

En-Bloc Cholecystectomy and Segmental Colectomy for Mirizzi Syndrome Type Va - Diagnosis and Management of a Rare Condition.

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Abstract

Introduction: Mirizzi syndrome is defined as hepatic duct obstruction by an impacted gallstone in the cystic duct or infundibulum. Classification is based on the degree of obstruction or fistulation between the cystic duct, hepatic duct, duodenum and colon. Diagnosis is challenging and often made intraoperatively. The absence of preoperative diagnosis increases the risk of bile duct injury up to 17%. This highlights the importance of preoperative diagnosis and planning.

Case Description: A 57-year-old female presented with right upper quadrant pain, nausea, fever, tachycardia on a background of gastric bypass for weight loss, with a 2-metre Roux limb. Imaging revealed biliary dilation with gallbladder wall thickening. MRCP revealed biliary obstruction with a large infundibular gallstone obstructing the hepatic duct. Colonoscopy and gastroscopy were performed to rule out gastrointestinal malignancy. Colonoscopy revealed bile stained mucosa confirming a cholecysto-enteric fistula, confirming Mirizzi syndrome type Va. The patient underwent open cholecystectomy, segmental transverse colectomy with primary repair, choledochotomy with removal of bile duct stones and bile duct repair over a T-tube with an uneventful postoperative course. Histology showed benign cholecystitis.

Conclusion: The diagnostic challenge in this patient was the presence of a 2-meter-long Roux limb from her previous gastric bypass, precluding ERCP, a key investigation in the diagnosis of Mirizzi syndrome. Colonoscopy was therefore invaluable in establishing the diagnosis and allowing for appropriate operative planning. This case demonstrates the diagnosis and management of a rare condition in a challenging diagnostic setting and highlights the importance of preoperative diagnosis and careful operative planning. The use of colonoscopy in the diagnosis of this condition presents a novel approach to the investigation of Mirizzi syndrome.

Key Words Hepatobiliary; Surgery; Planning; Mirizzi; Gallstone; Fistula

Introduction

Mirizzi syndrome was first described by Professor Pablo Luis Mirizzi, an Argentinian

surgeon, in 1948. It is defined as the obstruction of the hepatic duct by an impacted stone in the cystic duct or gallbladder infundibulum [1]. It is classified from types I-V (and their subtypes) based on the 2007 classification system by Csendez *et al.*, which was validated in 2008 by Beltran *et al* [2, 3]. Table 1 shows the details of the classification system.

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Table 1: A summary of Mirizzi syndrome based on the 2007 Csendes et al classification. Type I (a and b) describe compression of the hepatic duct by a gallstone in the cystic duct or infundibulum without fistulation between the ducts. Type II-IV describes cholecystochoal fistulae based on the fistula defect size as a proportion of the common hepatic duct. Type V and its subtypes describe Mirizzi syndrome with cholecystoenteric fistulae or gallstone ileus.

Type	Fistula	Description
Ia	No Fistula	Cystic duct not obliterated
Ib		Cystic duct obliterated
II	Cholecystochoal fistula	Fistula defect <33% of common hepatic duct
III		Fistula defect between 33% and 66% of common hepatic duct
IV		Fistula defect >66% of common hepatic duct
Va		Cholecystoduodenal or cholecystocolic fistula
Vb	Cholecystoenteric fistula in addition to Mirizzi I-IV	Combined cholecystoduodenal and cholecystocolic fistula
Vc		Presence of gallstone ileus

The diagnosis of Mirizzi syndrome under normal circumstances is challenging and often is made intraoperatively [1]. The absence of a preoperative diagnosis in these patients increases the risk of bile duct injury, possibly up to 17% [4]. This highlights the importance of making the diagnosis prior to proceeding to surgery.

Multiple imaging modalities are available to the clinician in the diagnosis of Mirizzi Syndrome. Beltran presents an evidence-based summary of these modalities in a paper published in 2012 as a review of current knowledge about Mirizzi syndrome [1]. To summarise, ultrasonography is useful in delineating biliary anatomy and

has a reported diagnostic accuracy of 29%. Computed Tomography (CT) findings are nonspecific but can be useful in excluding liver or porta hepatis malignancies. Magnetic Resonance Cholangiopancreatography (MRCP) is useful in showing hepatic duct obstruction or fistulation and to differentiate causes of biliary obstruction and has a diagnostic accuracy of ~50% for Mirizzi syndrome. Endoscopic retrograde cholangiopancreatography (ERCP) is a useful diagnostic and therapeutic tool in the diagnosis and management of Mirizzi syndrome, with a diagnostic accuracy of 55% to 90%.

