

Bouveret's Syndrome. Case Report and Literature Review.

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Citation

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Abstract

Cholecystoenteric fistula is a well-recognized complication of biliary lithiasis. We describe the clinical scenario in an 88-year-old male who presented to the emergency room at KPMC, Riyadh, with features of gastric outlet obstruction of four weeks duration. The patient's abdominal CT scan revealed pneumobilia and upper GI endoscopy revealed a tight stenosis in the pyloric area. The patient had no history of any surgical or endoscopic intervention in the past. ERCP demonstrated a fistulous tract between gallbladder and duodenum. The patient was managed with intensive proton pump therapy and in 10 weeks' time his symptoms resolved. The overall features were suggestive of Bouveret's syndrome. This rare syndrome is revisited with special reference to proton pump therapy.

INTRODUCTION

In 1896 Léon Bouveret described the first case of gastric outlet obstruction due to migration of gallstones. Grove in 1976 first described a case of pyloric obstruction due to a gallstone by gastroscopy. Since then, there have been several case reports of unique manifestations of Bouveret's syndrome, as well as reports of novel endoscopic treatment modalities. We report a case of Bouveret's syndrome and review the literature with special emphasis on the role of proton pump inhibitors alone in management of non-obstructive cases.

CASE REPORT

An 88-year-old male with no significant past medical history presented to the emergency department at KPMC, Riyadh, with chief complaints of postprandial fullness and on-and-off vomiting for one month. There was history of coffee-ground emesis for 3 days prior to presentation to our hospital. He had lost 5kg in one month despite of maintained appetite. The patient denied having had any abdominal surgery or endoscopic procedure prior to this presentation. On examination, the patient was mildly dehydrated with normal vitals. Abdominal examination revealed no surgical scar, fullness in the upper abdomen, soft, not tender abdomen and there was no organomegaly or ascites. "Succession splash" was positive. The rest of the exam was unremarkable. Laboratory investigations revealed a hemoglobin of 14.4gm/dl with normal white cell and platelet count. The patient had features of pre-renal azotemia which normalized after fluid therapy. His liver function tests were

normal. The patient's upper GI endoscopy revealed evidence of retained food and liquid in the stomach and approximately 1200 ml of fluid was aspirated. A tight stenosis was noted at the pylorus. Only a pediatric diagnostic scope could successfully negotiate the pylorus. The duodenal bulb was deformed, inflamed, and stenotic. An opening was noted at the junction of the duodenal bulb and the second part of the duodenum, with the ampulla lying in very close proximity to this opening. (Fig. 1)

Figure 1

Figure 1: Endoscopic view of duodenal bulb with an opening at the junction of the duodenal bulb.



The biopsy from the pre-pyloric area showed no malignancy. CT scan of abdomen and pelvis showed pneumobilia, along

with distended, large stomach, with evidence of incomplete gastric outlet obstruction and no definite mass or lymph nodes. There was evidence of dilated intra- and extra-hepatic ducts with pneumobilia (Figure 2).

Figure 2

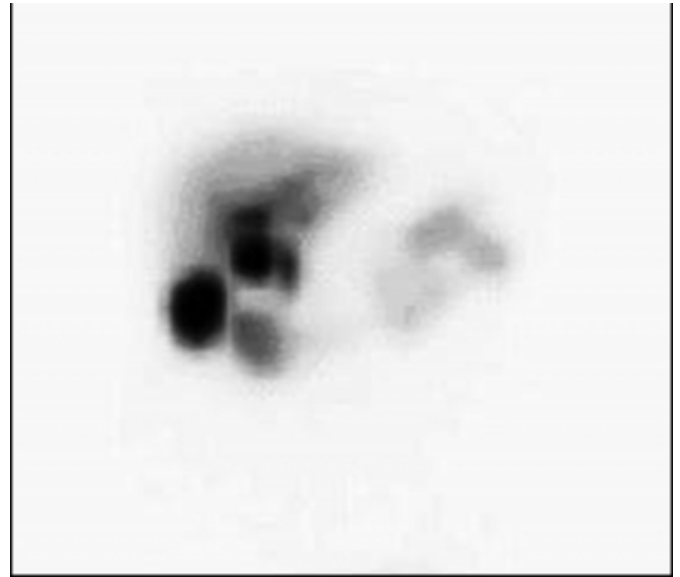
Figure 2: CT scan of the abdomen and pelvis revealing pneumobilia and normal viscera.



