



## Case Report

# Adult Choledochal Cyst Associated with Carcinoma Gall Bladder: A Rare Entity

Authors

**Dr Asmita Arya<sup>1</sup>, Dr Hira Lal<sup>2</sup>, Dr Ashok Kumar<sup>3</sup>**

<sup>1</sup>Senior Resident, Department of Radiology, Sanjay Gandhi Postgraduate Institute of Medical Science, Lucknow, 226014, India

Email: [dr.asmitasmita06@gmail.com](mailto:dr.asmitasmita06@gmail.com)

<sup>2</sup>Additional Professor, Department of Radiology, Sanjay Gandhi Postgraduate Institute of Medical Science, Lucknow, 226014, India

Email: [hiralal2007@yahoo.co.in](mailto:hiralal2007@yahoo.co.in)

<sup>3</sup>Associate Professor, Department of Surgical Gastroenterology, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, 226014, India

Corresponding Author

**Dr Ashok Kumar**

Associate Professor, Department of Surgical Gastroenterology

Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, 226014, India

Email: [drashok97@gmail.com](mailto:drashok97@gmail.com)

## Abstract

*Choledochal cysts usually present with abdominal pain, jaundice, abdominal lump and in various combinations, however, sometimes it may present in unusual manner like hepatolithiasis, portal hypertension, spontaneous perforation, and biliary tract malignancy. Here we report case of adult choledochal cyst with carcinoma gall bladder which is very uncommon. Cholangiocarcinoma is the most common malignancy associate in patients with the choledochal cyst; however, carcinoma gall bladder could also be associated with this entity due to the possible occurrence of anomalous pancreatobiliary ductal junction which leads to continuous reflux of activated pancreatic enzymes, amylase, and bile stasis contribute proliferation of bile duct wall in presence of choledochal cysts. A high index of suspicion is needed for early diagnosis and management of these two entities. Ultrasound is first line of investigation followed by MRI protocol including MRCP, T2WI, T1WI and contrast study is quite useful to delineate type of choledochal cysts, biliary anatomy, congenital anomalies and level of biliary duct infiltration / obstruction and extent & characterisation of tumor which is important for planning management.*

## Introduction

Choledochal cyst is a rare anomaly of bile duct characterised by abnormal dilatation of extraheptic bile duct and/or the intraheptic bile

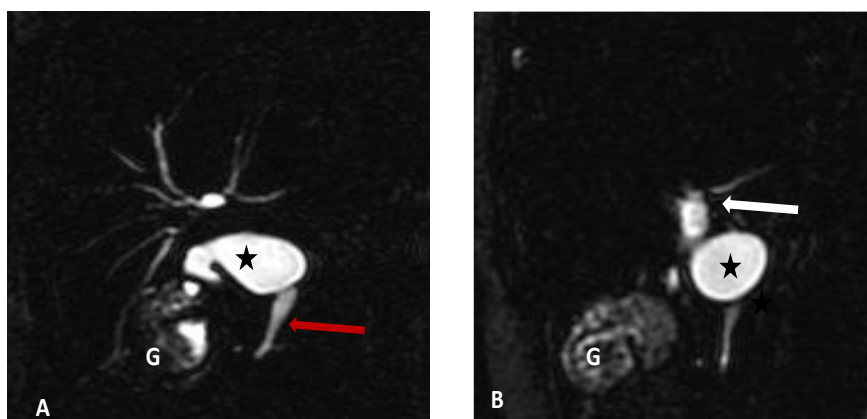
duct. It is common among Asian population. About 60 % cases of patients are presented during the first decade of life while 20% remain undiagnosed until adulthood. The risk of

malignancy increases with age and more risk in adult patients than childhood [1], [2]. Gall bladder malignancy is relatively uncommon in compare to cholangiocarcinoma in choledochal cysts patients, only limited cases are reported in literature. The etiology of development of malignancy in choledochal cyst is still unknown. Pancreaticobiliary reflex is one of postulated hypothesis behind the development of carcinoma [3]. Patients usually present with nonspecific symptom rarely it may associate with its other complications including malignancy. Early detection of malignancy is important for treatment plane and better prognosis [2], [4]. Magnetic resonance imaging (MRI) with magnetic resonance cholangiopancreatography (MRCP) is the non-invasive modality for diagnosis of choledochal cysts and associated malignancy which is able to delineate the anatomy and detection of choledochal cyst type accurately [5], [6]. Here we reported a case adult choledochal cyst associated with gall bladder malignancy.

