Agenesis Of The Gall Bladder And Cystic Duct: Laparoscopic Diagnosis

T Grandhi, S El-Rabaa

Citation

T Grandhi, S El-Rabaa. *Agenesis Of The Gall Bladder And Cystic Duct: Laparoscopic Diagnosis*. The Internet Journal of Gastroenterology. 2004 Volume 4 Number 1.

Abstract

Congenital absence of gall bladder is a very rare but well recognised congenital abnormality, the reported incidence ranging between 0.01 and 0.05%. To our knowledge, this is the 1st case reported from UK that was diagnosed by imaging modalities avoiding laparotomy. Laparoscopic cholecystectomy was attempted on a suspected case of chronic cholecystitis and was found to have congenital absence of gall bladder and cystic duct. The diagnosis was confirmed by CT Scan and ERCP post operatively. Standard investigations for chronic cholecystitis are misleading. Agenesis of gall bladder should be highly suspected whenever the gall bladder is not visualised on ultrasonography or at laparoscopy done on misinterpreted ultrasound. If the gall bladder is not seen at laparoscopy, further procedure should be avoided and agenesis should be confirmed by a combination of imaging modalities namely CT Scan, MRCP, laparoscopic or endoscopic ultrasound, if available. If the gall bladder is not visualised at preoperative ultrasound a combination of above imaging modalities should be used for diagnosis without recourse to laparoscopy / laparotomy.

DEPARTMENT WHERE WORK WAS DONE

Department of General Surgery
West Cumberland Hospital
Whitehaven
Cumbria CA28 8JG
United Kingdom

INTRODUCTION

Congenital absence of the gall bladder is very rare but well recognised condition, and the condition so far has only been diagnosed at laparotomy in a patient who has not undergone previous biliary tract surgery. The condition may be asymptomatic, present with symptoms suggestive of biliary tract disease or associated with congenital malformations. We report a case of a lady who was found to have agenesis of gall bladder at attempted laparoscopic cholecystectomy on the basis of misinterpreted ultrasound scan. To our knowledge, this is 1st case reported from UK that was diagnosed without laparotomy.

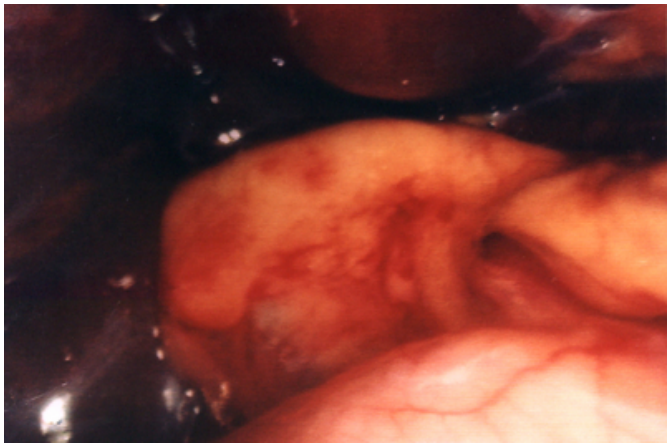
CASE REPORT

A 45-year-old lady was referred to our surgical unit at West Cumberland Hospital with right upper quadrant pain, which radiated to the back and precipitated by fatty meal. General examination and abdominal examination were unremarkable. Routine haematological and biochemical investigations were normal. Ultrasonography (USG) of the abdomen was reported to be showing lot of echoes arising from the region of the gall bladder bed consistent with gall bladder packed full of stones.

Laparoscopic cholecystectomy was attempted based on history and sonographic findings. Initial laparoscopy failed to visualize gall bladder. The biliary tree appeared normal (Fig. 1). Extensive and careful search did not reveal either ectopic gall bladder or any other abnormality in other upper abdominal viscera. No further procedure was performed. The patient improved symptomatically following the laparoscopy.

Figure 1

Figure 1: Laparoscopic photograph showing absence of gall bladder



Computerised Axial Tomography Scan (CTScan) of the abdomen performed post operatively, did not show gall bladder or calculi (Fig. 2). Other upper abdominal viscera were normal as identified on laparoscopy. Endoscopic Retrograde Cholangio Pancreatography (ERCP) confirmed the diagnosis showing a normal papilla and a normal common bile duct (CBD), but did not show either gall bladder or cystic duct (Fig. 3). The patient was symptom free at follow up.

