Bouveret’s Syndrome: An Extensive Summary of the Literature

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Abstract

Bouveret’s syndrome is a rare condition characterized by gastric outlet obstruction from an ectopic gallstone, facilitated by aberrant connection of the biliary and luminal gastrointestinal tracts. Establishing a diagnosis of Bouveret’s syndrome can be aided by clinical, radiologic, and endoscopic assessments and importantly relies on prompt diagnosis and management. Despite its rarity, the disease confers a high rate of morbidity and mortality. We therefore aimed to provide a review of the literature to enhance awareness of this phenomenon to broaden the differential diagnosis of gastric outlet obstruction and subsequent management of this particular—and peculiar—etiology.

Keywords: Bouveret’s syndrome; Gallstone ileus; Cholecystoenteric fistula; Gastric outlet obstruction; Cholelithiasis

Introduction

Cholelithiasis is a prevalent disease, affecting 7.1% of the United States population [1]; however, the vast majority of cases are asymptomatic and without consequence. It is estimated that less than one percent of patients develop complications, the most common of which include acute/chronic cholecystitis, cholangitis, cholecdocholithiasis, and pancreatitis. An exceptionally rare complication is gallstone-related luminal gastrointestinal (GI) tract obstruction, reported to occur at rates of 0.3-0.5%, and comprising no more than five percent of small bowel obstructions [2,3].

While most luminal obstruction from a gallstone occurs in the terminal ileum, as this is the narrowest part of the bowel, 1-3% of impactions occur proximally, leading to gastric outlet obstruction (GOO), a phenomenon known as Bouveret’s syndrome. Though initially described by Beaussier in 1770 and Bonnet in 1841, GOO from an ectopic gallstone derived its eponym after the French internist Léon Bouveret published two case reports of this condition in the Revue Medicale in 1896 [4,5].

Clinical, radiologic, and endoscopic signs of Bouveret’s syndrome demand prompt recognition to arrive at an early diagnosis. Esophagogastroduodenoscopy (EGD) can miss the gallstone in a third of the cases, particularly if the stone is deeply embedded in the mucosa [6,7]. The delay in diagnosis, along with the comorbidities commonly observed in the geriatric population most frequently afflicted by Bouveret’s syndrome, likely contribute to the high mortality rate—estimated to be 12-30%—observed in this condition [8]. Ninety-four percent of cases of Bouveret’s syndrome described in the literature are either case reports or editorial letters. This highlights the need for regular reappraisal and synthesis of the published accounts of Bouveret’s syndrome to help calibrate knowledge of clinical presentation, diagnostic and therapeutic modalities, and—ultimately—to maintain Bouveret’s syndrome’s position within the differential
diagnoses for GOO as an etiology that, despite its rarity, confers a high mortality that may be mitigated by prompt diagnosis and treatment [9].

**Pathogenesis and Presentation**

After an episode of acute cholecystitis, the ensuing inflammation and cholecysto-biliary adhesions, along with mechanical pressure exerted by gallstones on the gallbladder and bowel wall, may contribute to a pattern of ischemic injury of the apposed gallbladder and bowel wall. Such a set up creates potential for fistulization between the gallbladder and bowel through which the gallstone could pass [10]. Beyond acute cholecystitis, there have been a few reports of the development of a cholecystoenteric fistula secondary to a gallbladder malignancy [11-13]. Bilioenteric fistulas most frequently manifest as cholecystoduodenal fistulas and account for 68% of the cases. Less commonly, cholecystocolic or cholecystogastric fistulas comprise 17% and 5% of the cases, respectively [9]. It is probable that the cholecystogastric fistula is the rarest due the thickness of the gastric wall [14]. Nevertheless, even when a bilioenteric fistula is present, the majority of ectopic gallstones are eliminated through defecation or emesis. Clinically-relevant symptoms are more likely to arise with larger stones, as well as in the context of either a pre-existing stenosis or post-surgical anatomy of the GI tract [9,15].

