the bile duct wall erosion	□a-cholecystic-biliary fistula <50%of the bile duct diameter	□-cholecystic-enteric fistula <50%of the bile duct diameter	□-cholecystic-biliary fistula
		□-cholecystic-biliary fistula up to 2/3 of the bile duct wall destruction	□b-cholecystic-biliary fistula >50% of the bile duct diameter	□-cholecystic-enteric fistula >50% of the bile duct diameter	□-gallstones in the cystic duct and common hepatic duct confluence
		□-cholecystic-biliary fistula with complete destruction of the bile duct wall	□a-cholecystic-enteric fistula	Further subtypes for cholecystic -enteric fistula A-no fistula, B-fistula without gallstone ileus, C-fistula with gallstone ileus	□-stricture without stones
		□a-cholecystic-enteric fistula □b-cholecystic-enteric fistula with gallstone ileus	□b-cholecystic-enteric fistula with gallstone ileus		

Diagnosis of Mirizzi's syndrome

The ability to diagnose Mirizzi's syndrome preoperatively is important to prevent potential bile duct injuries when performing a cholecystectomy. Ultrasound can detect dilated common bile duct and intrahepatic ducts in addition to the presence of gallstones and inflamed gallbladder.

Ithas a sensitivity of 8% to 57%. Computerized tomography is useful to rule out other causes of obstruction of the biliary system like malignancy and it has a sensitivity of 31% to 50%. Magnetic resonance cholangiopancreatography (MRCP) can detect dilatation of the intrahepatic ducts, narrowing of the common hepatic duct and presence of gallstones. It has a diagnostic accuracy of 50% and a sensitivity of 77% to 100%. Endoscopic retrograde Cholangiopancreatography (ERCP) is considered the gold standard in the diagnosis of Mirizzi's syndrome. It can detect dilatation of the biliary system, compression of the biliary system and the presence of a cholecystic-biliary fistula. In addition to the diagnostic ability, it can treat the underling obstruction by application of a stent. The diagnostic accuracy is 55% to 90% and the sensitivity is 50% to 100%. The main disadvantage of endoscopic retrograde cholangiopancreatography (ERCP) is the risk of perforation, cholangitis and acute pancreatitis that can occur due to the invasive nature of the procedure(15–21).

Management of Mirizzi's syndrome

The management of Mirizzi's syndrome can be divided into endoscopic therapy, which is used to relieve biliary obstruction in patients with acute cholangitis and surgical therapy which would depend on the stage of Mirizzi syndrome. For patients with a Mirizzi type 1 an open or laparoscopic cholecystectomy can be performed, Patients with a type 2 and 3 Mirizzi syndrome, a partial cholecystectomy and removal of the underlying fistula is done followed by closure with a T-tube. For patients with a type 4 Mirizzi syndrome a chole- cystic enteric or a chole - cystohepatic jejunostomy is performed(22–24).When performing a cholecystectomy for a Mirizzi type 1 patient, a fundus first approach is often recommended due to the distorted anatomy. The impacted stone can than be removedand depending on the level of inflammation at the calot's triangle a partial or total cholecystectomy can be performed to limit the risk of injury to the bile duct(25–27).

Patients with type 2 and 3 Mirizzi syndrome are treated with a subtotal cholecystectomy and depending on the extent of the fistula, excision of the fistula followed by closure and placement of a T-tube to allow biliary drainage. A choledochal-plasty may be performed but if repair of the fistula is not possible then a choledochal-duodenostomy or a choledochal-jejunostomy may need to be performed. In some cases, a Roux-en-Y hepaticojejunostomy may be need if reconstruction is not possible(28).

Laparoscopic cholecystectomy has been attempted for patients with type 1 Mirizzi syndrome, but the inflammation at the calots triangle and the contracted nature of the gallbladder make mobilizing the gallbladder difficult. The risk of bile duct injury is also higher and hence the higher conversion rate to an open cholecystectomy(29–32).The use of preoperative investigations in the form of magnetic resonance cholangiopancreatography (MRCP) and

endoscopic retrograde cholangiopancreatography (ERCP) are essential to delineate the anatomy and stage the Mirizzi syndrome when performing a laparoscopic cholecystectomy. Laparoscopic cholecystectomy is often reserved for Type 1 Mirizzi's syndrome(33).

Laparoscopic management has also been attempted for patients with type 2 Mirizzi's syndrome, but this may require experience in performing a cholecystectomy with laparoscopic common bile duct exploration. The experience of placing a T-tube laparoscopically will be required and hence this type of procedure can only be performed in subspecialized centers(34,35).

A systemic review on the safety and feasibility of the laparoscopic approach for the management of Mirizzi's syndrome was conducted by Zhao et al.17 studies with 857 patients who were diagnosed with Mirizzi's syndrome, of which 440 underwent laparoscopic cholecystectomy. The conversion rate was 34.09% and the pre-operative diagnosis of Mirizzi's syndrome was 67.60%. This study concluded that the laparoscopic method for the treatment of Mirizzi's syndrome was safe and feasible for type 1 and 2 if done in the hands of experienced laparoscopic surgeons(36).Another systemic review on laparoscopic treatment of Mirizzi's syndrome was conducted by Antoniou et al.10 studies that included 135 patients of which 124 underwent laparoscopic treatment. The conversion rate was 40%, complication rate was 20% and the reoperation rate was 6%. This study did not recommend laparoscopic treatment for Mirizzi's syndrome as a standard procedure(37).

Table □ : Table showing the conversion rate for patients who underwent laparoscopic cholecystectomy for Mirizzi's syndrome

Study	Number of attempted laparoscopies	Number of completed laparoscopies	Conversion Rate (%)
Shirah et al	49	45	8.16%
Chui Y et al	65	23	64.62%
Kulkarni et al	17	5	70.59%
Paya-Llorente et al	16	6	62.50%

Conclusion

The diagnosis and treatment of Mirizzi's syndrome is a challenge for the general surgeons with most cases being diagnosed in only 50% of cases. Although it is a rare complication of gallstone disease, the intraoperative diagnosis of this condition makes it difficult, as in patients who are

undergoing a laparoscopic cholecystectomy may require a conversion to an open cholecystectomy to prevent injury to the common hepatic and bile ducts. There are various surgical methods that can be performed including the fundus first approach, opening the fundus of the gallbladder and removing the gallstones and performing a subtotal cholecystectomy. Laparoscopic cholecystectomy may be attempted for type 1 Mirizzi's syndrome but there should be a low threshold to convert to an open cholecystectomy if the operation is difficult. Patients with type 3 and 4 Mirizzi's syndrome are better managed in sub-specialized centers with hepato-pancreatic- biliary surgeons. Mirizzi's syndrome should always be borne in mind in patients with long-standing symptoms of biliary colic and pre-operative investigations in the form of computerized tomography and magnetic resonance cholangiopancreatography (MRCP) should be done.

Conflict of interest-there is no conflict of interest

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