Bouveret Syndrome: The Rare Challenging Case of the Day

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Abstract

Bouveret syndrome (BS) is an extremely rare disease. We report the case of a 54-year-old man who presented to our department with acute vomiting. An upper digestive endoscopy was first done, revealing an enclaved gallstone in the second duodenum. The correct diagnosis was then made using cross-sectional imaging.

Introduction:

During clinical practice, complications related to cholelithiasis such as pancreatitis, cholecystitis, common bile duct lithiasis, and gallstone ileus are not uncommon(1). During gallstone ileus, the stone is often blocked at the level of the terminal ileum and the ileocecal valve. The impaction of the calculus at the level of the distal stomach or the proximal part of the duodenum following a passage via a cholecysto-duodenal, cholecysto-bulbar or cholecysto-gastric fistula defines Bouveret syndrome. It’s a rare biliary complication that is seen in 1 to 3% of gallstone ileus patients with high mortality(2). Given the rarity of the pathology, diagnostic and therapeutic management are not well standardized. We wrote this paper to review current literature highlighting the pathogenesis and management of the disease.

Case Presentation:

A 54-year old male presented with abdominal pain and vomiting evolving over five days. His previous history was consistent with a diagnosis of acute necrotizing pancreatitis one year ago (Figure 1). Physical examination showed tenderness in the epigastric region and right upper quadrant without clinical signs of dehydration. The white blood cell count was 7,200/mm³ and the CRP was 30 mg/dL. The renal function was normal without hydroelectrolytic disorder. Total bilirubin and alkaline phosphatase were normal. Lipasemia was normal. In front of this clinical presentation, an upper gastrointestinal endoscopy was performed first and showed an obstacle in the second duodenum (Figure 2). Abdominal magnetic resonance imaging (MRI) revealed an enclaved gallstone of three centimeters in the second duodenum (Figure 3), gastric distension...
The surgical procedure consisted of cholecystectomy and extraction of the gallstone through a midline incision. Faced with the significant inflammatory reaction around the duodenum and the presence of a large fistula, we performed a duodenostomy, pyloric exclusion, and gastro-jejunal anastomosis. The patient was discharged seven days after the operation with a good clinical outcome. The duodenostomy tube was removed one month later. An abdominal scan was performed six months after the operation and was normal. During 12 months of follow-up, the patient was symptom free.

Discussion:

Bouveret syndrome is a rare complication of cholelithiasis. It was first described by a Parisian internist, Leon Bouveret, in 1896. Subsequently, only 315 cases were reported between 1967 and 2016. This rare pathology most often affects elderly subjects with several comorbidities and is burdened with a high mortality rate of 12 to 30%. The main risk factors for the development of BS are: cholelithiasis, female gender, age over 60 years, and stones larger than 20 mm. Our clinical case is particularly marked by the young age and the absence of comorbidities.

The pathophysiology of Bouveret syndrome is explained by the development of adhesions between the gallbladder and a close segment of the upper digestive tract following recurrent episodes of cholecystitis. Furthermore, the huge stone’s pressure induces necrosis, which leads to a biliointestinal fistula. The most frequent type of fistula is a cholecysto-duodenal fistula, while cholecysto-gastric fistulas are less common. The stones at the origin of Bouveret syndrome have, in the majority of cases, a size greater than 25 mm. The clinical presentation is not specific. BS can cause nausea, vomiting, abdominal pain, weight loss, anorexia, or gastric outlet obstruction. Digestive hemorrhage, related to erosion of the mucosa by the gallstone, can be observed in 13.8% of cases. Our patient presented with acute vomiting and abdominal pain.

An elevation of CRP and leukocytes can be observed due to inflammatory phenomena. In the case of severe vomiting, metabolic alkalosis and hypokalemia can be observed. Our patient had an elevated CRP with a normal blood cell count.

Oesogastroduodenal fibroscopy is generally the first examination to be performed. It may show gastric distension and a blocked stone at the level of the distal part of the stomach or the proximal part of the duodenum. However, the calculus is only found on endoscopy in 69% of cases. This can be explained by the food impaction upstream of the gallstone. For the biliointestinal fistula, it is only found in 13% of cases on endoscopy.

On abdominal X-ray, Rigler’s triad, which includes intestinal obstruction, pneumobilia (air in the biliary tract) and gallstone in the intestinal lumen, is only observed in 30 to 35% of cases. Ultrasonography, computed tomography (CT), and MRI are other helpful examinations in diagnosis. Abdominal CT is a good test for the positive diagnosis of Bouveret syndrome with high sensitivity and specificity. It makes it possible to objectify the blockage of the calculus in the digestive lumen, gastric distension, and biliointestinal fistula. Regarding MRI, it is especially interesting for better visualization of the biliary tract and for the diagnosis of isodense gallstones on the CT. In our presented case, MRI was superior to computed tomography in the study of the biliary tree before surgical treatment.

Several surgical and non-surgical treatment options have been described for BS. For non-surgical methods, we find endoscopic extraction of the stone and other techniques aimed at fragmenting the stone, such as endoscopic lithotripsy, electro-hydraulic lithotripsy, endoscopic laser lithotripsy, and extracorporeal lithotripsy. A study showed that these techniques were ineffective in 42% of cases. The main limitations of endoscopic treatment are: the migration of stone fragments with the risk of distant gallstone ileus; the impossibility of repairing the fistula; the size of the stone; and the operator’s experience. Not to mention that the gallstone is not found on endoscopy in 30% of cases. As a result, surgical treatment remains a reference for BS to this day. Surgical management is based on three components: stone extraction, fistula repair, and cholecystectomy. It can be performed in one or two steps. For the elderly with comorbidities, the
two-step procedure is safer. The one-step procedure is more commonly used in young patients. Recently, with the extensive application of laparoscopy, the laparoscopic approach has become a safe and effective alternative to open surgery(13). In the literature, the cases reported with duodenum exclusion and gastro-jejunal anastomosis have been mostly described in cases of unreparable duodenal lesions or difficult localisation of the gallstone, particularly at the level of D2/D3(14). In our case, the reason for performing gastrojejunal anastomosis was unfixable lesions of the duodenum. Surgical treatment was successful for our patient.

**Conclusion :**

Bouveret syndrome is a rare but scary complication of gallstones. Therefore, knowledge of its risk factors, symptoms, radiological and endoscopic findings is important for a rapid diagnosis to minimize morbidity and mortality. Treatment is based on surgery. However, endoscopic treatment remains promising and needs to be improved, especially for such vulnerable patients.

**Conflicts of Interest :**

The authors declare no conflicts of interest regarding the publication of this paper.

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![Axial CT slide one year before the onset of Bouveret’s syndrome showing pancreatic necrosis (blue arrow) and a large 3 cm stone in the gallbladder (green arrow)](image)

**Figure 1 :** Axial CT slide one year before the onset of Bouveret’s syndrome showing pancreatic necrosis (blue arrow) and a large 3 cm stone in the gallbladder (green arrow)
**Figure 2**: Demonstration of the stone embedded in the second duodenum before (A) and after washing (B) on endoscopy.

**Figure 3**: Axial MRI slide, sequence T2, at the portal phase, showing the stone wedged in the second duodenum in hypo-signal (red arrow) with a marked periduodenal inflammation (blue arrow).
**Figure 4**: Coronal section of abdominal MRI T2 sequence showing gastric distension (blue arrow) upstream of the calculus (red arrow)

**Figure 5**: Axial slice of an in-phase abdominal MRI showing pneumobilia (blue arrow)
Figure 6: Iconography of calculus after surgical extraction

References:


