OXFORD SURGICAL GROUP

Mirizzi's Syndrome

Overview:

Mirizzi's syndrome was first described by an Argentinean surgeon called Pablo Mirizzi in 1948. He described an unusual presentation of gallstones that, when lodged in either the cystic duct or the Hartmann pouch of the gallbladder, externally compressed the common hepatic duct (CHD), causing symptoms of obstructive jaundice.

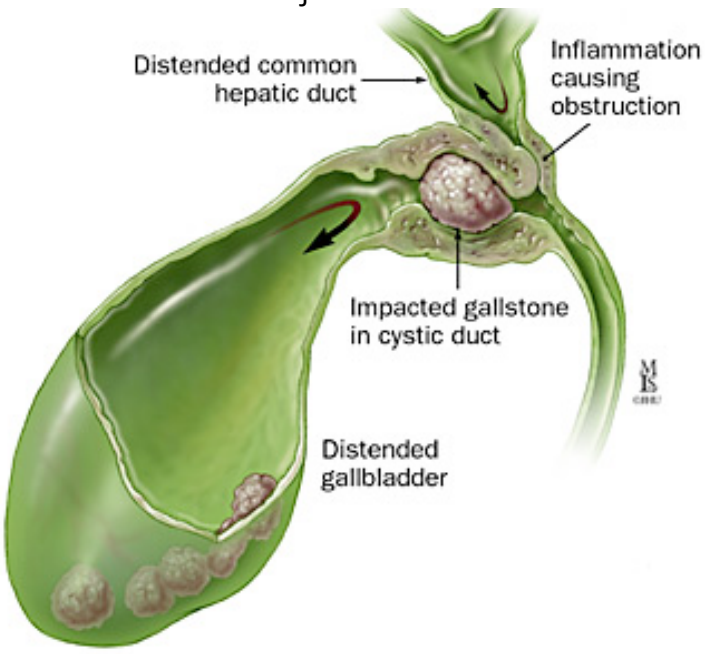
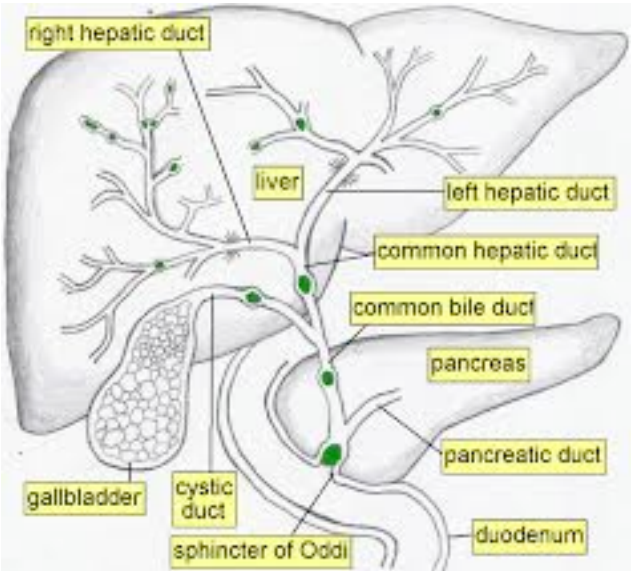
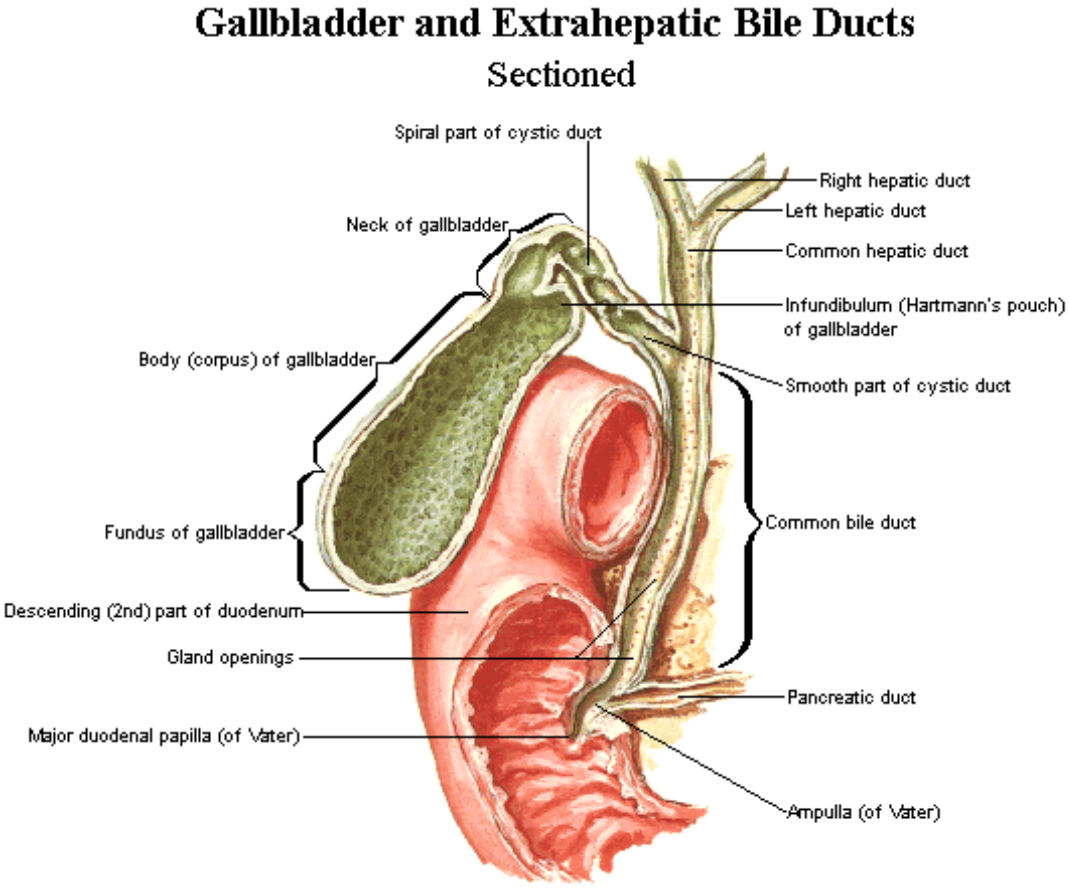


