CASE REPORT

Agenesis of the Gallbladder : Falacies of Ultrasonography

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Abstract

A case of the agenesis of gallbladder detected at laparotomy for symptomatoloy suggestive of gallbladder disease is described. The agenesis of gallbladder, diagnostic inaccuracies and fallacies of ultrasonography for gallbladder disease are discussed. The diagnosis of agenesis was substantiated on MRCP, done postoperatively.

Key Words

Agenesis, Gallbladder, Ultrasonography

Introduction

Gallbladder agenesis is a rare condition that results from the failure of the cystic bud to develop in the 4th week of intrauterine life. Agenesis is usually discovered at laparotomy for cholecystectomy since ultrasound examination of a patient with suggestive symptoms, not visualizing the gallbladder is compatible with chronic cholecystitis (shrunken gallbladder). Many authors assert that absolute certainty of diagnosis is impossible prior to laparotomy or autopsy. This is due to the fact that the various methods of radiological investigation of gallbladder disease have a less than 100% sensitivity.

We present here a case of an adult agenesis of gallbladder with false positive ultrasonographic report of cholecystitis and cholelithiasis.

Case Report

HC, a 30 year old man presented with a 6 month history of recurrent pain in right upper quadrant of abdomen and dyspepsia. There was no previous history of abdominal surgery. Physical examination showed mild tenderness in the right hypochondrium. Liver function tests were normal. During the symptomatic period this patient had got 3 ultrasonographic examinations done, before he reported to our outpatient department. Two of them were reported as contracted gallbladder showing calculi with shadowing, done within an interval of 3 months, by the same ultrasonologist (fig. 1 & fig. 2). Third ultrasonography (USG) was done after a gap of 2 months, which revealed non visualised gallbladder and a hyperechoic area with distal shadowing, suggesting a contracted and fibrosed gallbladder with cholelithiasis.

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Common bile duct in all three reports was reported as normal. The patient was taken up for elective cholecystectomy. To everybody's surprise, gallbladder was not found in its fossa. Thorough search was made for its presence at other known ectopic sites but could not be traced anywhere. Some thickening was palpated in the region of peripancreatic region, therefore biopsy was taken and abdomen was closed in layers. Peroperative cholangiography could not be arranged. Biopsy reported as reactive hyperplasia of lymph node. Postoperatively patient is asymptomatic for the last 10 months.

To confirm the diagnosis postoperatively and look for any ectopic site of the gallbladder, MRCP was done. It also diagnosed agenesis of the gallbladder (fig. 3).

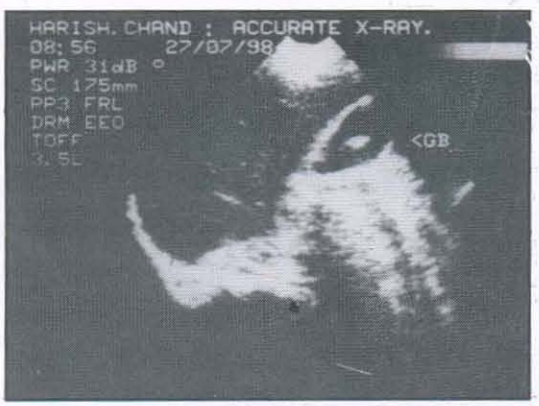

Fig. 1 : Ultrasound showing chronic cholecystitis with gallbladder stones.