Bouveret’s syndrome has a female predominance, ranging from 1.4:1 to 1.9:1, which is unsurprising in light of the well-established prevalence of cholelithiasis in women [8,16,17]. A history of known cholelithiasis or paroxysms of biliary colic are present in the majority of cases. The median age at presentation is 74 years [6,8,17]. Patients typically present with non-specific signs and symptoms of GOO, including nausea/vomiting (86%), abdominal pain (71%), hematemesis (15%), unintentional weight loss (14%), anorexia (13%), constipation (9%), melena (6%), early satiety (3%) and dysphagia (2%) [6]. Commonly, symptoms of GOO develop a week before medical advice is pursued [18]. Hematemesis may result from a Mallory Weiss tear or esophageal mucosal injury from the forceful expulsion of the stone during retching [9,18-20]; more rarely, such bleeding has been observed from stone-induced erosion into splanchnic arteries [9,18,21]. In a small subset of cases, emesis can be severe enough to induce Boerhaave syndrome [22]. Findings on physical examination are typically non-specific, and often include abdominal tenderness, abdominal distension, and signs of intravascular depletion [6]. Physical manifestations may become more severe and specific with particular complications of Bouveret’s syndrome, such as biliary or duodenal retroperitoneal perforations [23] or superimposed acute pancreatitis [24,25].

**Diagnosis**

GOO has a wide differential diagnosis, particularly in a geriatric patient. Sinister etiologies, such as malignancy, deserve thorough investigation. Though rare, Bouveret’s syndrome has a high mortality rate and thus deserves acknowledgement as a potential cause, especially in those with underlying neoplastic or inflammatory conditions that can predispose to bilioenteric fistulization.

Laboratory studies are non-specific and do not generally help in establishing a diagnosis of Bouveret’s syndrome. Results associated with intravascular depletion (elevated blood urea nitrogen, creatinine, and sodium) are typical but rarely narrow the differential diagnosis. One of the highly specific findings on imaging is Rigler’s triad: distended stomach, pneumobilia, and an ectopic gallstone. However, Rigler’s triad is observed in only half of the cases of Bouveret’s syndrome [26]. The initial screening step in diagnosing a patient with suspected GOO is typically an abdominal plain X-ray (XR) which has a sensitivity ranging from 40-70% and a specificity of only 21% for Bouveret’s syndrome [8]. Abdominal ultrasonography (US), while is the method of choice to detect cholelithiasis, is operator-dependent and often hampered by patient discomfort with abdominal pressure and gaseous/liquid distention of proximal bowel from the obstruction, all of which may limit the examination’s yield [27]. Combining an abdominal XR and an US increases sensitivity of detecting gallstone ileus to 74% [28].

The investigation of choice in detecting bowel obstruction is a contrast-enhanced computed tomography (CT) scan. The advantages of CT scans include higher sensitivity (90%), specificity (100%), and accuracy (99%) in detecting gallstone ileus [27]. Furthermore, a contrast-enhanced CT can provide a detailed anatomical outline (additionally delineating a fistulous tract and the size of the stone), identify complications of the syndrome (such as bowel wall ischemia or edema), and help evaluate for alternate etiologies of GOO [27]. Because oral administration of contrast would worsen symptoms of GOO and would not be easily interpreted due to the already fluid-distended bowel, intravenous contrast is preferred [29]. Furthermore, oral contrast administration would delay the CT imaging due to the anticipated length of transit time of the contrast through the bowel [30,31]. Interestingly, Haddad et al. reported the disimpaction and migration of a gallstone after ingestion of contrast. This was attributed to the hyperosmolality of the contrast, thought to increase the gut’s motility and reduce bowel edema [32]. While rarely encountered in this setting, if the patient presented with a bowel perforation, oral contrast administration could lead to a fatal peritonitis [23]. Nevertheless, CT scan with intravenous contrast is not without any limitations. A significant number of patients present with vomiting...
and intravascular depletion with concomitant acute renal injury, increasing the detrimental effects of the intravenous contrast [33]. Furthermore, up to 20% of the gallstones may be isodense, relative to the surrounding tissues and fluid, hampering the ability of CT scans to detect these stones [6,8,27,34,35]. This can be bypassed by doing magnetic resonance-based imaging techniques, which have been noted to detect Rigler’s triad in nearly all cases where it was present, compared to a detection rate of 78% by CT scan, though this adds considerable time and expense to the diagnostic work up instead [27,36,37].