Figure 25. Mirizzi's syndrome.

Generally Mirizzi's syndrome is treated with surgery. This involves a cholecystectomy with or without the insertion of a T-tube. In advanced cases an operation to reconstruct the damaged biliary tree with the aid a hepatico-jejunostomy is required. There is an extremely low mortality associated with these operations, however, preoperative diagnosis of Mirizzi's syndrome is preferable as it can change the surgical approach. Preoperative diagnosis of Mirizzi's syndrome is difficult as there are no pathognemonic clinical signs and often multimodal imaging techniques are implemented to confirm the diagnosis.

Anatomy:





The well known normal anatomy of the biliary system is shown in Fig 1.

The close proximity of the gallbladder/cystic duct (CD) with the common hepatic duct (CHD)/common bile duct (CBD) means that a large stone in one may impact on the other. Some variations in the anatomy within individuals can increase the chances of an individual acquiring Mirizzi's syndrome. These variations may be the presence of a long CD in parallel with the CHD or a low insertion of the CD into the CBD (1).

Epidemiology

Mirizzi's syndrome occurs in around 0.1% of all patients with gallstone disease. (6) Within the population of those who have gallstones, there is no bias of Mirizzi's syndrome towards a gender or race. There is an increased risk of developing Mirizzi's syndrome with age and the average age of patients with this condition is over 60 years (3,4,7). However, the risk of developing gallstones does differ in certain populations. Gallstones are more likely in females than males, as well as being higher in those of Hispanic origins and Northern Europeans compared to those from Asia and Africa.

Pathophysiology:

When a gallstone becomes impacted in the neck of the gallbladder, commonly in Hartmann's pouch, or in the CD, it can cause compression of the biliary tree. This obstruction can either be direct mechanical compression or secondary obstruction due to repeated instances of local inflammation caused by the gallstones. The initial result of this obstruction is cholestasis. Once cholestasis occurs the natural progression is to inflammation and sepsis if left unchecked. This in turn can lead to a biliary stricture or promote fistula formation.

Continual pressure of the impacted gallstone directly onto tissue e.g. the wall of the gallbladder can lead to erosion of the involved tissues (via inflammation and pressure necrosis). In turn this can lead to a subsequent stricture of the CHD /CBD or eventually lead cholecystobiliary fistula.

Classification:

Pablo Mirizzi described a syndrome of common hepatic duct obstruction in the setting of long-standing cholecystitis and cholelithiasis.

McSherry, in 1982, then devised a sub-classification of Mirizzi's original description into two types:

Type I: External compression by a calculus in the cystic duct, Hartmann's pouch or the neck of the gallbladder.

