Case report

A rare presentation of gastric outlet obstruction (GOO) — The Bouveret's syndrome

Dheer singh kalwaniya**, S.V. Arya, Soumya Guha*, Manigandan Kuppuswamy, Jasneet G. Chaggar, Lalmalsamwe Ralte, Rajkumar Chejera, Ashok Sharma

Dept. of General Surgery, Vardhman Mahavir Medical College and Safdarjung Hospital, New Delhi 110029, India

Highlights

- Bouveret's Syndrome is an uncommon cause of Gastric outlet obstruction (GOO) caused by a gall stone which has migrated into the duodenal bulb from a bilioduodenal fistula. Case of cholelithiasis with presentation of GOO must raise a high index of suspicion.
- We report a case of a 45 year old woman who presented to us with recurrent pain upper abdomen for last 8 years that progressed to develop symptoms of gastric outlet obstruction. Upper Gastrointestinal endoscopy revealed duodenal ulceration with stony hard lesion in the first part of duodenum. CECT abdomen done later confirmed the presence of cholecystoduodenal fistula and gall stone in proximal jejunum.
- Enterolithotomy and the pylorus preserving gastrojejunostomy with cholecystectomy was performed in this patient. We have tried to show that there is no gold standard principle laid down for the management of Bouveret's syndrome as it depends on the site of lodgement of stone, the pathological outcome due to its presence and the anatomical distortion.

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Abstract

Introduction: Bouveret's Syndrome is a rare cause of Gastric outlet obstruction (GOO) caused by a gall stone which has migrated into the duodenal bulb from a bilioduodenal fistula. It is an uncommon condition and only a few cases have been reported in the medical literature till date. Presentation of case: We report a case of a 45 year old woman who presented to us with recurrent pain in the upper abdomen for the last 8 years, and that progressed to develop symptoms of gastric outlet obstruction. An upper gastrointestinal endoscopy revealed duodenal ulceration with a stony hard lesion in the first part of duodenum. An contrast enhanced computed tomography (CECT) scan of the abdomen performed later confirmed the presence of cholecystoduodenal fistula and gall-stone in proximal jejunum. Discussion: Bouveret's syndrome is a case of gastric outlet obstruction caused by the impaction of a large gall stone in the duodenum as a result of a cholecystoduodenal fistula. It constitutes 1–3% of cases of gall stone ileus which in turn complicates only 0.3–4% cases of cholelithiasis. Bouveret's syndrome is managed surgically. Conclusion: Bouveret's syndrome being an uncommon condition may pose a difficulty in diagnosis and dilemma in managing such patients. An awareness is hence essential.

1. Introduction

Bouveret's syndrome is a rare cause of gastric outlet obstruction owing to the impaction of a large gall stone in the duodenal bulb after migration through a bilioduodenal fistula. It is a rare variant of a relatively rare disease (gall stone 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 gall stones in his- “Revue Medicale” [2]. Since then, only...
few cases of Bouveret’s syndrome have been reported in the worldwide medical literature. Owing to its uncommon and unpredictable symptomatology, Bouveret’s syndrome can pose as a diagnostic and therapeutic challenge for clinicians. The aim of reporting this case is to increase clinical awareness which will ultimately lead to early diagnosis and timely management of this rare syndrome.

2. Presentation of case

A 45 year old woman presented with complaints of recurrent mild pain in the upper abdomen for the last 8 years, often associated with multiple episodes of nausea and non-bilious vomiting. Pain started in epigastrium and spread to right hypochondrium, and it was not associated with fever. Patient did not have any history of jaundice or prior surgery. She did not have any other abdominal complaints. The patient also did not reveal any other significant personal or family history.

On examination, all the vital signs were within normal limits and the patient was afebrile and non icteric. Mild tenderness elicited in epigastrium and right hypochondrium, however there was no guarding or rigidity. No lump or organomegaly was present and the bowel sounds were normal. Laboratory investigations were all within normal limits including the liver function test. An ultrasonography of the abdomen revealed a partially distended thick walled gall-bladder with multiple calculi in the lumen suggestive of chronic cholecystitis. The patient was treated conservatively as a case of calculous cholecystitis and was planned for elective cholecystectomy.

