Pictorial case series review on Bouveret’s Syndrome.
Crucial imaging findings to identify.

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Authors: L. Vo, S. Dimmick; St Leonards/AU
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Learning Objectives

To identify imaging findings of Bouveret's Syndrome in order to facilitate early diagnosis, enabling prompt treatment, thereby reducing mortality and morbidity.
Background

Bouveret's Syndrome was first described in 1770 by Beaussier and named after French physician, Leon Bouveret who published the first case in 1896 [1].

Cholelithiasis is a relatively common health problem. 35% of women and 20% of men develop gallstones by the age of 75 [1]. The prognosis for most is good however, in small group (~6%), complications occur. Rarely, a fistula may develop secondary to adhesions, which form between the gallbladder and bowel wall as a consequence of inflammation, compromised arterial supply and impaired venous drainage [2, 3, 4]. Subsequent pressure necrosis and compression of a calculus against the wall of the gallbladder may result in the formation of a fistula. The cholecystoduodenal fistula is most common type of fistulous formation followed by cholecysto colic and choledochoduodenal fistulas [4]. Most calculi pass through cholecystoduodenal fistula and the duodenum without causing obstruction (85%) [2]. Spontaneous biliary-enteric fistulas are uncommon, over 90% of fistulas develop as a complication of cholelithiasis or choledocholithiasis [5].

In 15%, the calculus causes an obstruction [2]. Obstruction most commonly occurs in the terminal ileum (70%) [1, 3, 4] which is classified as a classic "gallstone ileus".

In 1-3% of cases, the gallstone becomes obstructed in the duodenum, resulting in gastric outlet obstruction. This phenomenon is called Bouveret's syndrome [1, 4]. The obstructing gallstone is usually greater than 2.5 cm [3]. Bouveret's syndrome is thus defined as gastric outlet obstruction secondary to gallstone impaction at the level of the stomach or duodenum as it passes through a biliary-enteric fistula. Bouveret's syndrome occurs most commonly in elderly women with a history of biliary disease [2] - classically, over 65 years of age [3].

Diagnosis of Bouveret's syndrome is important as mortality may be as high as 33% [2, 4]. The high mortality is attributed to advanced age of the patients who commonly present with Bouveret's, associated presence of comorbidities and elevated surgical risk of such candidates.

Diagnosis

The presenting signs and symptoms of Bouveret's Syndrome are non-specific. Most common symptoms include nausea and vomiting, abdominal pain and, uncommonly, haematemesis (secondary to duodenal erosions or erosion of the coeliac axis) [4], weight loss, anorexia [1, 8].
Examination findings may include abdominal tenderness, distension and dehydration [1]. There may be an obstructive pattern of deranged liver function tests with elevated bilirubin and alkaline phosphatase may be seen [4].

Diagnosis is historically based on endoscopy, demonstrating visualization of the calculus causing obstruction [1]. This is seen in 69% of cases [1]. In 31% of cases, no stone and or fistula can be identified. In the remaining cases, the stone cannot be visualized or is only partly visualized as it compresses the duodenal wall [1]. Imaging with radiographs, CT, MR or ultrasound are increasingly contributing to early diagnosis. Identification of the calculus in the distal stomach or duodenum is highly suggestive of a diagnosis of Bouveret's Syndrome. Detection of the fistulous tract is diagnostic.
The classic imaging finding triad of gallstone ileus is known as Rigler's triad - intestinal obstruction (partial or complete), pneumobilia and an ectopic gallstone within the stomach or duodenum. As stated previously, the ectopic gallstone is commonly larger than 2.5 cm [3]. In some clinical cases, classic imaging findings are present, while in other cases, the diagnosis should be suspected upon identification of indirect signs of Bouveret's. Familiarity with the variety of signs may facilitate correct diagnosis, enabling prompt management.

CT is diagnostic in approximately 60% [1]. It may be useful in identifying the precise level of obstruction, identification and location of the duodenal fistula and appearance of the gallbladder. In about 15-25%, the gallstone is not radiodense. In these cases, administration of oral contrast may facilitate identification of the stone by surrounding the hypodense, non-calcified calculus. Alternatively, magnetic resonance cholangiopancreatography (MRCP) may be of value for non-calcific stones. The size of the calculus is significant as many cases of failed endoscopic treatment are higher when the stone is larger than 2.5 cm [1]. The larger stones are more often associated with ischaemic ulceration of the duodenal wall. CT is useful in identifying the point of obstruction and position of the offending calculus, aiding planning for endoscopic or surgical intervention.

Other features to be identified on imaging include the presence of other intraluminal calculi. This may be a predictor of risk of recurrence and facilitates in decision for concurrent cholecystectomy.

The identification of a biliary enteric fistula may be difficult in the acute phase with the concurrent changes of inflammation. Indirect signs of biliary enteric fistula include intraluminal air in the gallbladder indicating erosion of the duodenal wall an obstructing gallstone. The presence of oral contrast in the gallbladder further herald's possible fistulous tract formation.

