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A Case of Cholecystitis Associated with Serosal Phrygian Cap: Importance of Preoperative Diagnosis

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ABSTRACT

Introduction: the Phrygian cap is a congenital anomaly with an estimated incidence of 1-6%. It is usually an accidental finding during surgery or in imaging studies. It can however cause cholecystitis as a complication. We report here a case of Phrygian cap with cholecystitis detected intra-operatively along with a mini review of literature. It was found to be serosal type and benign in nature as confirmed by histopathology. A pre-operative diagnosis of Phrygian cap is not warranted as it is considered pathologically quiescent; but it may mimic solid hepatic or adnexal mass in imaging studies and can be misdiagnosed. Multi-phase MRI and cholescintigraphy is the preferred imaging modality due to their higher sensitivity over conventional ultrasound and computed tomography scan. This report is highlighting the importance of pre operative diagnosis of this anomaly, which is not easily detected in routine evaluations but can be misdiagnosed and present as cholecystitis later.

Keywords: Phrygian cap; Gallbladder anomalies; Cholecystitis; Hepatobiliary imaging; Gallbladder imaging.

Introduction

The Phrygian cap is a relatively common congenital anatomical variation of the gallbladder fundus which belongs to the variety of anomalies associated with gallbladder septation[1]. The other known anomalies of the gallbladder development are agenesis, septate gallbladder, hypoplastic gallbladder, duplicated gallbladder, intrahepatic gallbladder and left-sided gallbladder[2]. Its reported incidence rate is approximately 1-6%[3]. Two types of this anatomical variation is recognised: the concealed type or retroserosal type and the visible or serosal type[3]. Phrygian cap is usually diagnosed either at surgery or through pre-operative radiological investigations. As it does not narrow the lumen of the gallbladder, it is not commonly found to be associated with gallbladder pathology most of the times[4]. Many known congenital and acquired anatomical variants of gallbladder have been described in the literature since the first cholecystectomy performed in 1882[5] careful thought, cadaver dissections and careful patient selection. Langenbuch's career was linked with the Lazarus Hospital in Berlin, to which he was appointed as Director at the age of 27. The hospital was then 4 years old and in a poor area of Berlin, and they grew in reputation together. Going through repeated troubled times, this hospital has withstood financial disaster, three wars, an occupation and the ambitions of administrators and developers. Treatment for cholecystitis before Langenbuch was stone extraction from spontaneous cutaneous biliary fistula or skin incision of near-pointing empyema. Cholecystostomy was carried out by Bobbs (1867. These anomalies are with regard to its shape, locationor numbers some of which are not known to be associated with pathologies and only observed accidentally. The occurrence of congenital anomalies is due to faulty embryonic development during the very crucial period of organogenesis. At 4th week of intrauterine life, the caudal portion of the ventral foregut bud gives rise to the liver, gallbladder, and biliary tree[6] The initial hepatic diverticulum develops further into gallbladder during 4th-5th week. Folding of the fundus during this embryonic period can result in the occurrence of Phrygian cap which is the most common congenital gallbladder congenital[7]. *Boy den* originally described this anatomical variation in 1935[8]. The name Phrygian cap has been derived from the conical headgear used by people of Phrygia now located in central Turkey whose top portion is pulled forward and encircles the head from all around. Though a common anatomical variant, it can mimic a hepatic or adnexal mass during hepatobiliary imaging misleading the clinicians. It also confuses clinicians while differentiating from duplication of gallbladder in imaging studies[1]. Primarily considered to be a benign entity with no known pathological association, it might sometimes be associated with cholecystitis. The conventional ultrasonography (USG) and computed tomography scan (CT scan) are not very sensitive. Therefore, it is imperative that more advanced and sensitive imaging modalities such as multiphase magnetic resonance imaging (MRI) and scintigraphy

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are preferred to accurately diagnose such a rare congenital variant. Prenatal USG has been found effective for diagnosing duplication of gallbladder in fetus, however no such record is available for Phrygian cap[9] who were referred to our Centers were included in this study. The diagnosis of duplication of gallbladder was based on the evidence of double gallbladder in the standard abdominal circumference plane using grey scale. Postnatal magnetic resonance cholangiopancreatography (MRCP. The present case reports an accidentally observed Phrygian cap in a middle-aged female with cholecystitis during surgery along-with a narrative review explaining significance of pre-operative diagnosis of Phrygian cap anomaly of gallbladder.

Case Report

A 41 years old woman presented with acute right upper abdominal pain accompanied by 5 episodes of vomiting. It was not associated with passage of loose watery stool, passage of blood in stool, loss of appetite, weight loss and her bowel and bladder habits were normal. Past medical history revealed essential hypertension for which she was on regular medication but without any obvious gastrointestinal issues. General physical examination including per abdominal and digital per rectal examination was insignificant. Abdominal USG revealed a solitary calculus in the gallbladder lumen with normal wall thickness.

