Agenesis of gallbladder – our experience and a review of literature

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Abstract

Background Agenesis of gallbladder is a rare congenital anomaly. Variable diagnostic approaches, perioperative management strategies and postoperative follow up protocols provide a dilemma to its correct diagnosis and management.

Methods We hereby present five patients with agenesis of gallbladder encountered in our institution between 1992 to 2008. Four out of five patients had symptoms suggestive of gallstones including upper abdominal pain and acid dyspepsia, which were further substantiated by ultrasound reports showing shrunken contracted gallbladder with calculi.

Results On laparoscopy the gallbladder was absent in all five patients leading to conversion to laparotomy in the initial three patients. With increasing experience, the next two patients were diagnosed conclusively by laparoscopy to have agenesis of gallbladder, without the need for conversion which was further confirmed by post operative imaging studies.

Conclusion Gallbladder agenesis is rarely encountered in clinical practice. With increased experience in laparoscopy, the condition no longer mandates conversion to laparotomy for confirmation of diagnosis. Symptomatic improvement occurs in all patients following surgical intervention.

Keywords Agenesis of gallbladder · Laparoscopic cholecystectomy

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Introduction

Agenesis of the gallbladder is a rare condition resulting from failure of development or canalisation of the cystic bud in the fourth week of intrauterine life. Most cases reported in literature are isolated case reports and therefore not much is known about this rare anomaly. Almost half of all patients with agenesis of gallbladder have symptoms suggestive of cholelithiasis. Preoperative diagnosis is difficult and mostly confirmed intraoperatively by complete dissection of the common bile duct (CBD) and exploration of other possible ectopic sites of gallbladder. Most of them have been in the past diagnosed after open exploration to conclusively prove agenesis. However with the advent of laparoscopy, it has been possible to diagnose such cases definitively without the associated morbidity of open surgery. We have retrospectively reviewed five patients in our institution who were diagnosed as cholelithiasis on abdominal ultrasound but were found intraoperatively to have no gallbladder.

Materials and methods

In our experience of over 28,000 laparoscopic cholecystectomies over a period of 16 years we have encountered five patients with agenesis of gallbladder (0.018%). There were three males and two females, mean age was 33 years (range 17–45 years). The patient demographics, clinical and radiological findings and postoperative follow up are as indicated (Table 1). After a diagnostic laparoscopy, a formal laparotomy was performed on the first three patients for confirming the diagnosis. In the next two patients however, agenesis of gallbladder was confirmed on laparoscopic exploration. This was later corroborated by postoperative radiological studies.

All our patients of agenesis of gallbladder had a preoperative ultrasound of the abdomen that reported

Table 1 Detailed clinical and radiological follow up of patients with agenesis of gallbladder

Patient	Age/sex	Symptoms	Pre-op USG	Intra-op findings	Intra-op course	Post-op investigation	Follow up
Patient No. 1	26/F	Pain RHC, nausea	Contracted gallbladder with calculi	No gallbladder, normal CBD	Converted to open	CT scan	Asymptomatic
Patient No. 2	45/M	Asymptomatic	Contracted gallbladder with posterior acoustic shadowing	No gallbladder, dilated CBD	Converted to open	CT scan	Lost to follow up after 12 months
Patient No. 3	45/F	Acid dyspepsia	Shrunken gallbladder with single calculus	No gallbladder, dilated CBD	Converted to open	CT scan	Asymptomatic
Patient No. 4	31/M	Pain RHC, nausea	Contracted gallbladder with an echogenic foci with posterior acoustic shadowing	No gallbladder, normal CBD	Thorough laparoscopic exploration	CT scan	Asymptomatic
Patient No. 5	17/M	Pain RHC, nausea	Contracted gallbladder containing small calculi	No gallbladder, dilated CBD	Thorough laparoscopic exploration followed by intra-op USG	MRCP + MRI	Asymptomatic

gallbladder calculi. Preoperative biliary imaging studies like magnetic resonance cholangiopancreatography (MRCP) or endoscopic retrograde cholangiopancreatography (ERCP) were not performed in any patient as none of them had any previous history suggestive of biliary obstruction. In these five patients postoperative biliary imaging by MRCP or CT scan was done to confirm agenesis of gallbladder.

The latest patient with agenesis of gallbladder encountered in our institution was that of a 17-year-old male patient who presented in the outpatient clinic with a history of recurrent dull aching pain in right upper quadrant region associated with nausea for the last 8 months. His physical examination was unremarkable. Biochemical and haematological examinations were within normal limits. Preoperative ultrasound (USG) reported a contracted gallbladder containing small calculi. The CBD was not dilated. Liver showed multiple scattered calcific foci (Fig. 1). A laparoscopic cholecystectomy (LC) was planned for the patient.