Figure 2

Figure 2: CT Scan showing agenesis of the gall bladder

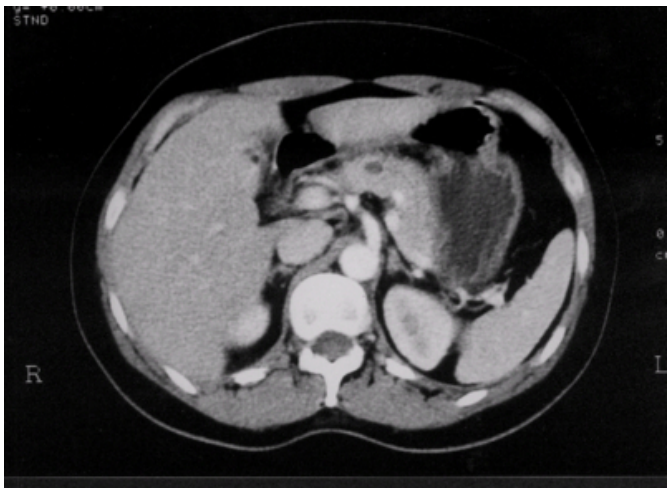
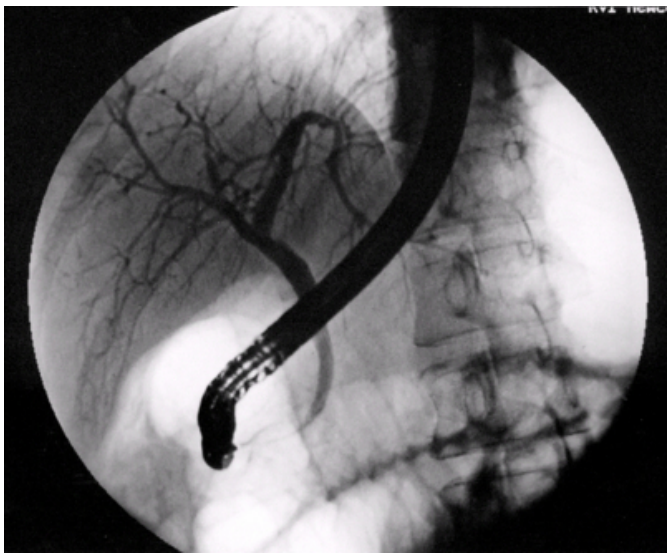


Figure 3

Figure 3: ERCP confirming agenesis of the gall bladder and cystic duct



EMBRYOLOGY

In 4th to 5th week of life, a 4-mm. human embryo develops a bud from the foregut that grows cephalad and the cranial portion becomes the liver and hepatic bile ducts. In the caudal portion of the growing bud, there develops a second bud, or diverticulum, and this is destinated to become the gall bladder and cystic duct [1]. Failure in organogenetic sequence at this point results in agenesis of gall bladder [2]. Theories for this condition include failure of gall bladder analage from the hepatic diverticulum, or failure of recanalisation following the solid phase. Gall bladder agenesis can be part of multiple foetal anomalies and the association has been attributed to deranged development between the paired ompheloenteric and umbilical veins and the sinus venosis cordis; this results in disturbed positional development of the cells of the upper umbilical position [3, 4].

DISCUSSION

Lemary first reported agenesis of gall bladder in literature in 1701 [5, 6]. The reported incidence in literature ranges between 0.01 and 0.05% [5]. The incidence is equal in both sexes from studies at autopsy where as 2-3 times more common in females in clinical cases [4].

Bennion et al described 3 categories of agenesis of gall bladder [7].

1. Multiple foetal anomalies (15-16%) - These patients invariably die in the perinatal period due

to associated anomalies and agenesis of the gall bladder was only recognised at autopsy. The most frequently encountered malformations were cardiovascular, gastro-intestinal/genitourinary, anterior abdominal wall, and central nervous system. In this group gall bladder agenesis is only a trivial anomaly.

2. Asymptomatic group (35%) - Agenesis of gall bladder was discovered either at autopsy, at laparotomy for unrelated diagnosis or by screening the family members of patients known to have agenesis of gall bladder. These patients do not have symptoms of biliary tract.

3. Symptomatic group (50%) - This major group present in 4th or 5th decades. This is usually an isolated anomaly.

In symptomatic group, patients undergo surgery for right hypochondrial symptoms only to find no gall bladder at surgery. Common symptoms include chronic right upper quadrant pain (90%), dyspeptic (30%), nausea and vomiting (66%), fatty food intolerance (37%) and jaundice (35%) [8]. The possible mechanisms of symptoms include primary duct stone, biliary dyskinesia or non-biliary disorder.

The usual preoperative investigations for biliary tract namely USG and oral cholecystography (OCG) are misleading. USG is highly operated dependant. Periportal tissues or sub hepatic peritoneal folds are usually focused and interpreted as thick, contracted, shrunken or scarred gall bladder [8]. Failure to see the gall bladder on scintigraphy and OCG is interpreted as non-functioning or diseased gall bladder [8]. Failure to outline the gall bladder at ERCP may be due to cystic duct block [5]. As a result preoperative diagnosis of agenesis of gall bladder is extremely difficult.