The patient underwent a cholecystectomy operation which was uneventful. Kocher's subcostal rooftop incision of approximately 5.5 centimeter length was made parallel to the sub costal margin. The abdomen was opened up in layers and a common bile duct was identified. Dissection of the Calot's triangle was done by blunt and sharp dissection and cystic duct and the cystic artery were identified. Gall bladder was separated from the gallbladder fossa and the gallbladder was removed. Cystic artery and cystic duct was ligated using 3-0 silk suture and cut using scissors. Haemostasis was achieved with diathermy and liver bed was double checked for residual bleeding. Abdomen was closed in layers with Polyglactin 910, 1-0 suture and finally skin incision was closed by nylon 2-0 suture. The Phrygian cap anomaly was observed intra operatively (Fig.1 and 2). The fundus of the gallbladder was folded into a cap-like appearance. Histopathological examination revealed minimal inflammatory changes in the gallbladder. The patient was discharged on the 3rd post-operative day. The intra-operative and peri-operative hospital stay was uneventful. The patient had no fresh complaints at initial follow up at 7th post-operative day and subsequent follow up at 1 month.

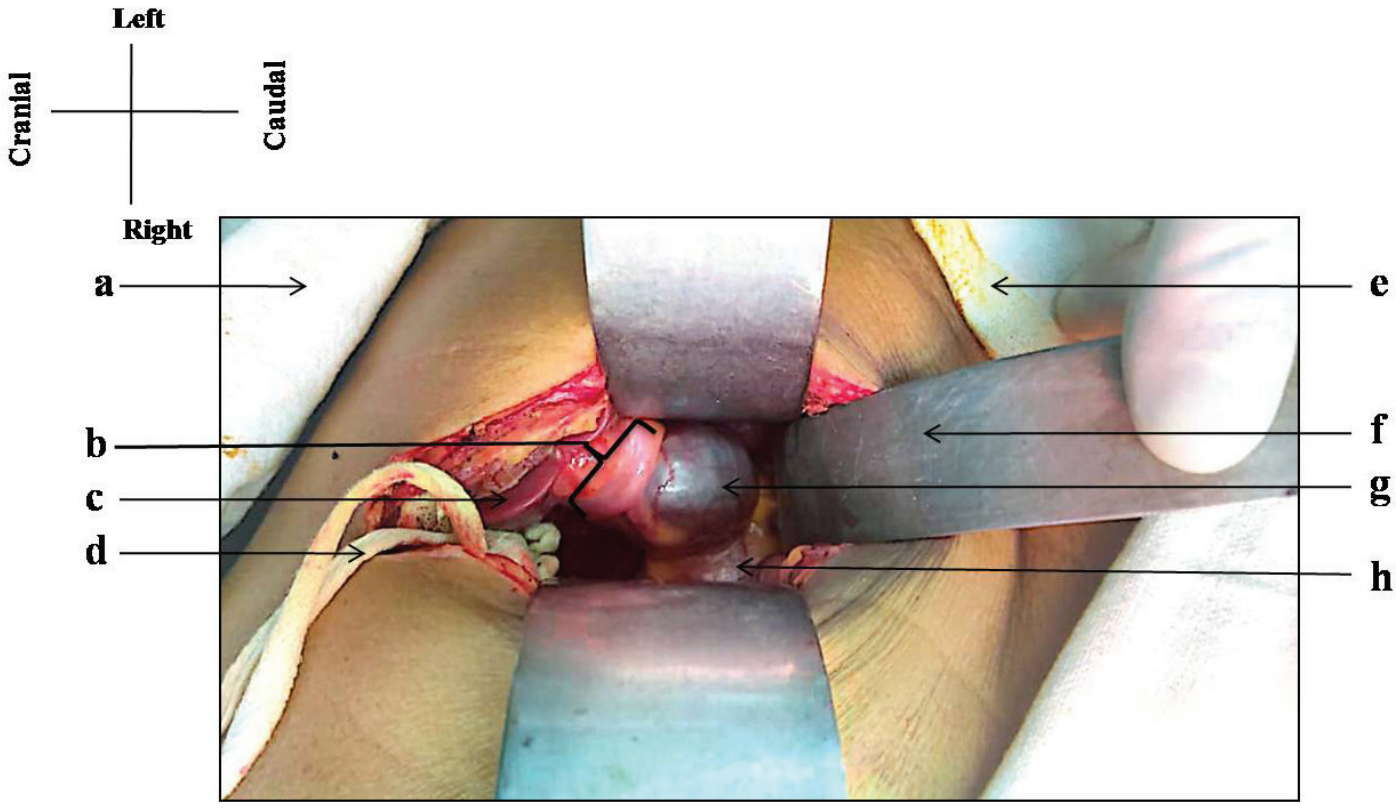

Figure 1. Perioperative image of the gallbladder with Phrygian cap which is seen to be folded like a cap. a-surgical drappings, b-Phrygian cap, c-inferior border of the liver, d- surgical towel, e-surgical drapping, f- Deaver retractor, g-fundus of the gallbladder, h-common bile duct.

