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A 42-Year-Old Woman with Recurrent Pancreatitis Associated with Gallstones and Phrygian Cap Gallbladder

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Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Patient: Female, 42-year-old
Final Diagnosis: Phrygian cap gallbladder • recurrent pancreatitis
Symptoms: Abdomen distension • recurrent pancreatitis
Clinical Procedure: —
Specialty: Surgery

Objective: Rare disease

Background: Gallbladder anomalies are rare congenital defects with an incidence rate of approximately 2% in the general population. Phrygian cap gallbladder is a common anatomical variant in which the fundus of the gallbladder folds on itself. Gallstone impaction is rare, and it can be associated with acute pancreatitis. This report describes a 42-year-old woman with recurrent pancreatitis associated with gallstones and Phrygian cap gallbladder.

Case Report: We report the case of a 42-year-old woman with acute biliary pancreatitis and a history of repeated hospitalizations for episodes of pancreatitis. A preoperative MRI was conducted, which revealed the presence of a Phrygian cap gallbladder that had not been previously reported in imaging studies. The patient underwent cholecystectomy surgery with a laparo-endoscopic approach (rendezvous technique). No intra- or postoperative complications occurred.

Conclusions: We report a case of acute biliary pancreatitis caused by stone migration and describe the anatomical variant of the Phrygian cap gallbladder with its clinical implications. The literature contains very few reports of cholecystitis or pancreatitis in patients with a gallbladder anomaly. Continuous reporting of anatomical variations of the gallbladder and biliary tract improves clinical knowledge, and knowledge of gallbladder anomalies is crucial to avoid injury to the biliary tract during laparoscopic cholecystectomy. This case emphasizes the importance of accurate preoperative evaluation to prevent serious surgical complications.

Keywords: Cholecystectomy, Laparoscopic • Cholestasis with Gallstone, Ataxia, and Visual Disturbance • Diagnostic Imaging • Gallbladder Diseases • Pancreatitis

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Introduction

To avoid intraoperative damage, surgeons need to be aware of the various gallbladder anomalies, which can include variations in number, location, and form [1]. The Phrygian cap is a common normal variation of the gallbladder form. It denotes folding of the fundus back upon the gallbladder body. The term Phrygian cap is derived from the appearance of this anomaly resembling the traditional hats worn by the inhabitants of the ancient region of Phrygia (modern-day Turkey) [2]. In a recent review on morphological variations of the gallbladder [3], the presence of a Phrygian cap gallbladder was found in 2.15% of the analyzed samples. A Phrygian cap, however, has no pathological significance and normally causes no symptoms [4]. The literature and case review indicated that misidentification of biliary anatomy was the major cause of bile duct injury [5]. For this reason, several clinical cases have been presented in the literature over the years, emphasizing the rarity and potential risk of not being aware of gallbladder and biliary tract anomalies. It is therefore important to use appropriate imaging techniques to diagnose anomalies such as Phrygian cap [6]. This report describes a 42-year-old woman with recurrent pancreatitis associated with gallstones and Phrygian cap gallbladder.

Case Report

A 42-year-old woman was admitted to our department with a diagnosis of acute pancreatitis. She had a past medical history of small bowel resection, hysterectomy with removal of adnexa, mesenteric lymph node removal, appendectomy, and omentectomy for neuroendocrine tumor (NET). She had also received treatment with somatostatin and had a ureteral stent placed due to recurrent kidney stones. She had a history of recurrent abdominal pain and was admitted to another hospital 3 months ago for acute biliary pancreatitis, prior to admission at our hospital. On clinical examination, the abdomen was tender in the right hypochondrium, without any other abdominal signs. Blood tests revealed white blood cell (WBC) count 12.65×10^3/uL, C-reactive protein (CRP) 0.57 mg/dl, and lipase 4230 U/l. Emergency abdominal computed tomography (CT) revealed the presence of microlithiasis in the gallbladder and biliary tract with thickened walls (**Figure 1**). In consideration of the likely presence of gallstones in the biliary tract not visualized by CT, we performed T2-weighted magnetic resonance cholangiopancreatography (MRCP), which showed cholecysto-choledochal gallstones (**Figure 2**). However, only 3D coronal reconstruction images raised the suspicion of an abnormal gallbladder (**Figure 3**).

