Mirizzi Syndrome: A Real Pain in the "Neck"

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Learning Objectives

1. Mirizzi syndrome is a rare biliary phenomenon whereby the hepatic duct becomes obstructed via extrinsic compression from an impacted stone in the neck of the gallbladder.
2. Mirizzi syndrome has been associated with an increased risk of gallbladder cancer and occurs in 0.05 to 4 percent of patients undergoing surgery for cholelithiasis.
3. In this case, we describe a patient who presented with cholangitic symptoms and MRCP findings concerning for Mirizzi Syndrome type 1a.

Patient Presentation

A 49-year-old female with a past medical history of colorectal carcinoma status post chemotherapy and radiation, HTN, HLD, obesity presented to the ED with complaints of fever, jaundice and right upper quadrant pain. Of note, patient was admitted two months prior for similar presentation and was found to have an obstructing common bile duct stone treated with ERCP, balloon extraction and sphincterotomy. She was discharged in stable condition but was soon readmitted due to intractable nausea, vomiting, and was found to have a mechanical large bowel obstruction secondary her known colonic mass. She underwent diverting loop colostomy, was subsequently cleared by surgery, and discharged in stable condition. The patient now presents with fever, jaundice, and RUQ pain.

Physical Exam: Vitals: Fever 101.4 F, Tachycardia HR 105

General: Uncomfortable appearing, frail

Eyes: Icteric conjunctivae, pallor

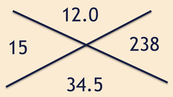
Skin: Jaundice of face and trunk

CVS: Tachycardia, normal S1/S2, no murmurs

Resp: Coarse breath sounds b/l

GI: Soft, mildly tender in RUQ, diminished bowel sounds

Lab Values

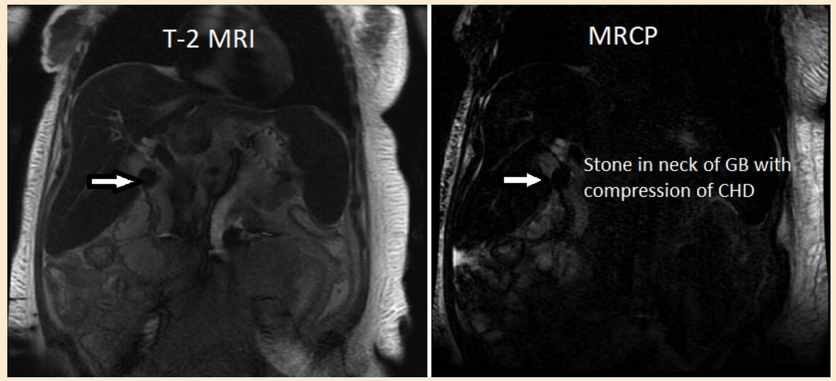


Tbili: 6.7
Dbili: 5.3
ALP: 1,553
AST/ALT: 125/108

RUQ US and MRCP Findings

RUQ ultrasound showed a contracted gallbladder with CBD measuring 14.5mm. **MRCP** showed a large stone impacted in the gallbladder neck with external compression of the common hepatic duct.

MRI/MRCP



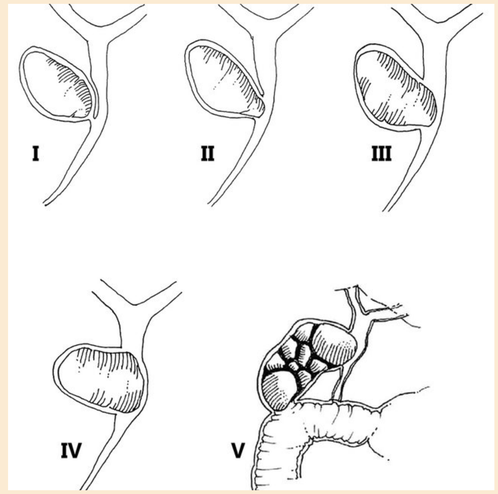
Hospital Course

- Patient was seen by Surgery and Gastroenterology who advised against surgery or endoscopic intervention given the patient's poor oncologic prognosis.
- The decision was made to undergo percutaneous cholecystostomy for biliary decompression.
- The patient's symptoms worsened, and her liver function tests continued to uptrend.
- The patient's family ultimately decided to pursue a Palliative Care and Hospice consultation to discuss goals of care.
- Given the severity of symptoms and overall poor prognosis, the patient was ultimately readmitted to inpatient hospice and passed away soon thereafter.

Mirizzi Syndrome Classification

- **Type Ia**: External compression of the common hepatic duct due to a stone impacted at the neck/infundibulum of the gallbladder or at the cystic duct (11% prevalence)
- **Type Ib**: External compression of the common hepatic duct but cystic duct is absent.
- **Type II**: Presence of cholecystobiliary fistula involves less than one-third of the circumference of the common bile duct (41%).
- **Type III**: Fistula involves between one-third and two-thirds of the circumference of the common bile duct (44%).
- **Type IV**: Destruction of the entire wall of the common bile duct (4%).
- **Type Va**: Any above type + cholecystoenteric fistula, without ileus
- **Type Vb**: Any above type + cholecystoenteric fistula, with ileus

Csendes Classification



Take Home Points

- Given the degree of obstruction and likelihood of cholangitis progression, Mirizzi syndrome must be treated as a surgical emergency.
- The mainstay of treatment involves surgical resection which permits direct removal of causal factors: both the gallbladder and impacted stone.
- If fistula is present, fistula closure is achieved with suture repair, T tube placement or choledochoplasty with remnant GB
- Type III and IV typically require bilioenteric anastomosis, either choledochoduodenostomy or choledochojejunostomy
- Mirizzi syndrome carries a high mortality rate and should always be suspected in patients presenting with fever, jaundice, and right upper quadrant pain.

References

1. Jones MW, Ferguson T. Mirizzi Syndrome. [Updated 2022 Apr 13]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK482491/
2. Mirizzi syndrome. (n.d.). *SpringerReference*. https://doi.org/10.1007/springerreference_136673
3. Umasjanker R, Smink, D. Mirizzi Syndrome. UpToDate. June 2021.
4. Veltchev, L. (2010). Mirizzi syndrome-rare cause of major biliary complications. /case Report/. *Journal of IMAB - Annual Proceeding (Scientific Papers), 15, book 1*(2009), 92–94. https://doi.org/10.5272/jimab.1512009_92