Journal of Community Hospital Internal Medicine Perspectives

Volume 12 | Issue 6 Article 12

2022

Mirizzi Syndrome: An Unusual Complication of Cholelithiasis

Shruti Jesani

Ana Lucia Romero
Trinitas Regional Medical Center Williamson Street Campus: Trinitas Regional Medical Center, luciaromero_182@hotmail.com

Subutay Berke Bozkurt

Abdel Azez Abu Samak

Jesus Romero

See next page for additional authors

Recommended Citation

Jesani, Shruti; Romero, Ana Lucia; Bozkurt, Subutay Berke; Samak, Abdel Azez Abu; Romero, Jesus; Sookdeo, Jonathan; and Naik, Arun (2022) "Mirizzi Syndrome: An Unusual Complication of Cholelithiasis," *Journal of Community Hospital Internal Medicine Perspectives*: Vol. 12: Iss. 6, Article 12.
DOI: 10.55729/2000-9666.1114
Available at: https://scholarlycommons.gbmc.org/jchimp/vol12/iss6/12

Mirizzi Syndrome: An Unusual Complication of Cholelithiasis

Authors

Shruti Jesani, Ana Lucia Romero, Subutay Berke Bozkurt, Abdel Azez Abu Samak, Jesus Romero, Jonathan Sookdeo, and Arun Naik

CASE REPORT

Mirizzi Syndrome: An Unusual Complication of Cholelithiasis

Shruti Jesani [a], Ana L. Romero [a,*], Subutay B. Bozkurt [a], Abdel A. Abu Samak [a], Jesus Romero [a], Jonathan Sookdeo [b], Arun Naik [a]

[a] RWJB Health/Trinitas Regional Medical Center, United States
[b] St. Georges University School of Medicine, Grenada

Abstract

Mirizzi syndrome is a rare condition caused by the obstruction of the common bile duct or common hepatic duct by external compression from multiple impacted gallstones or a single large impacted gallstone in Hartman's pouch. The condition can easily be confused with choledocholithiasis, bile duct stricture or cholangiocarcinoma due to the presence of obstructive jaundice hence may be overlooked due to the rarity of the condition. The incidence of Mirizzi syndrome among patients with gallstones is reported to range from 0.63 to 5.7%. Furthermore, it poses a differential diagnosis dilemma for the physician as well as radiologists because there are no clinical features or diagnostic procedures that have a 100% specificity and sensitivity. Laparotomy is the preferred surgical technique of choice. For the patients who are poor surgical candidate, mainstay of treatment is biliary stent placement for the restoration of normal biliary drainage. Due to low incidence of the Mirizzi syndrome, an elevated index of suspicion is required to diagnose this condition. At present, there are no well-developed, internationally recognized clinical guidelines for the management of this syndrome. Furthermore, the diagnostic procedures available still pose a barrier in the ability to confirm the diagnosis prior to surgical treatment, even though the diagnostic rate has increased dramatically.

1. Introduction

Mirizzi syndrome is a rare complication of chronic cholelithiasis within the neck or infundibulum of the gallbladder or cystic duct that causes extrinsic compression of the common bile duct (CBD) or common hepatic duct (CHD). The typical presentation is RUQ abdominal pain, jaundice, and fever. Management includes endoscopic retrograde cholangiopancreatography (ERCP) for biliary decompression followed by laparoscopic cholecystectomy or an open approach depending on the classification. We presented an 83-year-old male with a past medical history of seizure disorder and contracture found to have Mirizzi syndrome that was successfully managed by percutaneous trans-hepatic biliary drainage. ERCP and cholecystectomy could not be performed due to the patient's unfavorable gastrointestinal tract anatomy and poor surgical candidate status, respectively (see Fig. 1).

2. Case presentation

83-year-old male with a past medical history of seizure disorder resulting in contractures, presented to the hospital after being found to have elevated liver enzymes during a routine laboratory test performed at the nursing home. On presentation, the patient was asymptomatic, afebrile, and hemodynamically stable. On physical examination, he was jaundiced with icteric sclera. Pertinent labs include normal white blood cells, aspartate aminotransferase 77 U/L, alanine aminotransferase 91 U/L, alkaline phosphatase 430 U/L, total bilirubin 8 mg/dL direct bilirubin 5 mg/dL and an INR of 1. Right upper quadrant ultrasound was remarkable for a large gallstone with a contracted gallbladder and intrahepatic biliary dilatation. Abdominal CT with contrast revealed a 12 mm gallstone at the gallbladder neck with a contracted gallbladder, causing extrinsic compression of the common hepatic duct (CHD) with moderate intrahepatic bile duct

Received 24 June 2022; accepted 27 July 2022.
Available online 7 November 2022

* Corresponding author at:
E-mail address: luciaromero_182@hotmail.com (A.L. Romero).

