

Report on a “Double Phrygian Cap” and Phrygian prominence in a Nigerian population

Adam M. Afodun^{1,2}, Olayemi J. Olajide³, Airat A. Bakare⁴, Kofoworola K. Quadri⁵, Mustapha A. Masud⁶, Abdurrasheed O. Muhammed⁷, Taofeek O. Ayinde⁸

¹Department of Anatomy, School of Biomedical Sciences, Kampala International University, Western Campus, Ishaka, Uganda, ²Department of Radiology, Ultrasound and Doppler Unit, Crystal Specialist Hospital, Akowonjo-Dopemu, Lagos, ³Department of Anatomy, Faculty of Basic Medical Sciences, University of Ilorin, ⁴Department of Anatomy, College of Medicine, University of Lagos, ⁵Department of Physiology, University of Lagos, College of Medicine, ⁶Department of Human Anatomy, School of Health and Medical Sciences, State University of Zanzibar, Zanzibar, Tanzania, ⁷Department of Histopathology, Faculty of Medical Laboratory Sciences, Usmanu Danfodiyo University, Sokoto, ⁸Department of Physiology, College of Health Sciences, University of Ilorin

SUMMARY

It is debatable among anatomists whether the Phrygian cap of the gall bladder is an anomaly or anatomic variation with an incidence of 4.0% reported. A 2-fold increase of 7.9% during a 5-year population study was observed in Lagos, Nigeria. A 48-year old male with minimal (non-acute) epigastric discomfort was referred for ultrasound after suspicious cholecystitis. On radiological evaluation, a double “Phrygian cap” was found by coincidence. There is no recent emphasis on double Phrygian cap study, thus it is reported for its rarity and difficulty in arriving at conclusive diagnosis. Common imaging choice is mostly by multi-slice CT for biliary aspects and appendages. Most Phrygian caps are asymptomatic with little or no pathologic consequence. Surgical intervention (cholecystectomy) for a Phrygian cap is only indicated in case of recurrent symptoms. To our knowledge, sparse anatomic and ultrasound literature exists on the Phrygian cap, thus the need for this rare case report.

Key words: Phrygian cap – Ultrasound – Anomaly – Gall bladder – Cholecystectomy

Corresponding author: Adam M. Afodun. Department of Anatomy, School of Biomedical Sciences, Kampala International University, Western Campus, Ishaka, Uganda. Phone: +256754792056, +2348077295769. E-mail: afodunadam@yahoo.com

INTRODUCTION

The average pear-shaped gallbladder is a digestive organ about 7 cm in length and 3.2 cm in width, located underneath the liver (Hardy, 1993) at the right hypochondriac region. Cholecystectomy was introduced in the late 1800s (Severn, 1971) with many gallbladder variations. Kannan et al., (2014) stated; congenital abnormality of the gallbladder combined with Phrygian cap as 4%. Others (Lamah et al., 2001; De-Csepe et al., 2003) declared gall fundal folding during embryology as responsible for the genesis (Edell, 1978). Literature about Phrygian cap is outdated however, in 1935, Boyden described the fold because of its similarity to a 12th-century bonnet-cap worn by common folks (Brown, 1993) (Fig. 1). It is an established tradition that certain medical terms of historical importance or antique commemoration to be enshrined into relevance through eponyms. Boyden (1935) further made descriptions and classification about the gall bladder fold.

MATERIALS AND METHODS

Longitudinal Gall Bladder Procedural Study

Ultrasonography was used to perform this longitudinal study. We also report a 7.9% incidence of prominent Phrygian fold among male patients in Nigeria during a 5-year observation period. Our

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Fig 1. (Classical eponym) A marble bust of the Phrygian cap; named after conical headgear worn by Greek slaves as a symbol of liberation against tyranny and oppression. The cap is historically linked to ancient dwellers of Phrygia, close to present day Ankara (Turkey) during the antiquity around 1250 – 700 BC. Commemorative description of the bonnet cap was affirmed by the astute Anatomist Boyden in 1935.



Fig 2. Arrowed; observe the contrasting hepatic portal vein mimicking a gallbladder by the left. Gallbladder fundus characterized by “kinking” like a cap; it may be wrongly interpreted as an apparent septum within an otherwise normal gallbladder. This “deformity” may be suspected when its land-marks features are demonstrated on high-resolution sonography.

subjects were voluntary participants, no inducement or coercive influence was exercised by the sonologist, and informed consent was obtained in line with the 1975 Helsinki Declaration on Patients’ Right. To better enhance visibility and prevent artefacts during scheduled elective scans, medical fasting (minimum of 8hour food abstinence) was observed by patients for gall bladder turgidity.

Patient Series and Ultrasound Methods

General indications for gall sonography in patients include; recurrent symptoms of pelvic ulcer,

presence of right upper abdominal mass, subject history of jaundice, fever of unknown origin, and pain in the superior abdominal region. Prior to any coincidental discovery of anatomic variations, suspicion is high for cholecystitis and gallstones. Surprisingly, gallbladder wall thickness did not correlate with Phrygian deformity.