Common imaging findings of Bouveret's Syndrome on CT have summarised in Table 1.

Radiographs are less useful but may demonstrate pneumobilia and if the gallstone is calcified, it will be seen projected over the region of the stomach or duodenum. Subsequent radiographs may show changing position of the gallstone indicating that the stone lies outside the gallbladder.
Imaging findings of Bouveret's are difficult on ultrasound as the gallbladder tends to be collapsed, air-filled or both. When the gallbladder is not seen, the impacted duodenal calculus could be misinterpreted as an orthoptoic gallstone [4, 5].

Contrast swallow studies, may help to identify the presence and position of radio-opaque stones or delineate the fistulous tract when it is unclear on other imaging [4].

**Table 1. Imaging findings to identify and note in Bouveret's syndrome [1, 2, 3, 4].**

*Rigler triad*

- Intestinal obstruction - partial or complete
- Pneumobilia
- Ectopic gallstone

_Ectopic gallstone_ (in stomach or duodenum):

- Non radiodense ectopic gallbladder calculi may become visible on CT when surrounded by oral contrast
- Size?
- Position?
- Other ectopic intraluminal calculi?

_Collapsed gallbladder_

_Identification of biliary-enteric fistula_ (cholecystogastric or cholecystoduodenal, cholecystocolic and choledochoduodenal)

- Direct visualization
- Indirect signs
  - Intraluminal gallbladder air suggestive of erosion of the duodenal wall by the gallbladder
  - Oral contrast in gallbladder suggesting presence of biliary-enteric fistula
  - Obliteration of fat planes between the gallbladder and duodenum

_Treatment_

Bouveret's can be managed surgically or endoscopically [1, 3], the later being the preferred intervention due to the significant comorbidities associated with the typical patient group, despite the low success rate [1]. This includes endoscopic extraction, and endoscopic laser lithotripsy (ILL). Other alternatives to surgery include extracorporeal
shockwave lithotripsy (ESWL) and intracorporeal electrohydraulic lithotripsy (IEHL) [1]. These methods are more successful when the gallstone is impacted proximally. When endoscopic extraction has failed, surgery - enterolithotomy, duodenectomy or gastrotomy or stone extraction must be considered [1]. Large stones are comprised of a hard outer shell with soft inner core, rendering mechanical fragmentation with endoscopic forceps or laser more challenging [1].

The need for surgical repair of the fistula is contentious due to spontaneous closure in some cases. The argument for fistulous repair lies with some evidence that there is increased risk of recurrence of Bouveret's and elevated risk for development of gallbladder cancer [5].

Case series

A series of images is presented demonstrating the various features of Bouveret's syndrome by way of case report.

Case 1: A 71-year man who presented with upper abdominal pain and haematemesis on a background of cholelithiasis. Clinical examination demonstrated distended but soft and non-tender abdomen. The patient's vital parameters were normal. Blood investigation showed elevated C-reactive protein (162 g/L), moderate leukocytosis (WCC 20.5 x 10⁹/L) and mildly raised gamma-glutamyl transpeptidase (82 U/L). The remaining liver function tests were normal. The working diagnosis was gastritis but to exclude cholecystitis. An abdominal ultrasound was requested for further evaluation.

Ultrasound demonstrated a non-mobile 2.6 cm calculus in the gallbladder with thickening of the gallbladder wall and transducer tenderness, suggestive of acute cholecystitis. The gallbladder wall is thickened measuring up to 7 mm. Linear echogenic foci in the gallbladder wall were deemed suspicious for porcelain gallbladder. A CT was requested for further evaluation of the unusual appearance of the gallbladder wall.

CT demonstrated large 5.2 cm laminated calculus in the gallbladder with minor pneumobilia and pericholecystic stranding. Further 4.3 cm calculus was also identified in the third part of the duodenum with upstream dilation of the stomach and duodenum, consistent with Bouveret's syndrome (Fig. 1-7). Ultrasound images have been provided to demonstrate the difficulty in detecting Bouveret's on ultrasound alone (Fig. 8 & 9).

The patient underwent surgical management with subtotal cholecystectomy. At the time of the operation, a large fistulous hole was seen in the second part of the duodenum. The large stone from the third part of the duodenum was extracted through the fistulous
tract. The fistula was plugged using the neck of the gallbladder and reinforced using an omental patch. The patient had an uneventful recovery.

Case 2: (Figure 10-13) An 89 year old man presented with three-day history of malena and two large black vomits on background of cholecystitis. Clinical examination demonstrated a soft, non-tender abdomen with no blood on per rectal examination. Blood investigation demonstrated mild leukocytosis (WCC 15.4 x 10^9/L). The remaining blood tests were unremarkable. Specifically, the liver function tests were normal.