Therefore, we proposed a surgical laparo-endoscopic approach with the rendezvous technique. The technique consists of anterograde transcystic cannulation of the bile duct during laparoscopic cholecystectomy. The guidewire can subsequently

Figure 1. Preoperative examinations. Abdomen CT scan shows thickening of the gallbladder (arrow) walls and dilation of the common bile duct (triangle). Furthermore, the twisted appearance of the organ simulates the presence of 2 different chambers.

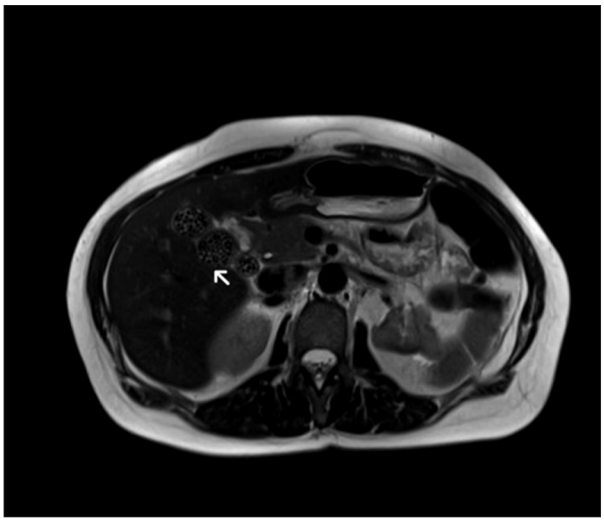

Figure 2. Preoperative examinations. In T1-weighted sequences, magnetic resonance cholangiopancreatography confirms the presence of cholecysto-choledochal gallstones.

be retrieved from the endoscope, thus facilitating retrograde cannulation of the bile duct. A wire sphincterotomy is then performed and standard maneuvers for endoscopic removal of common bile duct stones are performed. The procedure is then completed by cholecystectomy in a single procedure. The laparo-endoscopic rendezvous technique was developed to facilitate cannulation of the bile duct during endoscopic sphincterotomy while reducing the risk of failed endoscopic clearance of the common bile duct and pancreatitis after endoscopic retrograde cholangiopancreatography (ERCP) [7].

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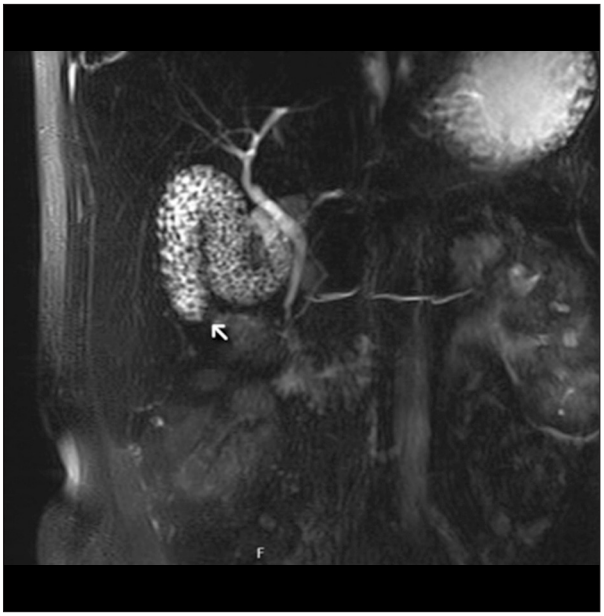

Figure 3. Preoperative examinations. In T2-weighted sequences, magnetic resonance cholangiopancreatography shows a Phrygian cap gallbladder (arrow) in the coronal plane with the typical twisted appearance.

