

# Mirizzi Syndrome and Gallstone Ileus: An Unusual Presentation of Gallstone Disease

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We discuss the case of a man with an unusual complication of gallstone disease. An 85-year-old patient presented to the emergency department with a 3-week history of abdominal pain in the right upper abdominal quadrant. Thoracoabdominal radiography demonstrated that the whole extrahepatic biliary tree, including the common bile duct, common hepatic duct, gallbladder, and left and right hepatic ducts, were visibly delineated by air. The operative findings revealed a small shrunken gallbladder, a fistula between the gallbladder fundus and the gastric antrum, and a cholecystohepatic fistula, corresponding to Mirizzi syndrome, type II. A large gallstone was found impacted in the jejunum. This patient seems to have developed initially a cholecystohepatic fistula. Due to the acute inflammatory process, the stone eroded through the gallbladder wall and into the gastric antrum, passing from the antrum into the small bowel, where it became impacted. We suggest that the natural history of Mirizzi syndrome does not end with a cholecystobiliary fistula but that the continuous inflammation in the triangle of Calot may result in a complex fistula involving not only the biliary tract but also the adjacent viscera.

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KEY WORDS: Mirizzi syndrome, gallstone ileus, complications

The most common complications of chronic gallstone disease are acute cholecystitis, acute pancreatitis, cholangitis, and a gangrenous gallbladder.<sup>1</sup> Other complications are extremely rare and include Mirizzi syndrome, cholecystocholedochal fistula, and gallstone ileus.<sup>1-10</sup> The late nineteenth century and early twentieth century surgical literature are rich in descriptions of bizarre complications of long-standing gallstone disease.<sup>11-20</sup> Those complications are seldom found today. The current knowledge of biliary disease and the widespread use of ultrasonography have led to early diagnosis and early treatment for those with gallstone disease. Usually patients with gallstone disease have only the most common complications associated with their disease.

External compression of the biliary tree resulting in obstructive jaundice was described by Kehr<sup>13</sup> in 1905, Ruge<sup>14</sup> in 1908, Levrat and Chayvialle in 1941<sup>15</sup>, and Mirizzi<sup>17</sup> in 1948. Puestow<sup>16</sup> first described a cholecystobiliary fistula in 1942; Behrend and Cullen<sup>18</sup> in 1950 and Mirizzi<sup>19</sup> in 1952 reported other cases. Courvoisier<sup>11</sup> initially described so-called gallstone ileus resulting from obstruction of the small

bowel by an impacted gallstone in 1890. In 1896, Bouveret<sup>12</sup> described a syndrome of gastric outlet obstruction caused by an impacted gallstone in the duodenal bulb after the migration of the stone through a cholecystoenteric fistula.

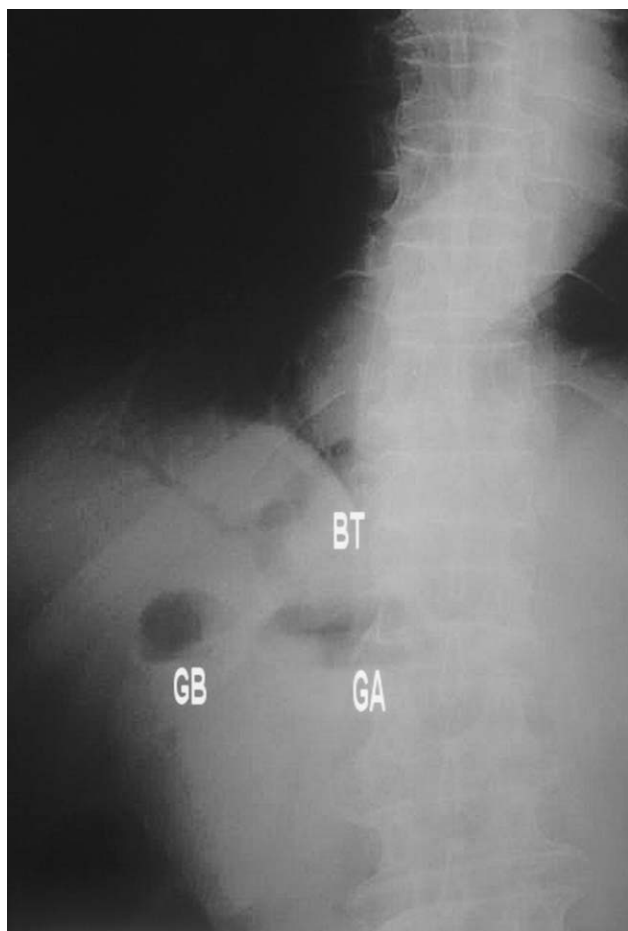
Until the early 1980s, these cases were considered as separate entities. The diagnosis and surgical approach were almost anecdotal.<sup>4,5</sup> In 1982 McSherry et al.<sup>2</sup> and Csendes et al.<sup>3</sup> in 1989 published seminal articles describing the physiopathologic process and classifying Mirizzi syndrome. These articles formed the basis on which the surgical approach to the Mirizzi syndrome was standardized.<sup>3,6,7,9</sup> We report herein an older patient who had both Mirizzi syndrome and gallstone ileus.