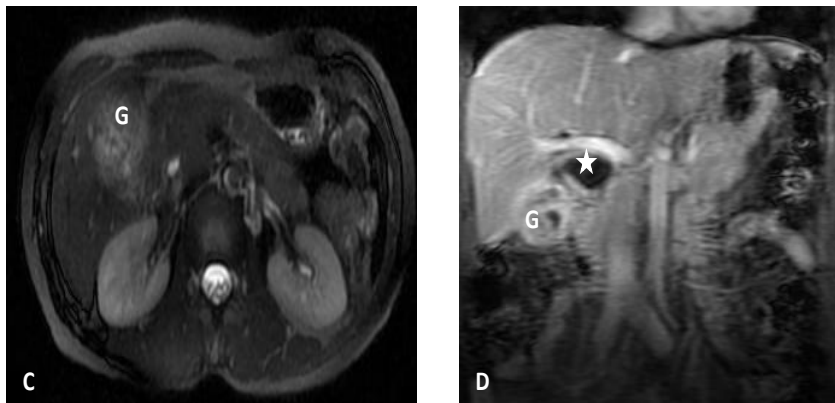
### Case Report

A 41 year old female presented with right upper quadrant abdominal pain for last 5 months and jaundice for 2 months, no associated history of nausea, vomiting and melena. Physical examination revealed icterus and tenderness in right hypochondrium. Total leukocyte count (TLC), serum bilirubin and alkaline phosphatase

were raised, however, liver enzymes were in normal limits. Ultrasound of abdomen showed ill-defined mass lesion in fundus and body of gall bladder with cholelithiasis and fusiform dilatation of common hepatic duct and cystic duct. Abdominal MRI with MRCP revealed the fusiform dilatation of common hepatic duct (10 mm) and cystic duct (17 mm) (fig.1), gall bladder showed heterogeneously enhancing mass arising from fundus and body with infiltration of adjacent liver segment. The mass was hyperintense signal intensity on T2-weighted image and hypointense on T1 weighted image with diffusion restriction on diffusion weighted imaging. Primary and secondary confluences were formed (fig. 2). No liver metastasis/ vascular invasion/ lymph nodal metastasis noted. On the basis of MRI findings the impression of type 1 choledochal cyst with gall bladder mass was made. Extended cholecystectomy with excision of choledochal cyst followed by Roux-en-Y hepatojeostomy was performed. MRI findings were consistent with operative findings. Histopathological examination showed choledochal cyst with well differentiated adenocarcinoma of gall bladder. No malignant cells were seen in excised choledochal cyst. No intra-operative or post operative complication was noted. On 7<sup>th</sup> postoperative day, patient was discharged. On follow up visits patient improved in her clinical symptoms. No recurrence detected after two year of follow.



**Fig 1:** Type 1 choledochal cyst in a case of carcinoma of gall bladder in 41 year old female. (A & B) 2 D breath hold coronal image shows a dilated and tortuous cystic duct (\*) with a wide neck insertion into the fusiform dilated common hepatic duct (white arrow). CBD distal to the opening of cystic duct is normal in caliber (red arrow).



**Fig 2:** (C) Axial T2-weighted fat-suppressed MRI at the level of body of pancreas showing hyperintense mass in gall bladder (G). (D) Post contrast LAVA coronal image shows heterogeneously enhancing mass lesion in the gall bladder fundus and body region.