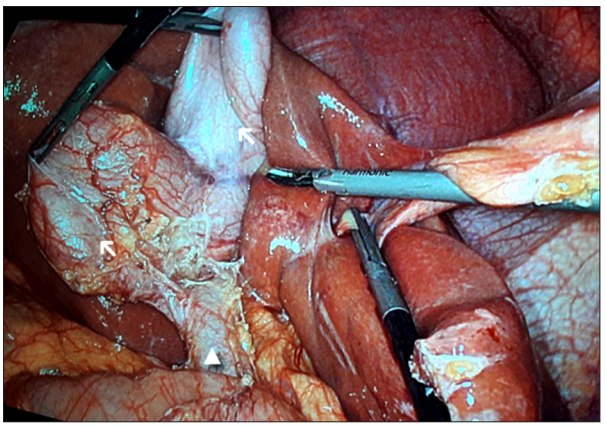

Figure 4. Intraoperative image. Phrygian cap gallbladder (arrows) and the common bile duct (triangle).

During the surgery, after a challenging adhesiolysis, a Phrygian cap gallbladder was identified (**Figure 4**) with a single cystic duct that joined the main bile duct. Before performing cholecystectomy, we performed a critical view of safety (CVS), which confirms the anatomy and shows cholecystectomy can be performed safely. The guidewire inserted into the cystic duct was retrieved by the endoscopist, and the bile duct was cleared of small stones after sphincterotomy. Postoperative blood tests showed WBC 9.61×10^3/uL, CRP 0.02 ng/mL, and lipase 47 U/l. The patient was discharged 3 days after surgery, with a 4-day hospital stay. Histological examination of the surgical specimen showed a gallbladder with multiple stones and moderate inflammation. Three-month follow-up showed no problems. The patient now continues routine oncological follow-up at our center.

Discussion

Anatomic abnormalities of the gallbladder are rare and estimated to occur in approximately 2% of the population [8-10]. The Phrygian cap usually is asymptomatic and without pathological significance. Despite this, knowledge and study of anomalies of the biliary tract and gallbladder are of fundamental importance to safely perform routine operations such as cholecystectomy**.** Congenital anomalies of the gallbladder and anatomical variations of their positions can be associated with an increased risk of complications after laparoscopic cholecystectomy, both elective and emergency. Gallbladder anomalies can be found completely randomly during cholecystectomy surgery or can be shown in preoperative imaging. Various gallbladder anomalies occur, including gallbladder diverticula, gallbladder fold, gallbladder duplication, common bile duct cyst, pericholecystic fluid, focal adenomyomatosis, intraperitoneal fibrous bands, Phrygian cap gallbladder, and gallbladder duplication. Regarding classifications for gallbladder duplications, the most well-known are the Boyden classification [11] and the Harlaftis classification [12]. According to Boyden's classification, the different types of gallbladder duplications are: 1) divided gallbladder (bilobed or bifid gallbladder, double gallbladder with common neck); 2) duplex gallbladder (double gallbladder with 2 cystic ducts); (i) Y- H-shaped type (the 2 cystic ducts joining before entering the common bile duct); and (ii) H-shaped type (ductular type, the 2 cystic ducts entering the biliary tree separately). In our patient, we observed a gallbladder anomaly known as Phrygian cap gallbladder. In a recent review on morphological variations of the gallbladder [3], the presence of Phrygian cap gallbladder was found in 2.15% of the analyzed samples. This type of anomaly is not described in the Boyden and Harlaftis classifications. However, based on preoperative imaging, we can categorize the anomaly in our case as a divided gallbladder according to Boyden's classification (**Figure 5**). According to Harlaftis's classification (**Figure 6**), the gallbladder in our case was a type 1 septated gallbladder. We believe there is confusion between the macroscopic appearance and imaging, leading to a lack of clear distinction between the duplicated gallbladder and the gallbladder with a Phrygian cap. Recognition of these anomalies is important for the surgeon's practice, as a lack of knowledge can lead to intraoperative complications [11,13]. Through reviewing these rare cases in the literature, it becomes evident that there is a low incidence of this specific type of anomaly. Further understanding and recognition of these anomalies can aid in avoiding potential complications during surgery. Various diagnostic methods are used to detect gallbladder anomalies. Ultrasound is the most used diagnostic tool. Although

