

A rare case of Mirizzi syndrome due to pure calcium carbonate stones (Limy Bile)

Nooman Gilani,¹ Muhammad Farooq Hanif,² Veronika Karasek³

Abstract

We report the first case of Mirizzi syndrome in a patient who presented with biliary obstruction caused by pure calcium carbonate stones. A 61 years old male with history of portal vein thrombosis presented with rash, nausea and jaundice. An ultrasound of biliary tree showed gallstones with dilatation of hepatic duct and intrahepatic biliary tree. There was suspicion of a stone in proximal CBD. CT scan showed an opaque gallbladder with dense radio-opaque material in its lumen. An ERCP was then performed revealing external common hepatic duct obstruction at the neck of the gallbladder. A plastic biliary stent was placed across the obstruction, followed by a cholecystectomy. Resected gallbladder specimen revealed thick whitish paste like material, and formed stones filling the gallbladder lumen. Laboratory testing showed this material to be composed of 100% calcium carbonate crystals.

Keywords: Limy bile syndrome, Mirizzi syndrome, Bile duct obstruction.

Introduction

Limy bile syndrome was first described in 1911 which continues to be a rare disorder with prevalence ranging from 0.1 to 1.7% of patients undergoing surgery for biliary lithiasis.¹⁻³ The gallbladder and rarely the common bile duct are filled with a thick, paste-like radio-opaque material mostly composed of calcium carbonate. Many theories have been suggested regarding the etiology of limy bile syndrome,⁴ however bile stasis including obstruction at the level of cystic duct or gall bladder neck is necessary for this disorder to develop. Under these circumstances of bile stasis, calcium carbonate can precipitate, forming crystals, but the exact mechanism and the time required for these biochemical changes have not been fully elucidated.⁵⁻⁸

Women are affected more than men at a ratio of 3:1. Recent studies show that calcium carbonate stones are more frequent in children which could be related to the

smaller size of their cystic ducts.^{5,6}

Most reported cases have identified only a thick whitish paste-like material in the gallbladder or biliary ducts. In a few cases, where formed stones were seen, these were either of pure cholesterol or a cholesterol core encased by calcium carbonate.^{5,7} There is only one reported case dating back to 1968 where gallstone was mostly composed of calcium carbonate except 5% of cholesterol and trace of bile pigments.⁸

Limy bile syndrome may rarely be associated with long-term use of total parenteral nutrition, hereditary spherocytosis, primary biliary cirrhosis, or primary hyperparathyroidism.^{3,4}

In this report, we present a case of Mirizzi syndrome (Limy bile) caused by pure calcium carbonate stones which is a rare presentation.

Case Report

A 61-year-old man with a medical history complicated by portal vein thrombosis presented with complaints of pruritus and rash in August 2010 at Phoenix VA Healthcare system, Arizona. Patient had noted

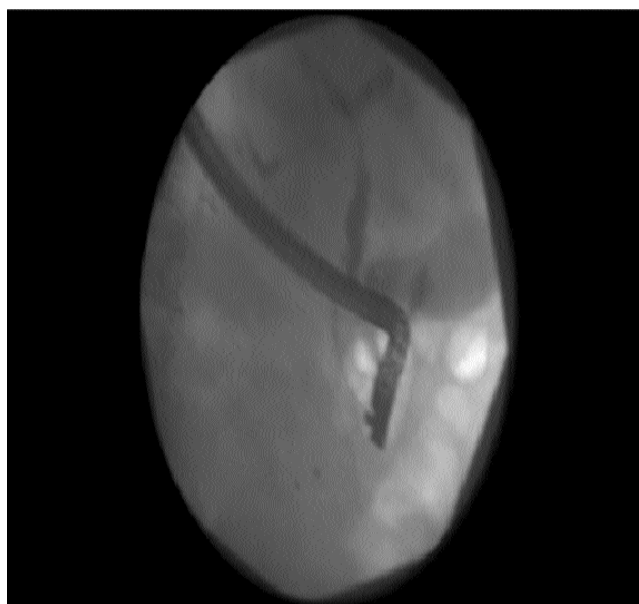


Figure-1: ERCP image showing compression of gallbladder neck on the common hepatic duct.

^{1,2}South Medical Ward, Mayo Hospital/King Edward Medical University, Lahore, Pakistan, ³Department of Gastroenterology and Hepatology, Phoenix VA Healthcare System/University of Arizona, Phoenix, AZ, USA.

Correspondence: Nooman Gilani. Email: ngilani@hotmail.com

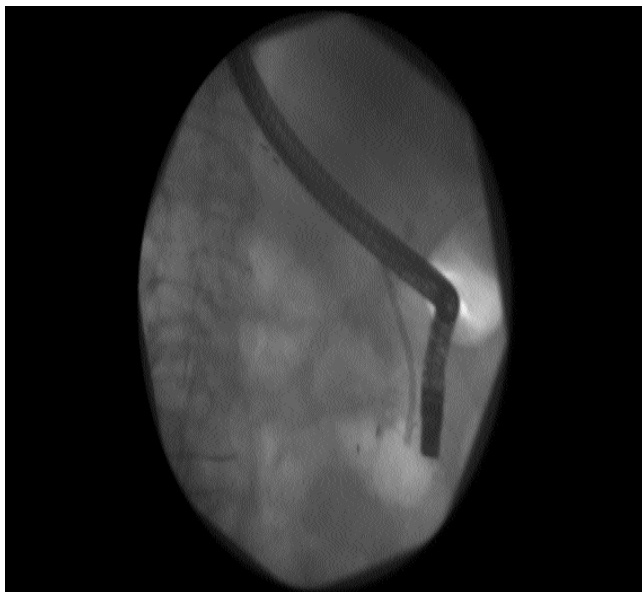


Figure-2: Fluoroscopic image post biliary ductal stent placement.

intermittent nausea with decrease in appetite, a reddish-brown discoloration of urine, and pale stools for the past 4 days prior to presentation. Physical examination revealed scleral icterus and mild epigastric area tenderness only.