When fluid was required by patient before scan, only water was given. Examination commenced with lying in the supine position. It may be necessary to turn patients unto the left side (decubitus) or examine them in erect position (when there was excessive gas, as sitting will not displace bowel gas). Sonar coupling agent (water soluble) was applied liberally to the right upper abdomen and patients were told to hold their breaths (for few seconds) with the abdomen “pushed-out” in full expiration; a precaution to minimize artefacts.

Scanning technique commenced by placing transducer centrally and proximal to the xiphoid angle before moving slowly towards the right. Anatomically, the gall bladder wall consists of a mucosal layer (hyperechoic), a smooth muscle layer (hypoechoic), a perimuscular connective tissue layer (hyperechoic) and a serosal layer (hypoechoic). To ensure all gall measurements were accurately obtained by ultrasound, it was taken from the long axis (anterior) of the sub hepatic plane.

CASE REPORT

A 48-year-old male with minimal epigastric pain reported for ultrasound at Crystal Specialist Hospital, Akowonjo-Dopemu, Lagos. The ultrasound was performed by a convex 3.5 MHz probe, using a Siemens Sonoline ultrasound machine (made in Germany), with knobology set in proximal “near-field-mode” (Figs. 2 and 3). No Murphy’s sign in epigastric region was felt by patient on probe/transducer contact and there were no other symptoms. Bilateral “folds” of Phrygian cap (Fig. 4) was observed on both the anterior and posterior wall. He had an average Body Mass Index (BMI); and no complaint of anorexia. Hospital medical records was free of gastrointestinal problems. Routine Liver Function Test (LFT) and haematology was within normal limits. The gallbladder measured 73 mm in length; with the sonogram printed on a longitudinal plane. No lodging of foci/stone was found in-situ (gallbladder) of the middle-aged man and the rest of the abdomen was normal. Sonographically, the gallbladder was found to be bilobed with a rare “double Phrygian cap” (Fig. 4, below) in both sides (anterior-posterior walls).

Summary Findings

Out of a sample of 334 examined gall bladders in patients, one noted case of double Phrygian cap 1 (0.29%); normal location in the right hypochondria (plane of inter-lobar fissure) was 310 (92.81%), 2 cases of gall location in the left lobe of liver, and no report of multiple septae or complete (extra-hepatic) gall ectopia. Phrygian cap kinking

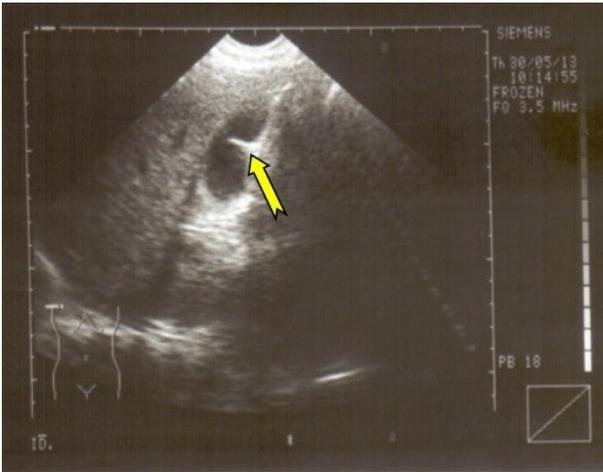


Fig 3. Rarely the fundus of the gallbladder is folded back (Phrygian cap). Near-gain field (knobology) demonstrates the dorsal region. The biliary texture is homogeneous with high liver fat content (normal fatty liver); contrasting to the sonopenic bladder. Note the posterior sonic acoustic shadow and attenuation at the bottom left area of the sonogram.

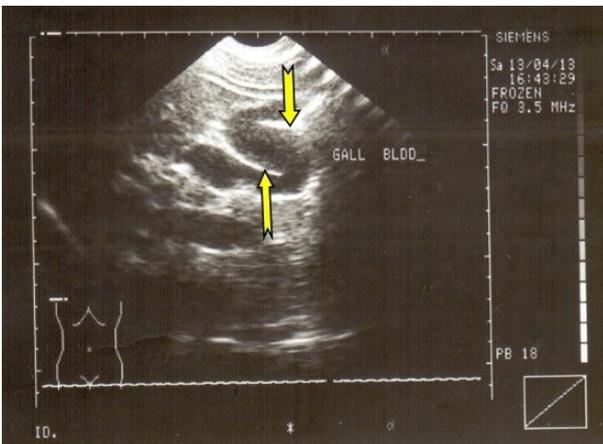


Fig 4. Double Phrygian cap in bilateral anterior and posterior wall (alternate); arrowed in the ventro-dorsal surfaces. This unique variant makes the bladder volume appear larger than conventional anatomical types (Figs. 2 and 3).

“deformity” accounts for 26.38 (7.9%) with 15 cases classified as prominently “long” Phrygian gall fold, while 3 patients (0.89%) had a “U-shaped” curved gallbladder. Gallbladder kinking (Fig. 3) is not suspected in the case report below (Fig. 4), because our patient was fasting (> 8 hours) before ultrasound examination as a standard procedure, to create a non-flaccid bladder. An error or “pitfall” that must be avoided is mistaken the gall bladder with a sectioned portal vein (Fig. 2).

