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CLINICAL VIGNETTE

Mirizzi Syndrome

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The patient is a 96-year-old female admitted to the hospital with jaundice. She had history of diabetes, chronic kidney disease and hypertension. She developed progressive fatigue followed by onset of jaundice with bilirubin greater than 20. She had noted worsening jaundice over several weeks prior to her admission. CT scan on admission revealed a 3 cm gallstone in a distended gallbladder and dilated extra hepatic biliary ducts without the presence of a pancreatic or hepatic mass. CT chest revealed multiple pulmonary nodules of unclear significance. Prior to her admission she was independent in activities of daily living although she lived with her daughter.

MRI cholangiogram revealed moderate to marked intra-hepatic ductal dilation up to 1.5 cm. There was an abrupt transition point at the level of the common bile duct or common hepatic duct. Respiratory motion limited interpretation. There was also questionable dilation of the cystic duct. No pancreatic or hepatic abnormality was noted. She reported one episode of dark emesis and one episode of melena. On admission INR was greater than four which corrected to 1.5 with an infusion of FFP. Her Hgb was 11.2 with a WBC of 11,200. Other chemistries included Bilirubin 21, AST 318, ALT 148, and the Alkaline Phosphatase was 964. The chemistries were consistent with cholelithiasis with bile duct obstruction. No stones were seen in the Common Bile Duct or intra-hepatic ducts. A large stone in the neck of the gallbladder appeared to impinge to the common bile duct at the level of the cystic duct consistent with a Mirizzi syndrome. ERCP was performed with endobiliary stent placement with subsequent reduction of the LFT's to normal.

Mirizzi syndrome results from extrinsic bile duct compression. The Obstruction of the common hepatic duct is from extrinsic compression from a large stone in the neck of the gallbladder or cystic duct. Patients generally present with jaundice, fever and RUQ pain.[1] It is reported in 0.06 to 2.7% of patients referred for cholecystectomy.[2] Diagnosis should be confirmed before laparoscopic cholecystectomy. The majority of patients with Mirizzi syndrome are women which may be due to increased incidence of gallstones in women. The syndrome may result in compression of the common hepatic duct or fistulization between the gallbladder and the common duct due to stone erosion.The classification of Mirizzi syndrome is based on the presence and extent of a cholecysto-biliary fistula.

Type I involves external compression of the bile duct due to an impacted stone. Type II involves a fistula occupying less than a third of the CBD circumference. Type III involves 1/3-2/3 the circumference of the common duct and Type IV involves the destruction of the common duct wall.

The initial workup should include an abdominal ultrasound to evaluate the gallbladder, common duct and the liver. Findings consistent with Mirizzi syndrome include dilation of the biliary system above the gallbladder neck; presence of an impacted stone in the neck of the gallbladder and an abrupt change in the width of the common duct at the level of the stone.

If these findings are present, MRCP has significant sensitivity in determining the presence of common duct stones or extrinsic compression of the common bile duct or the presence of a mass lesion in the pancreas or bile duct.[3]

ERCP is both diagnostically and therapeutically essential for diagnostic confirmation and possible placement of an endo-biliary stent to decompress the duct, prevent cholangitis and rule out the presence of common duct stones.[4]

Management of Mirizzi's syndrome is surgery. Laparoscopic surgery can be performed if there is no evidence of a fistula between the gallbladder and he common bile duct.[5]

Presence of Type II or Type III or Type IV Mirizzi syndrome will most likely result in an open procedure with a choledocho duodenostomy or choledocho jejunostomy. Surveillance for gallbladder cancer is also recommended as there is a high incidence of gallbladder cancer, 5-28% has been reported in patient with this syndrome.

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