



Case report

Concomitant hilar cholangiocarcinoma with choledochal cyst and cholelithiasis in an asymptomatic patient: A case report

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ABSTRACT

Introduction and importance: Choledochal cyst is an important risk factor for cholangiocarcinoma. Concomitant hilar cholangiocarcinoma with choledochal cyst with cholelithiasis is a rare finding.

Case presentation: A 52-year-old male presented with non-specific complaints of generalized weakness for 3 months. Transabdominal ultrasound showed dilated common bile duct with hyperechoic mass at hilar region; further evaluation with magnetic resonance cholangiopancreatography and contrast-enhanced computer tomography of abdomen and pelvis revealed concomitant intraductal hilar cholangiocarcinoma (Bismuth Corlette type 1) with Choledochal cyst (type IVa) with cholelithiasis. After optimization patient underwent left hepatectomy with common bile duct excision with cholecystectomy with Roux-en-Y hepaticojejunostomy. Histopathological examination confirmed it to be well-differentiated adenocarcinoma, intestinal type at the hilar confluence.

Clinical discussion: An asymptomatic male patient with the concomitant finding of perihilar cholangiocarcinoma (Bismuth Corlette type 1) with choledochal cyst type IVa with cholelithiasis is a rare finding. The patient was managed with surgical excision of the common bile duct with Roux-en-Y hepaticojejunostomy and cholecystectomy. Diagnostic evaluation should be proper not to miss or overlook such a synchronous lesion.

Conclusion: Incidental finding of concomitant perihilar cholangiocarcinoma with the choledochal cyst is rare. Proper evaluation of the patient with CECT abdomen and pelvis and MRCP is necessary for the diagnosis. Proper surgical resection with adequate lymph node removal is important for surgical clearance.

1. Introduction

Cholangiocarcinoma is a worrisome complication of unresected choledochal cysts, with an incidence up to 20 to 30% in early adulthood [1]. Cholangiocarcinoma is the most common malignant tumor associated with choledochal cyst. As per Todani classification, types I, IV, and V biliary cysts increase the risk for cholangiocarcinoma by 30-fold [2,3]. Because of the risk of cholangiocarcinoma, primary excision of extrahepatic choledochal cysts with biliary-enteric anastomosis is the treatment of choice in all cases, even in the absence of symptoms. The incidence of cancer in patients with the primary choledochal cyst is 9.9%, whereas the incidence after cyst excision is 0.6% [4]. Metachronous lesions after many years of choledochal cyst resection have been reported [5]. We report a case of a male patient diagnosed with concomitant perihilar cholangiocarcinoma (cT1N0M0) with choledochal cyst type IVa and cholelithiasis in an otherwise asymptomatic

patient which was diagnosed incidentally and properly managed with surgery. Few cases have been reported in the literature about such rare and synchronous findings. This case report has been reported in line with the SCARE Criteria [6].

2. Presentation of the case

A 52-year-old male had non-specific complaints of generalized weakness for 3 months. He had normal bowel and bladder habits with no history of abdominal, vomiting, yellowish discoloration of eyes, itching, weight loss, fever, or other constitutional symptoms. On examination, he was of average built with normal cardinal and vital signs as well as normal systemic and physical examinations.

The transabdominal ultrasound done in other center showed heteroechoic lesion of 2.9 × 2.1 cm in the hilar region leading to upstream dilation of Intrahepatic Biliary Ducts (IHBDs) more prominent in the left

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lobe of the liver with dilated proximal common bile duct (CBD) size 20 mm, with gallbladder concretion; features suggestive of hilar cholangiocarcinoma dilated CBD with cholelithiasis. After which he visited us and further evaluation was done with magnetic resonance cholangiopancreatography (MRCP) which showed 24×11.7 mm irregular enhancing endoluminal soft tissue intensity mass in common hepatic duct suggestive of intraductal hilar cholangiocarcinoma; dilated common bile duct of size 18.6 mm and dilated intrahepatic biliary ducts, contracted gallbladder with concretions and sludge. Contrast-enhanced computer tomography (CECT) abdomen and pelvis showed approximately $2 \times 1.9 \times 1.3$ cm heterogeneous enhancing soft tissue mass in hilar confluence with extension to the left hepatic duct, no complete occlusion of confluence, with the abutment of the left branch of the portal vein with maintained fat plane (features of hilar cholangiocarcinoma, Bismuth Corlette type I) with contracted gallbladder with dilated common bile duct of size 19 mm and dilated intrahepatic bile ducts (grade II) [7].