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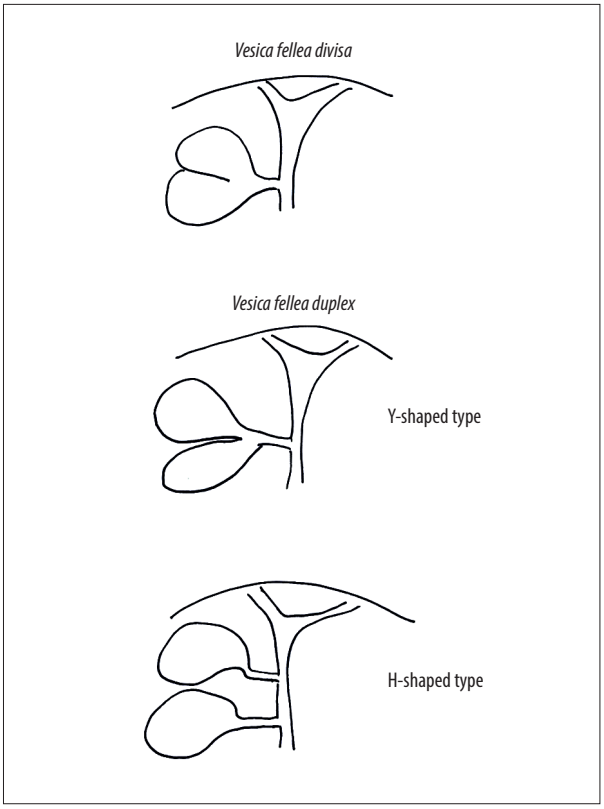

Figure 5. Boyden's classification. Anomalies described as vesica fellea divisa and vesica fellea duplex. 1) Vesica fellea divisa or divided gallbladder (bilobed or bifid gallbladder, double gallbladder with common neck). 2) Vesica fellea duplex or duplex gallbladder (double gallbladder with 2 cystic ducts), a) Y-H-shaped type (the 2 cystic ducts joining before entering the common bile duct), b) H-shaped type (ductular type, the 2 cystic ducts entering the biliary tree separately).

