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An Elderly Woman with Abdominal Pain: Mirizzi Syndrome

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Conflict of interest: None declared

Patient: Female, 80
Final Diagnosis: Mirizzi syndrome
Symptoms: Abdominal pain • fever
Medication: —
Clinical Procedure: Percutaneous cholecystectomy tube
Specialty: Gastroenterology and Hepatology

Objective: Rare disease

Background: Mirizzi syndrome is an uncommon but clinically important complication of gallbladder disease that occurs when there is extrinsic compression of the common hepatic duct from gallstones within the cystic duct or from within the gallbladder itself. Obstructive jaundice and cholangitis may ensue. In severe cases, bile duct erosion or gallbladder rupture occur.

Case Report: A demented 80-year-old woman presented to the Emergency Department (ED) with fever and right upper-quadrant abdominal guarding and tenderness. Computed tomography of the abdomen revealed a markedly dilated and thickened gallbladder with hyperdensity in the region of the gallbladder neck. The mass effect of these gallstones caused central intrahepatic biliary ductal dilatation from extrinsic compression of the extrahepatic biliary duct, consistent with Mirizzi syndrome. Additionally, there were 2 areas of focal rupture of the gallbladder wall. General Surgery recommended non-operative management and temporizing the patient with a cholecystostomy tube. She remained in the hospital on IV antibiotics and discharged to follow-up as an outpatient with General Surgery.

Conclusions: Significant morbidity and mortality can be associated with the disease states of Mirizzi syndrome, and it is imperative for the ED physician to promptly recognize and treat such clinical entities. In general, treatment requires a multidisciplinary approach, using the history and physical examination to guide appropriate consultation with General Surgery, Gastroenterology, or Interventional Radiology. The prognosis of Mirizzi syndrome is related to the degree of concomitant complications. Aggressive treatment is appropriate for most patients, with surgical intervention being individualized based on the stage and severity of the disease.

MeSH Keywords: Abdominal Pain • Gallbladder Diseases • Gastroenterology • Mirizzi Syndrome

Full-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/914642>

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Background

Mirizzi syndrome is an uncommon but clinically important complication of gallbladder disease that should be recognized promptly by the treating physician. In this condition, there is extrinsic compression of the common hepatic duct from gallstones within the cystic duct or from within the gallbladder itself. Obstructive jaundice and cholangitis may ensue. Differentiating this condition from other hepatobiliary pathology is essential to rule out potential etiologies such as cholangiocarcinoma. In severe cases, Mirizzi syndrome may be associated with bile duct erosion or gallbladder rupture. We present a case of an elderly woman who presented with right-sided abdominal pain and was found to have Mirizzi syndrome with gallbladder rupture. The interventions and multidisciplinary approach needed to care for such a patient with this rare condition are outlined in this case presentation.

Case Report

A severely demented 80-year-old woman presented to the Emergency Department (ED) after an unwitnessed fall from her bed the previous day. She was a resident at a long-term nursing facility. The staff at that facility reported the patient was experiencing right-sided abdominal discomfort, and they were concerned about a possible musculoskeletal or abdominal injury from the fall. The patient was unable to provide additional history secondary to her Alzheimer's dementia. A physical examination showed significant abdominal tenderness in the right upper quadrant with associated guarding. The patient had no focal bony tenderness, scalp hematoma, or other soft-tissue swelling to suggest sequelae from a fall.

Initial vitals were normal, without tachycardia or hypotension. Approximately 2 h into her ED course, however, a fever of 39.2°C was noted. Laboratory evaluation was significant for a leukocytosis to $23.3 \times 10^9/L$, urinalysis with large leukocyte esterase and 10^9 WBCs, and normal liver function tests with total bilirubin of 1.1 mg/dL, direct bilirubin of 0.4 mg/dL, ALT of 30 U/L, AST of 50 U/L, and alkaline phosphatase of 131 U/L. Computed tomography (CT) of the abdomen revealed a markedly dilated and thickened gallbladder with hyperdensity in the region of the gallbladder neck. The hyperdensity appeared consistent with gallstones, but the Radiologist also indicated that cholangiocarcinoma could not be excluded. The mass effect of these gallstones appeared to be causing central intrahepatic biliary ductal dilatation from extrinsic compression of the extrahepatic biliary duct. Also noted was an apparent cut-off of the common bile duct at its mid-level. Finally, there were 2 areas of focal disruption of the gallbladder wall with contained extraluminal contents in the adjacent hepatic parenchyma (Figures 1, 2). There was no evidence of pneumoperitoneum

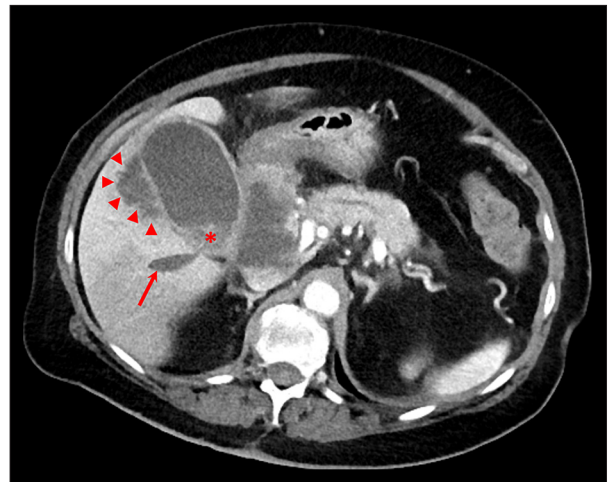


Figure 1. Axial computed tomography revealing intrahepatic biliary ductal dilation (arrow), focal contained gallbladder rupture (arrowheads), and stones/debris within gallbladder neck (asterisk).



