SUBMITTED 3 APR 24
REVISION REQ. 13 JUN 24; REVISION RECD. 17 JUN 24
ACCEPTED 9 JUL 24
ONLINE-FIRST: JULY 2024
DOI: https://doi.org/10.18295/squmj.7.2024.047

A Case of Mirizzi Syndrome with Pancreatic Divisum

A rare association

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Abstract

Mirizzi syndrome (MS), a rare complication of gallstones, refers to extrahepatic biliary compression by calculus in the cystic duct or Hartman's pouch and is usually associated with cystic duct abnormalities. Its association with pancreatic divisum(PD) is infrequent, the most common complication of PD being recurrent pancreatitis. We report a case of 39-year-old female who presented with acute abdominal pain to the department of general surgery, Chennai, Tamil Nadu, India in August 2022. Magnetic resonance cholangiopancreaticography (MRCP) showed calculous cholecystitis with a calculus indenting the cystic duct, causing luminal narrowing of the common hepatic duct (type I), which was associated with type II PD. The association of MS with PD has been rarely described. PD may be one of the factors responsible for bile stasis leading to calculous cholecystitis and its complications. Knowledge of MS and its associations helps in early diagnosis and selection of appropriate treatment management.

Keywords: Mirizzi's syndrome; Pancreatic Divisum; Gall stones

Introduction:

Mirizzi syndrome(MS) results from extrinsic compression of the extrahepatic biliary duct by the impacted gallstone in the Hartman's pouch or cystic duct. It is one of the rare complications of gallstones, with an incidence rate ranging from 0.6 to 5.7%. [1] The

compression can cause external obstruction, erosion, fibrosis, or fistula, leading to complex complications. [2] There is an association of cystic duct abnormalities in cases of MS, and knowledge of various anatomical variations of the cystic duct is important in planning appropriate treatment strategies. [3] The association of pancreatic divisum (PD) that occurs due to the failure of the fusion of dorsal and ventral pancreatic ducts with MS has rarely been described. However, thickened bile and Cholelithiasis have shown an association with PD. [4, 5] This report reviews a case of MS in a patient with PD.

Case Report:

A 39-year-old female patient presented to the general surgery outpatient department, Tamil Nadu, India, in 2022 with abdominal pain complaints lasting for two days. On examination, the Vitals were stable. Mild tenderness was noted in the right hypochondrium. Preliminary ultrasound evaluation showed multiple calculi in the gallbladder with mild wall thickening. The Total leucocyte counts (TLC) were mildly raised ~10,720/mm3. (Normal TLC count 4500 - 10500/mm3), and other laboratory investigations (liver function tests, serum amylase, lipase, and renal function tests) were within normal limits. The patient was subjected to magnetic resonance cholangiopancreatography (MRCP) for further evaluation. MRCP imaging showed multiple calculi in the gallbladder lumen with features of acute calculus cholecystitis [Figure 1]. One of the calculi was noted in the cystic duct, indenting the common hepatic duct and causing luminal narrowing; however, no biliary dilatation was noted [Figure 2].

The dorsal duct of the pancreas was seen draining into the minor papilla, and the ventral duct was not well delineated. A common hepatic duct was seen draining into the major papilla [Figure 3].

The diagnosis of type I MS with type II PD was made. The patient underwent cholecystectomy, and the post-procedure period was uneventful. The patient provided informed consent to the publication of this case.

Discussion:

Cholelithiasis is a relatively common disease entity with a female predilection. The formation of gallstones can be due to cholesterol supersaturation, excess bilirubin, gallbladder hypomotility, or impaired contractility. [6] Most are asymptomatic; one in five become

symptomatic, and presentation depends on the number, size, and location of gallstones. [7] Symptomatic Cholelithiasis can lead to acute or chronic cholecystitis, empyema, obstructive jaundice, perforation of gallbladder, pericholecystic abscess, Mirizzi syndrome, gallstone ileus, etc., MS is a rare complication that needs high clinical suspicion because of non-specific symptoms and imaging plays an important role in its diagnosis. Csendes classification for MS is widely used to describe its severity as mentioned in [Table 1]. [8] MS has shown an association with anatomic abnormalities of the cystic duct, like long cystic duct, tortuous course, and low insertion of the cystic duct. [3] Few cases have reported an association of pancreatic duct abnormalities with improper biliary drainage, thickened bile, and Cholelithiasis. [4] PD is the most common congenital anomaly of the pancreas that results from the abnormal fusion of ventral and dorsal PD during fetal development with the incidence of 3.6-5.8%. Patients with PD were found to have gallstone disease more frequently than other conditions. [9] Abnormal sphincter mechanism at ampulla can lead to bile stasis or could be due to dysfunction of bile excretion from the cystic duct. 85.7% of the patients with PD had demonstrated inflammation/cholesterolosis in the gallbladder. [10] Our case is a rare association of MS type I with type II PD. Familiarity with the imaging appearance, anatomic variants, related associations, and disease processes helps in accurate diagnosis and treatment planning, which prevent iatrogenic biliary injuries and complications.

