IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 13, Issue 11 Ver. V (Nov. 2014), PP 42-45
www.iosrjournals.org

Mirizzi Syndrome: Inflammation Mimiking Malignancy.

[1]Prashant U. Titare, [2]Bhawana D. Sonawane, [3]Pradip B. Rathod,
[4]Narendra G. Tembhekar
Radiology department, Government Medical College and Superspeciality Hospital, Nagpur,Maharashtra, India.

Abstract: *Mirizzi syndrome is an uncommon cause of obstructive jaundice characterized by impacted gallstone in cystic duct or gallbladder neck with associated inflammatory changes causing external compressive narrowing of common hepatic duct and proximal biliary dilatation. Inflammatory changes around the impacted gallstone form a soft tissue mass which sometimes mimic like malignant lesion on ultrasound. We are hereby reporting a similar case presentation in a 45 years Indian woman.*
Key words: *Gallstone, Inflammation, Malignancy, Mirizzi Syndrome, Obstructive jaundice.*

I. Introduction

Mirizzi syndrome is an uncommon cause of obstructive jaundice characterized by impacted stone or stones in the neck of gallbladder or the cystic duct leading to narrowing of common hepatic duct lumen. The impacted gallstone and surrounding inflammation initially causes external compression of the bile duct and subsequently erodes into the bile duct leading into cholecystocholedochal fistula or cholecystoenteric fistula. Acute cholecystitis, acute cholangitis, acute pancreatitis are the most common complication of chronic gallstone disease. Gallstone ileus and mirizzi syndrome are the unusual complications reported in gallstone disease.[1] Availability and widespread use of ultrasound led to early diagnosis and treatment of gallstone disease and avoiding the development of further complications like mirizzi syndrome.[2] We report here an uncommon case of mirizzi syndrome with associated inflammatory soft tissue lesion mimicking like malignancy on ultrasound.

II. Case Report

A 45 year old Indian woman, housewife by occupation presented in emergency department of our hospital for recent onset right hypochondriac pain associated with nausea, occasional vomiting, dark urine and gray colored stool. On clinical examination icterus was present. Her vitals were stable. Abdominal examination revealed diffuse tenderness in right hypochondriac region on deep palpation. Laboratory investigations showed total serum bilirubin 11.10 mg/dl, direct bilirubin 9 mg/dl, aspartate transaminase 68 U/L, alanine transaminase 187 U/L, alkaline phosphatase 370 U/L, γ- glutamyltransferase 301 U/L, lactate dehydrogenase 331 U/L and TLC- 13,400/cumm.

Abdominal ultrasound examination revealed a dilated thick walled gallbladder containing multiple gallstones of average size 8 mm (Fig. 1). Separate stone of approximate size 8mm was seen at the junction of gallbladder neck and cystic duct. Hypoechoic soft tissue of approximate size 17 x 17 mm was seen around the impacted stone. Common hepatic duct proximal to the lesion was dilated (9mm diameter) (Fig. 2). Intrahepatic biliary radicles and both hepatic duct were also dilated. Common bile duct was not dilated.

Endoscopic retrograde cholangiopancreaticography revealed narrowing of the distal common hepatic duct and dilatation of proximal biliary tree (Fig. 3). No mass lesion was detected. Distally common bile duct was normal in caliber. Fistula was absent. Biliary brush cytology was done. Plastic biliary stent was inserted across the narrowing into the biliary system.

Cytology did not revealed any evidence of malignancy. Subsequently she had interval open cholecystectomy and histopathology was negative for malignancy. She recovered well on follow up.

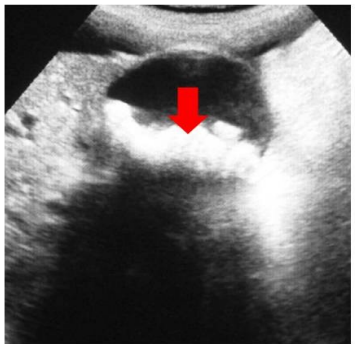

Figure 1: Ultrasound image of dilated gallbladder showing multiple stones (denoted by arrow) within the gallbladder lumen giving posterior acoustic shadowing.