Type II: A calculus has partially or totally entered into the CHD/CBD resulting in a cholecystocholedochal fistula.

Csendes et al 1989 (2) then further subdivided McSherry's type II classification to form the currently accepted system:

Type I – No fistula present

Type IA: External compression by a calculus in the cystic duct, Hartmann's pouch or the neck of the gallbladder. Cystic duct present and intact

Type IB: External compression by a calculus in the cystic duct, Hartmann's pouch or the neck of the gallbladder. Cystic duct obliterated.

Type II-IV – Fistula present

Type II: Cholecystobiliary fistula with an erosion of less than 33% of the circumference of the CHD/CBD

Type III: Cholecystobiliary fistula with an erosion of 33-66% of the circumference of the CHD/CBD

Type IV: Greater than 66% to total erosion and destruction of the CHD/CBD at the point of impaction.

When this classification system was first devised there was an incidence of roughly (2):

11% type I
41% type II
44% type III
4% type IV

However, this study was conducted in 1989 and some of the more recent data suggests that there is a decrease in the incidence of later stages (type III and IV) and a higher percentage of earlier presentations (type I-II).

Schafer et al in 2003 (3) reported that 87% of those in their study had type I and only 13% had a cholecystobiliary fistula (types II-IV). Other recent data, however, agrees more with the ratios originally described by Csendes. Aydin et al 2008 (4) reported rates of:

35% type I
53% type II
12% type III
0% type IV.

There could be a decrease in later stages due to improved diagnosis and intervention with modern imaging and surgery but there is a relative scarcity of data due to the infrequency of the syndrome.

Signs & Symptoms

As mentioned before, the diagnosis of Mirizzi's syndrome is difficult as there are no definite clinical signs or symptoms for the syndrome.

A combination of generalised symptoms point to a diagnosis and they may include:

Obstructive jaundice (76.9%)
Pruritis (77.8%)
Right upper quadrant pain (75.8%)
Pale stools (65.3%)
Vomiting (54.8%)
Nausea (38.4%)
Recurrent cholangitis

Acute presentations of Mirizzi's syndrome can be associated with pancreatitis or cholecystitis. Acute cholecystitis is much more frequent in type I, whereas types II-IV are associated with chronic cholecystitis due to the longer duration of the pathology process. Some patients only experience intermittent or mild signs due to the partial obstruction of the biliary system.

Investigations Haematological

All patients with Mirizzi's syndrome will have abnormal liver function tests at some point in the course of their disease. Some may present with frank obstructive jaundice. If the diagnosis is in question then tumour markers such as CA19-9, Ca 125 and CEA should be performed to try and exclude an early malignancy.

Investigations Radiological

The investigations used to aid a diagnosis of Mirizzi syndrome are often also aimed at excluding a diagnosis of bile duct cancer or gallbladder cancer. Mirizzi's syndrome may present like a biliary tree cancer and vice versa.

Ultrasonography (US), magnetic resonance cholangiopancreatography (MRCP), endoscopic retrograde cholangiopancreatography (ERCP) and computed tomography (CT) are the main imaging techniques employed in the diagnosis of Mirizzi's syndrome (5).

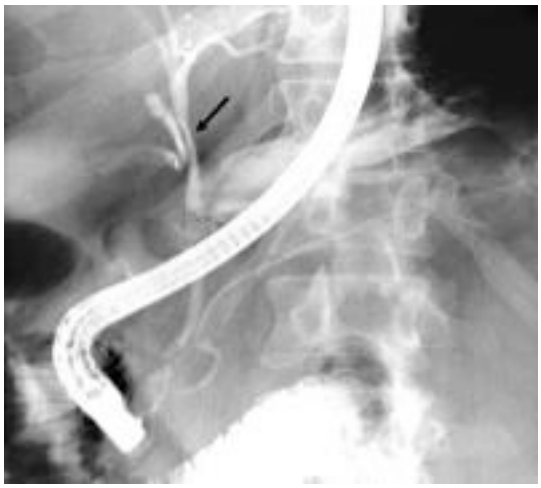
US is cheap, non-invasive and can readily demonstrate an impacted calculus in Hartmann's pouch or the cystic duct. A dilated common hepatic duct (>7mm) with intrahepatic biliary tree dilation is most often seen in type I Mirizzi's syndrome and can be easily seen on US as well as the narrowing at the level of impaction of the stone. Gallstones themselves may be visible with US imaging techniques and it is noted that they are always present in type I but may be absent in types II-IV due to fistula formation.

