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## A galling disease? Dieulafoy's lesion of the gallbladder

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## ABSTRACT

**INTRODUCTION:** Dieulafoy's lesion is a rare vascular anomaly characterized by an abnormally large and tortuous submucosal arteriole leading to an area of mucosal defect with minimal inflammation. It is most often seen in the stomach but could occur anywhere along the gastrointestinal tract. Only five cases of gallbladder Dieulafoy's lesion have been published so far.

**PRESENTATION OF CASE:** We report a case of Dieulafoy's lesion in the gallbladder in a 44 year-old patient who presented with calculous cholecystitis.

**DISCUSSION:** The clinical, radiologic and histologic findings are discussed in light of the existing literature on Dieulafoy's lesions of the gallbladder.

**CONCLUSION:** Gallbladder Dieulafoy's lesion has potentially serious complications and emergency surgery is often required. Due to the rarity of the entity, the diagnosis is often not considered.

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## 1. Introduction

The term “Dieulafoy's lesion” (DL) is attributed to Dr Paul Georges Dieulafoy, who in 1898, described a series of 10 patients who had massive hematemesis due to a lesion in the stomach that he called “exulceratio simplex” [1]. DL classically presents acutely as massive hemorrhage which is often recurrent and can manifest as hematemesis, melena, or hematochezia [2]. The lesions are more common in men (M:F = 2:1) and the elderly (50–70 years) [1]. Patients often have multiple co-morbidities though not necessarily previous history of gastrointestinal pathology [2]. Histologically, it is characterized by a normal vessel with an abnormally large caliber of 1–3 mm that runs a tortuous course within the submucosa [2]. Typically the vessel protrudes through a small mucosal defect with fibrinoid necrosis at its base.

The commonest location for DL is on the lesser curvature of the stomach [2]. In one case series involving 101 cases, 82% of the gastric DLs were found within 6 cm of the gastroesophageal junction [3]. In 1978, two “Dieulafoy-like erosions,” previously thought to be unique to the stomach, were reported in the jejunum [4]. Subsequently, examples of extragastric DL have been described throughout the gastrointestinal tract including the esophagus, duodenum, jejunum, ileum, cecum, colon, rectum and anal canal [2]. Of the gastrointestinal bleed caused by DLs, stomach accounts for 71% of the cases, followed by duodenum with 15% and esophagus

with 8% [2]. Bronchial DL, the first case of non- gastrointestinal DL, was not reported until 1995 [1].

Gallbladder DLs have rarely been reported in literature with only five previously published cases [5–10]. This report details another case of gallbladder DL in line with the SCARE criteria [11] and reviews the existing literature on the clinical presentation, investigations and treatment options for gallbladder DL.

## 2. Case presentation

A 44-year-old Caucasian man with known Crohn's disease, hypertension, ankylosing spondylitis and left hip replacement presented to the emergency department with a 5-h history of chest and epigastric achy pain. He had similar pains intermittently for a few months and was diagnosed with atypical non-cardiac chest pain.

On examination he had crepitation in the right lung base and mild palpation pain in the epigastrium. His blood tests showed normal troponin I and elevated white blood cell count (WBC) (Table 1). The liver function tests (LFTs) showed elevated alanine aminotransferase (ALT) only. The chest X-ray showed right-sided basal atelectasis. Despite oral amoxicillin treatment for possible community-acquired pneumonia, his epigastric pain was still not resolved and he remained pyrexial with worsening WBC and raised C-reactive protein. An abdominal ultrasound arranged to check for gallstone disease failed due to the patient's ankylosing spondylitis and body habitus. Instead, a computed tomography (CT) scan was performed and showed features in keeping with calculous cholecystitis. The patient was started on oral co-amoxiclav. Two weeks after the initial presentation, the patient had an elective laparoscopic cholecystectomy and uneventful recovery.