Direct visualization of an ectopic gallstone can be provided by EGD, which has the additional advantage of being therapeutic. Since the first endoscopic diagnosis of Bouveret’s syndrome in 1976, EGD has been viewed as a reasonable diagnostic tool in Bouveret’s syndrome; however, it has been reported to fail in visualization of the gallstone in nearly a third of cases [6,7,9]. This is suspected to result in the setting of a gallstone that has embedded deeply into the mucosa, with surrounding edema and inflammation that further obscures it. Experts therefore recommend a careful endoscopic examination of the gastric outlet’s mucosa if Bouveret’s syndrome is suspected and have further indicated that identifying a smooth, hardened, convex, non-fleshy mass are good clues for an embedded stone [6].

Management

There have been sporadic reports of spontaneous resolution of Bouveret’s syndrome after conservative management [38,39]. However, the chances of spontaneous resolution are less than 1% [6]. Moreover, a dislodged stone carries the risk of distal obstruction. As any delay in treatment of Bouveret’s syndrome increases the already high rate of mortality, patients should not be managed by watchful expectancy unless there are significant contraindications to endoscopic or surgical intervention [16].

Endoscopic management remains the first line of treatment of Bouveret syndrome, though it does enjoy a markedly diminished success rate compared to surgical intervention [43.0% vs. 94.1%, P < 0.001] [16]. The trend for initial endoscopic intervention is largely borne from the comparatively lower combined mortality and morbidity rates of endoscopy vs surgery [1.6% vs. 17.3%, P = 0.003] for Bouveret’s syndrome [7]. Prior to therapeutic endoscopy, decompression of the distended stomach by a nasogastric tube should be performed to reduce the risk of aspiration. With continued evolution and refinement of endoscopic techniques over the past two decades, more endoscopy-based treatment options have become available, with a paralleled improvement in success rates for Bouveret’s syndrome, rising from 13.6% - 18.0% to the current success rate of 43.0% [6,16,40,41]. Endoscopic extraction of intact stones by nets, graspers, snares, or baskets can be attempted. During extraction, a latex rubber or an esophageal overtube can be used to protect the esophagus.

Still, stones exceeding 2.5 cm in diameter require fragmentation by lithotripsy for safe removal. Mechanical lithotripsy [ML] is the most common method used to that end. Fluoroscopy may be used in conjunction to ensure that the instrument did not wrongfully enter the cholecystoenteric fistula rather than passing between the stone and the wall of the duodenum. The second most common method of lithotripsy is electrohydraulic lithotripsy [EHL]. Maximal EHL intensity may be needed due to the large gallstones commonly encountered in cases of Bouveret’s syndrome [42]. Caution must be taken to keep the tip of the probe adhered to the gallstone, as the shockwave can perforate the bowel [43]. It is also advised to keep the endoscopic lens above the water level to avoid disturbance of the view during fragmentation [44]. Laser lithotripsy [LL] and extracorporeal shockwave lithotripsy (ESWL) are other modalities of treatment, the former being more effective and successful in clearing the gallstones [45]. In addition to the higher failure rate, ESWL is burdened by the lack of endoscopic control. It is advised to utilize intraoperative ultrasound during ESWL to target the gallstones, as the intestinal gases usually impede plain XR [7,46]. However, no head-to-head trials have been made comparing the different forms of lithotripsy in managing Bouveret’s syndrome. While the endoscopic modality is at the discretion of the operator and institutional availability, ultimately it is preferable to manage the patient in a single session to avoid the migration of any stone fragments that can lead to re-impaction and distal ileus [47].