Frey laid strict criteria for the diagnosis of agenesis of the gall bladder [9]. Intra operatively, if the gall bladder is not visualised in its normal anatomical position, a thorough search should be carried out in the ectopic location namely intra hepatic, left sided, beneath the posterior inferior surface of liver, attached to the left lobe of liver, free floating with in the falciform ligament, between the leaves of lesser omentum, retro peritoneal, retro hepatic, retro pancreatic, retro duodenal and in the anterior abdominal wall [6]. If gall bladder is not found in all these sites peroperative cholangiography is mandatory. CBD exploration should be carried out only if the cholangiogram shows calculi in CBD or CBD is dilated more than 20 mm. Nothing should be done if no gall bladder is found in spite all these measures.

Extensive dissection is required to fulfil Frey's criteria for agenesis of gall bladder. This amount of dissection is associated with complications that can be detrimental to the patient. It is a well known fact that agenesis of gall bladder is a harmless condition on its own. It is not worth performing a major operation just for the sake of diagnosis. Recent literature suggests that when a case of agenesis of gall bladder is suspected on laparoscopy, further procedure should be abandoned and agenesis should be confirmed by diagnostic modalities such as CT scan of the abdomen and ERCP [2, 4]. Whenever available, laparoscopic ultrasound should be used to confirm the diagnosis of agenesis of the gall bladder, as it is an effective imaging modality [6]. Our case exactly fulfils these criteria as we have searched for the gall bladder in all possible sites and laparoscopy abandoned with a provisional diagnosis of agenesis of gall bladder. CT scan and ERCP then confirmed our diagnosis postoperatively. If the agenesis of gall bladder is suspected preoperatively, it should be confirmed by magnetic resonance cholangiopancreatography (MRCP) and / or endoscopic ultrasound before embarking on laparotomy. Frey's criteria have been postulated three and a half decades ago and should be replaced by the advanced imaging modalities namely CT scan, ERCP, MRCP, endoscopic and laparoscopic ultrasound. With a combination of these imaging modalities one should be able to diagnose this seemingly harmless condition with out recourse to laparotomy.

ACKNOWLEDGEMENTS

We are thankful to Dr L Huntley, Consultant Radiologist, West Cumberland Hospital, Whitehaven and Dr C Record, Consultant Gastroenterologist, Freeman Hospital, Newcastle for their help in investigating and confirming the diagnosis. We also thank Ms Janis Corrigan, Medical Illustrations Department for her valuable support in preparing the photographs.

CORRESPONDENCE TO

Mr Tagore M Grandhi 59 Regency Avenue King's Lynn Norfolk PE30 4UH United Kingdom Tel: +44 1553 775797 Fax: +44 1553 613466 E-mail: tmgrandhi@hotmail.com

References

1. Nadeau LA, Cloutier WA, Konecki JT, et al. Hereditary gall bladder agenesis - Twelve cases in the same family The

J of the Maine Med Ass 1972; 63:1-4
2. Sugrue M, Gani J, Sarre R, et al. Ectopia and agenesis of the gall bladder: A report of two sets of twins and review of literature Aust N Z J of Surg 1991; 61:816-18
3. Blechschmidt CM. Agenesis of the gall bladder - Borderline-case of normality? Anat Anz Jena 1982; 151:281-85
4. Singh B, Satyapal KS, Moodley J, et al. Congenital absence of the gall bladder Surg Radiol Anat 1999; 21:221-24
5. Toufeeq Khan TF, Baqai FU. Agenesis of the gall bladder with duplication cysts of the hepatic flexure - A case report and literature review Singapore Med J 1991; 34:181-82
6. Praseedom RK, Mohammed R. Two cases of gall bladder agenesis and review of the literature Hepato-Gastroenterology 1998; 45:954-55
7. Bennion RS, Thompson JE Jr, Tompkins RK. Agenesis of the gall bladder without extra hepatic biliary atresia Arch Surg 1988; 123:1257-60
8. Vijay Kumar, Hemant T, Kocher H, et al. Agenesis of gall bladder - A diagnostic dilemma J of Postgrad Med 1996; 42(3): 80-82
9. Frey C, Bizer L, Ernst C. Agenesis of the gall bladder American Journal of Surgery 1967; 114:917-26

Author Information

Tagore M. Grandhi, FRCS
Staff Grade (General Surgery), Queen Elizabeth Hospital

Saleem M. A. El-Rabaa
Consultant Surgeon, Kettering General Hospital