Conclusion:

MS is a rare complication of Cholelithiasis, and its association with PD is unusual. PD may be an additional factor, along with other associated risk factors for calculous cholecystitis and its complications. Management strategies mainly depend on patient symptoms and the presence of complications.

Authors' Contribution

DM contributed to the clinical data collection, drafting of the manuscript and manuscript review. AS contributed to the collection of radiological data and manuscript editing. All authors approved the final version of the manuscript.

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Table 1: Types of Mirizzi syndrome (Csendes classification)

Type	Description
I	Extrinsic compression of CHD by impacted stone a - in the cystic duct/Hartmann's pouch. b - absence of cystic duct.
II	Formation of cholecysto-choledochal fistula (<1/3rd wall circumference)
III	Formation of cholecysto-choledochal fistula (1/3rd - 2/3rd wall circumference)
IV	Formation of cholecysto-choledochal fistula (>2/3rd wall circumference)
V	Cholecystoenteric fistula with any other type of MS a - without gallstone ileus b- with gallstone ileus

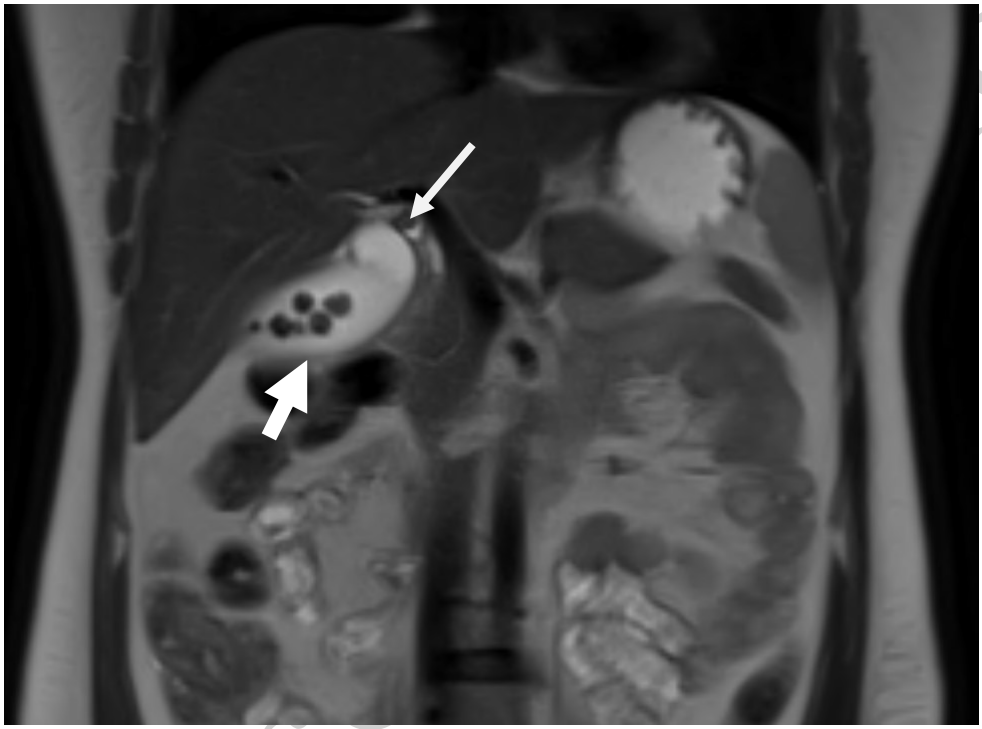

Figure 1 (MRCP T2 imaging coronal section) Gall bladder shows multiple calculi (short arrow) within the lumen, one calculus in cystic duct (long arrow)