His preoperative blood investigations were within normal range. Tumors markers carcinoembryonic antigen (CEA) was 6.69 ng/ml and cancer antigen 19.9 (CA 19.9) was 3 U/ml. The Patient was optimized for surgery. CT volumetric analysis was done with the plan for left hepatectomy; Total liver volume was 1324.1 cm^3 , Left Liver volume: 330.87 cm^3 & Functional Liver Remnant (FLR) was 998 cm^3 . The biliary duct was Huang Type A4 [8]. Portal vein was Nakamura et al. type A [9]. Hepatic veins and arteries were normal and free from tumor. The patient underwent left hepatectomy including caudate lobe with cholecystectomy with CBD excision with Roux-en-Y hepaticojejunostomy and perihilar and pericholedochal nodal clearance. Intraoperative findings were dilated common bile duct with choledochal cyst (type IVa), 2x1cm firm mass intraductal within the common hepatic duct (CHD), however, had no liver or peritoneal metastasis. The histopathological report of the surgical specimen revealed well-differentiated adenocarcinoma, intestinal type at the confluence of CHD with tumor invading beyond the wall of the bile duct to surrounding adipose tissue, no perineural and lymphovascular invasion, caudate lobe free from the tumor, chronic cholecystitis, gastroduodenal lymph node free of tumor, TNM staging pT2aN0 (AJCC Ca Protocol, 8th edition2020) [10].

Postoperatively, the patient experienced good overall recovery. Each day complete blood count, renal function test, and liver function test

(including PT/INR) were monitored, there were no features suggesting post hepatectomy liver failure. The patient was discharged on the 8th postoperative day. After multidisciplinary meeting, patient was referred to the oncologist and was on adjuvant chemotherapy. The patient was kept on regular follow up and there were no symptoms and signs of recurrence (See Figs. 1–5).

3. Discussion

Choledochal cysts are dilatations of bile ducts, which may be single or multiple. The incidence of biliary cysts is estimated to be 1:100,000 to 150,000 with higher incidence rates in some Asian nations. The female to male ratio is approximately 3:1. Cysts may be congenital or acquired; however, the exact pathogenesis is unknown [11]. Most of the choledochal cysts are diagnosed during childhood before the age of 10, but 20% of the choledochal cyst are diagnosed during adulthood [12]. Bile-duct cysts are an established risk factor for Cholangiocarcinoma. Type I (solitary, extrahepatic) and IV (extrahepatic and intrahepatic) bile-duct cysts have a higher incidence. It is presumed that prolonged reflux of pancreatic secretions into the biliary tract occurs in Todani types I and IV, which frequently present with abnormal pancreaticobiliary duct junctions [13]. Cholelithiasis is also a known risk factor for both intrahepatic and extrahepatic cholangiocarcinoma [14].

The lifetime incidence of Cholangiocarcinoma in these patients ranges from 6% to 30%. The risk of malignancy decreases after complete choledochal cyst excision; however, these patients are still at an increased risk of developing Cholangiocarcinoma compared with the general population [15]. Associated biliary malignant tumor should always be considered in patients with a choledochal cyst, especially in aged patients or patients with anomalous pancreaticobiliary ductal union or an elevated tumor marker level. Cholangiocarcinoma developing in a choledochal cyst usually has an adverse outcome because of late diagnosis and a low possibility of resectability [16].

In a retrospective multicenter survey from South Korea done by Seung Eun Lee et al. (2011), a total of 808 patients who underwent surgery for choledochal cyst; biliary tract malignant tumor was associated in 9.9%, & only 2 had synchronous gallbladder and bile duct cancer [17].

