Gallbladder duplication: evaluation, treatment, and classification☆,☆☆

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Abstract Duplicate gallbladder is a rare congenital anomaly resulting from abnormalities in embryogenesis during the fifth and sixth weeks of gestation. Approximately 210 cases have been described. Variations include duplicate, triplicate, and septated gallbladder. We encountered a 15-year-old girl with both a duplicated gallbladder and a duplicated cystic duct who underwent successful laparoscopic cholecystectomy. This combination is extremely unusual, and based upon our findings in this case and a review of the literature, we propose the Unified Classification of Multiple Gallbladders.

Duplicate gallbladder is a rare congenital anomaly of the hepatobiliary system with an incidence of 1 in 3800 [1]. Duplicate gallbladder and its variable anatomy was first described by Boyden in 1926, and there are approximately 210 published cases to date. In 1936, Gross [2] described congenital abnormalities of the gallbladder and classified them into classes A-F. The duplication typically occurs because of outpouchings from the normal extrahepatic biliary system during the fifth and sixth weeks of gestation. These outpouchings typically regress; however, their persistence results in formation of an accessory gallbladder [3]. Harlaftis et al [3] classified duplicate gallbladder anatomy into type 1 (split primordial gallbladders) and type 2 (accessory gallbladders) in 1977. Recently published data have described a modified Harlaftis classification that added a left trabecular variant to the type 2 classification [4]. How multiple gallbladders develop is not fully understood, and the full extent of the potential aberrant anatomy may be unknown.

Many of these patients will present with atypical symptoms for traditional biliary disease, and it is very important to recognize duplication of the gallbladder as a possible confounding issue. Overall, patients with aberrant anatomy are more likely to undergo open surgery vs laparoscopic surgery. Laparoscopic resections are reasonable and well described. Successful laparoscopic resection requires the surgeon to consider aberrant anatomy as a possibility when atypical situations are encountered. For this reason, identification of a dual duct as an anatomic variant is particularly important.

1. Case report

A 15-year-old girl was referred to the pediatric surgery clinic after 10 years of right upper quadrant abdominal pain. Laboratory evaluation was notable for normal total bilirubin
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and liver function. Imaging demonstrated cholelithiasis with no evidence of any other complicating factors. She was taken to the operating room where she underwent a difficult laparoscopic cholecystectomy because of extremely dense adhesions around the cystic duct and body. During this dissection, the cystic duct stump was observed and noted to have a dual lumen. The gallbladder was examined on the back table and found to have duplication of the gallbladder and 2 cystic ducts in continuity with the 2 gallbladders (Fig. 1). No further exploration was performed. Retrospective review of her preoperative right upper quadrant ultrasound demonstrated that there was in fact a septum dividing 2 gallbladders (Fig. 2). The patient did well, recovered uneventfully, and a postoperative MRCP demonstrated an otherwise normal hepatobiliary anatomy.

Microscopically, there was evidence of chronic cholecystitis and cholelithiasis. The duplicated gallbladders and cystic ducts had separate mucosal and smooth muscle layers. The separate smooth muscle layers were verified by a smooth muscle actin immunohistochemical stain. Both cystic ducts and both gallbladders were fused along their entire length by perimuscular connective tissue (subserosa). However, the most significant finding was a shared serosa (Fig. 3).

2. Discussion

Gallbladder duplication was described in ancient Roman text. It is seen in 0.026% of autopsy patients [1]. In multiple gallbladder anatomy, each gallbladder must have valves at the neck, a tunica muscularis, and the ability to concentrate bile. The Harlaftis classification is the most commonly used classification and is divided into 2 main groups based upon embryogenesis (Fig. 4). Type 1, or split primordial group, is subdivided into septated, V shaped, or Y shaped. Therefore,
when the cystic primordium splits during embryogenesis, both gallbladders share a common cystic duct. Type 1 septate duplicate gallbladder occurs when there is a single cystic duct and a septum that divides the 2 gallbladders. Type 2 describes accessory gallbladders that are ductular or trabecular, meaning that they arise from separate primordium from the biliary tree and have individual cystic ducts [3].

Preoperative imaging is very important in diagnosing duplicate gallbladder; however, this is limited by the type of aberrant anatomy in that type 1 gallbladders may be detected as a single gallbladder with folds. Successful preoperative diagnosis is noted in only one half of all cases [4]. MRCP has better diagnostic capability than ultrasound, and endoscopic retrograde cholangiopancreatography is considered the gold standard for diagnosis [4]. However, in our case report the patient had a duplicate septated type 1 gallbladder with 2 cystic ducts. There are 2 likely etiologies to account for this variation, the first is that this represents a new subtype of type 2 gallbladders that arise from separate primordium or that this is a variant of a type 1 septated gallbladder in which the septation extended all the way down to the level of the common bile duct. Postoperative MRCP was unable to clarify the anatomy any further (Fig. 2).

Triple gallbladders have been previously described in three anatomic variations [5]. These three types vary depending upon the embryologic development and occur in the same manner as duplicated gallbladders. Given this information, we propose a Unified Classification (Fig. 4). This includes triple

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**Fig. 3** Left, Histologic section at the level of the duplicated cystic ducts. Note the separate mucosal and smooth muscle layers (arrows) fused by perimuscular connective tissue (arrowhead) (hematoxylin-eosin ×2). Right, Histologic section at the level of the body. Note the separate mucosal and smooth muscle layers (arrows) fused by perimuscular connective tissue with no intervening serosa (arrowhead) (hematoxylin-eosin ×2).

**Fig. 4** Unified Classification of Multiple Gallbladders. Solid line is original Harlaftis Classification. Dashed line is Modified Harlaftis Classification.
gallbladders because these are an extension of duplicate biliary anatomy. The triple primordial group develops similar to the split primordial duplicate gallbladders and occurs when a triple split in the cystic primordium occurs during embryogenesis. The triple ductular group arises from 3 separate primordia similar to the ductular duplicate gallbladders. The new classification follows the Harlaftis classification based on embryology and adds a third group that occurs when there is a combination of types 1 and 2 anatomy. The triple combined group occurs from a split in one primordium and a second developmental primordium, and we classify this as type 3, the combined group. We classify our case as type 3 combined and named it septate B to remain consistent with the original classification. It is unknown if during embryogenesis there was a split in the cystic primordium very early in development or if 2 primordial arose from the same location. Histologically, the gallbladder did not fit into any of the currently described classification systems.

In general, the literature supports laparoscopic intervention for type 1 duplicate gallbladder and advocates open surgery for type 2 duplicate gallbladder. This is because of the increased theoretical risk of injury to the common bile and the right hepatic artery secondary to the high insertion of the second cystic duct and a more extensive dissection that is required [4]. In the case of a septated gallbladder with duplicated cystic ducts (type 3), laparoscopic intervention is feasible using traditional methods, although it will likely be very difficult. There is no substitute for meticulous and safe dissection beginning high on the gallbladder with known anatomy and identifying landmarks. In addition, cholangiography through each individual duct can clarify the anatomy. Inspection of the gallbladder specimen can also clarify the anatomy a great deal.

In conclusion, the Unified Classification of Multiple Gallbladders is useful in assessing each patient individually and in planning operative intervention. It is possible more variations on this classification system have yet to be reported. The Unified Classification should include the majority of duplicated gallbladders and a framework on which to classify further duplicate anatomy if it is discovered. It will also hopefully clarify the possibilities of aberrant anatomy for surgeons faced with highly atypical operative scenarios.

References