Gastroscopy demonstrated duodenal narrowing and circumferential ulceration. A CT was requested for investigation of possible mass in the duodenum. This demonstrated 2.8 cm low-density lesion in the first part of the duodenum, with stranding in the superior and descending duodenum, suspect for contained perforation. This lesion was later confirmed surgically to be a large (radio-opaque) calculus. On CT, the gallbladder was poorly identified. There was evidence of marked gastric outlet obstruction.

Surgery demonstrated an inflamed and contracted gallbladder, adherent to the duodenum. A large calculus at the junction of D1 and D2 was removed and a duodenectomy was performed.
Fig. 1: Bouveret's syndrome. Coronal portal venous image demonstrates a large calcified ectopic calculus in the horizontal part of the duodenum and within the gallbladder. Note the partly imaged nasogastric tube that has partly decompressed the stomach.

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**Fig. 2:** Axial non contrast images demonstrate pneumobilia, large laminated calculus in gallbladder and minor pericholecystic stranding. The pneumobilia is unusual for a case of simple acute cholecystitis. Bouveret's and gallstone ileus should be considered and a search for a fistulous tract and ectopic calculi should be undertaken.

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Fig. 3: Axial non contrast images through a second laminated obstructing calculus in a confirmed case of Bouveret's syndrome. This calculus is located in the third part of the duodenum. This is a crucial finding in Bouveret's.

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Fig. 4: Coronal portal venous images: A fistulous tract may be difficult to delineate. Ectopic gas (white arrow) and obliteration of fat planes between duodenum and gallbladder is highly suspicious for fistulous formation - a key finding in Bouveret's syndrome.

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Fig. 5: Bouveret's syndrome: (coronal portal venous phase). The fistulous tract may be difficult to visualise. Here, obliteration of fat planes between duodenum and gallbladder (white arrow) and the presence of ectopic gas (Fig. 4) should raise high suspicion for fistula.

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Fig. 6: Axial non enhanced images again demonstrating obliteration of fat planes between the second part of the duodenum and the gallbladder (white arrows), highly suspect for fistulous communication, raising the possibility that this is a case of Bouveret’s syndrome. This was confirmed surgically.

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Fig. 7: Further portal venous enhanced axial images demonstrating definite obliteration of fat planes between the second part of the duodenum and the gallbladder (white arrows). There is very likely a fistulous communication between the two structures. This was confirmed surgically.

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**Fig. 8:** Bouveret's syndrome, easily mistaken for cholecystitis. Ultrasound images from the same patient as in Fig. 1 to 7 have been provided to illustrate the difficulty in recognising Bouveret's on ultrasound alone. Here, there is evidence of gallbladder wall thickening and a large stone (Fig. 9). Lack of awareness of the disease entity of Bouveret's syndrome could easy result in acute cholecystitis as the final differential.
Fig. 9: Further ultrasound images provided to illustrate how easily Bouveret’s syndrome could be over-looked or misinterpreted as acute cholecystitis.

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**Fig. 10:** Case of Bouveret’s with non-radiodense calculus demonstrating the value of oral contrast. Round hypodense structure in the superior part of the duodenum is seen. Without oral contrast, this non-calcified ectopic gallstone could easily be overlooked. It would not be discernable on radiography. Note too the gastric outlet obstruction.

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**Fig. 11:** Further oral and IV contrast enhanced images of Bouveret's with non-radiodense calculus. Note the gas and hypodense change in the superior part of the duodenum. This could be confused for food debris. Note how the oral contrast surrounds this round hypodense mass rendering it recognisable as a possible non calcific stone. This was confirmed as a non-calcified stone post operatively. Gas in the gallbladder is suggestive of biliary-enteric fistula (black arrow), although, the fistula itself is easy to miss given the surrounding inflammatory stranding. Absent fat plane between gallbladder and duodenum (white arrow) is again suspicious. Again note gastric outlet obstruction (S).
**Fig. 12:** The biliary-enteric fistula may be difficult to identify in Bouveret’s syndrome. The combination of findings demonstrated in Fig. 10-11 and clinical history should alert the radiologist to raise the possibility that this is a biliary-enteric fistula. The fistula was confirmed at surgery. Again note gastric outlet obstruction (S).

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Fig. 13: The biliary-enteric fistula is often difficult to identify in Bouveret's syndrome however, with the other supplementary CT findings (Fig. 10-12) in addition to the clinical signs, the presence of pneumobilia with a tubular structure closely apposed to the duodenum and obliteration of fat planes (white arrow) is highly suspect for biliary-enteric fistula.

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Conclusion

Bouveret's syndrome is a rare variant of gallstone ileus. The syndrome is associated with a mortality rate of up to 33%. Early and accurate detection using non-invasive means such as CT or MRCP may help reduce associated mortality. Knowledge of Bouveret’s syndrome and the radiological findings will facilitate early diagnosis and prompt management.
Personal Information

Dr. Linh Vo
Royal North Shore Hospital, Sydney.

Dr. Simon Dimmick
Royal North Shore Hospital, Sydney.