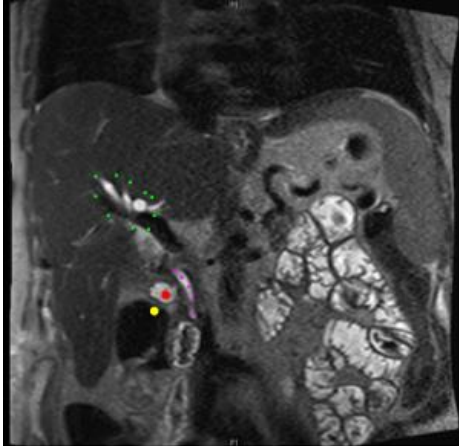


Figure 1: Coronal MR image showing a large stone impacted in the gallbladder neck (Red) in close proximity to the common bile duct (Pink) and hepatic flexure of the colon (Yellow). The green dots encircle a dilated intrahepatic biliary tree.

The incidence of complications of gallstones, including Mirizzi syndrome, are less than 1% in the developed world [5]. Mirizzi type V can be present in up to 29% of all other subtypes of Mirizzi syndrome [4]. Overall, this makes it a rare condition. In searching the MEDLINE, OVID and PMC databases, as well as gray literature, we found four case reports describing Mirizzi type Va and its management [6, 7, 8, 9].

Case Report

A 57-year-old female presented to the emergency department for 5 hours of worsening right upper quadrant pain, nausea and rigors. Past medical history included a Roux en Y gastric bypass one year prior with a 2-metre-long Roux limb, resulting in 100kg weight loss. There was no known history of gallbladder disease. She also suffered from type two diabetes and recently diagnosed atrial fibrillation for which she was anticoagulated.

On examination the patient was distressed, febrile and tachycardic. She had right upper quadrant abdominal tenderness and was positive for Murphy's sign.

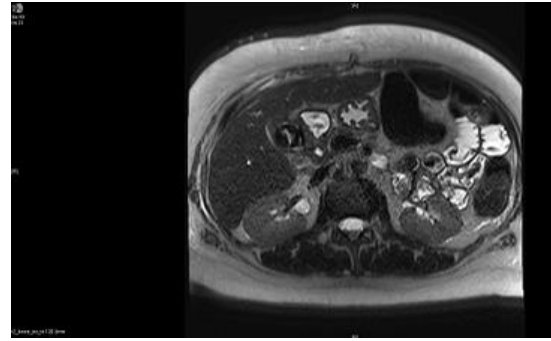


Figure 2: Axial MR image showing a large obstructing infundibular stone and gallbladder wall thickening

Initial investigation revealed a bilirubin of 48 umol/L, Gamma-glutamyl transferase 923U/L, Aspartate aminotransferase 961 U/L, Lipase 27 U/L. White cell count was $11.4 \times 10^9/L$ and C-reactive protein 18mg/L.

Computed tomography (CT) and ultrasound imaging revealed multiple gallstones with dilation of the intrahepatic and extrahepatic biliary tree proximal to the gallbladder and significant gallbladder wall thickening and heterogeneity.

The decision was made to proceed to open cholecystectomy. Preoperatively the patient was treated with intravenous piperacillin and tazobactam in lieu of intravenous fluid resuscitation with normal saline and analgesia. Intraoperatively a hard mass was found centered at the gallbladder. The stomach and colon were adherent to the mass. This raised the suspicion for malignancy. The procedure was abandoned to allow for further investigation and operative planning.

