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Case Report

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Distal cholangiocarcinoma arising in a Choi Type I septate common bile duct: a case report

Santosh Thorat[1,*], Mayur Baviskar[2], Dattatray Toradmal[2], Aniket Kuldipak[2], Nikita Gaikwad[2], Nikhil Gavhane[2]

[1]HPB and Liver Transplant Surgery, Head of Department, Surgery, PGI YCMH, Pune, Maharashtra, India
[2]Department of Surgery, PCMC PGI Yashwantrao Chavan Memorial Hospital, Sant Tukaram Nagar Rd, Sant Tukaram Nagar, Pimpri Colony, Pune, Pimpri-Chinchwad, Maharashtra 411018, India

*Corresponding author. Department of Surgery, PCMC PGI Yashwantrao Chavan Memorial Hospital, Sant Tukaram Nagar Rd, Sant Tukaram Nagar, Pimpri Colony, Pune, Pimpri-Chinchwad, Maharashtra 411018, India. E-mail: drsantosh308@gmail.com

Abstract

Septate common bile duct (CBD) is a rare congenital anomaly resulting from incomplete recanalization of the primitive biliary tract. Although often asymptomatic, it predisposes to bile stasis, chronic inflammation, and potential malignant transformation. Distal cholangiocarcinoma arising in association with a septate CBD is exceptionally rare. We report the case of an 84-year-old male presenting with progressive jaundice, right hypochondrial pain, anorexia, and 20 kg weight loss over 2 months. Examination revealed icterus and a palpable non-tender mass. Laboratory studies showed marked hyperbilirubinemia (total bilirubin 11 mg/dl). Ultrasonography demonstrated a 2.5 × 2.0 cm mass in the CBD/pancreatic head region causing obstruction. Magnetic resonance cholangiopancreatography (MRCP) identified a malignant distal CBD stricture with proximal dilatation and a longitudinal septum, consistent with Choi Type I septate CBD. The patient was diagnosed with distal cholangiocarcinoma and scheduled for pancreaticoduodenectomy. Recognition of rare biliary anomalies is crucial, with high-resolution MRCP guiding safe resection and reconstruction.

Keywords **septate common bile duct, distal cholangiocarcinoma, periampullary mass, biliary obstruction, Whipple procedure**

Background

Extrahepatic bile duct tumors are rare, with the majority of these being adenocarcinomas, which are mostly well-differentiated. Tumors that develop in the biliary tree, which are carcinoid tumors, are extremely rare, representing 0.2%–2% of all digestive tract carcinoids. Primary liver carcinomas are classified as follows: hepatocellular carcinoma, cholangiocarcinoma or cholangiocellular carcinoma, and a combination of the two [1].

Embryology of the biliary tree

The biliary tree arises from the hepatic diverticulum between the fourth and eighth weeks of gestation. The cranial portion differentiates into the hepatic parenchyma and intrahepatic ducts, whereas the caudal portion gives rise to the extrahepatic biliary system and gallbladder. Initially, the primitive biliary tract is a solid cord of endodermal cells that must undergo a process of vacuolization and recanalization to form a patent lumen. A septate common bile duct (Choi Type I) represents an embryological failure of this process, where incomplete coalescence of vacuoles results in a persistent longitudinal shelf of tissue that divides the lumen [2, 3].

While previously difficult to distinguish from other periampullary pathologies, high-resolution magnetic resonance cholangiopancreatography (MRCP) now allows for the precise localization of distal cholangiocarcinoma (dCCA) and the identification of coexisting anatomical variants [4]. In this case, MRCP clearly visualized the longitudinal septum (Figs 1 and 2), which was later confirmed on macroscopic pathological evaluation (Figs 3 and 4). dCCA typically manifests as a high-grade mechanical obstruction in the distal portion of the ductal system (Fig. 5). For these patients, surgical resection (pancreaticoduodenectomy) remains the definitive therapeutic standard, necessitating meticulous preoperative mapping of the ductal anatomy to ensure R0 margins and successful reconstruction [2].

Clinical significance of septate common bile duct

A septate common bile duct is an infrequent anatomical variation which is usually symptom-free but may cause biliary complications for the affected person. The recognition of this abnormality is very important as it may cause complications during endoscopic and surgical procedures, including bile duct resection and hepaticojejunostomy. This may result in accidental injury to the duct during surgery [5].