Laparoscopy was performed under general anaesthesia. Standard four ports for LC were made after achieving pneumoperitoneum of 12 mm of Hg. On elevation of the right lobe of the liver, the gallbladder was not visualised in its normal position. The CBD was identified and seen traversing from the porta hepatis to behind the duodenum. There were no adhesions in the region of the porta. A thorough search was made to rule out an ectopic gallbladder including the falciform ligament, the posterior inferior surface of the liver, the left lobe of the liver, the lesser sac, retroperitoneal, retrohepatic and retroduodenal spaces but was unsuccessful (Fig. 2).

A meticulous intraoperative ultrasound failed to locate the gallbladder and only revealed dense fibrous tissues

in the region of the Calot's triangle. The CBD although dilated, was clear up to the distal end. Intraoperative cholangiography is not standard practice in our institution. A diagnosis of agenesis of gallbladder was made and the procedure terminated.

An MRCP was done postoperatively for demonstrating the extrahepatic biliary apparatus (EHBA). It failed to reveal the gallbladder or any cystic duct (Fig. 3). The right posterior segment duct (RPSD) was found to be draining into the left main biliary channel. The CBD and the pancreatic duct were normal (Fig. 4). The patient was discharged the next day. Four months postoperatively, the patient continues to be asymptomatic.

Fig. 1 Ultrasound picture of patient five showing contracted gallbladder with posterior shadowing

Fig. 2 Intraoperative picture of the biliary anatomy in the same patient showing no gallbladder

Fig. 3 Transverse cut of MRCP through the porta hepatis showing the CBD with no gallbladder

In a similar case a 31-year-old male patient presented with pain in the right hypochondrium for 2 weeks. There was no fever or jaundice. He was a known case of atrial septal defect (ASD) and was awaiting treatment for it after cholecystectomy. USG reported a contracted gallbladder with an echogenic foci with posterior acoustic shadowing. A thorough diagnostic laparoscopy was similarly performed but no gallbladder was found even in the aberrant locations described. Agenesis of gallbladder was documented postoperatively by a CT scan of the whole abdomen with multiple slices taken at the porta hepatis [1].

Another 45-year-old male patient who was otherwise asymptomatic, was detected to have a contracted gallbladder with posterior acoustic shadowing on routine

Fig. 4 Longitudinal section of the biliary system in the same patient

health check-up. In this patient and in two other initial cases, conversion to laparotomy was performed to be able to explore completely the possible ectopic sites of gallbladder. As our experience with laparoscopy grew, we were able to convincingly complete the exploration laparoscopically in the next two patients.

Results

In our institution four out of five patients that we have encountered were symptomatic for cholelithiasis. We have found an almost equal sex incidence in our patients (M:F=3:2).

Preoperative ultrasound suggested cholelithiasis with contracted gallbladder in all our patients. None of our patients had abnormal liver function tests (LFT) or a history suggestive of CBD calculi. Preoperative ductal imaging with a MRCP or ERCP was therefore not performed in any of our patients.

Complete dissection of the entire biliary system along with exploration of the possible ectopic sites of gallbladder was in the past mandatory to conclusively diagnose agenesis [2, 3, 4]. In our initial three patients, conversion to laparotomy was needed for complete exploration. In the subsequent two patients we were able to perform a thorough exploration laparoscopically. Postoperative radiological studies were performed to document the rare abnormality and also to rule out CBD calculi. A CT scan was done in the initial four patients and an MRCP scan was done in the last patient, that confirmed agenesis of the gallbladder in all these patients. All the patients on follow up continue to be asymptomatic.

Discussion

Agenesis of the gallbladder is a rare congenital anomaly. Exact incidence is difficult to determine as most patients reported in literature are isolated case reports. Gallbladder agenesis without EHBA occurs in 0.01–0.02% [2] and are associated with other congenital anomalies including agenesis of lung and Tetralogy of Fallot.

Fewer than 400 cases [1] have been reported in literature since the first case was reported by Lemery in 1701. Bennion et al after a review of world literature described three separate groups of patients with agenesis of gallbladder. The symptomatic group constitute the majority (50%) and have symptoms suggestive of cholelithiasis. Of all the symptomatic patients most patients present with right hypochondriac pain (90%), dyspepsia (30%), nausea and vomiting (66%), intolerance to fatty food and jaundice. It is difficult to determine what causes the symptoms in these patients. A possible explanation is the associated biliary dyskinesia and the resultant rise in the sphincter of oddi pressure. These patients may have a dilated hepatic duct that takes up the function of bile storage. Cholestasis resulting from biliary dyskinesia and resultant infection might result in conditions favourable to CBD stones. Asymptomatic patients constitute 35% and the other 15–16% of the patients who have multiple congenital anomalies are usually incompatible with survival [2, 5].

Although literature suggests majority of patients with agenesis of gallbladder to be symptomatic for gallstones, in our institution four out of five patients that we have encountered were symptomatic. The total number of patients however is too small to be statistically significant. Patients diagnosed through operations present a female predominance of 3:1, although in autopsies the proportions are equal between the sexes [6, 7]. In our patients too we found an almost equal sex incidence.

