Hellenic Journal of Surgery 2012; 84: 5

Vascular Complications of Large Gallstones: Proposal of a First Classification

Review and Case Report

A. Marinis, I. Kalaitzis, P. Basioukas, S. Rizos
Received 16/04/2012 Accepted 25/07/2012

Abstract

Aim-Background: The development of large gallstones are responsible for disastrous complications, either by exerting pressure and erosion of adjacent bile ducts or a hollow viscus known as Mirizzi syndrome or by obstructing the gastric outlet named Bouveret syndrome. Moreover, a very rare complication of large gallstones is the involvement of adjacent vascular structures either by the development of a pseudoaneurysm or by rupture of the pseudoaneurysm or erosion of a vessel, which is manifested as haemobilia, upper GI haemorrhage and, more dramatically, as massive intraperitoneal bleeding. This manuscript proposes a first classification for this rare complication.

Methods: A review of the current literature using Pubmed search of the terms "Mirizzi syndrome", "Bouveret syndrome", "pseudo-aneurysm" and "vascular emergencies" was done. Discussion of clinical manifestations, diagnostic workup and therapeutic strategies in a differential diagnostic approach was accompanied by our personal experience presented as a case report.

Results: Vascular complications of large gallstones have rarely been reported: in this manuscript they are collectively used to propose a first classification, as follows: development of a pseudo-aneurysm (type I); intrabiliary haemorrhage - haemobilia (type II); upper gastrointestinal haemorrhage (type III) and intraperitoneal haemorrhage (type IV). This case report describes a 74-year-old female patient presenting with alternating intermittent jaundice and upper gastrointestinal haemorrhage, suggesting haemobilia. Computed tomography demonstrated a large gallstone without contrast extravasation in the biliary tree. A new episode of severe bleeding prompted an emergency laparotomy, during which the large gallstone was found firmly attached to and eroding the second portion of the duodenum. Gallstone extraction through the destroyed gallbladder's fundus was followed by sundden massive intraperitoneal haemorrhage. After initial haemostasis by Pringle's manoeuvre the site of haemorrhage was detected retroduodenally and included the gastroduodenal artery. Therefore, this was classified as a type IVb vascular complication.

Conclusions: Vascular complications of large gallstones are rarely seen in clinical practice. Clinical suspicion of these complications should be raised when a large gallstone is present and peculiarly combined signs and symptoms that question differential diagnosis.

Key words:
Mirizzi syndrome, Bouveret syndrome, Pseudo-aneurysm, Vascular emergencies

Introduction

Gallstones in the gallbladder can either be asymptomatic or manifest clinically. They vary from simple non-specific upper gastrointestinal (GI) complaints, usually confused with other diseases (e.g. peptic ulcer, gastritis, etc), to more common complications, including biliary colic, cholecystitis, empyema, pancreatitis, as well as more severe situations caused by large gallstones, such as pressure and erosion of adjacent bile ducts or hollow viscus (Mirizzi's syndrome) [1] and obstruction of the gastric outlet (Bouveret's syndrome) [2]. A very rare complication of large gallstones concerns the involvement of adjacent vascular structures, with the development or rupture of a pseudoaneurysm or the erosion of a vessel, which manifests as haemobilia, upper GI haemorrhage or, more critically as massive intraperitoneal bleeding during laparotomy. This report proposes a first classification for this rare complication, reviews the current literature and discusses clinical manifestations, diagnostic workup and therapeutic strategies in a differential diagnostic approach.