The reported success rate of endoscopic extraction in Bouveret’s syndrome is just 43%, and it is suspected to be even lower, as multiple case reports of successful surgical intervention may have neglected to mention a prior failed endoscopic attempt [7]. While endoscopy as a first line of treatment is agreed upon by the majority of clinicians familiar with Bouveret’s syndrome, its relatively lower chances of success compared to surgery have prompted clinicians to consider how to appropriately triage patients to the ideal therapy. Ong et al. observed that certain factors were more associated with successful endoscopic extraction. These included gallstone with diameter ≤ 4 cm, impaction of the gallstone in the proximal half of the duodenum, and the use of combination therapy (e.g. endoscopic retrieval via net/basket + a method of lithotripsy). These items were subsequently parlayed into a scoring system to predicting the chances of endoscopic success [16].
The surgical approach in treating Bouveret’s syndrome remains complex and provides rich substrate for debate. Considerations for surgery for Bouveret’s syndrome include access (enterotomy vs gastrotomy), the merits of fistula takedown, and the merits of cholecystectomy. The latter two elements have received particular attention in the literature as worthwhile discussion points, particularly in regards to timing (at time of surgical stone extraction vs delayed). To date, there is no consensus on these questions. Advocates of the one-stage procedure (lithotomy with cholecystectomy and fistula take down) argue that saving the patient from an additional operation is reasonable, and that one should endeavor to remove a fistulous gallbladder, which is prone to developing cholecystitis, cholangitis, cholangiocarcinoma, gallbladder adenocarcinoma or recurrent gallstone-related obstruction in the future. The chances of developing those complications may reach up to 17% and the risk of developing cholangiocarcinoma increases 15-fold in patients with cholecystoenteric fistula [48]. Furthermore, there is a 5% risk of gallbladder adenocarcinoma associated with cholelithiasis [18].

On the other hand, a United States nationwide study showed that performing a simultaneous cholecystectomy was independently associated with nearly a three-fold rise in mortality in patients with gallstone ileus [49]. This may be explained by the increased duration of surgery, as well as the technical difficulty of repairing a bowel defect in the setting of abundant inflammation, edema and tissue friability [48]. Moreover, cholecystoenteric fistulas tend to resolve spontaneously in half of the cases [48-50]. Delayed cholecystectomy and fistula take-down should be discussed with the patient and advised if malignancy is suspected or present [51].

### Variants of Bouveret’s Syndrome

As described in this review, Bouveret’s syndrome classically presents as GOO from an ectopic gallstone that was permitted through bilioenteric fistulization; however, there are case reports of Bouveret syndrome variants that warrant mentioning. Three reports have described variants of Bouveret’s syndrome in which the GOO was caused by the mechanical compression on the duodenum by a gallstone-distended gallbladder rather than a gallstone itself [52-54]. Indicators of this “pseudo-Bouveret’s syndrome” include the absence of pneumobilia or signs of cholecystoenteric fistula on imaging and the positioning of a distended gallbladder vertically against the duodenum along its longitudinal axis. These cases could be treated either by decompression of the distended gallbladder or a cholecystectomy. Similarly, another variant described by Park et al. illustrates a gallstone that migrated to the duodenum via a cholecystoenteric fistula, akin to a typical Bouveret’s syndrome; however, the chronically anchored large sized stone in the duodenum, rather than causing the obstruction directly, led to the development of a submucosal mass that obstructed the duodenum [55]. Mittal et al. presented an intriguing case of Bouveret’s syndrome in a patient who underwent a Billroth II gastrectomy with Roux-en-Y reconstruction. The deviation from a typical Bouveret’s syndrome is the absence of a fistula – the accrescent stone was hypothesized to pass via the duodenal papilla when it was small in size. The second deviation presented in this case was that the abdominal pain was not due to GOO, but rather due to obstruction of the duodenal stump. In such rare cases, additional means of management may warrant incorporation of endoscopic retrograde cholangiopancreatography [56].

### Conclusion

Considering the rarity of Bouveret’s syndrome, physicians are encouraged to enrich the published literature with cases that provide valuable teaching lessons, particularly that may provide clarity on the nuanced management of this disease. Bouveret’s syndrome is encumbered by the lack of head-to-head trials comparing endoscopic modalities, the debate of optimal surgical approaches outlined in this review, and various other knowledge gaps, which may be bridged by the on-going documentation and conversations surrounding its clinical encounters.

### Author Contributions

KTO, AW, AF and AMA reviewed the literature and wrote the manuscript. DBM reviewed and edited the manuscript.

### Conflict of Interest

The authors declare that there is no conflict of interest regarding this invited literature review.

### References


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