DISCUSSION

Given the wide availability and frequency ultrasonography of the biliary tree, radiologists must be aware of its malformations. However, cases of double gallbladder and cystic-duct duplication (with gall congenital anomaly) exists (Boyden, 1935), as literature search has not shown any re-

port of a rare combination of prominent (bilateral) double Phrygian cap in a human subject.

Concerning the bending, shape and folding of the gall bladder on its body, this may easily be identified on ultrasound. When the fundus of the gall bladder is flexed on its body, a “Phrygian cap” variation is formed. Thus, the radiologist must always inform the clinician about the existence of an aberrant Phrygian cap if any exists. The crux of this case report is that double Phrygian cap has to be differentiated from “hour-glass gallbladder” (Harlaftis et al., 1977), bi-lobed gallbladder (Hobby, 1970), “septated gall variants” (Mohan, 1965; Stringer, 2016), other similar diagnosis (Van Kamp et al., 2013) and congenital anomalies (Gross, 1936). Since differential diagnosis of gall duplication may involve Phrygian cap, bladder fold, peri-cholecystic fluid, choledochal cysts, our argument is that, irrespective of (rarity) pre-operative diagnosis in symptomatic cases, it can easily be missed or become difficult to identify in agreement with Goiney et al. (1985) and Lamah et al. (2001). Turgidity of the gallbladder after 3 h of filling will likely exclude a mass lesion (Dalal et al., 2013).

Careful examination of the gall organ across sonograms (Figs. 2, 3 and 4) excludes multi-septate gall bladder which is extremely rare. Characterized by septae mimicking Phrygian cap, it is found mostly in fundus, neck or the body and formed by incomplete cavitation gall bladder bud. Since the above observation is speculator, no concrete evidence about Phrygian cap causing pathological obstruction or blockage exists. The finding of this case report is contrary to the postulation of Edel (1978), who declared gallbladder folding as a common biliary anomaly. However, we found it to be of no serious clinical significance (Fig. 3). Apart from ultrasound, oral-cholecystography can also be used to identify a Phrygian cap. True gall bladder duplication is a rare congenital anomaly, more scarce is a double Phrygian presence (Fig. 4), in the interim; it appears prone to misdiagnosis and surgical problems (see Fig. 2 of look-alike portal vein mimicking gall Phrygian cap).

The “double-cap” folds (Fig. 4) may temporarily slow down bile movement and form a pseudo-partition in the Hartmann’s pouch. From the physiological and embryonic point of view, it may be considered as a pseudo-developed form of congenital septum; for the Phrygian to be symptomatic, connecting duct entrance would be narrow to obstruct bile flow. In agreement to a theory by Smergel and Maurer (1984) and Meilstrup et al. (1991), that Phrygian cap could impede cholescintigraphic procedure during filling or hepatobiliary imaging. If it is eventually classified as an anatomic variation (and not a “harmless” anomaly), incidence rate in South West Nigeria would be around 9.2%, contrary to the widely reported 4%. Based on the frequency, the folding of the fundal gallbladder like a cap is said (to be the commonest congenital gallbladder abnormality (Lamah et al., 2001; Csepel et al., 2003).

Surgical intervention when there is a potential for

biliary stasis caused by a double Phrygian cap as in this case-report (Fig. 4), a prophylactic cholecystectomy maybe warranted. As the debate in the anatomical “school-of-thought” whether it is a variation/abnormality continues, surgery is however indicated when a major deformity is present even without cholelithiasis (Gigot et al., 1997). Treatment based on patient complaint (symptomatic or asymptomatic) will also depend on the discretion of a gastroenterologist.

The folding gallbladder (Fig. 3) is not properly isolated in ultrasound at the right plane and angle can simulate a mass (Smergel and Maurer, 1984) or gallbladder duplication. As indicated in Fig. 4, on radiological examination it was diagnosed as a case of Phrygian cap duplication (note the opposing arrow-legends) of the gallbladder. Investigating embryological genesis of this condition (double Phrygian cap) at 4 weeks, the biliary tree, gall bladder and liver arise as a superior bud from the caudal part of the foregut (Rappaport and Wanless, 1993). Therefore, its (Phrygian cap) formation in retrospect and characterization by a “fold” intermediate between gall body and fundus can simulate a mass during biliary imaging as earlier stated. The “Phrygian cap” is considered empty at normal rate and plane.

Conclusion

Partial cholecystectomy is not indicated, since there is no severe symptom of pain. It is however of optimal clinical importance to use proper imaging to properly identify a Phrygian cap; which could easily pass for a calcified straw. If found sonographically, it could be conclusively declared as a benign mass and should not be mistaken for a pathology. This documentation is highly important for surgical purposes where numerous variations are seen, thus aiding differential diagnosis and preventing misdiagnosis.

Multi-slice Computer Axial Tomography (CAT) scan could be obtained to exclude a lurking mass (in both folds of cap) on classification; in-situ gallbladder if necessary. This variation or “deformity” with its distinct characteristic landmark is suggested for high resolution Magnetic Resonance Imaging (MRI) for further elucidation.

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