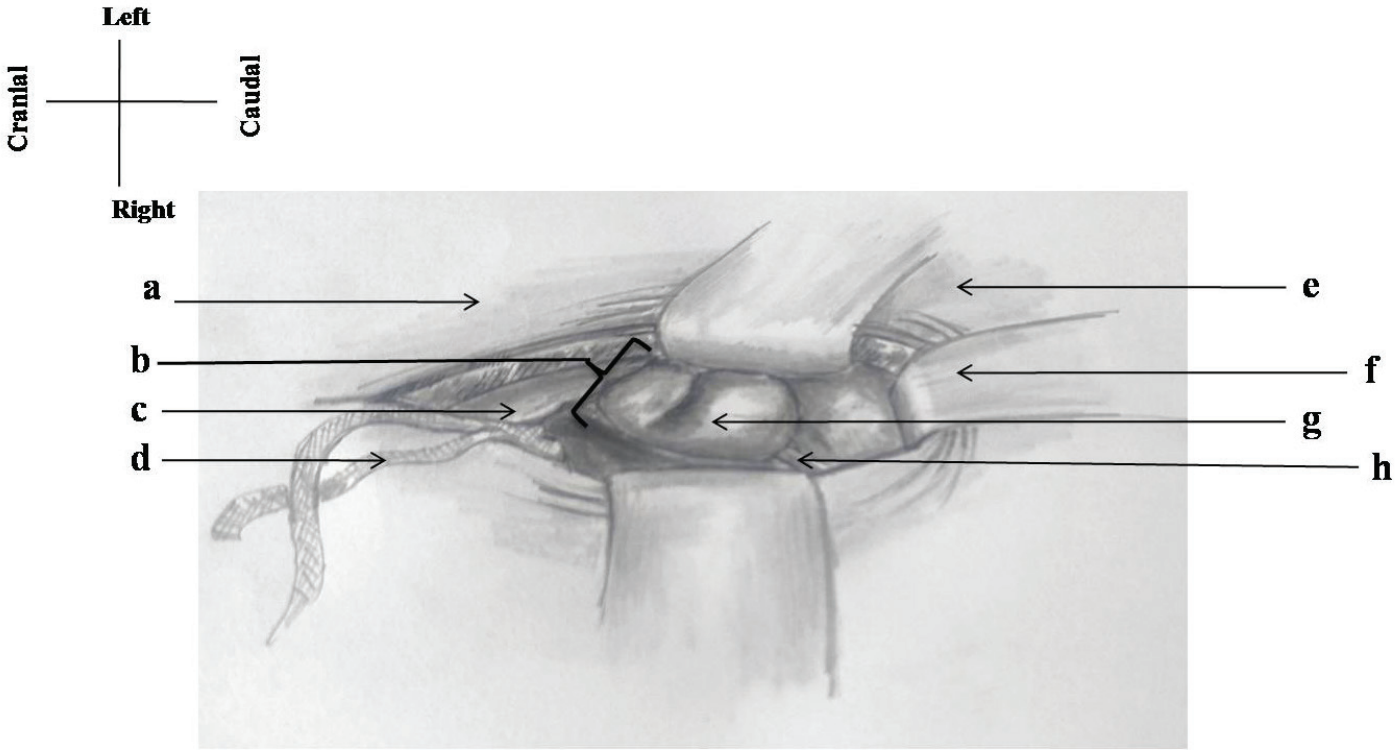

Figure 2. Sketch showing perioperative image of the gallbladder with Phrygian cap which is seen to be folded like a cap. a-surgical draping, b-Phrygian cap, c-inferior border of the liver, d- surgical towel, e-surgical draping, f- Deaver retractor, g-fundus of the gallbladder, h-common bile duct (This sketch is drawn by Kaushik Bhuya Keot, the operating surgeon on this case)

