Common Bile Duct Stone With Mirizzi’s Syndrome: Another Exception To Double Duct Sign And Courvoisier’s Law?
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Citation

Abstract
The combined dilatation of pancreatic and common bile ducts is known as double duct sign and occurs most commonly in malignancy of the head of pancreas. Courvoisier's law states that, in a patient with obstructive jaundice, if the gallbladder is distended and palpable, it is not due to common bile duct stones. We present our case, a 62-year old female patient who presented to our department with obstructive jaundice. She was found to have a large common bile duct stone exhibiting both double duct sign and Courvoisier's sign. We discuss the current literature pertaining to these signs and highlight their significance in day-to-day clinical practice.

CASE REPORT
A 62-year-old female presented with 8 days’ history suggestive of obstructive jaundice. She had right upper quadrant discomfort, dark urine and clay stool. She denied any similar history in the past. She also had loss of appetite and weight. Clinically, she was icteric and abdominal examination revealed palpable liver and gallbladder and there was no other mass lesion. She was worked up with the possibility of obstructive jaundice due to a mitotic disease.

Liver function tests confirmed obstructive jaundice with a total bilirubin of 4.2 with increase in direct bilirubin. Alanine and asparte aminotransaminase levels were normal but serum alkaline phosphatase was elevated (305 IU/l; normal range, 20-140 IU/l).

She underwent ultrasound and CECT of the abdomen which revealed the following: Hepatomegaly with dilatation of intrahepatic biliary radicals in both lobes and grossly distended gallbladder with a stone in the dependent portion. The common bile duct measured 2.5cm with a solitary stone of 2.5 x 2.0cm in the distal common bile duct and dilated main pancreatic duct (Figures 1, 2 & 3). There was no demonstrable mass lesion either in the head of pancreas or in the periampullary region. Both end and side-viewing endoscopies were done to rule out periampullary pathology.
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Figure 2
Figure 2. Axial CT scan showing combined dilatation of common bile duct and main pancreatic duct: ‘double duct sign’

She was optimized and taken up for surgery. At surgery, she was found to have a grossly distended and thick-walled gallbladder with a stone, Type II Mirizzi’s syndrome with a cholecysto-choledochal fistula, and a 2.5-cm common duct with a solitary mobile stone. There was no malignancy anywhere. Careful dissection of Calot’s triangle was done and the site of the cholecysto-choledochal fistula was visualized. As the fistula was broad, she underwent subtotal cholecystectomy and the gallbladder stump was closed with 00 vicryl without any compromise of the common bile duct lumen. Choledochotomy was then done, the CBD stone was extracted and the procedure completed with a cholecchochoduodenostomy using 000 vicryl (Figure 4, 5, 6). She had an uneventful recovery and is on regular follow-up.

Figure 4
Figure 4. Intraoperative picture showing cholecysto-choledochal fistula (Mirizzi syndrome - arrow) with dilated common bile duct (forceps pointing to the stone inside)

Figure 3
Figure 3. Axial CT scan showing a large common bile duct stone in the distal common bile duct

Figure 5
Figure 5. Retrieved common bile duct stone
DISCUSSION

In 1976, Freeny et al., in their study of endoscopic retrograde cholangiopancreatography (ERCP) in pancreatic carcinoma patients, reported that an irregular encasement or obstruction of pancreatic duct occurred exclusively in carcinoma patients [1]. They stated that the accuracy increased if it is associated with dilatation of common bile duct that is the presence of 'double duct sign', which is probably its first description. Subsequently, the ‘double duct sign’ was used to describe the combined dilatation of bile duct and pancreatic duct in other modalities of imaging like ultrasound, CT scan and MRCP [2][3].

The most common causes of 'double duct sign' were pancreatic adenocarcinoma and ampullary adenocarcinoma for which this sign was initially described [4]. However, numerous further reports also identified this to be present in chronic pancreatitis. In these patients, the duct is usually irregular with intraductal calculi unlike the smooth uniform enlargement in pancreatic head malignancy [5]. Other pancreatic causes are lymphoma and metastasis arising in the head region. Anecdotal causes reported were sphincter of Oddi dysfunction, primary retroperitoneal fibrosis and Kaposi sarcoma [6][7][8].

A ductal stone producing ‘double duct sign’ is rarely described. It appears as a solitary case in a series of 77 patients described by Edge et al. in 2007 [9]. In a study of 10 patients with dilated pancreatic duct, Fishman et al., in 1979, discussed about a patient (not reported) with impacted ampullary stone producing double duct dilation [5]. They concluded that this sign will be more useful in localising the level of obstruction to the head region rather than identifying the etiology. Our patient exhibited this first peculiar feature of a 'double duct sign' due to a common bile duct stone. Though it presented with double duct sign, the stone was a mobile one which could be easily removed through supraduodenal choledochotomy.

The second peculiar feature presented by this patient is the palpable gallbladder in the presence of a common bile duct stone. In 1890, Ludwig G. Courvoisier observed in his 187 patients with common bile duct obstruction, that gallbladder dilatation seldom occurred in stone obstruction of the duct [10]. This gradually came to be represented in the literature and taught as Courvoisier's law or sign. This is attributed to the contraction and fibrosis of the gall bladder due to recurrent cholecystitis. However, as it happens with any rule in medicine, there are exceptions. Those are impacted stone at Hartmann's pouch, chronic pancreatitis, autoimmune pancreatitis, biliary ascariasis, AIDS-related cholangiopathy, choledochal cysts, common hepatic duct obstruction and double pathology like distal malignancy with xanthogranulomatous cholecystitis as listed by Fitzgerald et al. in 2009 [11].

Thus, our patient with a palpable gallbladder and calculous bile duct obstruction is yet another exception to this 'law.' In 1999, Munzer reported a series of 86 patients with distended gallbladder: 83% resulted from a distal malignant obstruction while 15% were due to bile duct stones [12]. This throws light on the fact that the presence of Courvoisier's sign need not always mean a distal malignancy. However, the reverse is true that the gallbladder may not be palpable in all cases of malignant distal bile duct obstruction.

CONCLUSION

Traditionally, Courvoisier's sign and double duct sign are viewed as indicators of underlying malignancy and help differentiate from common bile duct stone. However, our case and the accumulating evidence suggest that they may not be accurate in predicting malignancy and can even be present in ductal stone. Hence we conclude stating that these signs should be interpreted in conjunction with other findings in a given patient to arrive at a clinical or radiological diagnosis.

References

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