Complications of Large Gallstones

a. *Compression or/and erosion of the common hepatic duct (Mirizzi syndrome type I-IV):* Mirizzi syndrome

A. Marinis (Corresponding author), I. Kalaitzis, P. Basioukas, S. Rizos - First Department of Surgery, "Tzaneion" General Hospital, Piraeus, Greece
e-mail: drmarinis@gmail.com

(MS) is attributed to Pablo Mirizzi who was the first to describe it as a benign condition during which a stone in the cystic duct or in the Hartmann's pouch impinges on the CBD, leading to mechanical obstruction by the stone itself or by secondary inflammation [1]. He termed this phenomenon as "functional hepatic syndrome". In 1982, McSherry et al. classified MS into two types based on ERCP findings [3]: Type I MS when there is extrinsic compression of the common hepatic duct and Type II MS when the gallstone erodes the common hepatic duct wall, resulting in a cholecystocholedochal fistula. In 1989, Csendes et al proposed a new classification in order to clarify the definition of cholecystobiliary fistulae and further establish an appropriate surgical treatment according to the underlying pathology [4]. Thus, he defined the following four categories:

- **Type I:** external compression of the common bile duct due to a stone impacted at the neck of the gallbladder or the cystic duct (62.5%).
- **Type II:** presence of a cholecystobiliary fistula (cholecystohepatic or cholecystocholedochal) due to erosion of the anterior or lateral wall of the CHD or CBD, respectively, with the fistula involving less than one-third of the circumference of the common bile duct (12.5%).
- **Type III:** presence of a cholecystobiliary fistula with erosion of the wall of the common bile duct that involves up to two-thirds of its circumference (12.5%), and
- **Type IV:** presence of a cholecystobiliary fistula with complete destruction of the entire wall of the common bile duct (12.5%).

b. *Erosion of an adjacent hollow viscus with or without gallstone ileus (Mirizzi syndrome type V):* In 2007, Csendes et al suggested that Mirizzi syndrome may not just implicate a cholecystobiliary fistula but may further complicate the clinical picture with a superimposed cholecystoenteric fistula [5]. Thus, a modified classification was proposed as follows: any aforementioned type (I-IV) plus a cholecystoenteric fistula without gallstone ileus (Type Va) [6], or with gallstone ileus (Type Vb).

c. *Gastric outlet obstruction (Bouveret's syndrome):* In 1896, Leon Bouveret reported the first two cases of gastric outlet obstruction due to gallstone disease [2]. Bouveret's syndrome is defined as obstruction at the level of the gastric outlet by one or more large gallstones that enter into the duodenal bulb through a cholecystogastric or cholecystoduodenal fistula [7]. It represents an uncommon form of gallstone ileus, comprising only 1-3% of cases [8].

d. *Vascular complications - proposed classification (Fig. 1):* Adjacent vascular structures may rarely be affected by the presence of a large gallstone. Complications include either the development of a pseudoaneurysm which may exert space-occupying effects on the adjacent bile duct, or the occurrence of haemorrhage which may be insidious or abrupt, presenting as an abdominal emergency. The most dreaded complication is intraoperative massive haemorrhage, occurring after pulling out the impacted large gallstone. These complications have been classified as follows:

i. Development of a pseudoaneurysm (type I)
The mechanical pressure exerted by the presence of a large gallstone along with the concomitant development of subsequent inflammation is responsible for the development of a pseudoaneurysm. This may include all the adjacent vessels: the cystic artery [9], hepatic artery [10-12] and gastroduodenal artery [13]. The pseudoaneurysm commonly exhibits space-occupying behaviour, externally compressing the adjacent bile duct, thereby causing the development of jaundice.

ii. Intrabiliary haemorrhage - Haemobilia (type II)
Rarely, a pseudoaneurysm may rupture in the gallbladder [11] or the biliary tree [14], resulting in haemobilia which usually presents as biliary colic, jaundice and UGIH or melenae (all three constitute Quincke's triad, observed in 32-40% of patients) [15].

iii. Upper gastrointestinal haemorrhage (type III)
Even more rarely, a large gallstone may cause UGIH by several mechanisms: a) erosion of the adjacent duodenal wall from the gallstone and mucosal haemorrhage, b) rupture of a pseudoaneurysm in the duodenal lumen or c) erosion of a vessel and the adjacent duodenal wall. The few reports that have been published are mostly in the context of Bouveret's syndrome [16, 17].

iv. Intraperitoneal haemorrhage (type IV)
This is a feared complication which may present, either (a) as a rupture of a pseudoaneurysm in the free peritoneal cavity, or (b) as massive haemorrhage which occurs intraoperatively after the removal of the impacted large gallstone, and is the result of erosion of an adjacent vessel from the large gallstone. This rare type is described in the following case report, which is the first in the current literature.