In a literature review by Richards et al 27% of patients with agenesis were jaundiced and in 18% of them CBD stones were found [8, 9]. All our patients however had normal LFTs and no CBD calculi were detected, confirmed by conversion in the initial three patients and by postoperative radiological studies in the next two patients. In all patients with agenesis of gallbladder preoperative or postoperative investigations are needed to rule out CBD calculi for successful treatment of the same and prevention of complications.

Gallbladder agenesis is usually detected intraoperatively. Preoperative diagnosis is almost impossible. During ultrasound thick peritoneal folds in the region of the Rouviere's sulcus might appear to the radiologist as thickened contracted gallbladder [7, 9], and in some cases might give rise to echogenic shadows suggesting calculi. In our case, liver calcifications were seen on ultrasound in the 17-year-old boy. These liver calcifications are a common incidental finding and are usually of no clinical significance

but the presence of calcific foci in the region of the porta could have led to misinterpretation by the sonologist as gallbladder calculi.

ERCP can be more helpful in such situations but again is not a routine examination in the preoperative evaluation of such patients. Non-visualisation of the gallbladder in ERCP can be attributed to a variety of reasons and gallbladder agenesis would be the least common explanation [8].

Frey suggested complete dissection of the biliary ducts and confirmation of the operative findings by cholangiography before issuing a diagnosis of gallbladder agenesis [3]. Intraoperative cholangiogram however needs a choledochotomy and a possible resultant postoperative bile leak and its morbidity. Many surgeons have questioned the ability of laparoscopy alone to completely visualise the possible ectopic sites in such cases. This might warrant conversion to open procedure. This is where the importance of intraoperative ultrasound comes in. Laparoscopic intraoperative ultrasound (LIOU) using powerful, flexible tip ultrasound probes is an effective way of assessing the CBD and neighbouring structures. In the last two patients that we have encountered in our institution, after meticulous and careful diagnostic laparoscopy we have managed to completely explore and rule out an ectopic gallbladder, which was later documented on postoperative radiological studies.

The possibility of an ectopic gallbladder missed on laparoscopy and discovered on postoperative imaging studies exists. In such an unlikely event, the surgeons still has the option of performing a definitive surgery at a later date.

Although patients with agenesis undergo a negative exploratory laparotomy or diagnostic laparoscopy they become asymptomatic postoperatively for unknown reasons. A probable explanation is the lysis of periportal and right hypochondrial adhesions at the time of surgery. All the cases that we have seen so far in our institution continue to be asymptomatic. Further follow up is needed to know whether such patients have recurrence of symptoms or develop common duct calculi in the long-term.

In most of the cases reported in literature conversion to an open procedure was done to prove the absence of gallbladder [10]. However we believe that laparoscopy can conclusively prove gallbladder agenesis with aid of tools such as intraoperative ultrasound in almost all cases in experienced hands despite the rarity of the condition.

Conclusion

Gallbladder agenesis is a rare clinical entity that is most often diagnosed intraoperatively. Although almost half of the patients present with symptoms of gallstones and are intraoperatively detected to have no gallbladder, most patients become asymptomatic postoperatively. After metic-

ulous and careful diagnostic laparoscopy, most patients with agenesis of gallbladder can be diagnosed without the need for conversion to laparotomy and the associated morbidity.

Conflict of interest The authors do not have any disclosable interest

References

1. Chowbey PK, Singh G, Sharma A, Khullar R, Soni V, Baijal M (2002) Case report *Agenesis* of the *gallbladder*, Indian Association of Gastrointestinal Endosurgeons, IAGES
2. Bennion RS, Thompson JE Jr, Tompkins RK (1988) Agenesis of the gallbladder without extrahepatic biliary atresia. *Arch Surg* 123:1257–1260
3. Frey C, Bizer L, Ernst C (1967) Agenesis of gallbladder. *Am J Surg* 114(6):917–926
4. Azmat N, Francis KR, Mandava N, et al. (1993) Agenesis of the gallbladder revisited laparoscopically. *Am J Gastroenterol* 88:1269–1270
5. Vijay KT, Kochar HH, Koti RS, Bapat RD (1996) Agenesis of gallbladder – a diagnostic dilemma. *J Postgrad Med* 42: 80–82
6. Jaques Waisberg, et al. (2002) Agenesis of the gallbladder and cystic duct. *Sao Paulo Med J* 120(6) São Paulo
7. Fikret A, et al. (2008) Agenesis of gallbladder diagnosed unexpectedly during a laparotomy for cholecystectomy. *Marmara Medical Journal* 21(3): 252–256
8. Trompetas V, Panagopoulos E, Ramatanis G (2004) Gallbladder agenesis presenting with obstructive jaundice and elevated CA19-9. *Acta Chir Belg* 104:347–349
9. Richards RJ, et al. (1993) Agenesis of gallbladder in symptomatic adults. A case and review of the literature. *J Clin Gastroenterol* 16:231–233
10. Fisichella Piero MA, et al. (2002) Isolated agenesis of the gallbladder: Report of a case, ISSN 0941-1291. *Surgery Today* 32:78–80