ultrasound is widely used, its diagnostic power is lower compared to computed tomography (CT), magnetic resonance imaging (MRI), magnetic resonance cholangiopancreatography (MRCP), and endoscopic ultrasound (EUS). MRI is particularly useful in detecting anatomic anomalies of the gallbladder and extrahepatic bile ducts [6]. Preoperative imaging is essential, especially in cases where there is a suspicion of anatomical abnormalities on ultrasound. Furthermore, imaging is essential in centers where intraoperative methods such as indocyanine green or intraoperative cholangiography are not available. In this case report, the patient had undergone CT and MRI scans. Interestingly, none of the radiologists reported any gallbladder abnormalities, indicating a lack of understanding of gallbladder anatomical anomalies and associated radiological findings. Images are crucial for adequate preoperative planning. The preoperative detection of gallbladder anomalies can prevent severe complications [14]. Therefore, correctly defining the anatomy of the biliary tree is crucial in reducing the risk of biliary or vascular injuries, including injuries to the common bile duct. In fact, the most common cause of biliary injury is misidentification [15]. The most frequent error is mistaking the common bile duct for the cystic duct. Performance of CVS has standardized the technique to avoid biliary injuries [16]. In this case, the gallbladder consisted of a single chamber with a single cystic duct and a single hepatic artery (**Figure 4**). CVS was achieved before clipping the cystic duct and cystic artery. Laparoscopic cholecystectomy was completed successfully. The operation was performed using 5 port placements. Intraoperatively there were dense adhesions of the omentum to the gall bladder, which were released carefully; the gall bladder was grasped at the fundus and retracted caudally to the right shoulder. The Phrygian cap anomaly with significant adhesions could have presented difficulties for the surgeon. However, the surgeon was aware of the anatomical anomaly thanks to preoperative imaging. In most reported cases in the literature, the presence of the Phrygian cap anomaly was detected incidentally during surgery performed for other reasons [4,17,18] or as a collateral radiological finding [19-21]. The Phrygian cap anomaly can be mistakenly identified as a thickening of the gallbladder walls, leading to unnecessary surgery [22]. Additionally, there are reported cases of acute cholecystitis in patients with gallstones and Phrygian cap anomaly [23,24]. Only 1 case in the literature described acute cholecystitis without gallstones [25]. Recurrent acute pancreatitis [26,27] and cholangitis [14] have also been described in patients with the Phrygian cap anomaly. No surgical complications were reported in the cases described, although we believe that complications are underestimated due to the limited available data. There are no guidelines addressing the indication for prophylactic cholecystectomy in patients with gallbladder abnormalities. However, routine treatment of asymptomatic gallstones is not recommended in current guidelines, mainly due to the low annual incidence of symptoms and complications, as well as the costs and risks of surgery [28]. Prophylactic cholecystectomy should be considered only in patients with hereditary spherocytosis and sickle cell disease and concomitant asymptomatic gallstones at the time of splenectomy. Darnis et al [29], in a systematic review of the anatomical findings of multiple gallbladders, concluded that cholecystectomy is indicated for symptomatic stone disease. Thus, we can conclude that prophylactic cholecystectomy is not recommended in cases of gallbladder anomalies. Laparoscopic cholecystectomy is the preferred approach for symptomatic gallbladder anomalies. Gallbladder malformations are considered a risk factor for iatrogenic bile duct injuries. In fact, a review by Pesce et al [30] shows that anatomical anomalies of the gallbladder are considered a risk factor for iatrogenic bile duct injury. In situations where an unexpected anomaly of the biliary tree is identified, intraoperative cholangiography can help avoid iatrogenic injuries. The routine use of "intraoperative cholangiography (IOC)" has been proposed to improve the visualization of biliary anatomy, detect silent common bile duct