The patient was managed with intravenous omeprazole 80mg IV stat followed by an 8mg/hour infusion for 3 days. He was gradually started on liquid diet and oral esomeprazole 40mg twice daily, which was continued for another 3 months. The patient was continued on liquid diet, which was later advanced to soft diet with good tolerance. He underwent a HIDA scan which demonstrated continuous and rapid emptying of HIDA from the CBD into the duodenum suspicious of choledocho-duodenal fistula. (Figure 3)

Figure 3

Figure 3: The HIDA scan demonstrating continuous and direct rapid emptying of HIDA from the CBD into the duodenum, suggestive of choledocho-duodenal fistula



Repeat EGD after 10 weeks showed no food residue and the pyloric channel this time permitted an adult diagnostic EGD scope. A catheter was introduced through the previously visualized opening of the choledochoduodenal fistula, and the cholangiogram showed dilated CBD, CHD, and IHD, without any filling defect. The gallbladder was distended without any filling defect. This opening only permitted access into the bile duct. (Fig. 4 and 5) Through the opening at the ampulla, only a pancreatogram could be obtained which was normal. Through the ampulla, the bile duct could not be accessed. Though gallstones could not be documented in the patient, the clinical course of acute gastric outlet obstruction, presence of choledochoduodenal fistula, and resolution of symptoms established the diagnosis of Bouveret's syndrome.

Figure 4

Figure 4: Catheter introduced through opening of the choledochoduodenal fistula

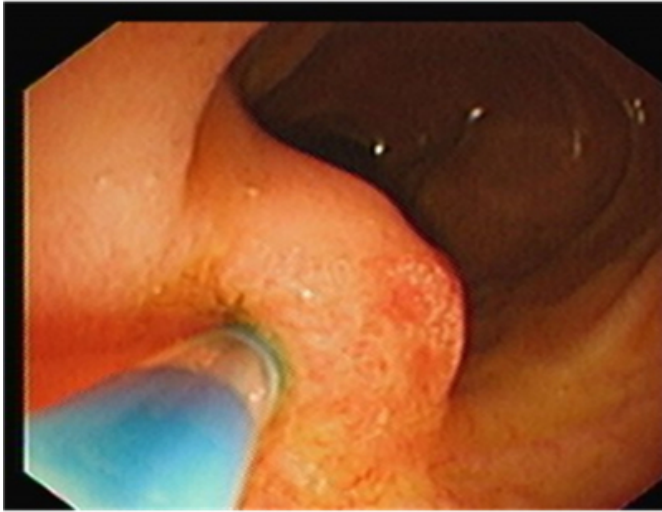


Figure 5

Figure 5: Cholangiogram through the choledocho-duodenal fistula revealing dilated intrahepatic biliary ducts with no filling defect.



DISCUSSION

Cholecystoenteric fistula is a well-recognized complication of biliary lithiasis. Fistulous tracts from the gallbladder are associated with gallstones in 90% of cases (1, 2). However, peptic ulcer disease, abdominal trauma, Crohn's disease, and malignancies of the biliary tract, bowel and head of pancreas have also been implicated as causes (3, 4, and 5). The duodenum is the most common portion of the intestine involved.

Bouveret's syndrome is defined as gastric outlet obstruction

caused by duodenal impaction of a large gallstone which passes into the duodenal bulb through a cholecystogastric or cholecystoduodenal fistula. The first published report of Bouveret's syndrome (1896) is attributed to Leon Bouveret who reported on two patients with the disease (6). Grove, in 1976, first described a case of pyloric obstruction due to a gallstone by gastroscopy (7). Since then, there have been several case reports of unique manifestations of Bouveret's syndrome, as well as reports of novel endoscopic treatment modalities (8).

In 85% of patients with Bouveret's syndrome the fistula communicates with the duodenum and the stones will pass spontaneously without causing bowel obstruction, whereas in 15% of patients the clinical features of bowel obstruction develop. In descending order of frequency, the gallstone can be lodged in the terminal ileum, proximal ileum, distal jejunum, colon, and duodenum or stomach (9). Obstruction most commonly occurs in the terminal ileum (90%) and less often in the duodenum (3%) (10).

Early diagnosis is important because mortality is historically high at 33 % (11), though it has decreased to 12% in recent years (12, 13).

Preoperative diagnosis is not easy (14). The disease is frequently encountered in elderly females with history of biliary disease. In most cases, the presenting signs and symptoms of Bouveret's syndrome are nonspecific (8). In a recent systematic review, Cappell and Davis described the most common symptoms of patients with Bouveret's syndrome as nausea and vomiting (86%), and abdominal pain (71%); less commonly, patients present with hematemesis, weight loss, and anorexia (15). On physical exam, patients often have abdominal tenderness, abdominal distention, and dehydration.