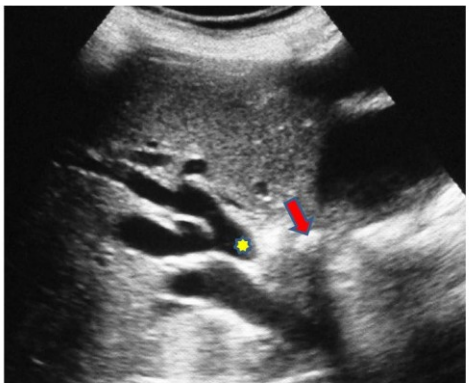

Figure 2: Ultrasound image showing thick walled gallbladder and impacted stone (denoted by red arrow) at the junction of gallbladder neck and cystic duct with associated hypoechoic soft tissue around the stone. Dilatation in proximal part of common hepatic duct (denoted by yellow *) is also shown.

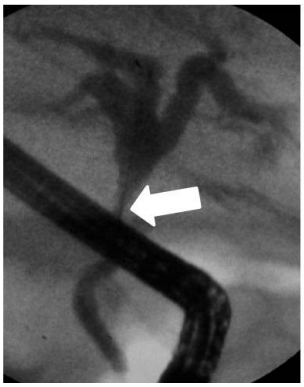

Figure 3: ERCP image showing luminal narrowing in the distal part of common hepatic duct (denoted by arrow) with proximal biliary dilatation.

III. Discussion

Impacted gallstone and associated inflammatory changes causing obstruction of the bile duct was first reported by Kehr in 1905 and Ruge in 1908.[1] This condition was first published in 1948 by Mirizzi and thereafter this condition was popular as Mirizzi syndrome.[1],[3] In 1942 Puestow had reported first cholecystocholedochal fistula as a complication of long standing gallstone disease. In 1950 Behrend had described double fistula between the gallbladder and common hepatic duct/duodenum.[1]

Incidence of mirizzi syndrome is reported to be <1% a year in western developed countries and 4.7 to 5.7% in underdeveloped countries particularly in latin America.[1] Recent studies showed mirizzi syndrome in 1 to 2 % of patients with symptomatic cholelithiasis.[1],[4] Associated gallbladder carcinoma is reported in 5.3 to 28% cases of mirizzi syndrome.[1] Mirizzi syndrome can occur at any age group. Mean age of presentation varies from 53 to 70 years. This syndrome is commonly seen in females (70%). It can present in acute or

chronic form. The most common form of clinical presentation is obstructive jaundice (60-100%), right upper quadrant abdominal pain (50-100%) and fever. The most common laboratory finding in mirizzi syndrome is hyperbilirubinemia. Increased aminotransaminase levels and leukocytosis had been frequently reported.[1] Similar findings were observed in our case.

Ultrasound is noninvasive, safe, cheap and easily available imaging modality generally used as preliminary investigation in patients of jaundice. Ultrasound not only differentiates between medical and surgical jaundice, but also detect the level and cause of obstruction in obstructive jaundice.[5] Diagnostic accuracy of ultrasound in mirizzi syndrome is about 29% and sensitivity varies from 8.3% to 27%.[4],[6],[7] Ultrasound findings in mirizzi syndrome includes contracted or dilated gallbladder with thick or thin walls with single large or multiple small gallstones impacted in the infundibulum with dilated extra and intrahepatic portion of hepatic ducts and normal size common bile duct below level of obstruction.[1] In our case we found a stone impacted at the junction of gallbladder neck and cystic duct with multiple stones in gallbladder. Hypoechoic soft tissue which was seen in our case was suspected to be inflammatory or neoplastic lesion.

ERCP not only confirm the presence of mirizzi syndrome with or without fistula but also for therapeutic purpose by stent placement, stone retrival etc. It is an invasive procedure with diagnostic accuracy of around 55-90%.[4],[7] ERCP finding in mirizzi syndrome includes a narrowing or curvilinear extrinsic compression of distal common hepatic duct with proximal biliary dilatation and normal distal caliber.[1] In our case we found similar finding without any mass lesion. Brush cytology came negative for malignancy and temporary stenting was done for biliary drainage.

