

Bouveret's Syndrome

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ABSTRACT

Gastric Outlet Obstruction (GOO) due to impaction of a gallstone in the duodenum after migration through a bilioduodenal fistula is known as Bouveret's syndrome. Its clinical symptoms are entirely vague and nonspecific. Because of its rarity, insidiousness and unpredictable symptomatology, Bouveret's syndrome is never thought of in the differential diagnosis as aetiology of gastric outlet obstruction. Recent advances in fiberoptics technology, advent of modern imaging modalities and minimally-invasive techniques like endoscopy and laparoscopy has brought a great revolution in the management of Bouveret's syndrome and have tremendously decreased morbidity and mortality associated with this rare clinical entity.

Key words: *Cholelithiasis. Bilioenteric fistula. Gastric outlet obstruction. Bouveret's syndrome.*

INTRODUCTION

Bouveret's syndrome is a rare cause of gastric outlet obstruction resulting from impaction of a large gallstone in the duodenal bulb after migration through a bilioduodenal fistula. It is a rare variant of a relatively rare disease (gallstone ileus) with an overall incidence of 1-3%.¹ It was first described in 1896 by a French physician, Leon Bouveret, who reported two cases of gastric outlet obstruction due to gallstones in his "Revue de medicine", unluckily both of them died.² Since then, only few cases of Bouveret's syndrome have been reported in the worldwide medical literature.³ To-date, no such case has been reported in Pakistan. Because of its rarity and unpredictable symptomatology, Bouveret's syndrome can pose diagnostic and therapeutic dilemma for the clinicians. The aim of reporting this case is to enhance clinical awareness among the healthcare professionals for early diagnosis and timely management of this rare clinical syndrome.

CASE REPORT

A 65 years old male, known case of diabetes mellitus, hypertension, ischemic heart disease and cholelithiasis, presented with the history of vague upper abdominal pain and repeated forceful vomiting of 5 days duration. Vomitus was coffee-ground in colour. General physical examination revealed marked pallor and features of hypovolemia (dehydration, hypothermia, hypotension, tachycardia and oliguria). Systemic examination was

unremarkable. Abdominal examination showed mild epigastric tenderness with no abdominal distension, guarding or rigidity. No abdominal mass or visceromegaly was detected. Bowel sounds were normal in intensity and frequency. Apart from mild prostatomegaly, no abnormality was detected on digital anorectal examination. Laboratory investigations showed marked leucocytosis (WBC: 22000/ul) and iron-deficiency anemia (Hb: 8.5 g%). Liver and renal function tests were normal. Electrolyte analysis showed: sodium: 128 mmol/l, potassium: 3.1mmol/l, chloride: 90mmol/l and HCO₃: 35mmol/l. ABGS showed acute hypochloremic metabolic alkalosis revealed old anterolateral MI changes. Chest X-rays showed mild cardiomegaly with clear lung fields. Abdominal X-rays displayed ground glass haziness in the lower abdomen and few non-specific loops of small gut in the upper abdomen and no free pneumoperitoneum. Ultra-sonography depicted fluid-filled distended stomach, features of chronic cholecystitis, and an acoustic shadow (2 x 2 x 3 cm) in the area of first part of the duodenum. CT scan abdomen revealed gastric outlet obstruction and a low-attenuating filling defect surrounded by a rim of high-attenuating contrast material in the duodenum, just distal to duodenal bulb (Figure 1). Upper GI endoscopy confirmed the presence of a huge gallstone impacted in the duodenum, just distal to duodenal bulb. The stone was retrieved endoscopically with the help of Dormia basket. Unluckily, in the distal one-third of the esophagus, the Dormia basket was broken. Rigid esophagoscopy was arranged and gallstone along with broken Dormia basket was extracted (Figure 2). Postextraction, ERCP showed normal biliary passages and presence of pneumobilia and bilioduodenal fistula. After endoscopic lithotomy, patient improved steadily and was discharged after 5 days.

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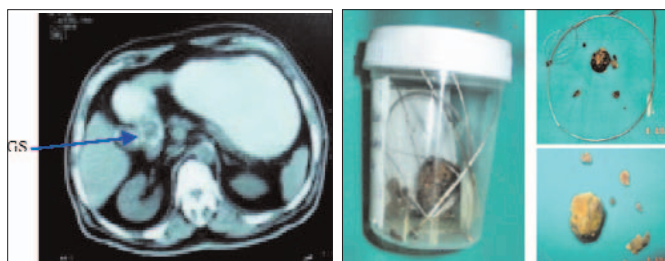


Figure 1: CT scan showing gallstone in the duodenum with

Figure 2: Gallstone with broken Dormia basket.

