AGENESIS OF THE GALL BLADDER WITH DUPLICATION CYSTS OF THE HEPATIC FLEXURE - A CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

A 24-year-old lady with recurrent upper abdominal pain, underwent surgery for cholelithiasis based on imaging diagnosis by ultrasound scanning. At laparotomy, the gall bladder could not be found either in its normal or ectopic locations. The diagnosis of agenesis of the gall bladder was confirmed by operative cholangiography. Duplication cysts of the hepatic flexure were discovered in the position normally occupied by the gall bladder. The stony hard faeces in the cysts were probably interpreted as gallstones on ultrasound. This rare condition is discussed and the importance of intraoperative cholangiography is stressed.

Keywords: *gall bladder agenesis, colonic duplication cysts, diagnosis.*

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INTRODUCTION

Agenesis of the gall bladder without extrahepatic biliary atresia is a rare but well recognised congenital anomaly (incidence of 0.01 to 0.04%)[1,2]. It is impossible to predict and is recognised either at laparotomy, by radiology or at autopsy. We report a 24-year-old female with right upper abdominal symptoms, attributed to cholelithiasis by ultrasound. At operation, agenesis of the gall bladder confirmed by a cholangiography and colonic duplication cysts were discovered.

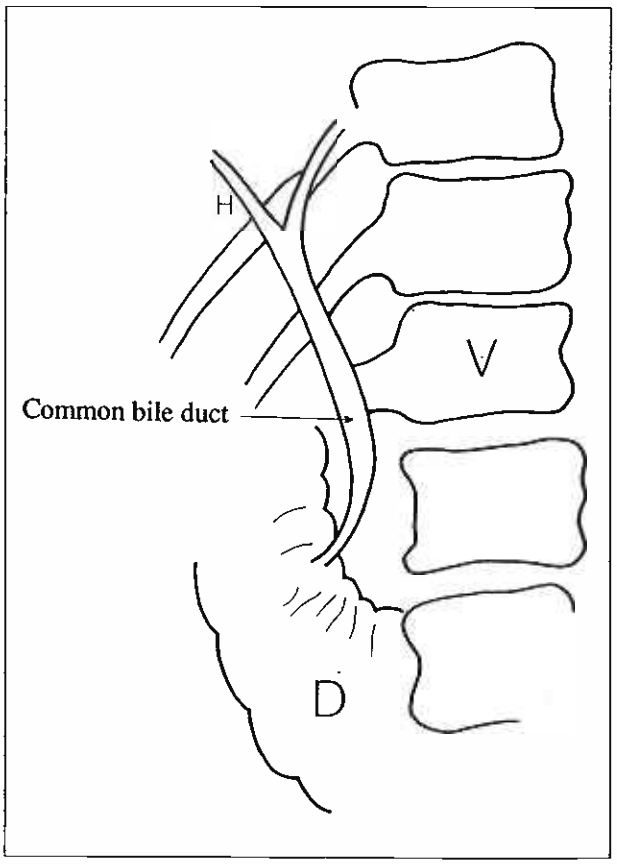
To the best of our knowledge, the finding of colonic duplication with agenesis of the gall bladder has not been reported in the English literature.

CASE REPORT

A 24-year-old newly-married female presented on 17 August 1988 with recurrent post-prandial right upper quadrant pain, occasional vomiting and a bloated feeling for the last 6 months. There was nothing remarkable on examination. All her blood investigations were normal and the oral cholecystogram failed to outline the gall bladder, indicating cystic duct block. Ultrasound examination revealed multiple calculi in the gall bladder.

With a diagnosis of symptomatic cholelithiasis she underwent laparotomy. The liver looked normal but the gall bladder could not be found in its normal position. It was then looked for in the gastrohepatic omentum, the hepatoduodenal ligament and its posterior aspect. After a Kocher manoeuvre, the retroduodenal and the area posterior to the pancreas was also explored. The liver was then aspirated in the regions of segment 4 and 5 for an intrahepatic gall bladder. As there was no cystic duct stump a butterfly needle was inserted into the common bile duct for operative cholangiography. The cholangiogram outlined a normal extra and intra hepatic ductal system and confirmed the absence of the gall bladder (Fig 1).

Fig 1 - Line drawing of the radiograph in depicting details. D (duodenum), V (vertebrae), H (hepatic ducts).



The surrounding structures were examined. The posterior surface of the hepatic flexure contained multiple cysts containing hard faecal material. The largest cyst was 5 cm by 6 cm and communicated with the colon. These were interconnected but the communications were blocked by the contents. All the cysts were excised and the colonic defect was closed. Colonic resection was not required. A search was also made in

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the small bowel mesentry and the remaining large bowel for other duplications.

A barium swallow was undertaken 2 weeks later to exclude oesophageal duplication. She has been followed up for three years now with complete relief of her symptoms. The histology showed a normal intestinal wall with considerably thinned-out muscle. All her immediate family members had normal gall bladders on ultrasound scanning.

DISCUSSION

Agenesis of the gall bladder was first reported by Lemery in 1701. This is a rare anomaly, with an incidence of one in 6,000 births and a total of 381 cases are reported in the literature till 1989[1]. There are various etiological theories of agenesis of the gall bladder. The important ones are: failure of the ventral cholecystic bud to develop and failure to recanalize after the solid phase.

It may present as an isolated anomaly or with other associated malformations affecting various systems. The significant anomalies associated with agenesis are rectovaginal fistulae and imperforate anus[1]. Bennion et al[1] had classified patients with gall bladder agenesis into 3 groups. The multiple foetal anomaly group in which the diagnosis can only be suspected, the asymptomatic group discovered at autopsy and the symptomatic group who have symptoms related to the biliary tract. In the latter group the symptoms are similar to the postcholecystectomy state[2] which needs to be investigated to exclude primary bile duct stones, biliary dyskinesia and anatomically related non-biliary pathology.

A strong familial incidence has been reported[3,4]. Colonic duplication cysts with gall bladder agenesis has not been previously reported though these duplications are associated with other malformations, especially the genito-urinary tract[5]. The drug, thalidomide, when used during pregnancy has been known to cause agenesis[6].

There is non visualisation of the gall bladder on oral cholecystogram which suggests cystic duct block causing the biliary colic. Ultrasound scanning is not diagnostic because the same picture is seen in contracted gall bladders in patients with right upper abdominal symptoms. In this case, the inspissated faecal material in the cysts situated in the subhepatic area led the radiologist to conclude that it was cholelithiasis. Failure to outline the gall bladder at ERCP may be due to cystic duct block. Non visualisation of the gall bladder by ultrasonography and ERCP enabled Sullivan et al[7] to successfully diagnose their case preoperatively as agenesis. Hida scan will fail to outline the gall bladder in agenesis, but this also happens in acute cholecystitis.

During surgery the surgeon has the responsibility of proving agenesis and this involves a meticulous search in both intra and extra hepatic locations[8]. Operative cholangiography is mandatory and is the only reliable way to confirm agenesis or locate an ectopic gall bladder.

CONCLUSION

A 24-year-old lady with agenesis of the gall bladder and duplication cysts of the hepatic flexure is reported. The location and the contents of these cysts was mistakenly interpreted as cholelithiasis by ultrasound. Agenesis of the gall bladder was confirmed by operative cholangiography and led to the discovery of the duplication cysts connected to the hepatic flexure. This association has not been reported before in the English literature.

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