Patient returned after 15 days with complaints of abdominal pain and distension post meals associated with recurrent episodes of vomiting. On examination, the patient had distension over the epigastrium with tenderness. There was no rebound tenderness or guarding. Succussion splash was positive and bowel sounds were sluggish. Digital rectal examination did not reveal any abnormality.

An upper Gastro-Intestinal endoscopy was performed which revealed duodenal ulceration with a stony hard lesion in first part of duodenum. The scope could not be negotiated beyond first part of duodenum. Multiple Biopsies were taken from the ulceration site and histopathological examination revealed only inflammatory changes. A contrast enhanced computed tomographic (CECT) scan of abdomen depicted a choledocho-duodenal fistula (in the first part of duodenum) with a gall stone in proximal jejunal loop, sludge and concretions in the gall-bladder with a distended fluid filled stomach with pneumobilia (Figs. 1 and 2).

Exploratory laparotomy was planned with a working diagnosis of gastric outlet obstruction (GOO). Operative findings were: a pulled up pylorus densely plastered at porta-hepatis with no palpable mass or scarring in duodenum or pylorus. A 5 × 4 cm calculus extracted from jejunum, 15 cm distal to duodeno-jejunal junction (Figs. 3–5). Gall bladder was contracted with sludge and concretions and a fistulous tract from gall bladder to first part of duodenum. Cholecystectomy with enterolithotomy and closure of the fistulous tract with gastro-jejunostomy was performed under general anesthesia. Post operative period was uneventful and the patient was discharged on 7th day. The patient is on a regular follow up. Patient had an uneventful recovery.

3. Discussion

Bouveret’s syndrome is an uncommon cause of GOO owing to intraluminal duodenal obstruction by gall stone which has migrated through a bilio-enteric fistula. Gall stone ileus is the more commonly heard complication of cholelithiasis (0.3–4%) [3]. The most common level of obstruction is the terminal ileum (60–70%), followed by the proximal ileum (25%), distal ileum (10%), jejunum (9%), colon (4%), rectum (2%), and lastly the duodenum (1–3%) [3].

This syndrome was first described by Beaussier in 1770 but it has been named after the French physician Leon Bouveret after his
Around 1% of gallstone cases develop bilio-enteric fistulas [4]. Types of bilio-enteric fistulas include cholecystoduodenal (60%), cholecystocolic (17%), cholecystogastric (5%), and choledochoduodenal (5%) [4]. Fistula formation is thought to occur as a result of adhesions between the gall bladder and the bowel wall secondary to chronic inflammation, impaired arterial blood supply, and decreased venous drainage [5]. Ensuing fistula formation can occur from pressure necrosis and compression of the gall stone against the gall bladder wall. The subsequent passage of the gall stones can occur via the fistula and the stone may get enlarged due to accumulation of salts and fecal matter on its surface.

Risk factors for Bouveret’s syndrome are age more than 70 years, female gender, gallstones more than 2.5 cm in size, post-surgical altered GI anatomy [6]. Symptoms include vomiting (87%), abdominal pain (71%), hematemesis (15%), recent weight loss (14%), anorexia (13%) [7]. Common signs encompass abdominal tenderness (44%), signs of dehydration (31%), abdominal distension (26%) [7]. Extrinsic duodenal compression by a massively enlarged gall bladder containing multiple calculi may also cause GOO (pseudo-Bouveret’s syndrome) [8]. Bouveret’s syndrome results in complications such as duodenal perforation, hematemesis (Mallory-Weiss tear), distal oesophageal rupture (Boerhaave’s syndrome), gastric bezoar [7].