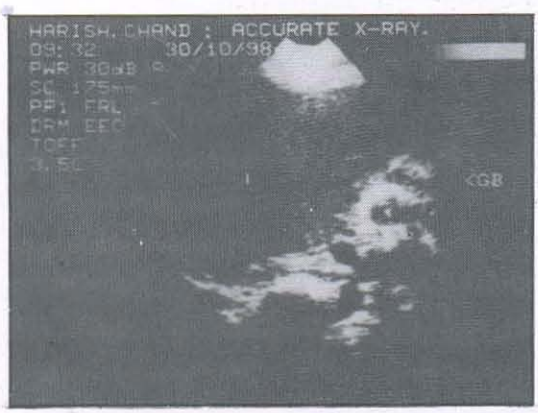

Fig. 2 : Ultrasound showing chronic shrunken gallbladder with gallstones

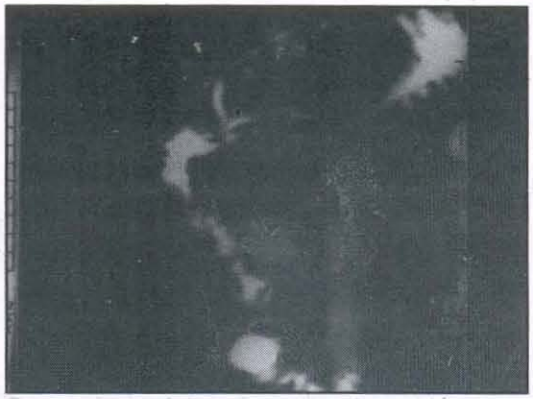

Fig. 3 : MRCP showing absent gallbladder.

Discussion

Agenesis of the gallbladder without extra-hepatic biliary atresia is a rare disorder (1). First described in 1701 by Leery, gallbladder agenesis is a rare but well recognized condition (2,3). The gallbladder develops during the 4th week of intrauterine life from the caudal part of the hepatic diverticulum and failure of the cystic bud to develop results in isolated gallbladder agenesis (4). Although the female : male ratio of incidence in postmortem studies has been reported as equal, clinical studies show 3 : 1 preponderance for the female gender (3). Still a general review of the autopsies reported in the literature shows an incidence of about one case in 6334 live births. The overall incidence of gallbladder agenesis is said to be approximate 0.01% to 0.04% (2). Although the sex distribution of those cases discovered at surgery is predominantly female (3 : 1), autopsy survey accounting for somewhat greater numbers, shows a 1:1 ratio (2).

Congenital absence of gallbladder may occur as an isolated defect or may be associated with other congenital anomalies; imperforate anus, cleft palate, tracheo-oesophageal fistula, skeletal deformities, polycystic kidneys, ventricular and atrial septal defects and pulmonic stenosis (5).

A review of the literature indicates that familial agenesis of the gallbladder has been studied on only three

previous occasions. In 1972, Nadeau et al (6) reported a family with two proved cases and 10 others with nonvisualization.

Dixon and Lichtman (7) stated that the post-cholecystectomy syndrome is physiologically comparable with the symptomatology of the patient with agenesis of the gallbladder. Presumably, causes of pain shared by the two conditions, primary bile stones, biliary dyskinesia, or nonbiliary conditions. Toouli et al (8) speculated that dyskinesia of the sphincter of Oddi, as measured manometrially, favours stasis, and therefore, the development of calculi. Meshkinpour et al (9) demonstrated that patients with an otherwise normal pancreaticobiliary tree who suffer from right-upper-quadrant abdominal pain have a higher sphincter of Oddi resting pressure.

Usually it is very difficult to diagnose agenesis of the gallbladder pre-operatively. Although ultrasonography has an accuracy of 95-98% in the diagnosis of gallstones, hyperechoic images in the right upper quadrant may be mistakenly interpreted as representing a gallbladder full of stones when the walls or lumen of the gallbladder cannot be visualized (10). Ultrasound examination of a patient with fever and right upper quadrant abdominal pain, in which the gallbladder is not visualized, might pre--indicate the presence of acute cholecystitis. Shrunken gallbladder, is usually taken as an expression of chronic cholecystitis. However, this is probably the predominant cause of diagnostic inaccuracy pre-operatively (3). The most likely cause of the false positive sonographic finding was the visualization of a small bowel loop in the area of the gallbladder (3).

The sonographic patterns of cholelithiasis have been well-described (11). Nonvisualization of the gallbladder lumen with or without detection of strong echoes with shadowing in the region of the gallbladder fossa, has been reported to be associated with gallbladder disease in over 90% of the cases (12). In clinical practice, however, lack of visualization may not be accepted as definitive for gallbladder disease because the image is nonspecific and may be simulated by other condition, including poor technique.