Erosion of the Gastroduodenal Artery Presenting as Intraoperative Bleeding (Type IVb) - A Case Report

A 74-year-old female patient was admitted to our Surgical Department complaining of haematemesis and melenae which had started 3 days earlier. Past

medical history included diabetes and hypertension under appropriate therapy, total abdominal hysterectomy (for the management of uteral myofibroids) and several episodes of upper gastrointestinal haemorrhage (UGIH) during the last year which were attributed to a duodenal ulcer. Clinical examination revealed icteric sclera and right upper quadrant (RUQ) tenderness on palpation. Gastroduodenoscopy showed a bleeding ulcer in the posterior duodenal wall which could not cease endoscopically due to a peculiar intraluminal mass of unspecified tissue in the same region. Interestingly, whenever jaundice increased, bleeding stopped, and vice versa (alternating intermittent jaundice and UGIH), suggesting the development of haemobilia. However, the contrast-enhanced abdominal computed tomography (CT) did not demonstrate contrast extravasation into the bile duct, excluding a possible haemobilia. Additionally, a large gallstone produced moderate dilatation of the intra- and extrahepatic biliary tree. However, the patient was not able to undergo MRCP due to a serious recurrence of bleeding which caused hypovolemic shock. After initial haemodynamic resuscitation with blood transfusion, colloids and crystalloids, the patient underwent an unsuccessful attempt of endoscopic haemostasis and was transferred to the operating theatre. Intraoperatively, a seriously inflamed gallbladder containing a large gallstone (7x5cm) (Fig. 2) was firmly attached to and had eroded the second portion of the duodenum, while the deformed duodenum was firmly attached to liver's hilum. Blunt dissection of the gallbladder from the duodenum revealed a large 3-4cm erosion of the lateral wall of the second portion of the duodenum (Fig. 3). Gallstone extraction through the destroyed gallbladder's fundus was followed by a sudden, massive haemorrhage. After initial haemostasis with a Pringle manoeuvre, the site

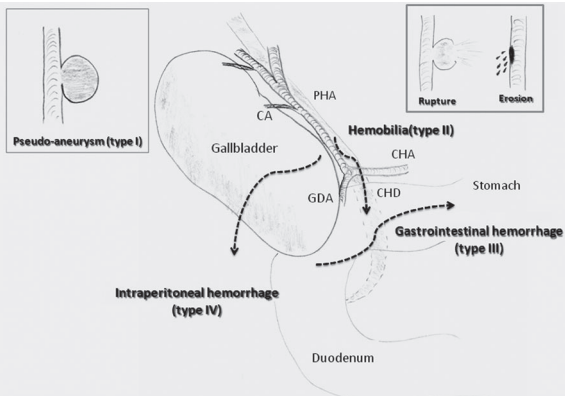

Fig.1 Vascular complications of large gallstones: development of a pseudoaneurysm (type I) and haemorrhage due to pseudoaneurysm rupture or erosion of adjacent vessels (type II-IV). Abbreviations: CHD (common hepatic duct), CHA (common hepatic artery), GDA (gastroduodenal artery), PHA (proper hepatic artery), CA (cystic artery)