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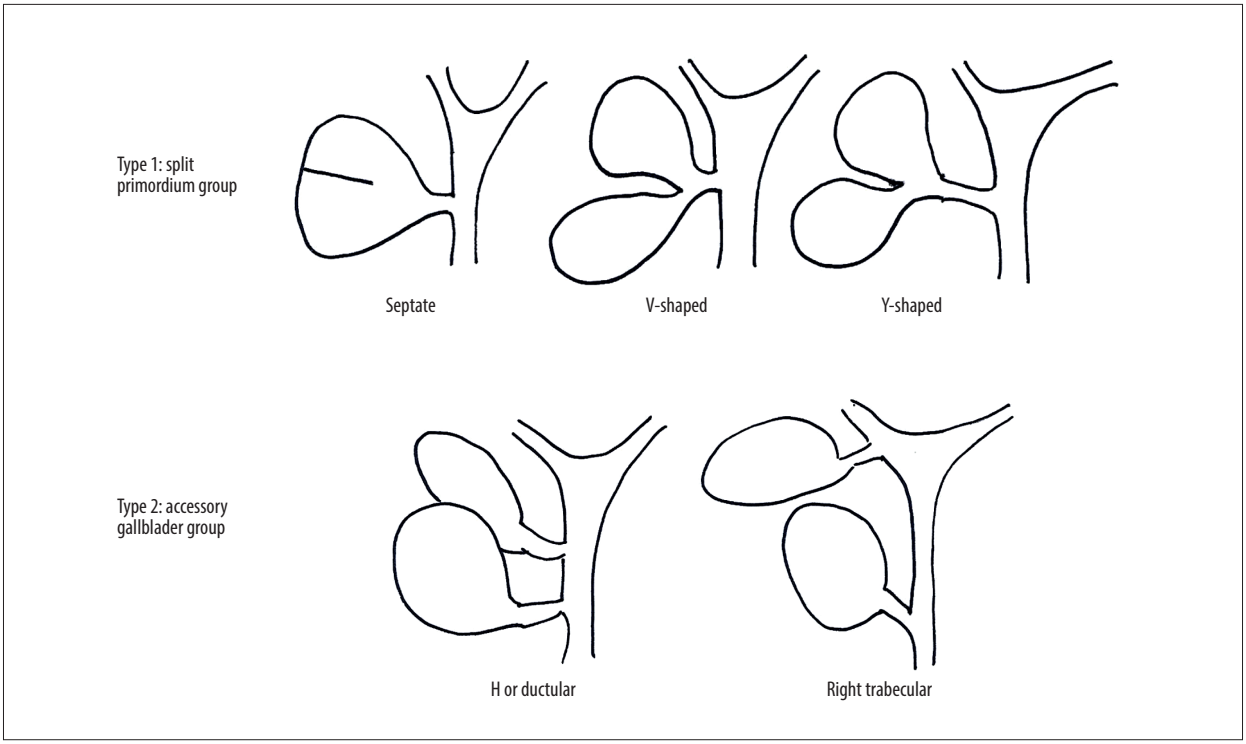

Figure 6. Harlaftis's classification. Anomalies divided into type 1 (split primordium group) and type 2 (accessory gallbladder group). Subsections of Type 1: septate, v-shaped (bilobed), y-shaped. Subsections of Type 2: h-type (ductular) and trabecular type.

stones, and reduce the incidence of iatrogenic bile duct injury. Its routine use is still debated. Studies in the literature show that the routine addition of IOC to laparoscopic cholecystectomy does not increase the rate of residual bile duct stones or bile duct injuries or lengthen operative time [31,32]. However, a recent systematic review [33] showed that routine IOC has benefits, such as choledocholithiasis detection and bile duct variation visualization, leading to further exploration, reduced postoperative complications, and improved outcomes. IOC should be used cautiously, considering clinical indications, such as dilated common bile duct (CBD), elevated liver enzymes, pancreatitis, jaundice, and the surgeon's skill level. One of the innovations in minimally invasive technology is fluorescence image-guided surgery; "near-infrared fluorescent cholangiography (NIRF-C)" is an innovative intraoperative imaging technique that provides real-time enhanced visualization of the extrahepatic biliary tree through fluorescence. NIRF-C can be very useful in elective laparoscopic cholecystectomy for preoperatively identified anomalies. Unfortunately, not all hospitals are equipped with the necessary technology for IOC and/or NIR-C. Consequently, preoperative evaluation of gallbladder abnormalities should be performed whenever possible. Preoperative imaging is sometimes the only means available. In emergency or elective situations, imaging evaluation before surgery is the first step to correctly addressing a laparoscopic cholecystectomy.

Conclusion

This report presents a case of acute biliary pancreatitis associated with a Phrygian cap gallbladder anomaly, emphasizing the importance of recognizing these anomalies. Recognition of such anomalies preoperatively, particularly using high-resolution MRI, is important to avoid surgical complications. In our patient, MRI allowed us to identify the anomaly before surgery, allowing us to perform cholecystectomy safely. It is crucial for surgeons to be aware of these abnormalities and approach a cholecystectomy after proper preoperative imaging study. Continuous reporting of anatomical variations improves clinical knowledge, contributing to safety during operations. Avoiding serious complications during cholecystectomy is crucial, and studying these anomalies can help achieve this objective.

Department and Institution Where Work Was Done

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Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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