Figure 2. Coronal computed tomography revealing intrahepatic biliary ductal dilation (arrow) and focal contained gallbladder rupture (arrowheads).

or free fluid. Patient was noted to have bilateral parapelvic renal cysts but no other masses or lesions.

Given the patient's normal bilirubin and alkaline phosphatase levels, Mirizzi syndrome was favored over hepatobiliary malignancy. Furthermore, trended labs during the hospitalization

revealed total bilirubin declining to 0.5 mg/dL, normal AST/ALT, and alkaline phosphatase levels consistently in the 120 U/L range. Given that CT imaging was consistent with severe perforated cholecystitis, broad-spectrum antibiotic coverage was initiated. The patient remained hemodynamically stable throughout her ED course. She was admitted to the Internal Medicine service who consulted the General Surgery service. Given her age and comorbidities, General Surgery recommended non-operative management and temporizing the patient with a cholecystostomy tube. On hospital day 2, Interventional Radiology placed a cholecystostomy tube and aspirated purulent material from the gallbladder during the procedure, which was sent for culture.

The patient's course was complicated by multi-drug resistant *E. coli* isolated from her gallbladder aspirate. As cultures and sensitivities became available, antibiotic coverage was modified to appropriately manage the patient's infection. She remained in the hospital on IV antibiotics until hospital day 7, at which time she was discharged in good condition with instructions to follow-up as an outpatient with General Surgery. At 2-week follow-up, after a discussion about the risks and benefits of surgery, the patient's family did not want to pursue any additional aggressive surgical interventions.

Discussion

Mirizzi syndrome is an uncommon but clinically important complication of gallbladder disease. Extrinsic compression of the common hepatic duct from gallstones or debris within the cystic duct or from within the gallbladder itself causes intrahepatic ductal dilatation. Obstructive jaundice and cholangitis may ensue. Significant morbidity and mortality can be associated with these disease states, and it is imperative for the ED physician to promptly recognize and treat such clinical entities. This condition must be differentiated from other hepatobiliary pathology to rule out potential malignant etiologies such as cholangiocarcinoma. In severe or long-standing cases, Mirizzi syndrome may be associated with bile duct erosion or gallbladder rupture [1]. Our patient developed a severe complication of Mirizzi Syndrome with gallbladder rupture. Interestingly, the patient in this case did not develop hyperbilirubinemia or transaminitis. In a series of 64 Mirizzi syndrome patients, the bilirubin was elevated in only 29.7% of patients, and the liver enzymes were elevated in only 37.5% (AST) and 39.1% (ALT) of individuals [2]. We surmise that the lack of rise of bilirubin in our case may be reflective of the acuity of the obstruction, and the subsequent prompt invasive therapy with percutaneous cholecystostomy tube placement prevented any further rise in levels on successive laboratory checks.

Mirizzi Syndrome remains quite rare. In a surgical series, the frequency of Mirizzi syndrome was 0.18% over a 23-year period

of performed cholecystectomies [3]. Two general classifications of Mirizzi syndrome have been suggested based on endoscopic retrograde cholangiopancreatography findings: McSherry type I is limited to external compression of the common hepatic duct by a gallstone impacted in the cystic duct or Hartmann pouch, whereas McSherry type II is associated with erosion between the gallbladder and the common bile duct with subsequent cholecystobiliary fistula [4]. Although discussion of the exact surgical approach for each type is beyond the scope of this case report, laparoscopic cholecystectomy may be appropriate in some patients with type I disease. In contrast, an open surgical approach remains the standard of care for those with type II Mirizzi syndrome, primarily because of the erosion of stones into ductal structures, development of adhesions, anatomic changes, and the potential need for biliary drains [3]. The patient in our case would be categorized as McSherry type II based on the presence of gallbladder rupture. Subsequent classification systems have been suggested in the literature for Mirizzi syndrome. Csendes et al. proposed a system that predicted operative mortality according to the severity of the lesion [5]. In this scheme, type I lesions are associated with external compression of the common bile duct, similar to McSherry type I. Type II Csendes is present with less than one-third ductal circumference erosion from a cholecystobiliary fistula. Type III Csendes involves up to two-thirds of the duct circumference, while type IV involves complete destruction of the bile duct. In our patient, the reading Radiologist did not report specific measurements in reference to erosion within the bile duct, thereby precluding classification of our case according to the Csendes categorization.

Treatment of Mirizzi syndrome often requires a multidisciplinary approach, using the history and physical examination to guide appropriate consultation with General Surgery, Gastroenterology, or Interventional Radiology. The prognosis of Mirizzi syndrome is related to the degree of concomitant complications. Aggressive treatment is appropriate for most patients, with surgical intervention being individualized based on the stage and severity of the disease [3,5].

Conclusions

Gallbladder disease can manifest in rare cases with extrinsic compression of the common hepatic duct and is called Mirizzi syndrome in such instances. We present a case of Mirizzi syndrome with depiction of acute gallbladder rupture and intrahepatic biliary ductal dilation in a set of CT images suggestive of severe, complicated Mirizzi syndrome. The treating clinician should consider other causes as well, such as cholangiocarcinoma, when Mirizzi syndrome is seen. Treatment is usually with aggressive surgical intervention, but must be individualized based on the patient's clinical picture and severity of comorbid disease.

Conflicts of interest

None.

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