Abdominal X-ray can show Rigler’s triad (pneumobilia, small bowel obstruction and gall stones) in 30–35% cases [9]. Ultrasonography abdomen is helpful in 60% of cases and can reveal fluid-filled distended stomach, features suggestive of cholecystitis, ectopic gall stone, pneumobilia. CECT abdomen has a sensitivity of 93%, specificity of 100% and diagnostic accuracy of 99% [10]. It can demonstrate the exact site of obstruction, fistula and gallbladder status [11]. MRCP has main utility in patients intolerant to oral contrast. It can differentiate between gallstone and surrounding bile (CT scan cannot in 15–25% cases) [11]. UGI endoscopy is both diagnostic and therapeutic. The common findings are stone causing obstruction (69%), obstruction without visualized stone or fistula (31%), partially visualized through duodenal wall (<1%), inflammation, edema or ulcer at impacted site, retained food or fluid in stomach [7].

Treatment options available are Endoscopic, Open Surgery and Laparoscopic Approach. Endoscopic approaches include endoscopic extraction, endoscopic laser lithotripsy, extracorporeal shockwave lithotripsy and intracorporeal electrohydraulic lithotripsy. The first successful endoscopic extraction has been described in 1985 by Bedogni et al. [12]. Subsequently, a number of case reports have been published describing successful endoscopic management of Bouveret’s syndrome [3]. Endoscopic management often requires the use of different sized and shaped snares, grasping forceps, retrieval baskets and nets, biliary balloons, and sometimes even a side-viewing endoscope and hence can be technically challenging, time-consuming, and success rates in case series have been previously reportedly to be less than 10% [3]. While the majority of patients tolerate attempted endoscopic treatment, pulseless electrical activity (PEA) can occur during mechanical retrieval due to the gall stone getting lodged in the esophagus. The PEA abruptly resolved when the stone was pushed back into the stomach [13]. Indications for open surgery [3,14] are stone size greater than 2.5 cm, residual stones in GB, multiple stones in intestinal lumen, sepsis, perforation, stricture and failure of endoscopic approach. Open surgery involves enterolitotomy with cholecystectomy with fistula repair. Different authors have recommended different approaches for management of Bouveret’s syndrome as shown in Table 1. Laparoscopy is also an additional option for surgical treatment. However it is difficult and has a long learning curve. Sica et al. reported, in 2005, the first case of uneventful stone removal and cholecystectomy by laparoscopy [15]. Usually enterolitotomy is performed at the site of impaction. A study by Capell & Davis in 2006 showed bile leak and...
duodenal leak in patients underwent enterolithotomy alone or enterolithotomy + cholecystectomy. Keeping this in mind, instead of doing enterotomy at the site of impaction which may have underlying mucosal ischemia, certain case reports mention milking the stone down along the jejunum and enterolithotomy under local anesthesia, certain case reports mention doing enterotomy at the site of impaction which may have led to gastroparesis. The ulcer noted in this patient was young and vitally stable with no comorbidities, the pylorus was pulled up and plastered at the porta hepatitis. This resulted in an alteration in the normal anatomic outflow of gastric contents and would have eventually led to eventual lead to gallstones. In order to spare the patient from a future surgery for GOO (this time due to a different cause) which would significantly increase his morbidity, we did a pyloric bypass via gastro-jejunostomy. As the patient was young and vitally stable with no comorbidities, enterolithotomy with cholecystectomy with fistula repair was done in single sitting.

4. Conclusion

- High degree of suspicion is required to diagnose Bouveret syndrome pre-operatively. Deviation from the classical presentation of choledolithiasis with a tilt towards GOO should be taken as a red flag sign in this disease which definitely has a high incidence with the following features if present:
  - gall stone more than 2.5 cm in size.
  - pneumobilia and fluid filled distended stomach in ultrasonogram.
  - incidental endoscopic finding of obstruction in later part of duodenal & proximal jejunum.
- GOO can encompass a band of etiologies varying from benign to malignant tumors to the rarer Bouveret’s syndrome and gall stone ileus. A case of choledolithiasis with presentation of GOO must raise a high index of suspicion towards Bouveret’s syndrome. There is no gold standard principle laid down for the management of Bouveret’s syndrome as it depends on the site of lodgement of stone, the pathological outcome attributed to the presence of the stone and the anatomical distortion. The most common treatment offered is enterolithotomy with cholecystectomy with fistula repair in a stable patient, whereas in this particular case report the operative findings warranted a deviation, which included incorporation of added gastro-jejunostomy. Hence, this case report not only presents red flags but also this sort of anatomical presentation, requiring a variance of operative management from the standard.