https://doi.org/10.55729/2000-9666.1114

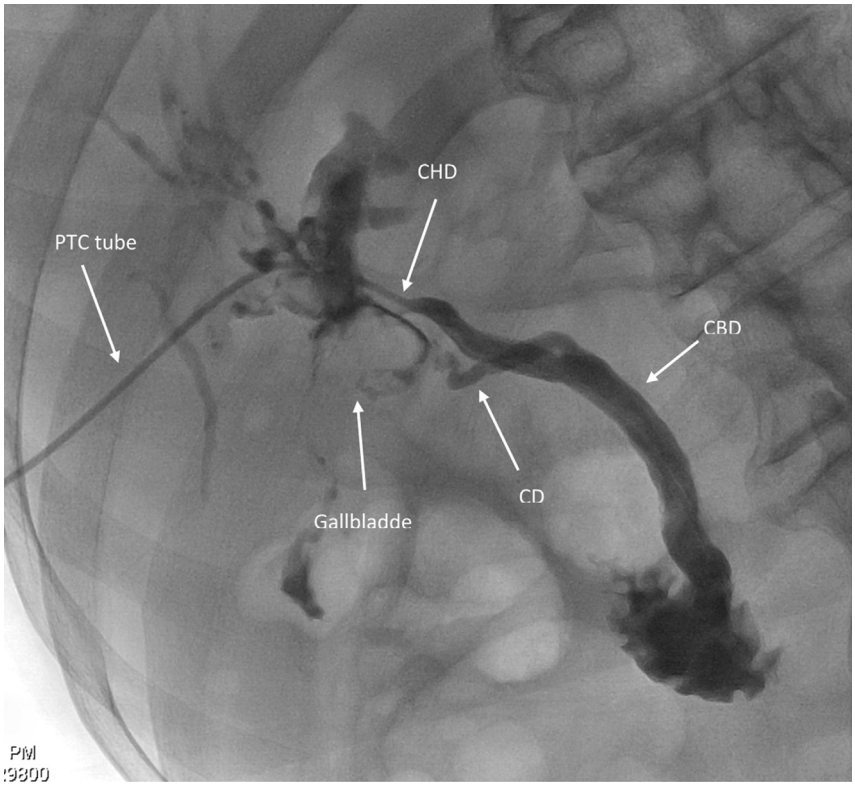

Fig. 1. Fluoroscopic image illustrating common hepatic duct obstruction and narrowing by external compression from gallbladder. PTC (Percutaneous transhepatic cholangiography) tube, CHD (common hepatic duct), CBD (common bile duct), CD (cystic duct).

dilatation, suspicious for MS. An ERCP was attempted with the hope of retrieving the gallstone. Attributed to his unfavorable gastrointestinal tract anatomy, the ampulla could not be localized, and the CHD could not be cannulated. Pre-operational evaluation for possible cholecystectomy concluded that the patient was a poor surgical candidate due to age and the contractures. Hence, he underwent percutaneous transhepatic cholangiogram which confirmed the diagnosis of MS, as the large gallstone caused a mass effect on the common hepatic duct. Internal/external biliary drainage catheter was placed by interventional radiology which was eventually replaced by a metallic stent placed across the narrowed portion of CHD, after which his elevated liver enzymes down trended. He was discharged back to the nursing home.

3. Discussion

Mirizzi syndrome is a product of chronic cholelithiasis, where the impaction of a single or multiple stones within the neck or infundibulum (Hartmann's pouch) of the gallbladder or cystic duct produces extrinsic compression of other biliary structures such as the common hepatic and common bile duct. Over time, inflammation and biliary stasis due to obstruction cause erosion of the gallbladder and cystic duct walls, which may lead to the formation of cholecystohepatic or cholecystocholedochal fistulas. In rare cases, some may present with cholecystoenteric fistulas.[1] Even though the most widely used classification system in literature (Csendes) categorizes compression of the common bile duct as a type I form of Mirizzi syndrome, the incidence of the condition is rare, occurring in only 1–4% of patients undergoing cholecystectomy, and in 0.05–4% of patients undergoing surgery for cholelithiasis.[2]

Common clinical manifestations of the disease include right upper quadrant pain, signs of obstructive jaundice, and acute cholangitis.[1,3,4] All three symptoms however are only present in about 44–71% of patients.[4] Other symptoms included nausea, vomiting, and anorexia.[3,5] One study reported that 50% of patients exhibited a positive Murphy's sign.[3] They can also present with acute cholecystitis, pancreatitis, and choledocholithiasis.[5] Between 3.7 and 17% of patients are asymptomatic.[3] Diagnostic findings are non-sensitive and non-specific, and a high index of suspicion is needed when arriving at a diagnosis. Elevated bilirubin and alkaline phosphatase are the most common findings, seen in more than 90% of

patients. Leukocytosis is seen in patients with accompanying cholangitis or cholecystitis. Other findings include elevated liver function enzymes and gamma glutamyl transferase. There has been a substantial association between Mirizzi syndrome and gallbladder carcinoma, with an incidence among patients with Mirizzi syndrome as high as 28%.[1,6] Few case reports have also shown elevated CA19-9 in the absence of malignancy. Imaging is almost always needed to arrive at a preoperative diagnosis.[1,5]