Results of laboratory tests were as follow: Hb 14.8 g/dl (ref=13-16g/dl); haematocrit 43.7% (ref=38-52%); white blood cell count 6.5×10^3 (ref=4-11 $\times 10^3$); platelets $187,000 \times 10^3$ (ref=150-450 $\times 10^3$); total bilirubin 8.3mg/dl (ref= upto 1.2mg/dl); direct bilirubin 4.9; alkaline phosphatase 276U/L (ref=30-120U/L); AST 74U/L (ref=10-40U/L); ALT 130U/L (ref=10-40U/L).

An abdominal ultrasound showed multiple gallstones, and common hepatic duct and intrahepatic biliary tree dilatation. The common bile duct (CBD) measured 8.2 mm and there was a suspected 7.5 mm calculus in mid CBD. Computed tomography showed gallbladder opacification with very dense radio-opaque material within the gallbladder lumen. Endoscopic retrograde cholangiopancreatography (ERCP) was successful in cannulating the CBD, but a guide wire could not be passed above the common hepatic duct due to a stricture (suspected to be an external compression) at the proximal common bile duct/ hepatic duct junction (Figure-1). A Percutaneous trans-hepatic cholangiogram was performed with placement of a guide wire in the extra-hepatic bile duct. A cholangiogram showed a focal hepatic duct stenosis at the level of the cystic duct insertion. A second ERCP was performed, and a 10F, 7 cm plastic biliary stent was

placed across the obstruction. Bilirubin started to decrease, however, 10 days later, patient noticed worsening of jaundice. Total bilirubin had risen to 27. A third ERCP was performed demonstrating stent migration with dilatation of common hepatic duct, suggestive of Mirizzi syndrome again. A 10F, 7cm stent relieved bile duct obstruction (Figure-2). One week later, patient underwent an open cholecystectomy. Gallbladder was markedly swollen containing 2 large, white and flaky stones in the infundibulum causing external compression of common hepatic duct. Biochemical analysis revealed that stones were composed of 100% calcium carbonate weighing around 0.663 grams.

At 1, 3 and 6 months follow-up interval, our patient did not have symptoms of jaundice or abdominal pain, and LFTs had completely normalized.

Discussion

The presence of limy bile in the gallbladder can be an incidental finding on plain abdominal X-rays of patients with no symptoms at all. Biliary symptoms are, however, present in most patients with limy bile syndrome including epigastric and right upper quadrant abdominal pain.⁹ Limy bile presenting as obstructive jaundice is reported in a handful of cases only.^{4,9,10} Complications such as acute cholecystitis or acute pancreatitis can also be present.^{9,10}

Furthermore, Mirizzi syndrome is an unusual complication of gallstone disease and occurs in approximately 1% of all patients with cholelithiasis.¹⁰ This syndrome can result in significant morbidity and mortality for which an open cholecystectomy is the standard therapy.

Conclusion

Our case is unique in a way that Mirizzi syndrome caused by formed and pure calcium carbonate stones in an older adult has not been reported previously.

Disclosure: The article or part of the article has not been published in any other journal.

Conflict of Interest: None

Financial Support: None

References

1. Ballas KD, Alatsakis MB, Rafailidis SF, Psarras K, Sakadamis AK. Limy bile syndrome: Review of seven cases. *ANZ J Surg.* 2005; 75:787-9.
2. Dhote S, Agarwal A, Govindan A, Korath MP, Jagadeesan K. Limy bile syndrome: Report of an unusual case. *Bombay Hospital Journal.* 2006; 150-2.

3. Onghena T, De Waele JJ, Vereecken L, Van Loon C. Limy bile and laparoscopic cholecystectomy. *Acta Chir Belg.* 2001; 101:31-4.
 4. Takatori Y, Yamauchi K, Negoro Y, Noro K, Yoshida A, Nanamiya W, et al. Limy bile syndrome complicated with primary hyperparathyroidism. *Intern Med.* 2003; 42:44-7.
 5. Stringer MD, Soloway RD, Taylor DR, Riyad K, Toogood G. Calcium carbonate gallstones in children. *J Pediatr Surg.* 2007; 42:1677-82.
 6. Sayers C, Wyatt J, Soloway RD, Taylor DR, Stringer MD. Gallbladder mucin production and calcium carbonate gallstones in children. *Pediatr Surg Int.* 2007; 23:219-23.
 7. Taylor DR, Crowther RS, Cozart JC, Sharrock P, Wu J, Soloway RD. Calcium carbonate in cholesterol gallstones: polymorphism, distribution, and hypothesis about pathogenesis. *Hepatology.* 1995; 22:488-96.
 8. Cooke M. Limy bile. *Proc R Soc Med.* 1968; 61:1110-2.
 9. Itoh H. Management of Limy Bile Syndrome: No Therapy, Laparotomy or Endoscopic Treatment. *Intern Med.* 2003; 42:1-2.
 10. Safioleas M, Stamatakos M, Safioleas P, Smyrnis A, Revenas C, Safioleas C. Mirizzi syndrome: an unexpected problem of cholelithiasis. Our experience with 27 cases. *Int Semin Surg Oncol.* 2008; 5:12.
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