Ethical approval

As this is not a research study but a case reporting with no mention of patient’s personal details anywhere in the report, ethical approval has not been sought.

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Author contribution

Conception and Design of case report — Dr. S.V. Arya, Dr. Dheer Singh Kalwaniya.

Drafting of article and revising it for important intellectual content — Dr. Soumya Guha.

Dr. Manigandan G.

Other contributors — Dr. Radte, Dr. Jasneet, Dr. Ashok Sharma, Dr. Rajkumar Chejara.

Conflicts of interest

None.

Guarantor

Dr. Dheer Singh Kalwaniya.

Dr. Soumya Guha.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

References

[1] Heneghan HM, Martin ST, Ryan RS, et al. Bouveret’s syndrome: a rare presentation of gall stone ileus. Ir Med J 2007;100:504–5.
[2] Bouveret L. Stenose du pylore adherent a la vesicule. Rev Medicale 1896;16:1–15.[Paris].
[3] Lowe AS, Stephenson S, Kay CI, May J. Duodenal obstruction by gallstones (Bouveret’s syndrome): a review of the literature. Endoscopy 2005 Jan;37(1):82–7.
[4] Vasileios KM, Dimitrios IM, Nikolaus KE, Ioannis DK. Bouveret syndrome—The rarest variant of gallstone ileus: a case report and literature review. Case Reports Surg 2013;2013. article ID 839370:6.
[5] Langhorst J, Schumacher B, Deseaers T, Neuhaus H. Successful endoscopic therapy of a gastric outlet obstruction due to a gallstone with intracorporeal laser lithotripsy: a case of Bouveret’s syndrome. Gastrointest Endosc 2000;51(2):209–13.
[6] Koulaouzidis A, Moschos J. Bouveret’s syndrome. Narrative review. Ann Hepatol 2007;6(2):89–91.
[7] Cappell MS, Davis M. Characterization of Bouveret’s syndrome: a comprehensive review of 128 cases. Am J Gastroenterol 2006;101:2139–46.
[8] Doody O, Ward E, Buckley O, Hogan B, Torreggiani WC. Bouveret’s syndrome variant. Digestion 2007;75:126–7.
[9] Masannat YA, Caplin S, Brown T. A rare complication of a common disease. Bouveret’s syndrome: a case report. World J Gastroenterol 2006;12:2620–1.
[10] Yu CY, Lin CC, Shyu RF, Hsieh CB, Wu HS, Tyan YS, et al. Value of CT in the diagnosis and management of gallstone ileus. World J Gastroenterol 2005;11(14):2142–7.
[11] Trubek S, Bhamla JK, Lamki N. Radiological findings in bouwret syndrome. Emerg Radiol 2001;8(6):335–7.
[12] Bedogni G, Contini S, Meiner M, Pedrazzoli C, Piccinini GC. Pyloroduodenal obstruction due to a biliary stone (Bouveret’s syndrome) managed by endoscopic extraction. Gastrointest Endosc 1985;31(1):36–8.

[13] Moschos J, Pilpalidis I, Antonopoulos Z, Paikos D, Tzilves D, Kadis S, et al. Complicated endoscopic management of Bouveret’s syndrome. A case report and review. Romanian J Gastroenterology 2005;14(1):75–7.

[14] Doycheva I, Limaye A, Suman A, Forsmark CE, Sultan S. Bouveret’s syndrome: case report and review of the literature. Gastroenterology Res Pract 2009;2009:4.

[15] Sica GS, Sileri P, Gaspari AL. Laparoscopic treatment of Bouveret’s syndrome presenting as acute pancreatitis. J Soc Laparoendosc Surg 2005;9(4):472–5.

[16] Costil V, Julles MC, Zins M, Loriau J. Le syndrome de Bouveret: l’ileus bilaire n’est pas toujours là ou on pense. J de Chir Viscerale August 2012;149(4):320–2.