DISCUSSION

Bouveret's syndrome is an uncommon cause of gastric outlet obstruction resulting from intraluminal duodenal blockade by a huge gallstone migrated through a bilioenteric fistula. Common bilioenteric fistulae include cholecystoduodenal (>60%), cholecystocolic, cholecystogastric and choledochoduodenal.^{1,5} After migration through a fistula, majority of the gallstones pass asymptotically per rectum. Rarely, a stone may get enlarged due to accretion of salts and fecal matter on its surface and may cause gallstone ileus by blocking the gut lumen. The commonest site of stone impaction is terminal ileum (> 60%), followed by proximal ileum, jejunum and colon.^{2,3} Seldom, stone impaction occurs in the duodenum resulting in gastric outlet obstruction. Apart from intraluminal blockade, extrinsic duodenal compression by a hugely distended gallbladder containing multiple calculi may also result in gastric outlet obstruction. This is known as pseudo-Bouveret's syndrome.⁴

Bouveret's syndrome commonly occurs in the elderly females unlike our patient, with an average age of 68.9 years.⁵ Its clinical features are entirely vague and non-pathognomonic. Cappell and Davis,⁶ after a comprehensive review of 128 cases from the global medical literature, described the characteristic features of Bouveret's syndrome. According to their analysis, the prominent symptoms included nausea and vomiting (87%), abdominal pain (71%), haematemesis (15%) recent weight loss (14%) and anorexia (13%). The prominent signs were abdominal tenderness (44%), signs of dehydration (31%) and abdominal distension (26%). Apart from gastric outlet obstruction, an impacted gallstone may cause duodenal perforation due to pressure necrosis, haematemesis due to Mallory-Weiss tear, distal esophageal rupture due to repeated forceful vomiting (Boerhaave's syndrome) and gastric bezoars formation.^{6,7}

The diagnosis of Bouveret's syndrome is often overlooked due to insidiousness, unpredictability and non-specificity of its clinical features.^{2,5,6} However, modern imaging modalities have made diagnosis of Bouveret's syndrome relatively easy and straight forward. Plain radiography still remains the investigation

of first choice in all patients of acute intestinal obstruction. Plain abdominal X-rays may demonstrate gastric dilatation, pneumobilia and ectopic calcified gallstone in 30-35% of cases (Rigler's triad).^{1,2,6} Ultrasonography is helpful in 60% of the cases for showing fluid-filled stomach, features of chronic cholecystitis, ectopic calcified gallstone, pneumobilia and double-arch sign. Contrast-enhanced CT abdomen⁷ certainly has proved as a promising imaging modality in diagnosing Bouveret's syndrome. Features of Rigler's triad are clearly picked-up on CT scan. It effectively delineates the presence of gastric distension, pneumobilia, curvilinear bilioenteric fistula, and intraluminal low-attenuating filling defect in the duodenum (ectopic stone) surrounded by rim of high-attenuating contrast material (Figure 1). However, CT scan fails to differentiate isoattenuating stones from surrounding bile in 15-25% of the cases. Under such settings, MRCP is the most effective noninvasive imaging modality.⁷ It clearly delineates isoattenuating stones from surrounding bile and beautifully depicts the presence of bilioenteric fistula. It is especially useful in the patients who are intolerant to oral contrast. Upper GI Endoscopy⁷ has played a pivotal role in unveiling the exact underlying cause of gastric outlet obstruction in virtually all cases. It shows the impacted gallstone as hard, black, convex, non-friable and non-fleshy mass protruding through the pylorus into the gastric lumen. Gastrograffin series may also display gastric outlet obstruction, intraluminal duodenal filling defect, pneumobilia and outline of bilioenteric fistula.^{7,8}

Currently, different therapeutic options to deal with Bouveret's syndrome include endoscopy, laparoscopy or open surgery. Endoscopic stone retrieval⁸ with or without lithotripsy has become a preferred therapeutic option, especially in high-risk patients. A densely adherent gallstone in the duodenum may need *in-situ* fragmentation with the help of laser, mechanical or Extra-corporeal Shock-wave Lithotripsy (ESWL) before endoscopic attempts at retrieval. One inherent drawback associated with lithotripsy is that a partially-fragmented stone may move distally and may convert Bouveret's syndrome into distal gallstone ileus with definite need for surgery.⁹ Therefore, such patients must be kept under close observation and followed-up for, at least, few days after endoscopic stone extraction. Laparoscopic enterolithotomy¹⁰ is a minimally-invasive technique and is quite helpful and successful in elderly patients having multiple co-morbid states. Since the advent of minimally-invasive techniques, there has been a remarkable change in the management of Bouveret's syndrome and open surgery does not remain a desirable therapeutic option. However, open surgery becomes indispensable after failure of repeated endoscopic attempts at stone-retrieval, distal migration of gallstone after dislodgement or non-availability of laparoscopic expertise and facility. Open surgery^{1,3,10}

consists of laparotomy, longitudinal duodenotomy, stone retrieval and transverse closure of duodenotomy in the form of pyloroplasty. Cholecystectomy and repair of bilioenteric fistula should not be performed simultaneously as there is a high incidence of spontaneous closure of such fistulae, especially in the presence of a widely patent cystic duct and absence of residual stone in the gallbladder.^{2,9,10}

Because of rarity and insidious clinical manifestations, Bouveret's syndrome can pose diagnostic and therapeutic challenges for the clinicians. A high index of clinical suspicion is highly demanded when an elderly patient, known case of gallstones, presents with upper GI obstruction.

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