Similarly in meta-analysis done by ten Hove A et al. (2018), overall

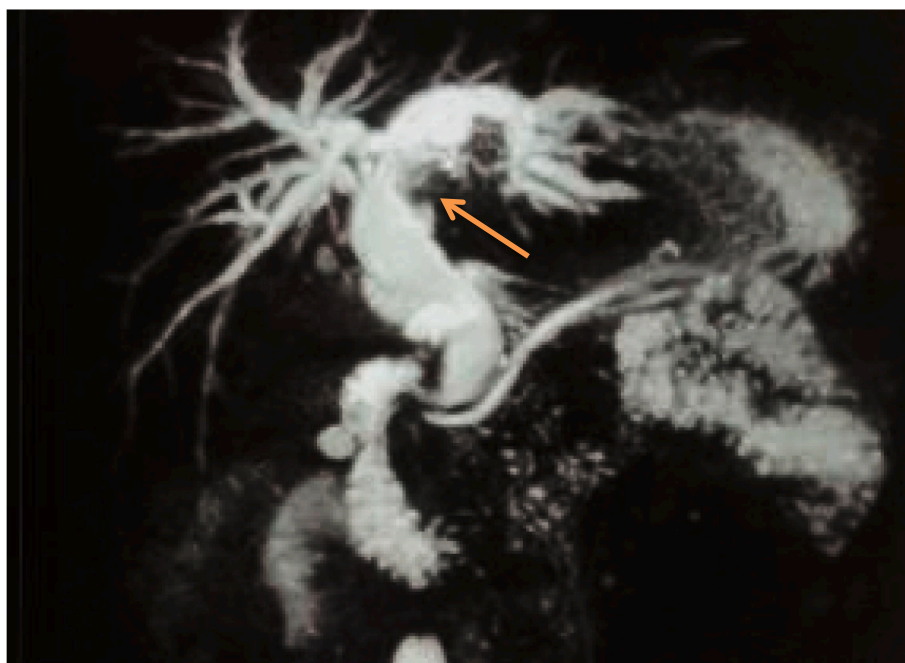


Fig. 1. MRCP showing dilated extrahepatic and left intrahepatic bile duct with filling defect (arrow) at the hilar confluence.

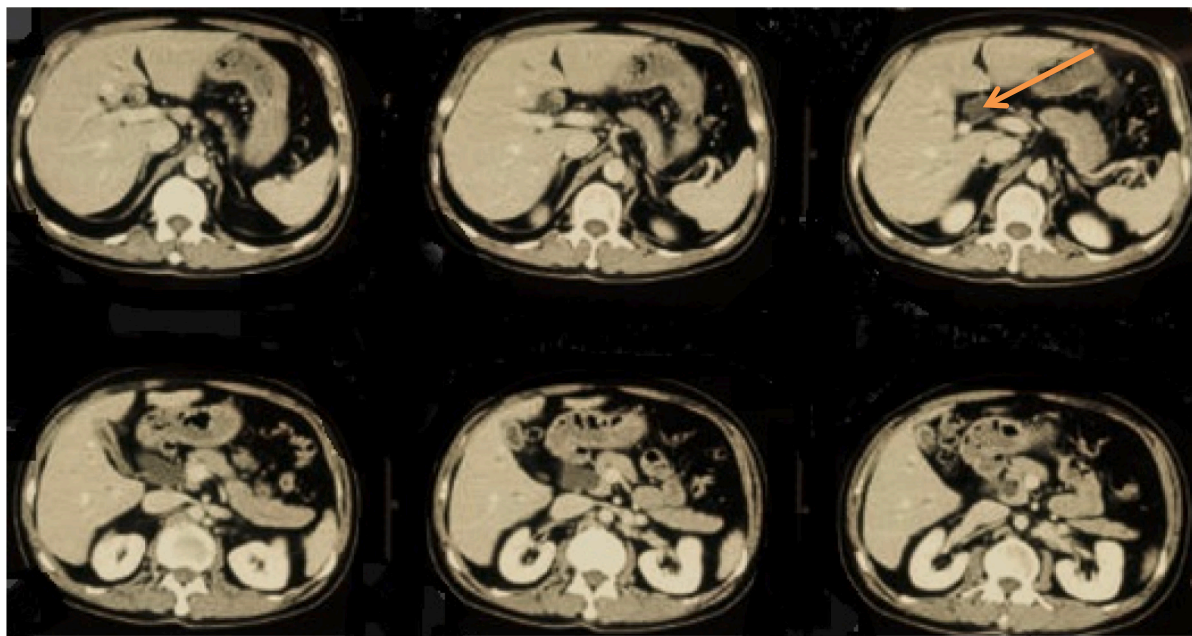


Fig. 2. CECT abdomen showing intraductal mass at hilar region (arrow) with dilated left hepatic duct.