The patient was further investigated with MRCP to better delineate biliary anatomy. She also underwent colonoscopy and gastroscopy to confirm the presence of a cholecystoenteric fistula and rule out a gastrointestinal primary malignancy. MRI revealed a large stone at the gallbladder neck causing proximal biliary dilation with close relation of the colon to the gallbladder neck (Figure 1, 2, and 3). Gastroscopy was

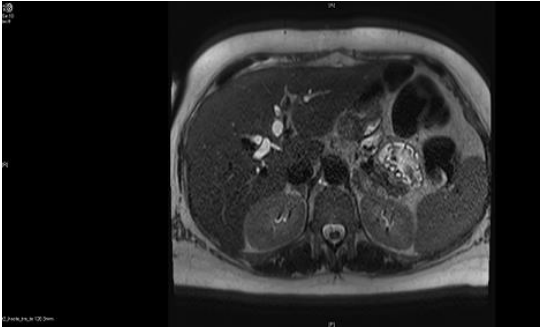


Figure 3 Axial MR image showing marked intrahepatic biliary dilation

unremarkable. Colonoscopy revealed bile within the ascending and transverse colon and was otherwise unremarkable (Fig 4). This confirmed a fistula between the gallbladder and colon. The diagnosis of Mirizzi syndrome type Va was hence established.

The patient proceeded to have an open cholecystectomy via a Kocher incision, segmental transverse colon resection with primary repair, choledochotomy with removal of common bile duct stones and repair of the common bile duct over a T-tube. The aforementioned intravenous antibiotics were continued for another 5 days and then changed to oral amoxicillin and clavulanate for a further week.

The patient had an uneventful postoperative course. Histology reported benign inflammatory tissue in keeping with cholecystitis.

Discussion

Mirizzi syndrome poses a significant clinical and diagnostic challenge, with a remarkable impact on patient outcomes based on the timing of diagnosis. The diagnostic challenge in this patient was further heightened due to the presence of a 2-meter-long Roux limb from her previous gastric bypass. This precluded ERCP, which is one of the key diagnostic and therapeutic tools for this condition. Colonoscopy was therefore invaluable in establishing the

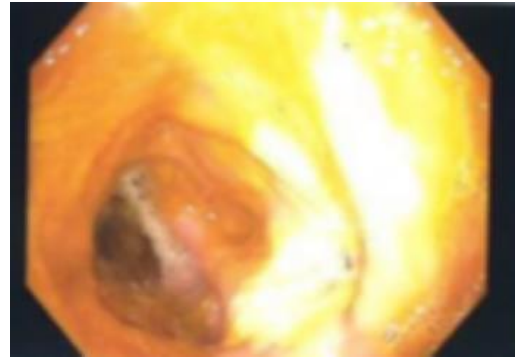


Figure 4: Colonoscopic imaging of the descending colon showing bile stained mucosa.

diagnosis and allowing for appropriate operative planning.

As mentioned in the introduction, four case reports of Mirizzi syndrome type V were found in our literature search. Of these, three described cholecystocolic fistulae and one described a cholecystoduodenal fistula. None of the patients underwent colonoscopy as part of the diagnostic process, nor is colonoscopy described as part of the routine investigation for this condition. Additionally, we found no reports describing the investigation and management of this condition in individuals with prior proximal gastrointestinal surgery that precludes ERCP.

Conclusions

This case demonstrates the diagnosis and management of a rare condition in a challenging diagnostic setting, and highlights the importance of preoperative diagnosis and careful operative planning. The use of colonoscopy in the diagnosis of this condition presents a novel approach to the investigation and operative planning for cholecystectomy in patients with suspected type V Mirizzi syndrome.

Learning Points

1. Familiarising with the definition and classification of Mirizzi syndrome

2. Understanding the diagnostic process and tools available for the diagnosis of this rare condition
3. Highlighting the importance of accurate diagnosis and preoperative planning
4. Use of a common diagnostic tool, colonoscopy, in a novel way not previously described for Mirizzi syndrome

Conflicts of Interest

None to declare

Authors' contributions

SMJT: Literature review, case data review, writing of the body of the manuscript, case submission and editing to fit journal criteria

YF: Patient care, diagnosis and clinical decision making. Performing surgery. Review of the manuscript and recommendations for editing. Clinical and theoretical guidance.

MP: liaising interstate and accessing further imaging and patient records to complete this manuscript after editor's comments were made

Consent

Written patient consent was obtained prior to writing this case report for use of de-identified case information and images

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No further

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