Discussion

Understanding of the hepato-biliary tract begins with the appreciation of its embryological development. At the beginning of 4th week of gestation, distal foregut gives rise to the liver bud. With formation of the hepatic parenchyma, the cells of the ventral mesentery of the foregut intervening the region between hepatic bud and distal foregut differentiate to form the progenitor bile duct. During 4th–5th week of gestation, the caudal portion of this progenitor bile duct sprouts off the gallbladder primordium which lies close to the developing ventral pancreatic bud[10]. The gallbladder primordium eventually differentiates into gallbladder and cystic duct. Various diagnostics tests can readily detect this anomaly with variable sensitivity. The commonly employed imaging studies are USG, CT scans, oral cholecystograms, cholescintigraphy and the multiphase MRI etc. Though USG and CT scans are the common initial imaging modality, they are not sensitive enough to detect it all the time. Moreover diagnostic confusion might occur with a solid hepatic or adnexal mass lesion, which has led to the wider use of advanced techniques such as cholescintigraphy, multidetector CT scans and multiphase MRI[2]. Phrygian cap has been imaged in non-human mammals also including mice in live animal MRI studies[11]. Phrygian cap is commonly mistaken with duplications of gallbladder in imaging studies. Duplication of gallbladder is a rare condition with a more predilection for its occurrence in males. It is not easy to clearly distinguish the two cavities of gallbladder in Phrygian cap even in the presence of inflammation unless when sludge, stone or contrast material is exclusively present in the primary cavity[12]. Therefore there can be many possible other differential diagnosis of Phrygian cap provided in imaging studies viz. folded gallbladder, duplicated gallbladder, gallbladder diverticulum, localized adenomyomatosis, choledochal cyst and vascular or fibrous bands etc. which need to be correlated clinically[2]. It was once considered to be representing the transverse congenital septum associated with pathology. This rare congenital anomaly has often perplexed clinicians. Phrygian cap cholecystitis has been reported across literature by many authors[13,14]. Double Phrygian cap has also been reported to be associated with cholecystitis[13] . A concise review of the studies reporting Phrygian cap and associated anomalies is summarized (Table 1). Currently the preferred method of hepatobiliary imaging is multiphase MRI or multidetector CT scans which can effectively differentiate between a gallbladder anomaly and a solid hepatic or cystic mass lesion. In our case, the preoperative USG missed the anomaly, which could be found later only during surgery. Prophylactic cholecystectomy is not recommended in all cases since a Phrygian cap is pathologically quiescent. Nevertheless cholecystectomy is suggested in symptomatic cases[15].

Table 1. A comprehensive literature review of the Phrygian cap and related gallbladder anomalies across different population groups.

Authors	Study Population	Sample size	Significant findings
Alexander A *et a*l. 1986	British	4	Congenital gallbladder septa presenting with acute cholecystitis
SeiderS *et al*. 1987	South African	1	Acquired gallbladder septum after cholecystojejunostomy
MazziottiS *et al*. 2001	Italian	1	MRI can accurately detect duplication of gallbladder
Pomp A *et al*. 2003	American	1	Gallbladder kinking confirmed on radionuclide scanning
Enid Y *et al*. 2009	American	1	Multiseptate gallbladder occurrences
Park S H *et al*. 2009	Korean	1	Pseudodiverticulosis of gallbladder mimicking cholelithiasis with multiseptate gallbladder
Anilkumar V R *et al*. 2013	Indian	1	Phrygian can mimicking choledochal cyst
Al-AshqarM *et al*. 2013	British	1	Phrygian cap cholecystitis
Jean-Pierre Raufman J P *et al*. 2012	American	-	First report of Phrygian cap in murine gallbladder.
Marie-JanneS *et al*. 2013	Dutch	1	Incidental Phrygian cap during surgery
Kannan N S *et al*. 2014	Indian	1	Congenital bilobed gallbladder with Phrygian cap with cholecystitis
Santos V M dos *et al*. 2014	Brazilian	1	Incidental Phrygian cap during surgery
Rafailidis V *et al*. 2014	Greek	1	Co-occurrence of Phrygian cap in ectopic gallbladder
Afodun A M *et al*. 2017	Nigerian	1	Double Phrygian cap during surgery
Kokilavani J *et al*. 2017	Indian	1	Ultrasonic findings of Phrygian cap
Mao F *et al*. 2021	-	-	Murine study showing Slc10a1 deficiency associated with Phrygian cap deformity
Kerr L *et al*. 2021	Australian	1	Phrygian cap cholecystitis

Conclusion

Phrygian cap presents only occasionally with symptoms suggestive of cholecystitis. Therefore, a strong clinical index of suspicion is required not to misdiagnose it. The accurate diagnosis of this congenital anatomical variant will avoid unwanted complementary examinations and mismanagement. A Phrygian cap should be considered in the differential diagnosis if the gallbladder appears smaller than the gallbladder fossa or if a mass is visible in the fundus on hepatobiliary imaging. In addition, multiphase images from an MRI or CT scan should be obtained whenever possible to rule out a mass lesion. Knowledge of this rare variant combined with the newer imaging studies will help clinicians to avoid misdiagnosis.

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Mini Curriculum and Author's Contribution

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