### Discussion

Choledochal cysts usually present with abdominal pain, jaundice and abdominal lump in various combinations, however, sometimes it's may present in an unusual manner like hepatolithiasis, portal hypertension, and spontaneous perforation and rarely with biliary tract malignancy. A Choledochal cyst is more common in female and involves both adult and children age groups equally.<sup>[1],[2]</sup> The most common type of choledochal cysts is type I followed by type IV A [7],[8],[9],[10]. Lee et al showed types I in 68.2 % cases, while type IV A in 28.4 %<sup>[11]</sup>. Presenting complaint of adult choledochal cyst is usually different from complains which occurred in children. Triad of abdominal pain, right quadrant lump, and jaundice are common in children than adult (85 % vs 25 % respectively). Adult patients usually present as abdominal pain, jaundice, cholangitis, pancreatitis, and rarely may associated with malignant changes. Biliary malignancy is seen in 9.8 % - 30 % of cases<sup>[2],[7],[11]</sup> and it has been found that incidence increase with older age groups of patient. Cho et al, showed cholangiocarcinoma as most common malignancy associated with choledochal cyst followed by gall bladder carcinoma. Etiological factors in pathogenesis of carcinoma includes chronic inflammation, cholestasis with less protective mucin-secreting glands of the bile duct causes hyperplasia, metaplasia in cyst wall and finally carcinoma<sup>[8],[9]</sup>. Histopathologically

adenocarcinoma is the most common malignancy followed by undifferentiated carcinoma, squamous cell carcinoma, and adenosquamous carcinoma<sup>[1],[2]</sup>. Early detection and extent of malignancy is important for surgical planning and prognosis<sup>[2],[4]</sup>. Due to late presentation and early invasion, the overall 5 years survival rate reduced up to of 28.6% in associated gall bladder malignancy<sup>[2]</sup>. CT and MRI are effective modality for diagnosis of choledochal cysts and their complications. MRI feature of malignancy have been well described in literature<sup>[4],[13]</sup>. In our case, MRI accurately shows type of cyst and associated gall bladder mass which was consistent with surgical and histopathologically findings. Irregular wall thickening, polypoidal mass with post contrast heterogeneous enhancement is seen in gallbladder carcinoma and these features are delineated on MRI. Malignancies in choledochal cysts cases show similar mentioned features. Bile duct calculi, liver abscess may associate with choledochal cysts<sup>[4],[8],[9]</sup>. Detection of associated biliary abnormalities like AUPBD (Anomalous Union of the Pancreaticobiliary Duct) is important. AUPBD is more common in adult choledochal cysts (59.6%–76.2%)<sup>[1],[2],[10],[11]</sup>. MRCP is a non-invasive imaging technique for evaluation of the biliary tree that has become a preferred alternative to standard invasive procedures for diagnostic indications. The accuracy of MRCP has been increased by the use of phased-array imaging coils and breath-hold

imaging techniques. MRCP also provides excellent anatomic delineation of the gallbladder and surrounding soft-tissue structures and biliary anatomy. However, it is not useful for AUPBD detection in large choledochal cysts and calculi, like in our case, we were not able to delineate Pancreaticobiliary junction. Our patient was presented with vague complains which was not characteristics of malignancy changes. Thus, this is important to diagnose these cases in early age of patient to prevent its complication including malignant changes. Close follow up is recommended in these patients. As these two entities are rare association, so that high index of suspicion is required in patient with complaints of upper abdominal pain, jaundice and weight loss. Early detection of malignancy is important for treatment plane and better prognosis. Ultrasound is initial first line of investigation and Magnetic resonance imaging (MRI) with magnetic resonance cholangiopancreatography (MRCP) is the all in one modality for diagnosis of choledochal cysts and associated malignancy.

### Conclusion

Carcinoma gall bladder in association with choledochal cyst is very uncommon and it is also very challenging to find out any association of malignancy with choledochal cyst in early stage. However, associated carcinoma gall bladder should be included in differential diagnosis of choledochal cyst for patients of adult age group and MRCP can help in its preoperative diagnosis.

**Conflicts of interest:** All authors declare no conflicts of interest.

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