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The ectopic gall bladder: A review

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Abstract---Various congenital anomalies of the gall bladder related to its location, number, shape and size have been described in the literature but the most common among them is the variation in its position. The other anomalies are not common. An ectopic gallbladder is a rare congenital anomaly of its position, which is usually missed on a preoperative workup and detected mostly during surgery. The entity is important because it causes difficulty during the procedure, and a knowledge can prevent serious post-operative complications including bile duct injury. Safe operating techniques including careful dissection of the gall bladder from the cystic plate, and division and lifting of the falciform ligament, reduces the chances of post-operative complications.

Keywords---congenital anomalies of the gall bladder, true left sided gallbladder, safe operating tips, falciform lift and division, bile duct injury.

Introduction

Congenital malformations of the gall bladder can be categorized by their location, size, number, and shape with the most common congenital anomaly of the gallbladder being a variation in its location. Other anomalies like agenesis and

duplication of the gallbladder are less common. Agenesis is very rare and is usually associated with other congenital syndromes like trisomy 18 and the Klippel-Feil syndrome.(1) A multiseptated gallbladder, an anomaly of shape, is also extremely rare. It may exist as an isolated anomaly or coexist with other biliary system anomalies such as hypoplasia or a choledochal cyst.(2)(3)(4). The various ectopic locations of the gall bladder are:

- Inside the liver
- Within the lesser omentum on the left side
- In the falciform ligament, above and below the liver
- Behind the pancreas and duodenum
- In the abdominal wall.(5)

The anomalous position of the gall bladder was published for the first time by Hochstetter in 1986 and about 150 cases have been reported in the literature. The incidence of the ectopic positions of the gall bladder is <1% in the general population.(6)(7)(8) It is more common in men with a reported incidence of 2:1. Most patients are in the 5th-6th decades, and the majority of case series have been reported from Japan and Australia.(9). In this review we discuss this rare anomaly and review the available literature on the subject. We further elaborate on the relevant embryology, other associated anomalies, diagnostic challenges and important tips for the safe surgery.

Embryological basis

The hepatic diverticulum originates from the foregut endoderm during the 4th week of gestation and develops into hepatobiliary system including the liver, gall bladder and the bile ducts. It has two parts - a cranial portion is known as the pars hepatica and a caudal end, the pars cystica. The pars hepatica develops into the liver and bile ducts, while the pars cystica forms the gall bladder and cystic duct. The position of the gall bladder depends upon the position of the liver, but it may sometimes be ectopically placed while the liver remains in its normal position. The gall bladder lies under the right lobe of liver, but an ectopic location is mostly detected under the left lobe, and it is hence called a left sided gall bladder. (10). There are various explanations regarding the development of a left sided gall bladder.

- The gall bladder develops in its normal position from the hepatic diverticulum, but it moves and becomes attached under the left lobe of liver. Since all the portal structures develop normally, the cystic duct becomes longer, courses in front of the bile duct from medial to the lateral side and enters the duct from the right.
- The gall bladder fails to develop normally but develops from the left duct directly. In this type, the cystic duct enters the bile duct on its left side.
- The gall bladder's location is normal, but it appears abnormally placed on the left side, because of an anomaly of the falciform ligament. There are two falciform ligaments in early life during the 6th week a right and a left. The right ligament atrophies and the left persists as the adult falciform ligament. In some cases, the left ligament atrophies and the right becomes dominant, so the normally placed gall bladder appears left sided. (11)

Classification and associated anomalies

Normally the gall bladder is located in its eponymous fossa, between segment IV and V under the right lobe of the liver, on the von Rex-Cantlie line, which is a surgical plane containing the middle hepatic vein. This plane divides the liver into the right and the left lobe.(12) A gall bladder found in any other position is called an ectopic or aberrant gall bladder. The old classification for the position of the gall bladder described four abnormal positions; left sided, transverse, retro displaced and intrahepatic, but the new classification divides it into two categories i.e. with or without situs inversus. In a patient without situs inversus an ectopic gall bladder may be of two types

- A true left sided gall bladder
- It appears left sided because of an abnormally developed falciform ligament(9)(13)

A true left sided gall bladder is also known as a sinistroposition and is the more common anomaly.(12) It occurs when the gall bladder is placed under the segment III of the left lobe of liver, and attached to the left of the ligamentum teres and falciform ligament in a patient without situs inversus.(14)(15) Figure 1



Figure 1. Sinistroposition of gall bladder visible on laparoscopy

A recent study regarding left sided gall bladders without situs inversus reported incidence rates of 83% and 16% between true left sided gall bladders and those due to an abnormally placed falciform ligament.(9). Five variations of the cystic and common duct union have been described in the literature. The commonest is the cystic duct joining on the right side after taking a hairpin bend in front of the common duct. The other variants are a union of the cystic duct on the left side, in the left hepatic duct, right hepatic duct or in a branch of the right duct.(15) The cystic duct takes a hairpin bend and enters the bile duct from the right side in the majority of cases (77.3%) while in the rest, it enters from the left side.(9) This anomaly of position may be associated with other congenital anomalies of the biliary system and portal vein. The most common of which is an atrophied caudate lobe of the liver.(9)(16)(17)(18)(19)