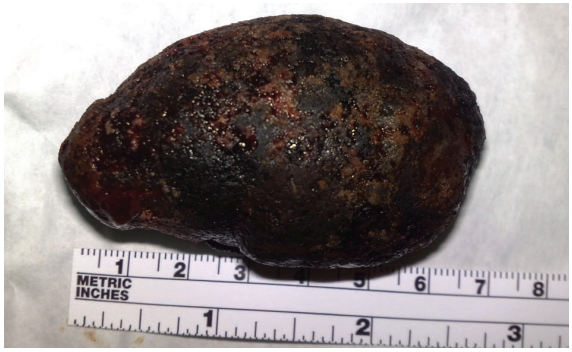

Fig.2 Photo of the large gallstone extracted from the gallbladder, measuring 7cm

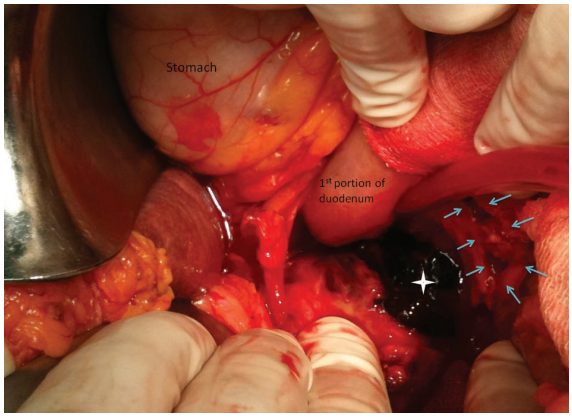

Fig.3 Intraoperative photo demonstrating the duodenal defect (arrows), the Hartmann's pouch (star) after the removal of the large gallstone and haemostasis

of haemorrhage was detected retroduodenally and included the gastroduodenal artery. After achieving haemostasis by ligating the vessel with a figure of eight prolene 2/0 suture, partial cholecystectomy was performed. The deficit of the second part of the duodenum was then debrided and sutured primarily in two layers with an absorbable suture (Vicryl) 3/0), and a Foley catheter was inserted into the duodenum for drainage. Postoperatively, the patient's condition was complicated by duodenal leakage and a concomitant necrotic infection of the surgical wound which, a month later, led to fulminant sepsis and death in the ICU. *Written informed consent has been obtained from the patient's son.*

Differential Diagnosis and Treatment of Large Gallstone Complications (Table 1)

a. *Mirizzi syndrome:* Intermittent jaundice, abdominal pain and alternating liver function tests can suggest Mirizzi syndrome in 80% of cases. Ultrasonography and abdominal CT disclose a dilated common bile duct above the gallbladder neck, an abrupt disruption of the common hepatic duct and a decompressed gallbladder with stones [18]. MRCP with MRI is a useful

method for the diagnosis of MS as it can demonstrate with precision the presence and degree of dilatation of the biliary tree. Additionally, it can also reveal extrinsic narrowing of the common hepatic duct, as well as anatomical variations and malformations. ERCP remains the gold standard for preoperative diagnosis in jaundiced patients, although it cannot often document the presence of a fistula. ERCP provides indirect signs such as lateral filling gap of the common hepatic duct, central dilatation of the biliary tree and insertion of the cystic duct at the point of obstruction, and/or complete obliteration of the cystic duct [19]. The usual surgical treatment for type I MS is laparoscopic or open cholecystectomy and exploration of the common bile duct. In cases of type II MS, most studies favour open techniques, although Binnie et al reported successful laparoscopic treatment for type II MS after previous endoscopic biliary stenting. Possible surgical approaches include choledochoplasty with gallbladder flap, cholecystoduodenostomy and Roux-en-Y hepaticojejunostomy [20]. Partial cholecystectomy with primary closure using the gallbladder remnant and T-tube insertion though the fistula opening is yet another option.

b. *Bouveret's syndrome:* In a large review of 128 cases [21], common symptoms were nausea and vomiting in 87%, abdominal pain in 71%, haematemesis in 15%, recent weight loss in 14%, and anorexia in 13% of patients. Prominent signs were abdominal tenderness in 44%, signs of dehydration in 31%, and abdominal distention in 26% of patients. Abdominal x-rays, upper gastrointestinal series and computed tomography may demonstrate Rigler's triad (upper GI obstruction, ectopic gallstones and pneumobilia) and contribute to the diagnosis in up to 26% of cases. However, gastroduodenoscopy will reveal gastroduodenal obstruction in virtually all cases and may show the obstructing stone in 69% of cases. Non-surgical treatment includes extracorporeal shock-wave lithotripsy, endoscopic stone retrieval or mechanical lithotripsy and is successful in about 9% of cases [22]. Endoscopic treatment should be preferred as it reduces emergency surgical procedures, thus minimizing morbidity and mortality.