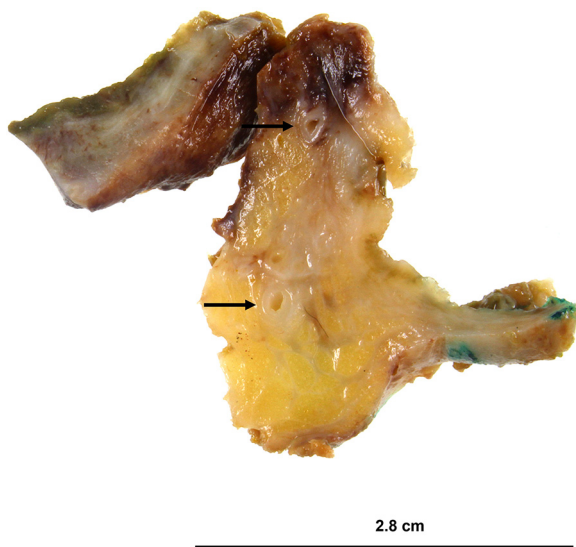
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**Table 1**

Blood test results. Hb: hemoglobin, WCC: white cell count, U&E: urea and electrolytes, CRP: C-reactive protein, ALT: alanine aminotransferase, Alk phos: alkaline phosphatase, N/A: not available.

Reference range	On admission	3 days post admission
Hb (130–180 g/L)	179	172
WCC (4–11 × 10 <sup>9</sup> /L)	13.5	13.7
U & E	normal	normal
CRP (0–10 mg/L)	N/A	242
ALT (0–45 U/L)	58	N/A
Bilirubin (0–21 μmol/L)	14	N/A
Alk phos (40–130 U/L)	99	N/A
Albumin (30–50 g/L)	41	N/A



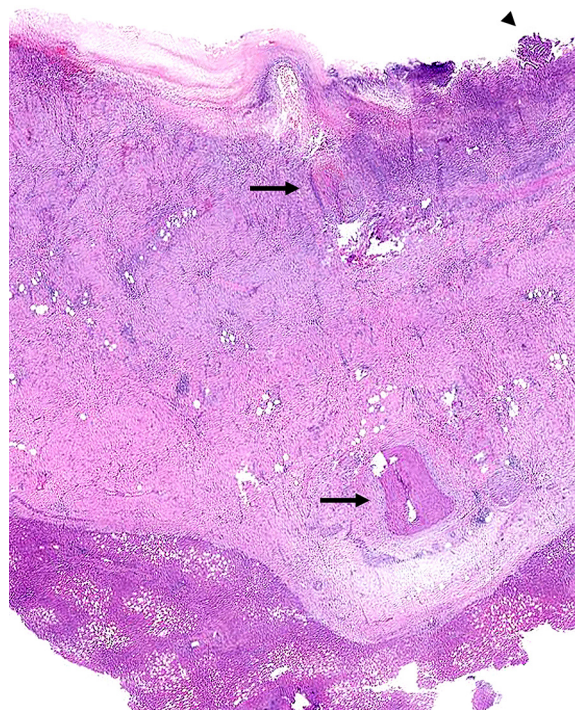
**Fig. 1.** A section of the gallbladder neck with pericholecystic fat showing thickened wall and a prominent vessel (arrows).

**2.1. Histologic findings**

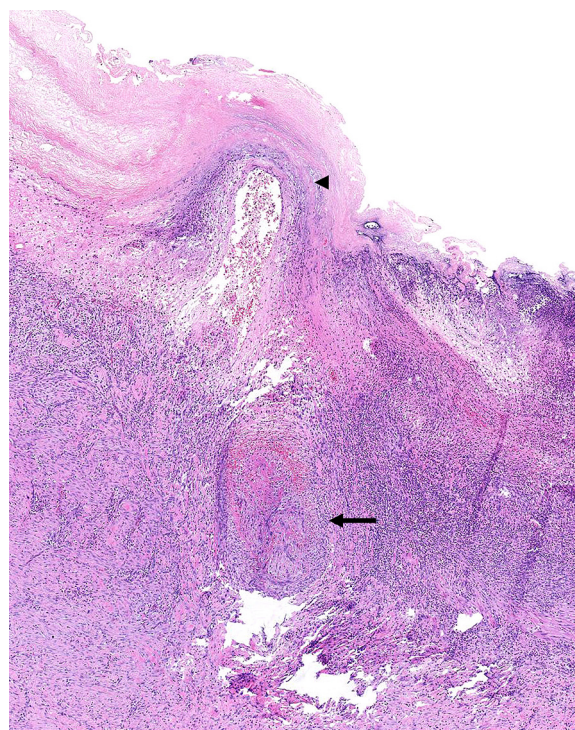
Macroscopic inspection of the 80 × 40 mm gallbladder showed the presence of stones and roughened, hemorrhagic serosa. The mucosal surface was mostly velvety with some areas having a smoother surface and brownish discoloration. The wall was thickened and measured up to 8 mm (Fig. 1). On microscopic examination, focal severe ulceration was found along with chronic mucosal inflammation, submucosal fibrosis, muscle hyperplasia and Rokitsansky-Aschoff sinus formation. In addition to these features of chronic cholecystitis, a large tortuous muscular arteriole measuring 1.2 mm in diameter was noted running through the pericholecystic fat and penetrating into the muscular wall and submucosa of the gallbladder (Fig. 2). Ulceration was seen where the vessel approached the mucosa with very mild surrounding inflammation and extensive fibrosis. Organizing thrombus was present at the site of vessel rupture at the ulcer base. The appearances are those of a DL (Fig. 3).