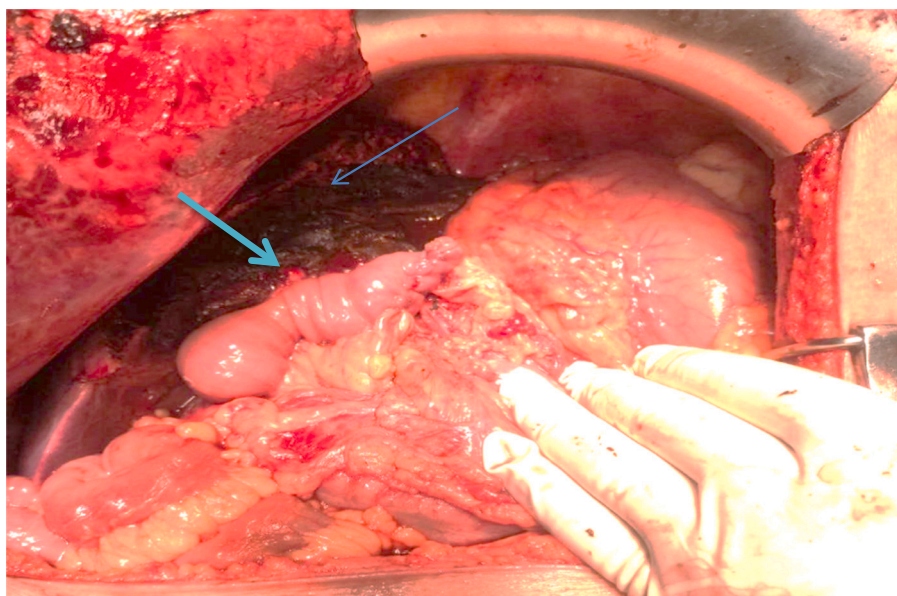


Fig. 3. Intraoperative image showing cut section of liver parenchyma (thin arrow) with the anastomotic site of Roux-en-Y Hepaticojejunostomy (thick arrow).

312 of the 2904 patients (10.7%) developed a malignancy out of which 7.3% had tumor present during primary surgery of choledochal cyst, and 3.4% had tumors that developed during follow-up [18].

The first scanning method of the biliary tree is transabdominal ultrasound. It is a non-invasive examination accessible quickly and inexpensively. Computer tomography (CT scan) and MRI are indicated to assess the lesion and to study the relations with the neighboring structures, especially when one suspects a malignant transformation of the choledochal cyst [19].

Cholangiocarcinoma occurring concomitantly with choledochal cyst and cholelithiasis is rare. In our case, despite the presence of perihilar cholangiocarcinoma, choledochal cyst, and cholelithiasis, the patient was almost asymptomatic. The patient had nonspecific complaints of generalized weakness for which he underwent investigations that revealed the incidental concomitant biliary lesions. After the

transabdominal ultrasound, the CECT abdomen and pelvis and MRCP revealed the presence of perihilar cholangiocarcinoma and concomitant choledochal cyst with cholelithiasis. Tumor markers were normal too. The patient underwent left hepatectomy with cholecystectomy and CBD resection with Roux-en-Y hepaticojejunostomy with perihilar and pericholedochal nodal clearance to have an adequate future liver remnant (FLR) for such findings. It is known that perihilar cholangiocarcinoma necessitates a major hepatic resection to achieve both longitudinal and radial margins negative for tumor. Routine *en bloc* lymphadenectomy of the perihilar and pericholedochal nodal basins along with R0 resection is of utmost importance and has been linked to improved survival [20]. Diagnostic evaluation should be proper not to miss or overlook such a synchronous lesion.

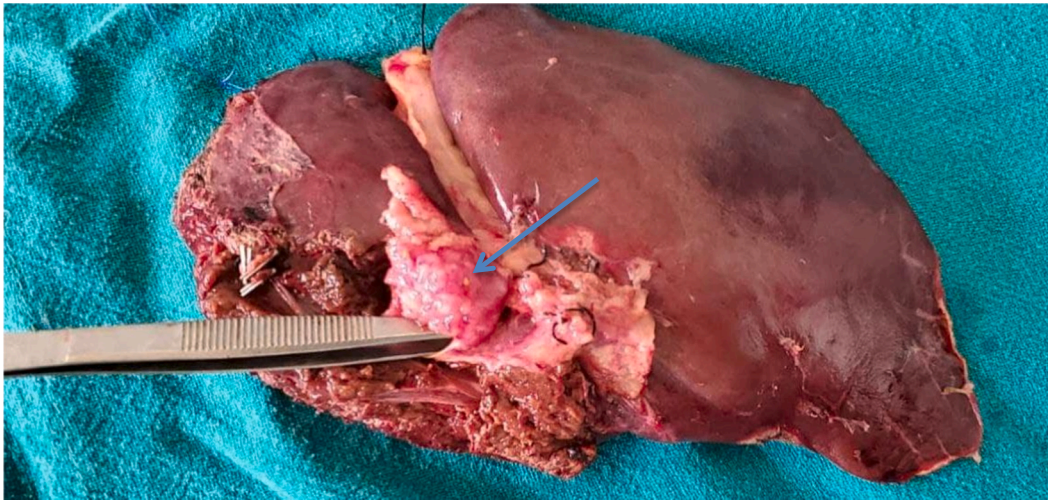


Fig. 4. Surgical specimen showing firm, exophytic intraductal mass at the hilar confluence (blue arrow) [forceps used for traction to reveal the tumor].

