Hepatic hilum-type II choledochal cyst masquerading as gallbladder duplication

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SUMMARY

Choledochal cyst and gallbladder duplication are rare congenital anomalies. They typically are surgical problems of infancy or childhood but rarely may present in adults also. Despite high resolution imaging, the differentiation of type II choledochal cyst from gallbladder duplication often causes the diagnostic dilemma; which may result in high risk for intraoperative iatrogenic injury. Operative management of choledochal cyst is the definite treatment because of its malignant potential. A type II choledochal cyst arising from the hepatic hilum presenting as gallbladder duplication on imaging has not been reported earlier in the literature and here we present a case report of the same which was managed successfully.

BACKGROUND

Hepatobiliary anomalies in close proximity to the gallbladder fossa such as choledochal cyst, gallbladder duplication, duodenal duplication or gallbladder adenomyomas needs to be distinguished accurately prior to surgery with high resolution imaging like MRI.[1] Despite high resolution imaging, the differentiation of type II choledochal cyst from gallbladder duplication often causes the diagnostic dilemma.[2] Laparoscopy is the best tool for diagnosis and operative management. These patients often associated with other hepatobiliary pathology, high risk for intraoperative iatrogenic injury. A type II choledochal cyst arising from the hepatic hilum presenting as gallbladder duplication on imaging has not been reported earlier in the literature was managed successfully. It is also imperative that resection of the choledochal cyst should be done to prevent its transformation into future cancer.[3]

CASE PRESENTATION

A 32-year-old woman hailing from North India presented with recurrent dull aching pain abdomen in right upper quadrant for 3 years. There was no history of jaundice, nausea and fever. She also complained of occasional dyspepsia post meal, for which she took no treatment.

INVESTIGATIONS

Sonography was done which revealed chronic cholecystitis with dilated proximal bile duct. Her lab investigations were within normal limit except with haemoglobulin level 11.2g/dL, white blood cell count of $6.8X10^9$/L. Serum alkaline phosphatase was 154 IU/L, slightly in near higher limit of our institute values, but total and conjugated bilirubin were normal 0.7 and 0.3 mg/dL, respectively. Magnetic Resonance Cholangiopancreatography (MRCP) showed cholelithiasis and a 1.4×1.3 cm cystic structure superior to gallbladder communicating with common hepatic duct via a linear duct and differential of duplicated gallbladder or type II choledochal cyst was made (figure 1A). However, there was no evidence of abnormal pancreatobiliary junction on reviewing of MRCP.

TREATMENT

She was taken up for surgery and on laparoscopy the findings were distended gallbladder, long cystic duct with normal insertion, calot's triangle dissected and cholecystectomy was done. There was single long cystic duct and artery. After cholecystectomy, another cystic structure filled with mucus arising from the hepatic hilum was noted and diagnosis of type II choledochal cyst was made (figure 1B,C). During excision of the cyst, there was an iatrogenic traction injury at hilum with bile effluent from pin point opening. The procedure was converted to open in view of technical difficulties and repaired primarily. Intraoperative cholangiogram was not done as the injury was readily identified and repaired without any technical difficulty. The reason for mucus filled cyst despite communication with hilum was due to acute angulation between the point of the origin and the cyst. The postoperative period was uneventful. Histology had features of chronic cholecystitis in the gall bladder specimen while the cyst showed flattened lining epithelium with absence of submucosal glands or muscle layer suggesting choledochal cyst.

OUTCOME AND FOLLOW-UP

She did well in postoperative period. Her oral feed was started in postoperative day 1 which was tolerated well and was discharged on day 2. Currently she is in follow-up in outpatient department and is doing well.

DISCUSSION

Choledochal cysts are congenital anomaly of biliary ducts, typically are surgical problems of infancy or childhood; however, in 20% of patients the diagnosis is delayed until adulthood.[4] Bile duct cysts account for approximately 1% of all benign biliary disease.[5] Incidence ranges from 1 in 100 000 to 1 in 150 000 individuals in western countries to 1 in 13 000 in certain parts of Asia.[6] Choledochal cyst and gallbladder duplication both are rare congenital anomalies of the biliary tree primordium and are difficult to differentiate clinically and radiologically. Incidence of duplicated gallbladder reported