**3. Discussion**

Paraf first raised the possibility of Dieulafoy’s disease of the gallbladder in a correspondence in 1996 [12]. Truong et al. had reported a case of hemoperitoneum caused by rupture of the gallbladder secondary to arterial ulceration by a calculus [5]. Paraf observed that the microscopic description and the figure in the report showed an artery with abnormal caliber deep to the ulcerated area of gallbladder with no mention of inflammatory infiltrate, similar to those



**Fig. 2.** Low-power view showing a prominent vessel (arrows) traversing through the gallbladder submucosa to an ulcerated area. A small focus of residual mucosa (arrowhead) can be seen in the right upper corner.



**Fig. 3.** High-power view revealing disrupted arteriole wall (arrow) within the submucosa. Ulceration (arrowhead) can be seen overlying the vessel.

seen in Dieulafoy’s disease. However, Truong et al. did not confirm this diagnosis.

Review of the literature reveals subsequent reports of Dieulafoy’s disease of the gallbladder [6–10]. These reports, along with Truong et al.’s case and the current case are summarized in Table 2. The 6 patients with gallbladder DL were aged 45–84 years with a

**Table 2**  
Summary of presentation of gallbladder DL. GB: gallbladder.

Year	Author	Age	Sex	Presentation	Radiology	Histology	Treatment
1996	Truong et al.	69	M	right upper quadrant and epigastric pain	US – peri-hepatic and splenic effusion, GB with hyperechoic areas; CT – subhepatic hemoperitoneum, gallstones	GB perforation, thrombus adherent to mucosal defect, ruptured artery, hemorrhagic dissection of the wall, chronic cholecystitis	emergency operation
2007	Hashimoto et al.	56	M	severe epigastric, upper abdominal guarding	US – distended GB containing high-echo material	an ulcer with a tortuous enlarged artery in the submucosa, inflammatory changes consistent with mild chronic cholecystitis	percutaneous transhepatic gallbladder drainage; emergency laparoscopic cholecystectomy
2011	Sevilla Chica et al.	73	M	severe epigastric and right upper quadrant pain, vomiting	US & CT – ruptured GB with cholehemoperitoneum	large caliber arteriole in area of mucosal defect, no inflammatory infiltrate	emergency laparoscopy; cholecystectomy
2014	Moszkowicz et al.	63	F	progressive abdominal pain, nausea and vomiting 7 days post-thoracoabdominal aneurysm repair	CT – distended GB filled with hyperdense hemorrhagic bile, 8 mm hypervascular lesion	normal vessel with abnormally large diameter, protruding through a small mucosal defect which has fibrinoid necrosis at its base, ischemic cholecystitis	emergency laparoscopy; transcystic drainage
2016	Tarini and Yeo; Stanes and Mackay	84	F	lower abdominal pain and jaundice post-anterior resection for diverticular disease; subsequent melena and hematemesis	US, CT, MRCP – complex gallbladder with an enhanced mural nodule/hypervascular blush; gallbladder distension and fluid extending into Rutherford-Morrison's pouch	multifocal mucosal ulceration with a tortuous medium-sized caliber artery at the base of one of the ulcers	cholecystectomy
present case		45	M	chest/epigastric achy pain	US – unsuccessful due to patient habitus; CT – calculus cholecystitis	large tortuous artery in submucosa, associated ulceration and mild inflammation, chronic cholecystitis	elective cholecystectomy

**Table 3**  
Blood test results for patients with gallbladder DL. Hb: hemoglobin, WCC: white blood cell count, Bili: bilirubin. AST: aspartate aminotransferase, ALT: alanine aminotransferase,  $\gamma$ GT: gamma-glutamyltransferase, N/A: not available.