ERCP can demonstrate an obstruction or erosion of the biliary tree. It is possible during an ERCP to extract any stones within the biliary tree or insert a stent to temporarily decompress the obstructed system.

Typical MRCP findings of Mirizzi syndrome can include impacted stones in the gallbladder neck, compression of the CHD/CBD, dilatation of the biliary system above the level of impaction and a contracted gallbladder with abnormal wall-thickening. Additional sequences should be used to exclude the presence of malignancy.

CT scans may supplement the other imaging techniques and are primarily used to exclude the possibility of malignancy. Occasionally laparoscopic ultrasound may be necessary to further delineate the biliary tree pathology and exclude any tumour dissemination.

Yun et al measured the preoperative diagnostic accuracy of MRCP and CT for Mirizzi syndrome. They found that for combined modality (MRCP and CT), the overall sensitivity was 96% (42% for CT alone); specificity was 93.5% (98.5% for CT alone); positive predictive value was 83.5% (93% for CT alone); negative predictive value was 98.5% (83.5% for CT alone); and overall accuracy was 94% (85% for CT alone) (8)



Due to the difficulty in diagnosing the syndrome preoperatively, an exact diagnosis is often made on surgical exploration.

Differential diagnosis

Due to the ambiguous signs of this disease, differential diagnoses may include:

Cholecystitis
Choledocholithiasis
Bile duct strictures
Pancreatic cancer
Gallbladder cancer
Cholangiocarcinoma
Primary sclerosing cholangitis (PSC)
Caroli's Disease.

Treatment

There are different treatment plans recommended for the different classifications of Mirizzi's syndrome. The general theme of the treatment is a surgical approach involving cholecystectomy and removal of the offending calculus. However, due to the potential invasion into the CHD/CBD it can be a complicated cholecystectomy.

Type I Mirizzi syndrome involves a laparoscopic cholecystectomy plus choledochotomy and CBD exploration if necessary. If there is chronic inflammation of the area, anatomical changes can occur which make the surgery more difficult and conversion to open surgery is relatively common (50%-71%) (3,4). The normal conversion rate, in uncomplicated gallstone disease, from a laparoscopic to open cholecystectomy is 2-5% and up to 7% if there has been associated inflammation or interventional procedure. There are a proportion of patients (20.5%) with a calculus impacted in the gallbladder (Type I Mirizzi syndrome) who do undergo a straightforward cholecystectomy. (4).

Insertion of a T-tube, through a separate choledochotomy, for external biliary drainage, which is often removed around 4-6 weeks postoperatively, is also common, especially when there is an inflamed common hepatic duct wall. This can prevent future strictures.

The majority of patients with type II –IV classifications undergo cholecystectomy with the excision of the extrahepatic bile ducts. This then requires reconstruction of the biliary tree involving either a choledochoduodenostomy or Roux-en-Y hepaticojejunostomy. The latter is by far the more common procedure these days.

Risks of Surgery

Infection and bleeding are common risks of any surgery. The most common risk of laparoscopic surgery is the conversion to open surgery. The remaining risks are related to the cholecystectomy and reconstruction of the biliary tree needed to treat the Mirizzi syndrome.

At any join (anastamosis) of the reconstructive operation there can be a leak. In this setting an anastamotic leak would result in a bile leak. This would require drainage and a possible stent placement in the first instance using imaging techniques, however it may require a further operation.

After any reconstructive operation on the biliary tree there is always the risk of the formation of a biliary stricture. This often presents sometime after the initial operation. It can be managed with balloon dilation, often multiple times, either endoscopically or percutaneously. If the balloon dilatations are unsuccessful then a repeat reconstructive operation is required which is often a hepaticojejunostomy(3).

The mean hospital stay for these operations has been reported as 12 days for type I and 18 days for type II.

There is a reported association of Mirizzi's syndrome with bile duct or gallbladder cancer and one study reported an incidence of malignancy in patients with Mirizzi's syndrome as 27%. This is considerably higher than in patients with long standing cholelithiasis (2%) (7). It is therefore important to send any removed tissue to the histopathology laboratory to exclude malignancy.

Summary

Mirizzi's syndrome is a rare complication of chronic gallstone disease that can present with non specific symptoms and signs. Although rare, it should be considered in the differential diagnosis of complications of chronic gallstone disease.

Preoperative diagnosis with the aid or blood tests and imaging is difficult but important to avoid complications and damage to the biliary tree during surgery.

References

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