Concise Review

Bouveret’s syndrome. Narrative review

Anastasios Koulaouzidis;1 John Moschos2

Abstract

Obstruction at the level of the gastric outlet by a gallstone is defined as Bouveret’s syndrome. It is an uncommon form of gallstone ileus. A single gallstone of at least 2.5 cm in diameter is the most common underlying cause of Bouveret’s syndrome. Diagnosis is based on the clinical manifestations, existence of pneumobilia, visualization of lithiasis and demonstration of duodenal obstruction. Enterotomy or gastrotomy with or without cholecystectomy and fistula repair is the most common surgical therapy. It has high success rate, with acceptable surgical morbidity and mortality. Heightened awareness of this syndrome may lead to decreased morbidity and mortality.

Key words: Gallstones, gallstone ileus, Bouveret’s syndrome.

Gallstone disease is common, with 10% prevalence in the United State and Western Europe. However, it is symptomatic in only 20-30% of patients. The most common complications are acute cholecystitis, acute pancreatitis and ascending cholangitis. Less frequent complications include Mirizzi’s syndrome, cholecystocholedochal fistula and gallstone ileus.1 Gallstone ileus is uncommon, comprising only 1-4% of cases of intestinal obstruction7 and refers to small bowel obstruction resulting from the impaction of one or more gallstones after they have migrated through a cholecysto-enteric fistula.1 Less than 1% of gallstones migrate into the gut, causing 25% of non-strangulated small bowel obstructions in elderly population. The level of obstruction is usually at the terminal ileum. Obstruction at other scarce levels of distal jejenum occurs in 9% of cases, colon in 4%, rectum in 4% and at the duodenum in only 1-3%.3 As a general rule, the larger the stone the more proximal the obstruction.4 Gallstone ileus incidence peaks in elderly women and has high mortality rate. The reported higher rate in women is explained by the higher incidence of gallstones in women, attributed to the cholestatic effects of female sex hormones and prior pregnancies.5 Most patients have many other concomitant diseases that may increase the operative risk.6

Obstruction at the level of the gastric outlet by a gallstone is defined as Bouveret’s syndrome. In 1896, Leon Bouveret reported the first two cases of gastric outlet obstruction because of gallstones.7 Bouveret’s syndrome is an uncommon form of gallstone ileus, comprising only 1–3% of cases.8

In gallstone ileus, it is common for the stone to obstruct the distal small intestine where the lumen is narrowest and rarely the duodenum. Hence, Bouveret’s syndrome presents as a clinically distinct form of gallstone ileus because of the proximal site of obstruction. The mean patient age with Bouveret’s syndrome is 74.1 ± 11.1 years and the female-to-male sex ratio is 1.86.9 A single gallstone is the most common underlying cause of Bouveret’s syndrome. It is usually necessary for a stone to be at least 2.5 cm in diameter to cause obstruction. Nonetheless, multiple gallstones can be the cause of small bowel obstruction.4,10

The large size of gallstones in Bouveret’s syndrome is consistent with the known pathophysiology. Small gallstones typically pass via the cystic and common bile ducts through the gastrointestinal tract, whereas large gallstones cannot pass through these ducts and in Bouveret’s syndrome erode through the gallbladder or choledochal wall into the duodenal lumen, where they obstruct.9 The size of the gallstone, the duodenal-biliary inflammatory process and cholecysto-duodenal fistulas are some of the factors that may cause a gallstone to become impacted in the duodenum.

The clinical picture of Bouveret’s syndrome is nonspecific and sufficiently uncommon for the diagnosis to be overlooked. Diagnosis is based on the clinical manifestations, the existence of pneumobilia, the demonstration of duodenal obstruction and visualization of lithiasis by radiography or ultrasonography.11 The insidious clinical presentation and the lack of specific signs of biliary disease are responsible for the delayed preoperative diagnosis which leads to an overall high mortality rate. Common symptoms are nausea and vomiting, abdominal
pain or discomfort, haematemesis, recent weight loss, anorexia, constipation, melena and early satiety. Common signs of the syndrome are abdominal tenderness, signs of dehydration, abdominal distention, obstructive jaundice and pyrexia.