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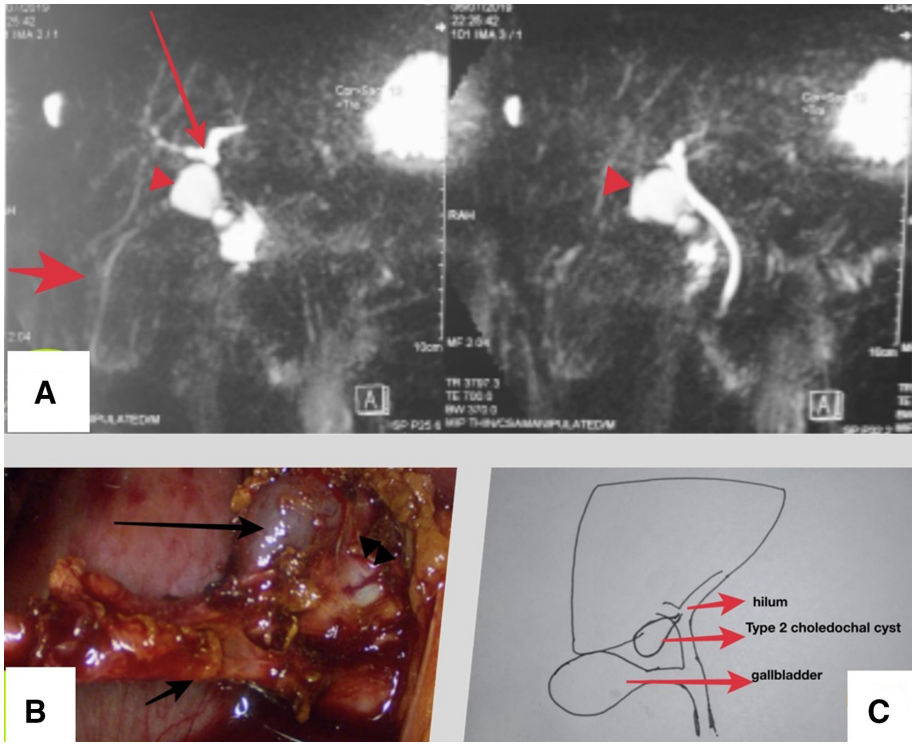

Figure 1 (A) Magnetic resonance cholangiopancreatography showing cholelithiasis and 1.4×1.3 cm cystic structure superior to gallbladder communicating with common hepatic duct via a linear duct. (B) Intraoperative laparoscopic view showing dissected calots triangle, gallbladder and a small cystic lesion located at the hilum. (C) Diagrammatic representation of intraoperative finding (courtesy—Dr Amaresh Aruni coauthor).

is 1 in 12 000 cholecystectomies and 1 in 4000 autopsies.[1] Type II choledochal cyst is rare with an incidence of 2.3%, Quaissi *et al* classified type II cysts according to the location in relation to the hepatoduodenal ligament as upper, middle and lower with incidence reported 58%, 21% and 21%, respectively.[7] A type II choledochal cyst arising from the hepatic hilum presenting as gallbladder duplication on imaging has not been reported earlier. The aetiology of type 2 Choledochal Cysts was suggested by Hayes *et al* as that the diverticulum is a remnant of an earlier stage of bile duct development when it represents network of cells than tubular ducts.[8] Ultrasonography is the initial investigative modality, however, MRCP has become the gold standard method of imaging the bile ducts and their anomalies. MRCP has a 96%–100% detection rate for bile duct cysts, 53%–100% rate for diagnosing anomalous Pancreatico-biliary Juction.[9] Laparoscopy is very helpful in evaluation of normal anatomy, anomalies, clears the diagnosis and proceeding with definitive procedure, however, histopathology confirms the diagnosis. The management of isolated type II choledochal cyst is surgical excision. Cholecystectomy is the procedure of choice for symptomatic gallbladder duplication. Minimally invasive approaches are used increasingly for the biliary disease and gallbladder pathologies due to superior visualisation of the structure around the cyst, gallbladder, hepatic hilum and associated anomalies. The magnification conferred by laparoscope helps assess the structures in detail, fine dissection and anastomosis at the hilar area.[10] However, it must be emphasised that laparoscopic excision of bile duct cysts is technically challenging and requires experience in both complex biliary operations and advanced laparoscopic surgery.[11] Caveats of performing surgery include higher risk of biliary injury due to variant anatomy.[12] Malignant transformation of choledochal cyst is a serious complication and risk increases with age, excision is most desirable in children and adults whenever it is suspected.[3 13]

Learning points

- Choledochal cyst and gallbladder duplication are rare congenital anomalies, though advancement in imaging one should proceed with certain differentials before surgery.
- Choledochal cyst if identified should be treated operatively, due to its malignant potential.
- Biliary ductal system is associated with anatomical anomalies and should be dealt preoperatively by imaging, intraoperatively by careful dissection and cholangiogram if required.

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