Preoperative diagnosis is generally difficult in mirizzi syndrome and can be made in 8% to 65% of patients.[4],[7],[8],[9] Inadequate recognition of this syndrome frequently led to high morbidity and mortality. In absence of preoperative diagnosis, the incidence of bile duct injuries in patients operated for mirizzi syndrome could be as high as 17%.[10] More than 50% cases of mirizzi syndrome are diagnosed during surgical exploration.[4] The characteristic surgical findings in mirrizi syndrome include a dilated gallbladder with thick wall or shrunken gallbladder with distorted wall and stone (single or multiple) impacted at gallbladder neck or infundibulum, obliterated Calot's triangle, dense fibrotic changes at Calot's triangle and dense adhesions subhepatic space.[1] In our case diagnosis of mirizzi syndrome was confirmed on surgical exploration and managed surgically.

Differential diagnosis includes other causes of obstructive jaundice such as cholangiocarcinoma, pancreatic carcinoma, carcinoma gallbladder, sclerosing cholangitis, metastasis and others.[1] In our case above mentioned differentials were excluded by radiological and surgico-pathological findings.

In 1989 Csendes et al classified the mirizzi syndrome into four types and in 2007 he added one more type to this classification (Table: 1).[11],[12]

Table 1: Classification of Mirizzi syndrome.

S.N.	Type	Subtype	Description
1	I	-	External compression of the bile duct by a gallstone impacted in cystic duct or gallbladder infundibulum.
2	II	-	Cholecystobiliary fistula involving less than one third of bile duct circumference.
3	III	-	Cholecystobiliary fistula involving upto two third of bile duct circumference.
4	IV	-	Cholecystobiliary fistula and complete bile duct wall destruction with fusion of gallbladder and bile duct forming a single structure without any dissection planes between them.
5	V	a	Cholecystoenteric fistula with any other type of Mirizzi syndrome without gallstone ileus.
6	V	b	Cholecystoenteric fistula with any other type of Mirizzi syndrome with gallstone ileus.

Mirizzi syndrome is mainly treated by surgery. Surgery in mirizzi syndrome is generally a challenge to the surgeon because of multiple reasons such as lack of preoperative diagnosis, inflammatory edematous tissue, dense adhesions, fistula and risk of bile duct injury during Calot's triangle dissection.[1] In 2009, more simplified classification was proposed by Solis-Caxaj.[13] They classified Mirizzi syndrome into three types and according to this recent classification and worldwide surgical experience different treatment is proposed for different types (Table: 2).

Table 2: Classification and treatment of Mirizzi syndrome proposed by Solis-Caxaj.

S.N.	Type	Subtype	Description	Treatment
1	I		External compression of the bile duct by a gallstone impacted at the Hartmann pouch or at gallbladder infundibulum and a acute or chronic inflammatory process.	Open cholecystectomy Open subtotal cholecystectomy Laparoscopic cholecystectomy
2	II	a	Cholecystobiliary fistula < 50% of the bile duct diameter.	Open cholecystectomy Open subtotal cholecystectomy
3	II	b	Cholecystobiliary fistula > 50% of the bile duct diameter.	Open subtotal cholecystectomy Biliary enteric anastomosis

4	III	a	Cholecystobiliary fistula with cholecystoenteric fistula without gallstone ileus	Simple closure of fistula. Treatment of gallbladder according to the presence of Mirrizi I, IIa or IIb.
5	III	b	Cholecystobiliary fistula with cholecystoenteric fistula with gallstone ileus	Treatment of gallstone ileus. Treatment of gallbladder according to the presence of Mirrizi I, IIa or IIb.

IV. Conclusion

We conclude that the inflammatory soft tissue around impacted gallstone in case of mirizzi syndrome may mimic like malignant lesion on ultrasound. Our work will help the reader to understand that the soft tissue seen on ultrasound is not always neoplastic, sometimes inflammatory soft tissue can mimic like malignant lesion as shown in our case report. ERCP, surgery and histopathology is essential for confirmation and appropriate management to reduce morbidity and mortality.

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