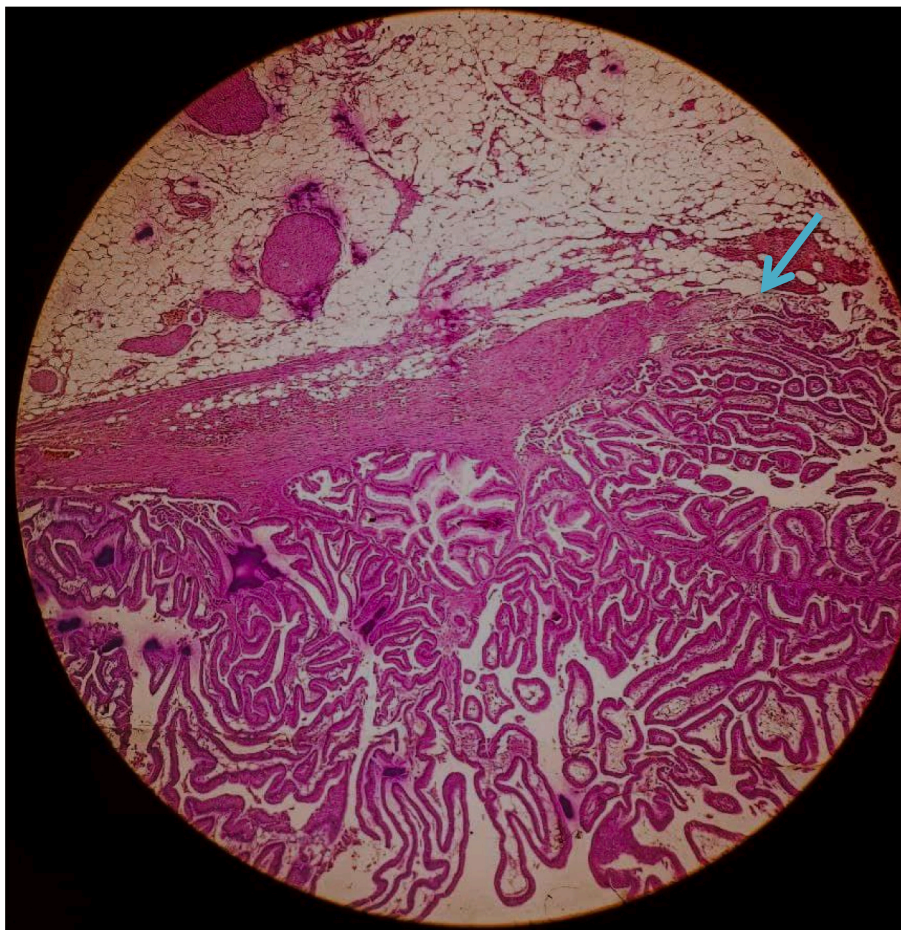


Fig. 5. Histopathological slide showing tumor cells (adenocarcinoma) invading into the adipose tissue (arrow).

4. Conclusion

Incidental finding of concomitant perihilar cholangiocarcinoma with the choledochal cyst and cholelithiasis is rare. Proper evaluation of the patient with CECT abdomen and pelvis and MRCP is necessary for the diagnosis. Proper R0 surgical resection with adequate lymphadenectomy is important for surgical clearance and for improving overall

survival.

Ethical approval

Nothing to declare.

Consent

A written informed consent was obtained from the patient and patient's family member for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Sushil Gyawali, Sujan Shrestha, Ramesh Singh Bhandari: Study concept, Data collection, and surgical therapy for patient.

Sushil Gyawali, Gagan Adhikari: Writing and original draft preparation.

Sushil Gyawali, Sujan Shrestha: Editing and writing.

Ramesh Singh Bhandari and Sumita Pradhan: senior authors and manuscript reviewer.

All the authors read and approved the final manuscript.

Registration of research studies

1. Name of the registry: –
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Guarantor

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Authors' contribution

Sushil Gyawali, Sujan Shrestha: Study concept, data collection, and surgical therapy for the patient.

Sushil Gyawali, Gagan Adhikari: Writing- original draft preparation.

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None.

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