The gold standard for diagnosis is ERCP with a sensitivity ranging from 76 to 100%.[7] It offers the best visualization of the extra-hepatic bile ducts and can show compression by impacted gallstones in the common bile duct with resulting proximal biliary dilatation as was the case in our patient. Furthermore, ERCP can accurately determine the presence of fistulas. Therapeutic options such as stenting, nasobiliary drainage, or stone removal may also be performed during the procedure, helping to relieve obstruction preoperatively.[8] One study has demonstrated that pre-op stenting also aids in identifying the common bile duct during surgery in patients with abnormal Triangle of Calot and biliary tree anatomy.[3] Complications may arise from ERCP which include infection and (1.4%) and perforation (0.6%).[9] Other common modalities include ultrasound, CT, and MRCP. Abdominal ultrasound may demonstrate dilation above the level of the gallbladder neck with the presence of an impacted stone, accompanied by changes to the width of the common bile duct. Current literature reports a diagnostic accuracy of 29%, with a sensitivity between 8.3% and 27%. Some studies have shown sensitivities as high as 72%. Ultrasound however is unable to distinguish whether the cause of bile duct narrowing is due to metastatic invasion or gallstone compression.[7] The sensitivity and specificity of abdominal CT scans vary widely between 42 and 99%. Its ability to distinguish Mirizzi syndrome from hepatic infiltration of metastatic tumors, particularly in patients with fistulas, is a major strength. Lastly, MRCP demonstrates sensitivity across studies at about 50%, however, it is limited due to its inefficiency in localizing fistulas.[7] If ERCP is not an option, using multiple imaging modalities can greatly increase the chance of preoperative diagnosis. One study showed that MRCP combined with CT yielded a diagnostic accuracy as high as 98% vs. 84% for CT alone.[10]

At present, Mirizzi syndrome is managed without well-developed, internationally recognized clinical guidelines. Furthermore, advancement in diagnostic techniques has not made it easier for a confirmed diagnosis to be made before surgery, even though diagnostic rates have improved markedly.[7] Laparotomy is usually chosen as the mode of treatment. This usually involves total or partial cholecystectomy of the fundus and neck with repair of compromised biliary structures.[1,5,7] Due to the association with gallbladder carcinoma, all excised tissue should undergo pathologic examination to rule out unsuspected carcinoma. The insertion of a T-tube or nasobiliary decompression is usually done pre and postoperatively to prevent further biliary leaks, strictures, or the development of cholangitis.[5] While the laparoscopic approach was avoided in the past due to high variability in the hepatobiliary anatomy seen in these patients leading to bile duct injury and high rates of conversion to open laparotomy, newer evidence suggests that the laparoscopic approach after ERCP identification and therapeutic management is a reliable option in patients with a pre-operative diagnosis.[7,8,11]

For patients who are poor medical candidates for surgery, treatment presents a medical conundrum that revolves around alleviating the acute symptoms at presentation and maintaining long-term patency hepatobiliary tree. For patients with type I Mirizzi syndrome, such treatment involves therapeutic ERCP for sphincterotomy and stone extraction via basket or balloon, dissolution therapy, or lithotripsy.[12] Percutaneous stone removal is usually reserved for patients who have failed endoscopic access and who are unsuitable surgical candidates. Stone removal tends to provide significant relief and is usually done after decompression via biliary drainage either from transhepatic percutaneous drainage or endoscopic nasobiliary tube.[8,12,13] Coupled with antibiotic therapy and decompression, endoscopic therapies can resolve initial presenting symptoms of cholangitis and obstructive jaundice. While the removal of impacted stones is desirable in non-surgical endoscopic therapy, there are cases where complete stone removal cannot be achieved, either due to anatomical issues or to the stone being impacted above the cystic duct, such as in Hartmann's pouch. In these cases, biliary stenting may provide a long-term option for relief.[12,13] One case report where the authors used a biliary stent in an elderly patient whose stone was left in situ demonstrated no complications at 2–5 years follow-up.[14] In a 2006 study, 6 patients who were determined to be poor surgical candidates underwent definitive endoscopic treatment. Outcomes showed low post-procedure morbidity, with only one patient dying due to stent migration leading to duodenal perforation. The rest of the patients were doing well upon follow-up which ranged from 1 to 7 months.[13] The most common complication of endoscopic

biliary stenting as a long-term option is recurrent cholangitis. Due to the risk of complications that may occur post-procedure, the use of endoscopic therapies as the definitive treatment for patients with Type I Mirizzi syndrome should be reserved for patients with severe comorbidities, poor surgical candidates, or short life expectancies.[12] These patients should also be followed up closely to monitor for any complications that may arise, with stents being evaluated and replaced as needed to maintain adequate biliary patency. Nevertheless, this treatment option allows for successful medical management in patients deemed poor candidates for surgery and may be used in this population as a potential long-term solution.

