CLINICAL VIGNETTE

Mirizzi Syndrome

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Mirizzi syndrome is an uncommon condition caused by the obstruction of the common hepatic duct by external compression from multiple small or a single large impacted stone in the cystic duct or in the gallbladder. It is difficult to diagnose and preoperatively the diagnosis is often missed. We describe a case of Mirizzi's Syndrome and discuss its presentation and treatment.

Presentation

A 27-year-old female was admitted to the hospital with a two-week history of right upper quadrant, epigastric, and lower chest pain. A few days before admission she had been evaluated by her primary physician who prescribed sulfamethoxazole/ trimethoprim and hydrocodone/acetaminophen for her symptoms. However, her pain had persisted, prompting emergency room evaluation and hospital admission. On admission, she denied any fever, chills, nausea or vomiting. She had no history of cardiac or gastrointestinal disease, but did report a prior ED visit for lower chest pain about 5 months before admission. She was evaluated in the emergency room, but no cause of her chest pain was found. Her past surgical history was remarkable for cesarean section and appendectomy.

On admission, the patient was afebrile. Physical examination was remarkable for right upper quadrant and epigastric tenderness. Murphy's sign was negative. Her laboratory tests were remarkable for WBC of 8.6, total bilirubin of 1.3, AST 461, ALT 336 and alkaline phosphatase 98. Ultrasound revealed hepatomegaly with fatty infiltration and sludge in the gallbladder, without wall thickening. The common bile duct measured 4 mm in diameter,

The following day, her abdominal pain persisted and bilirubin increased to 3.1. On MRCP, her common hepatic duct was dilated to 5 mm in diameter, but distal to the cystic duct there was a sudden change in the caliber of the common bile duct (CBD) which was only 2 mm in diameter. There was gallbladder sludge, but no choledocholithiasis.

ERCP was performed the same day revealing smooth tapering of the distal CBD and no choledocholithiasis. There was smooth extrinsic compression of common hepatic duct. Her cystic duct was not opacified and her gallbladder was not visualized. Mirizzi syndrome was suspected and this was communicated to the surgeon.

The next day, her bilirubin increased to 5.8. She was taken to surgery for laparoscopic cholecystectomy for presumed cholecystitis with possible Mirizzi syndrome. At surgery, she was found to have "significant amount of inflammation and fibrosis and the neck of the gallbladder appeared to transition into the common bile duct". She was converted to open cholecystectomy for better visualization and to minimize the risk of complications. Pathology report on the gallbladder stated "gallbladder covered with diffuse hemorrhagic fibrous adhesions" and "contained several yellow mulberry-like calculi".

On postoperative day one she complained of postoperative pain but her bilirubin had dropped to 1.7.On postoperative day four, she continued to complain of abdominal pain and her bilirubin was elevated to 5.0. Another ERCP revealed mild stricture of the CBD at the level of the cystic duct but no contrast extravasation to suggest biliary leakage.and an endobiliary stent was placed across the stricture. On postoperative day 6, her abdominal pain had resolved and her bilirubin dropped to 0.9 and she was discharged home.

Discussion

Mirizzi syndrome is defined as common hepatic duct obstruction caused by extrinsic compression from an impacted stone in the cystic duct or the infundibulum of the gallbladder. This was first described by Pablo Luis Mirizzi in 1948.[1] It is estimated to occur in 0.05 to 4 % of patients undergoing surgery for cholelithiasis.[2] It is often difficult to diagnose preoperatively, and 50% of cases are diagnosed intraoperatively.[3] It has been associated with gallbladder cancer. It was postulated that the recurrent inflammation and biliary stasis may predispose to both conditions. In a retrospective review of 4800 cholecystectomy cases, Prasad et al reported 2.8% of the patients had Mirizzi syndrome, and of those with Mirizzi syndrome, 5.3% was found to have gallbladder cancer, compared to 1% without Mirizzi syndrome.[4]

The neck of the gallbladder is connected to the cystic duct, which empties into the common bile duct. Gallstones can get impacted in the neck of the gallbladder causing inflammation with recurrent episodes of cholangitis, and obstruction of the CBD. Chronic inflammation may lead to bile duct wall necrosis and erosion into the CBD forming cholecystobiliary fistula.[5]

Csendes et al classified Mirizzi syndrome based on the presence and extent of a cholecystobiliary fistula, which was helpful in guiding surgical therapy.[6]

Type I: external compression of the common hepatic duct due to stone impacted at the neck or cystic duct. There is no fistula. 11% of individuals with Mirizzi syndrome.

Type II: fistula involves less than 1/3 of the circumference of the CBD. 41% of individuals with Mirizzi syndrome.

Type III: fistula between 1/3 to 2/3 of the circumference of the CBD. 44% of individuals with Mirizzi syndrome.

Type IV: disruption of the entire CBD. 4% of individuals with Mirizzi syndrome.

The presenting symptoms of Mirizzi's syndrome are right upper quadrant pain, fever, and jaundice. All three symptoms are present in 44-71% of the patients.[7] Pain is the most common symptom, follows by jaundice and cholangitis.

It is difficult to distinguish Mirizzi syndrome from choledocholithiasis or cholangitis. Therefore, one has to rely on imaging studies including ultrasound, abdominal CT scan, and magnetic resonance cholangiopancreatogram (MRCP). The diagnosis of Mirizzi syndrome requires the presence of the following:

Dilatation of the biliary system above the level of gallbladder neck.

The presence of a stone impacted in the gallbladder neck.

An abrupt change to a normal diameter of the CBD below the level of the stone.

MRCP has a high sensitivity for Mirizzi syndrome. It provides the extent of pericholecystic inflammation and differentiates from gallbladder malignancy. ERCP is useful in confirming the diagnosis of Mirizzi syndrome and to determine if a cholecystobiliary fistula is present. It allows for biliary decompression by internal stenting in patients with obstructive jaundice or cholangitis.[2] Endoscopic removal of the CBD stone may obviate the need for common duct exploration at the time of surgery.

For Mirizzi Syndrome type I, surgery is necessary for the removal of the inflamed gallbladder and the impacted stone. Laparoscopic cholecystectomy can be difficult due to dense adhesion and inflammation around the CBD. Laparoscopic cholecystectomy was successful in 59% of the patient with Mirizzi syndrome in one systematic review.[8] The surgeon should have a high index of suspicion for gallbladder cancer, and a frozen section biopsy should be performed on any suspicious areas of the gallbladder.[5] For type II, suture repair, T tube placement or choledochoplasty with remnant gallbladder are recommended. For type III, choledochoplasty or bilioenteric anastomosis are needed. Choledochojejunostomy is often required in type IV as there is total duct destruction.[9] ERCP can provide definitive treatment for type I and II patients who are poor surgical candidates. Tsuyuguchi et al reported 12 of 13 patients with no residual gallstone, remained asymptomatic. Of the 6 patients with residual stones, 4 had cholangitis, and 2 died from biliary causes.[10]

Conclusion

Our patient presented with biliary colic, and likely had biliary symptoms 5 months prior to her admission when she had complained of lower chest pain. Even though Mirizzi's Syndrome is uncommon, it should be suspected in all patients presenting with right upper quadrant pain, jaundice, and fever and MRCP evidence of dilatation of the biliary system above the level of the gallbladder neck, impacted stone in the gallbladder neck, and an abrupt change to a normal diameter of the common duct below the level of the stone. As in this case, the surgeon needs to be aware of possible diagnosis of Mirizzi Syndrome to be prepared for open cholecystectomy to minimize biliary injury and other complications from surgery.

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