Table 1 Clinical signs, diagnostic modalities and management of large gallstone complications

	Clinical signs	Radiology	Endoscopy	Angiography	Treatment
Mirizzi's syndrome	Intermittent jaundice, abdominal pain, bowel obstruction (in type Vb)	Abdominal x-ray: Rigler's triad (pneumobilia, bowel obstruction, ectopic gallstones), US & CT: Dilated biliary tree and large gallstone, M/ERC: Dilated CHD above cystic duct insertion, Biliary-enteric fistula	Erosion of the duodenum	Not necessary	Cholecystectomy, suture duodenal defect, biliary-enteric anastomosis (according to the type)
Bouveret's syndrome	Nausea and vomiting, abdominal pain, haematemesis,weight loss	Abdominal x-ray: Rigler's triad	Gastroduodenal obstruction by the gallstone	Not necessary	Endoscopic stone retrieval and, if unsuccessful, operative intervention (cholecystectomy, suture defect and possibly gastrointestinal bypass)
Vascular complications i)Pseudoaneurysm	Progressive jaundice, history of cholecystitis	Large gallstone	No specific findings	Aneurysm demonstration and localization	Selective angiographic embolization
ii-iii)Pseudoaneurysm rupture or Vascular Erosion	Alternating intermittent jaundice due to haemobilia and/or UGIH	Large gallstone, intravenous contrast inserting in the biliary tree	Blood exiting ampulla of Vater	Erosion identified and localized	Selective angiographic embolization or surgery, if angiography not available
iv) Intraoperative haemorrhage	Extraction of gallstone followed by massive intra-abdominal haemorrhage Useful manoeuvres: temporary manual compression of the bleeding site/ Pringle's manoeuvre may help				Suture- ligate the vessel

c. *Vascular complications:*

a. Type I: The combination of a large gallstone demonstrated by ultrasound or CT and insidious development of jaundice in a patient with a history of inflammatory attacks of the right hypochondrium should raise suspicion of a pseudoaneurysm compressing the bile duct. Selective angiography is the diagnostic method of choice and selective vascular embolization with coils is the preferred therapeutic modality [13].

b. Types II-III: An alternating clinical picture between intermittent jaundice and intermittent upper GI haemorrhage in a patient with a large gallstone, as demonstrated radiographically, should prompt the clinician towards the diagnosis of gallstone-induced aneurysm rupture or vascular erosion. Endoscopy cannot usually identify the source of bleeding and, in rare circumstances, may show blood escaping from the ampulla of Vater (haemobilia). If the patient stabilizes haemodynamically, interventional selective angiography is considered as the gold standard diagnostic and therapeutic method, i.e. demonstration and localization of the injured vessel is clearly elucidated and embolization performed. In haemodynamically unstable patients without access to angiographic embolization, immediate laparotomy and surgical haemostasis is required.

c. Type IV: the sudden occurrence of massive intra-peritoneal haemorrhage, due to rupture of the pseudoaneurysm or after removal of a large gallstone, is a surgical emergency. Immediate compression of the bleeding site along with a Pringle manoeuvre may temporarily stop bleeding, allowing a more specific effort to establish definite haemostasis by suturing the injured vessel.

Conclusions

Large gallstones may cause several known serious complications, due to pressure or erosion of the common bile duct (Mirizzi's syndrome types I-IV), formation of enterobiliary fistulae (Mirizzi's syndrome type V) or obstruction of gastric outlet (Bouveret's syndrome); however, adjacent vascular structures can be injured as well, ranging from the development of a pseudoaneurysm (type I), the rupture of a pseudoaneurysm or erosion of a vessel presenting as haemobilia (type II) or upper GI haemorrhage (type III), to intraoperative massive bleeding after rupture of the pseudoaneurysm or after extraction of the stone (type IV). The presence of a large gallstone and unusually combined signs and symptoms that question differential diagnosis should raise clinical suspicion of such complications.

Conflict of interest

The authors declare that they have no conflict of interest.