The findings of Rigler’s triad (small bowel obstruction, ectopic gallstones – presence of a calculus in its great dimension projected transversely to the right margin of T12-L1, and air in the biliary tree-pneumobilia) are observed on plain abdominal films. Subsequent abdominal plain radiographs may be useful to demonstrate the migration of the gallstone. Rigler’s triad can also be identified on abdominal CT images. The fistula may be seen if the tract is enhanced by positive oral or air contrast material. A secondary sign that may be useful is the identification of oral contrast material within the gallbladder. Pickhardt et al. described the use of MR cholangio-pancreatography for diagnosis of Bouveret’s syndrome with isoattenuating stones, and this may especially be true in a patient unable to tolerate oral contrast material. A combination of investigations is required before the diagnosis is made; radiological examinations, such as upper gastrointestinal contrast series and abdominal radiography, contribute to the diagnosis in 26% and 20% of cases, respectively. Although gastroscopy may lead to a more definite diagnosis in Bouveret’s syndrome, a direct abdominal x-ray is still the first step in the approach to these patients, so that, surgery can be performed as early as possible. Important findings on upper gastrointestinal series are a filling defect or mass in the duodenum, duodenal or pyloric obstruction, cholecysto-duodenal fistula and pneumobilia.

The first diagnosis of Bouveret’s syndrome by gastroscopy was made by Grove in 1976, with endoscopic retrieval achieved by Bedogni et al. in 1985. Endoscopically, the appearances can be bizarre with fibrotic-looking ulcers. Gastroscopy will reveal gastro-duodenal obstruction in virtually all cases and may show the obstructing stone in 69% of cases. The fistulous stoma was visualized in only 13% of examinations. In nearly all the other cases, the gallstone was not recognized because it was deeply embedded within mucosa. In this situation, the endoscopic diagnosis should be suspected when the observed mass is hard, convex, smooth, non-friable and non-fleshy; all these are characteristics of a gallstone. This finding may improve the sensitivity of endoscopy.

Ultrasonography may reveal gallstones in 75% of patients with Bouveret’s syndrome, of which 23% in the duodenum, and pneumobilia in 45% of cases. Ultrasound is usually difficult, due to overlying bowel gas, but if successful it depicts the “double-arch sign” which is considered pathognomonic. Also suggestive is a fluid-filled, distended stomach. Other ultrasonographic signs that have been described in patients with Bouveret’s syndrome are contracted gallbladder, thickened gallbladder wall and dilated duodenum. Major findings on abdominal CT are pericholecystic inflammatory changes extending into the duodenum, gas in the gallbladder, pneumobilia or cholecysto-duodenal fistula, filling defects (gallstones), thickened gallbladder wall, contracted gallbladder, and a gastro-duodenal mass or lesion. Therefore, although Bouveret’s syndrome is an atypical variant of a relatively rare disease, a timely diagnosis with appropriate imaging is possible and important.

Enterolithotomy or gastrotomy, with or without cholecystectomy and fistula repair, is the most common surgical treatment. They have a high success rate, with acceptable surgical morbidity and mortality. Although most data in the literature confirm the need for a conservative surgical approach, several authors report the one-stage repair of the cholecysto-duodenal fistula. The importance of diagnosis and early treatment is emphasized to improve prognosis.

In the low-risk patient, staged laparoscopic management of gallstone ileus and the associated cholecysto-duodenal fistula is feasible and appears to be safe. In such patients, imaging of the biliary tree is essential to detect silent choledocholithiasis which can be managed concomitantly and safely by the laparoscopic approach. Fistula repair is considered unnecessary because the fistula may spontaneously close, especially if the cystic duct is patent and residual gallstones are not present. In the most severe cases, enterolithotomy alone is usually advisable and subsequent cholecystectomy may not be required. However, a large retained gallstone in the gallbladder is an indication for cholecystectomy.

Nine percent of patients have successful non-surgical treatment, including endoscopic retrieval and endoscopic laser or mechanical lithotripsy and only 5% respond to extracorporeal shock-wave lithotripsy. Minimally invasive therapies, such as endoscopic retrieval, have a relatively low success rate; endoscopic removal of the gallstone should be attempted to minimize the necessary surgical procedure, whenever possible, but it is difficult and sometimes complicated. Distal gallstone ileus, due to migration of partially fragmented stones, can occur as a possible complication of laser lithotripsy treatment of Bouveret’s syndrome and it may require urgent enterolithotomy. Overall and despite a low success rate, endoscopic retrieval or other minimally invasive techniques are useful alternatives - when clinically appropriate- because of low morbidity and negligible mortality.

Conclusively, Bouveret’s syndrome has been associated with significant morbidity and mortality. Nowadays, earlier diagnosis by endoscopy and a combination of ultrasonography, abdominal CT and abdominal radiography (the signs of Rigler’s triad) and the advent of minimally invasive techniques have reduced the mortality to 12%. Heightened awareness of this syndrome may lead to additional decreased morbidity and mortality.
References