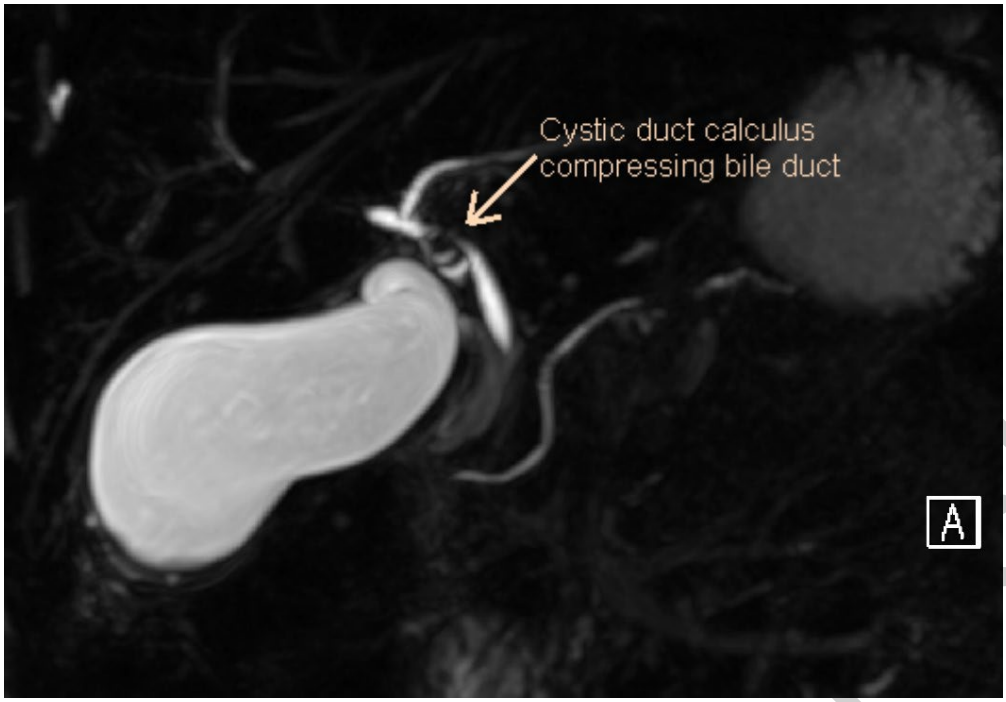

Figure 2 (MRCP MIP images) Calculus in cystic duct indenting the common hepatic duct.

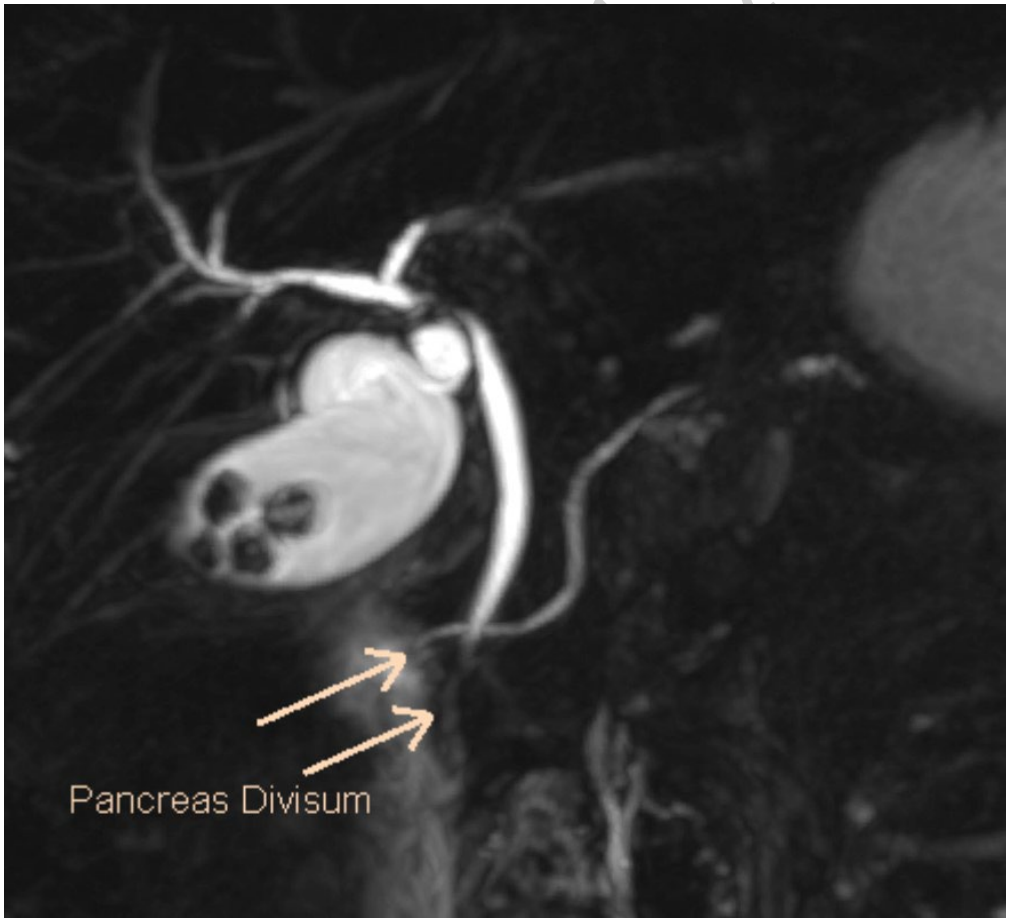

Figure 3 (MRCP MIP images) The dorsal duct of the pancreas drains into the minor papilla, and the ventral duct is not well delineated. Common bile duct is draining into major papilla.