Upper gastrointestinal hemorrhage and hematemesis as a presenting symptom of Bouveret's syndrome are well reported (16, 17, and 18). Heinrich et al. (19) have described massive arterial bleeding from the eroded cystic artery due to Bouveret's syndrome in a patient who ultimately required surgery. It is quite possible that our patient had a small stone and there was no cystic artery erosion. Migration of a small stone may have caused superficial duodenal erosions and local edema resulting in reversible obstruction. Minor bleeding may have resulted in coffee-ground emesis without any significant hemoglobin drop. Acid suppression by proton pump inhibitors reduced inflammation and relieved the obstruction.

Diagnosis may be made with radiological (abdominal X-ray, ultrasound, computed tomography or magnetic resonance imaging) and endoscopic techniques. Abdominal and chest radiographs should be performed in all cases suspected to have the syndrome looking for an evidence of aerobilia, bowel obstruction and ectopic gallstones (20). Air in the biliary tree is present in as many as 50% of patients (21) and the stone may be visualized in 69% of cases. A fistulous tract without stone can be seen in 31% of cases. Barium studies may demonstrate the passage of contrast into the biliary tree. Endoscopy is the cornerstone in diagnosis. Endoscopic findings include inflammation, edema or an ulcer at the impacted site. ERCP is the most important diagnostic tool in order to identify the presence of biliary-intestinal fistula. If the cystic duct is obstructed, gallbladder and fistula will not be visualised by ERCP (as in our case). Biliary nuclear scintigraphy may delineate the fistula but false-positive and false-negative studies have been reported (22).

Historically, the diagnosis usually has been made with endoscopy and the imaging diagnosis relied primarily on conventional radiography and contrast-enhanced fluoroscopy (23), but CT and more recently magnetic resonance (MR) cholangiopancreatography are becoming more useful for the diagnosis (24, 25, and 26).

CT likely represents the single best imaging technique for the diagnosis of Bouveret's syndrome (24). CT is more comprehensive because it effectively evaluates pneumobilia, the impacted ectopic gallstone, and the cholecystocolic fistula. The increasingly use of CT scan in emergency departments for abdominal pain, resulted in diagnosis of this unusual condition more frequently. Some authors advocate the early utilization of computerized tomography to confirm the diagnosis (27).

Ultrasound could be useful in diagnosis of Bouveret's syndrome assessing the level and identifying the cause of the obstruction. Few authors reported diagnosis of the syndrome on ultrasound (28, 29, and 30), others found that

Ultrasound may suggest the diagnosis but often present a confusing diagnostic picture. Although the gallstone will be sufficiently large to be apparent, it may be difficult to distinguish a duodenal location from an orthotopic location with a contracted gallbladder wall. If the fistulous tract is filled with fluid or air, the fistula may also be seen but can be confused with the common duct. Pneumobilia and a dilated stomach may also be seen with ultrasound (31).

Management is usually endoscopic and surgery should be reserved for complicated cases, including impacted stone, and once endoscopic methods fail. Endoscopic removal is often possible. Because the affected patients are often elderly and poor surgical candidates, conservative treatment for the gallbladder and fistula has been advocated, with some reports of success (32). Endoscopy should be considered as the first line of treatment in these frequently debilitated patients. Usually help of various accessories like grasping forceps, retrieval baskets, nets etc. are required. Use of extracorporeal and intracorporeal lithotripsy (33) in the management of this syndrome has been described. Surgery is usually carried out once endoscopic removal fails and surgical options include fistula repair, enterolithotomy and cholecystectomy (34).

In vitro studies (35) have shown that proton pump inhibitors promote the cell restitution rate which is independent of pH.

We observed progressive healing of local duodenal and pyloric edema which helped in resolution of gastric outlet obstruction in a period of around 3 months without any endoscopic or surgical intervention. Proton pump inhibitors, however, may not be enough if the stone has caused a large fistula or when the stone has been impacted. Endoscopy offers another method of treatment in these cases. Endoscopic treatment was first described by Bedogni et al. in 1985 followed later by multiple reports of endoscopic management (36, 37).

It will not be out of place to mention that surgery is associated with significant mortality and morbidity (38) and all efforts should be focused on conservative management.

The mortality is historically high at 33 %, though it has decreased to 12% in recent years. The high mortality may be related to the advanced age of the typical patient as well as to other co-morbidities or relatively weak disease resistance. The decrease in mortality in recent years likely represents the impact of endoscopic treatment options in place of surgery as well as early diagnosis with noninvasive imaging, such as CT and MRI (3).

CONCLUSION

Bouveret's syndrome should be suspected in old debilitated persons with de-novo pneumobilia. Hematemesis is an uncommon presentation of this syndrome and spontaneous resolution of gastric outlet obstruction in a set of patients may be achieved by proton pump inhibitors without any endoscopic or surgical intervention.

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