## CASE REPORT

An 85-year-old man presented to the emergency department with a 3-week history of right upper abdominal quadrant pain. He was anxious and had shortness of breath. The pain was associated with nausea and protracted vomiting that had debilitated

him. He could not eat, and he was very weak. The physical examination revealed a malnourished pale man who was febrile, sweating, and delirious. The arterial pressure was stable but he had a rapid pulse. The abdomen was tender, principally in the upper right and left quadrants, and he had rebound sensitivity. The laboratory examinations revealed a hemoglobin of 12.2 g/dl, a white blood cell count of  $13.6 \times 10^9/L$ , a total bilirubin of 0.69 mg/dl, and a direct bilirubin of 0.07 mg/dl. The alkaline phosphatase level was 103 U/L.

Plain radiographs of the thorax, abdomen, and pelvis were obtained, along with an abdominal ultrasound. The thoracoabdominal radiograph demonstrated a curious finding. The entire extrahepatic biliary tree, including the common bile duct, the common hepatic duct, the gallbladder, the left and right hepatic ducts, and some of the smaller intrahepatic radicals, were clearly visible, delineated by air. The gastric antrum was also visible (Fig. 1). This



**Fig. 1.** The complete biliary tract is clearly delineated by air, including the gallbladder (GB), the common, right, and left hepatic ducts, the common bile duct (BT), and some intrahepatic radicals. The gastric antrum (GA) also contains air.

unusual finding was interpreted as a biliodigestive fistula. A water-soluble contrast radiograph was subsequently taken, and it demonstrated a large gallstone lodged 60 cm distal to the ligament of Treitz (Fig. 2).

About 24 hours later, the patient underwent surgery. The operative findings were a small shrunken gallbladder, a fistula between the gastric antrum and the gallbladder fundus, and a cholecystohepatic fistula corresponding to Mirizzi syndrome type II, as described by Csendes et al.<sup>3</sup> A large gallstone, 45 mm in diameter, was found in the jejunum, impacted 60 cm from the ligament of Treitz. The stone was milked proximally into a dilated healthy area and retrieved via a longitudinal enterotomy.

The biliodigestive fistula was divided, and the opening was closed with a 4-0 polyglycolic acid suture. The cholecystohepatic fistula was sutured over a cuff of gallbladder with the same material. A T tube was placed into the common bile duct, and an intraoperative cholangiograph was taken. Two drains were left in place, and a nasojejunal tube was inserted for postoperative feeding.



**Fig. 2.** Water-soluble contrast radiography showing the outline of a large gallstone (GS) impacted in the jejunum.

The postoperative course was uneventful, and the patient recovered. He was discharged on postoperative day 7, on oral feeding, and without drainage from his abdominal drains. A postoperative cholangiograph through the T tube demonstrated only a dilated bile duct, no stones or other anomalies. The T tube was removed 60 days after the operation.

## DISCUSSION

The original description by Puestow<sup>16</sup> in 1942 of a cholecystobiliary fistula and the report of Mirizzi<sup>17</sup> in 1948 of a functional obstructive syndrome, both as complications of longstanding gallstone disease, led some surgical investigators to relate the two processes.<sup>2,3</sup> The physiopathologic process was elucidated after almost 40 years, in part due to the relative difficulty in the diagnosis that it represents<sup>1-9</sup> and to the low incidence reported.<sup>2,3,6-9</sup>

As stated by Csendes et al.,<sup>3</sup> the so-called Mirizzi syndrome and the cholecystobiliary fistula are different evolving stages of the same disease process. The concepts that an impacted stone in close contact with an inflamed mucosa develops first ischemia and then necrosis and that, because of the associated inflammation of the gallbladder wall and the hepatic or common bile duct wall, the impacted stone erodes through them and eventually forms a fistula are applicable to other biliary fistulas, such as cholecystoduodenal, cholecystogastric, and cholecystocolonic fistulas.<sup>3,6-10</sup>

This particular patient seems to have developed initially a cholecystohepatic fistula. After the last 3 weeks of his disease, due to the acute inflammatory process, the large stone found in the jejunum eroded through the gastric antrum wall, passing into the small bowel, where it became impacted. That could be the reason why we found this complex fistula, which appeared on the thoracoabdominal roentgenogram (Fig. 1).

The classic radiographic signs of gallstone ileus were first described by Rigler<sup>21</sup> in 1941 and included signs of intestinal obstruction, pneumobilia, aberrantly located gallstone, and change in location of the previously identified stone on serial examinations. Our patient presented with pneumobilia as well as two adjacent air-fluid levels in the right upper quadrant. This sign, as described by Balthazar<sup>22</sup> in 1978, is an additional helpful sign. The medial collection is located in the duodenal bulb and the lateral in the gallbladder (Fig. 1). It should be noted that even though the stone was large, it did not become impacted at the pylorus, so this patient did not develop Bouveret syndrome.<sup>10,12</sup> Instead the stone

migrated through the pylorus, the duodenum, and the first 60 cm of the jejunum until it became impacted (Fig. 2), causing the characteristic syndrome of intestinal obstruction known as gallstone ileus.

## CONCLUSION

We may consider the torpid evolution of this patient's complication as a lesson in advanced biliary pathology. We also suggest that the natural history of Mirizzi syndrome may not end with just a cholecystobiliary fistula. The continuous inflammation in the triangle of Calot area may result in a complex fistula involving not only the biliary tract but also the adjacent viscera.

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