Although the echo-shadow image is non-specific because of lack of visualization of the gallbladder lumen, it is an accurate, indirect indicator for gallbladder disease, provided there is strict adherence to impeccable technique. It was associated with cholelithiasis. On the other hand, nonvisualization is an unreliable sign for gallbladder disease. Only seven (67%) of 11 patients scanned with static scanner proved to have cholelithiasis, while the other four had normal gallbladder on oral cholecystography (13). Dynamic scanning decreased the number of patients with such indeterminate results (nonvisualization) by 45%, from 11 to six patients, only two (33%) of whom had cholelithiasis. These results support the recommendation, oral cholecystogrphy for all patients with nonvisualization on cholecystosonography (13).

Many authors assert that absolute certainity of diagnosis is immpossible prior to laparotomy or autopsy (14). This is due to the fact that various methods of radiologic investigation of gallbladder disease have a less than 100% sensitivity. The accuracy of ultrasound examination may be reduced by obesity, raised bowel gas and the inexperience of the interpreter (14). Likewise, the ultrasonic evaluation of a contracted gallbladder full of stones may be difficult. Failure to visualize the gallbladder at oral cholecystography, radionuclide scanning, percutaneous transhepatic cholangiography or endoscopic retrograde cholangiopancreatography (ERCP) is often interpreted as being evidence of cystic duct obstruction rather than an absent gallbladder (14).

The operative strategy at laparotomy where gallbladder appears to be absent should include careful search in all likely ectopic sites, peroperative cholangiography and exploration for non-biliary cause

of the patients symptoms. The usual ectopic sites described are attached to the left hepatic lobe, falciform ligament free-floating, retroperitoneal and location with the lesser omentum or anterior abdominal wall (14).

With laparoscopic cholecystectomy receiving greater attention (15), cases of absent gallbladder will soon be diagnosed by laparoscopy. As a completely intraheptic gallbladder is believed to be exceptional rarity (14), cases of absent gallbladder may be diagnosed by laparoscopy. Where the question of an intrahepatic gallbladder is seriously contemplated or in cases where doubt persists about the cause of symptoms, ERCP or CT or MRCP may clarify the situation. As was done in our case, where postoperative MRCP also diagnosed an absent gallbladder.

Gallbladder agenesis may not be as rare as was previously thought. A surgeon must be aware of its occurrence and maintain a high index of suspicion. Patient with clinic features compatible with gallbladder disease but equivocal imaging of the gallbladder should undergo laparosocpic exploration (14).

For some unexplained reasons, most of the symptomatic patients described in the literature became symptom-free after the operation (16). Extensive postoperative workups usually are not indicated. In case of pain and symptom recurrence, institution of oral smooth muscle relaxants and analgesic therapy may be indicated (14). Invasive procedures including sphincter ablative procedures should be restricted only in those patients not responding to conservative therapy (14).

Long term implications of absent gallbladder is not known, but indirect evidence can be obtained by studying the postcholecystectomy patients.

Upto 40% of patients continue to experience symptoms after cholecystectomy, only a minimum of which is attributable to unsuccessful surgery. Symptoms after cholectectomy may include persisting pain or dyspepsia (17).

Loss of the resevoir function of the gallbladder results in disturbance of bile metabolism and alteration in the dynamics of bile release. Bile flow into duodenum changes, from intermittent meal related to continuous, and its composition also changes, becoming more damaging to gastric and oesophageal mucosa. This leads to increased duodenogastric reflex which is aggravated further by the impaired function of the antropyloric motor unit that appears to results from direct effect of bile on the duodenum (17). Subsequent symptoms have been related to the increased incidence of gastritis described in up to 50% of patients due to alkalne duodenogastric reflux, and to the qualitative change in bile composition (17). The bile salt pool is reduced by 50%, which may result in subclinical fat metabolism and postcholecystectomy diarrhea, which is attributed to bile catharsis (17). The presence of fat in the colon is known to excite the release of gastrointestinal hormones, most notably peptite YY that affects the motility of the antrum and pylorus (11) and which may have an effect on the lower oesophagus. More bile is in circulation at any one time and is subject to the effects of bacterial degradation, leading to the formation of secondary bile acids which have been implicated in colonic neoplasia following cholecystectomy (17).

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