	Hb	WBC	Bili	AST	ALT	$\gamma$ GT
Truong et al.	10.8 g/dL	114000/mm <sup>3</sup>	19 $\mu$ mol/L	normal	normal	1.6x upper normal limit
Hashimoto et a.	9.9 g/dL	↑	↑	↑	↑	N/A
Sevilla Chica et al.	N/A	N/A	N/A	N/A	N/A	N/A
Moszkowicz et al.	9.6 g/dL	normal	65 $\mu$ mol/L	185 UI/L	304 UI/L	N/A
Tarini and Yeo; Stanes and Mackay	↓	33.8 × 10 <sup>9</sup> /L	N/A	N/A	N/A	N/A
Present case	17.9 g/dL	13.5 × 10 <sup>9</sup> /L	normal	normal	58 UI/L	normal

male predominance (67%). They all complained of abdominal pain. Four of the patients had pain focused in the epigastrium and right upper quadrant. One patient had a rigid abdomen on presentation [6]. Some blood test results were available for 5 of the patients (Table 3). Of those, 83% had decreased hemoglobin. All showed a degree of liver function derangement. Both ultrasound and CT were utilized to investigate the cause of abdominal pain. Only one case revealed a hypervascular lesion in the gallbladder on CT which might raise suspicion for DL [8]. Four patients required emergency cholecystectomy.

This review illustrates the difficulty of diagnosing gallbladder DL in the acute setting. Physical examination can localize pain and reveal acute abdomen. However, there are a multitude of causes for right upper quadrant and epigastric pain with overlapping signs and symptoms. Elevation of WBC and derangements in LFTs might help focus attention on hepatobiliary causes abdominal pain. Decreased hemoglobin level could point to a bleed but this is not seen in all patients. None of the patients reviewed had overt signs of bleeding such as hematemesis or melena.

Ultrasound is an excellent screening tool for suspected acute cholecystitis. Bleeding and perforation of the gallbladder can also

be detected with ultrasound [13]. Of the three patients who had an abdominal ultrasound, all showed either a ruptured gallbladder or hyperechogenic material within the gallbladder to indicate bleeding. However, the underlying cause of hemorrhage into the gallbladder was not identified. CT can detect high attenuation blood products and has the added advantage of being able to evaluate other intra-abdominal pathologies at the same time [13]. In one case the CT scan demonstrated a 8 mm hypervascular lesion leading to immediate surgery [8]. However, radiology did not contribute to the diagnosis of gallbladder DL for the remaining cases. CT angiography is the modality of choice for suspected hemobilia because of its ability identify the source of bleeding [14]. But unlike other DLs in the gastrointestinal tract, the treatment of choice for gallbladder DL is cholecystectomy rather than endoscopic clipping, sclerotherapy or embolization, rendering the angiography moot in an emergency situation.

Potential complications associated with gallbladder DL include hemobilia, ruptured gallbladder and hemoperitoneum. Sixty-seven percent of the patients reviewed required emergency surgery. Whilst cholecystectomy is in general a safe procedure, it is well recognized that morbidity and mortality rates are significantly higher

for emergent cholecystectomy compared to elective cholecystectomy [15]. In this review, all six patients had uneventful recovery after their cholecystectomy.

#### 4. Conclusion

This case provides further evidence of the existence of DL in the gallbladder. Given the rarity of the entity, the diagnosis is often not considered. However, gallbladder DL has potentially serious complications and emergency surgery is often required. Clinicians should be aware of this potentially lethal but easily curable disease. A major limitation of this study is its small patient population. Further case reporting of gallbladder DL could help better characterization of the disease.

#### Conflicts of interest

None.

#### Funding

None.

#### Ethical approval

Ethical approval is exempt by our institution for case reports.

#### Consent

Patient consent has been obtained and is available for review by the Editor-in-Chief.

#### Author contribution

Both authors have contributed to acquisition of data, data analysis and interpretation and writing of the article.

#### Guarantor

Dr Abed M Zaitoun.

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