Radiological Imaging

An ultrasound (USG) examination, which is the only investigation done in the majority of uncomplicated cases of gall stone disease, is usually unable to detect the anomaly, and advanced investigations like a Computed Tomographic (CT) scan and Magnetic Resonance Cholangio Pancreatography (MRCP) are seldomly performed. USG is only able to detect abnormalities of position in about 16.3% cases before operation.(9) and the majority (over 80%) of cases are discovered during surgery. A systematic review had reported that the CT abdomen had the highest positive predictive value (60%) to detect the anomaly, while USG had the lowest (2.7%).(15) MRCP may be more informative because it provides a clearer visualization of the relationship between the cystic duct, ectopic gall bladder, and common hepatic duct. The use of a Tc-99m labeled HIDA scan is also recommended whenever there is a doubt of the presence of an ectopic gall bladder on ultrasound examination. The role of fluorescent cholangiography with Indocyanine green has also been advocated to delineate biliary anatomy and possible anatomical variations.(20) Studies have reported that intra-operative cholangiograms have been used in up to 39.1% of cases when there is a doubt of aberrant anatomy.(9) However the role of intra-operative cholangiography is debatable and it should be selectively used e.g. to confirm about the correct plane of dissection, and to document the other associated biliary abnormalities.(15)

Tips for safe surgery

A sound knowledge about the abnormalities of position, associated biliary and vascular anomalies and safe operating techniques improves the rate of safe and successful surgery in this situation. It poses more challenges during surgery if a liver resection or transplant is planned.(21)(22). Laparoscopic cholecystectomy can be safely performed in about 79.6% cases but the rate of complications including biliary injuries is higher with the incidence of bile duct injury being reported to be up to 7.3% in these cases.(9) This high rate is due to poor dissection techniques, an obliquely placed gall bladder, a hairpin bend of the cystic duct and a difficult dissection of the cystic plate.(15) Figure 2

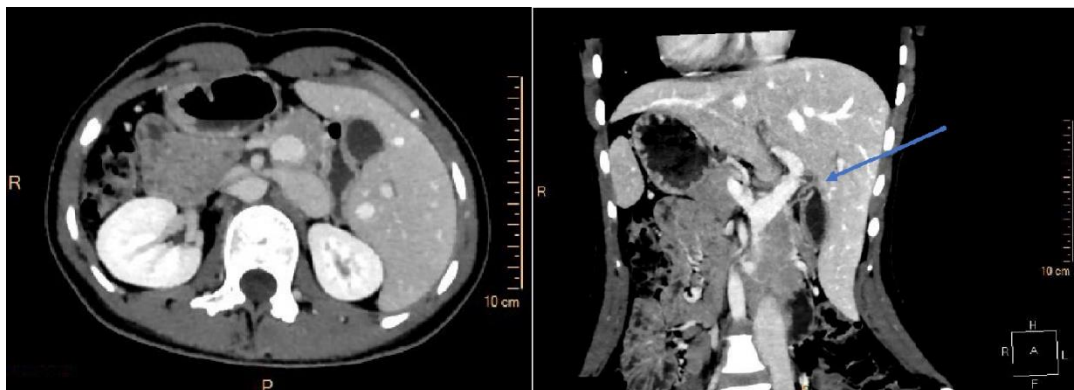


Figure 2. (a). CT showing ectopic gall bladder with situs inversus (b); Oblique position of gall bladder with hair pin bend of cystic duct (*arrow*)

Laparoscopic dissection becomes more difficult in cases with acute inflammation associated with an abnormality in position.(23) Two operative technique variations have been reported in the literature for safe laparoscopic cholecystectomy - a change in the port positions and a division/lift of the falciform ligament. A change in the port positions with creation of an epigastric port in the left hypochondrium and more medial placements of the retracting ports, have been suggested. Further adopting a French position (i.e. the surgeon standing between the legs) along with medial shifting of the grasping and dissecting ports have also been described.(24) A falciform lift(25) or division before starting the cholecystectomy has been reported which gives more room for the laparoscopic dissection.(11)(18)

The most important aspect is a safe dissection technique, one of which is described by Connor et al. The gall bladder is first dissected from the cystic plate and a critical view of safety is achieved.(26)(27)(28) Both the techniques of antegrade and retrograde dissection have been reported but the majority of studies have favoured antegrade techniques with only a few reporting fundus first techniques (16.7%).(15)(29)(30)(31). Studies have reported that antegrade dissection, intra-operative cholangiography, careful dissection of the Calot's triangle, inspection of any tubular structure before division, limited use of energy sources, and division/lift of the falciform ligament may reduce the chances of bile duct injuries in cases with abnormal position of the gall bladder.(32)(31)(33)

Conclusion

A true left sided gall bladder is usually missed on the pre-operative ultrasound examination which is the only imaging investigation performed in the majority of patients with uncomplicated gall stone disease. Patients with suspicion of this anomaly should be investigated further with advanced imaging techniques like MRCP and a CT scan. Safe operating techniques including a careful dissection of Calot's triangle and clear inspection of any tubular structure before clipping, may reduce the chances of bile duct injury. Prior division of falciform ligament before dissection of calot's triangle is also helpful in creating more room for dissection.

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