References

1. Mirizzi PL. Hepatic duct syndrome. Cir Cir. 1964; 32: 641-64

2. Bouveret L. Stenose du pylore adherent a la vesicule. Revue Medicale (Paris) 1896;16:1–16

3. McSherry CK, Ferstenberg H, Calhoun WF, Lahman E, Virshup M. The natural history of diagnosed gallstone disease in symptomatic and asymptomatic patients. Ann Surg 1985;202:59-63

4. Csendes A, Carlos Diaz, Burdiles P, Maluenda F, Nava O. Mirizzi's syndrome and cysto-biliary fistula: a unifying classification. Br J Surg 1989;76:1139-43

5. Csendes A, Munoz C, Alban M. Sindrome de Mirizzi—Fistula colecistobiliar, una nueva clasificacion. Rev Chil Cir 2007;59(Suppl):63–4

6. Lampropoulos P, Paschalidis N, Marinis A, Rizos S. Mirizzi syndrome type Va: A rare coexistence of double cholecysto-biliary and cholecysto-enteric fistulae. World J Radiol 2010;2:410-3

7. Doycheva I, Limaye A, Suman A, Forsmark CE, Sultan S. Bouveret's syndrome: case report and review of the literature. Gastroenterol Res Pract 2009;91:49-51.

8. Langhorst J, Schumacher B, Deselaers 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;51:209-213

9. Saluja SS, Ray S, Gulati MS, Pal S, Sahni P, Chattopadhyay TK. Acute cholecystitis with massive upper gastrointestinal bleed: a case report and review of the literature. BMC Gastroenterol. 2007;7:12

10. Lin SZ, Tseng CW, Chen CC. Hepatic artery pseudoaneurysm presenting with Mirizzi syndrome and hemobilia. Clin Gastroenterol Hepatol. 2009;7:e73

11. Akatsu T, Hayashi S, Egawa T, Doi M, Nagashima A, Kitano M, Yamane T, Yoshii H, Kitajima M. Hepatic artery pseudoaneurysm associated with cholecystitis that ruptured into the gallbladder. J Gastroenterol. 2004;39:900-3

12. Anderson O, Faroug R, Davidson BR, Goode JA. Mirizzi syndrome associated with hepatic artery pseudoaneurysm: a case report. J Med Case Reports 2008;2:351

13. Blomley MJ, Jackson JE. Case report: a gastroduodenal artery pseudoaneurysm presenting with obstructive jaundice and treated by arterial embolization. Clin Radiol. 1994;49:715-8

14. Sibulesky L, Ridlen M, Pricolo VE. Hemobilia due to cystic artery pseudoaneurysm. Am J Surg 2006;191:797-8

15. Sousa HT, Amaro P, Brito J, Almeida J, Silva MR, Romaozinho JM, Leitao MC. Hemobilia due to pseudoaneurysm of the cystic artery. Gastroenterol Clin Biol 2009;33:80-2

16. Heinrich D, Meier J, Wehrli H, Bühler H. Upper gastrointestinal hemorrhage preceding development of Bouveret's syndrome. Am J Gastroenterol 1993;88:777-80

17. Jones TA, Davis ME, Glantz AI. Bouveret's syndrome presenting as upper gastrointestinal hemorrhage without hematemesis. Am Surg 2001;67:786-9

18. Lakhtakia S, Gupta R, Tandan M, Rao GV, Reddy DN. Mirizzi's syndrome: EUS appearance. Gastrointest Endosc 2006;63:322-3

19. Hazzan D, Golijanin D, Reissman P, Adler SN, Shiloni E. Combined endoscopic and surgical management of Mirizzi syndrome. Surg Endosc 1999;13:618-20

20. Gomez D, Rahman SH, Toogood GJ, Prasad KR, Lodge JP, Guillou PJ, Menon KV. Mirizzi's syndrome - results from a large western experience. HPB (Oxford) 2006;8:474-9

21. Koulaouzidis A, Moschos J. Bouveret's syndrome. Narrative review. Ann Hepatol 2007;6:89-91

22. Lowe AS, Stephenson S, Kay CL, May J. Duodenal obstruction by gallstones (Bouveret's syndrome): a review of the literature. Endoscopy 2005;37:82-7