4. Conclusion

Gallstones are a prevalent medical condition that might result in long-term complications like Mirizzi syndrome. Given that the clinical and biochemical manifestations of MS resemble other medical disorders, establishing pre-operative diagnosis requires a high index of suspicion. This syndrome can be managed by percutaneous trans-hepatic biliary drainage when surgical intervention and ERCP are deemed to be unfeasible. Although such management carries risks of re-obstruction and/or infection, relieving the obstruction in the acute settings in such patient is deemed necessary to prevent morbidity or mortality.

Conflicts of interest

All authors declare that they have no conflicts of interest.

References

1. Beltrán MA. Mirizzi syndrome: history, current knowledge and proposal of a simplified classification. *World J Gastroenterol*. 2012;18(34):4639–4650. https://doi.org/10.3748/wjg.v18.i34.4639.
2. Kamalesh NP, Prakash K, Pramil K, George TD, Sylesh A, Shaji P. Laparoscopic approach is safe and effective in the management of Mirizzi syndrome. *J Minimal Access Surg*. 2015;11(4):246–250. https://doi.org/10.4103/0972-9941.140216.
3. Klekowski J, Piekarska A, Góral M, Kozula M, Chabowski M. The current approach to the diagnosis and classification of Mirizzi syndrome. *Diagnostics*. 2021;11(9):1660. https://doi.org/10.3390/diagnostics11091660.
4. Ibrarullah M, Mishra T, Das AP. Mirizzi syndrome. *Indian J Surg*. 2008;70(6):281–287. https://doi.org/10.1007/s12262-008-0084-y.
5. Valderrama-Treviño AI, Granados-Romero JJ, Espejel-Deloiza M, et al. Updates in Mirizzi syndrome. *Hepatobiliary Surg Nutr*. 2017;6(3):170–178. https://doi.org/10.21037/hbsn.2016.11.01.
6. Prasad TL, Kumar A, Sikora SS, Saxena R, Kapoor VK. Mirizzi syndrome and gallbladder cancer. *J Hepato-Biliary-Pancreatic Surg*. 2006;13(4):323–326. https://doi.org/10.1007/s00534-005-1072-2.
7. Chen H, Siwo EA, Khu M, Tian Y. Current trends in the management of Mirizzi Syndrome: a review of literature. *Medicine*. 2018;97(4), e9691. https://doi.org/10.1097/MD.0000000000009691.
8. Han W, Yue Q, Liu K, Ke JJ, Meng LY, Liu YH. Endoscopic nasogallbladder drainage combined with laparoscopic surgery for type I Mirizzi syndrome with acute cholecystitis: a Case series report. *Gastroenterol Res Pract*. 2020:2417539. https://doi.org/10.1155/2020/2417539.
9. Andriulli A, Loperfido S, Napolitano G, et al. Incidence rates of post-ERCP complications: a systematic survey of prospective studies. The. *Am J Gastroenterol*. 2007;102(8):1781–1788. https://doi.org/10.1111/j.1572-0241.2007.01279.x.
10. Yun EJ, Choi CS, Yoon DY, et al. Combination of magnetic resonance cholangiopancreatography and computed tomography for preoperative diagnosis of the Mirizzi syndrome. *J Comput Assist Tomogr*. 2009;33(4):636–640. https://doi.org/10.1097/RCT.0b013e31817710d5.
11. Senra F, Navaratne L, Acosta A, Martínez-Isla A. Laparoscopic management of type II Mirizzi syndrome. *Surg Endosc*. 2020;34(5):2303–2312. https://doi.org/10.1007/s00464-019-07316-6.
12. England RE, Martin DF. Endoscopic management of Mirizzi's syndrome. *Gut*. 1997;40(2):272–276. https://doi.org/10.1136/gut.40.2.272.
13. Gomez D, Rahman SH, Toogood GJ, et al. Mirizzi's syndrome–results from a large western experience. *HPB : Off J Int Hepato Pancr Bil Assoc*. 2006;8(6):474–479. https://doi.org/10.1080/13651820600840082.
14. Adam A, Roddie ME, Benjamin IS. Case report: Mirizzi syndrome–treatment with metallic endoprosthesis. *Clin Radiol*. 1993;48(3):198–201. https://doi.org/10.1016/s0009-9260(05)80283-1.