Surgical implications

The presence of a septate common bile duct (CBD) is known to affect the overall management of distal cholangiocarcinomas. Although rou-

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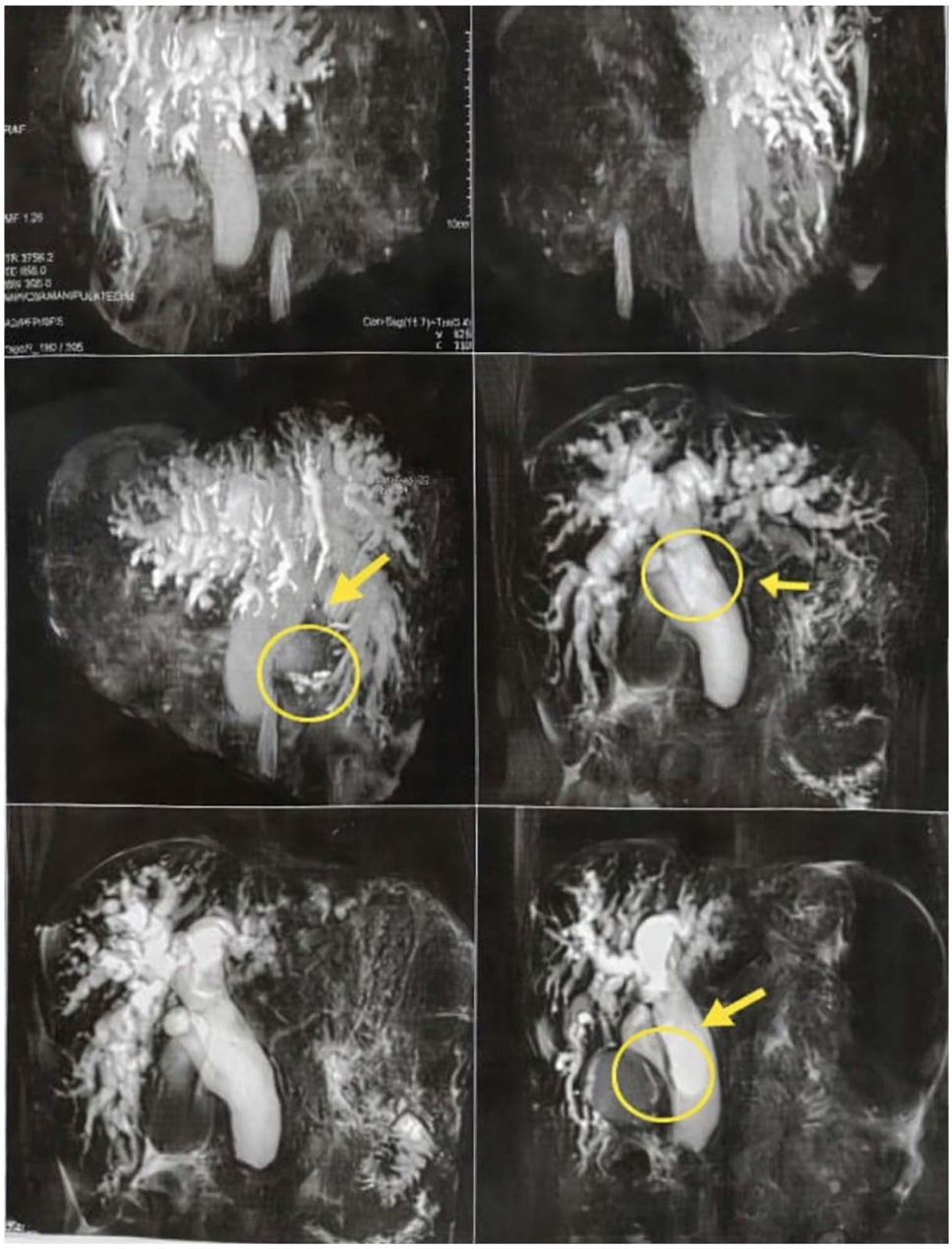

Figure 1 MRCP (coronal view)-dilated CBD with septum.

tine management modalities for distal cholangiocarcinomas include procedures like pancreaticoduodenectomy or excision of the distal CBD, a septate CBD may require additional procedures like bile duct resection or even creation of two hepaticojejunostomies [6]. Preoperative evaluation of such patients using modalities like MRCP or CT scans is critical in preventing intraoperative complications like bile leaks or failure of tumor resection [6].

Septate common bile duct is a rare but clinically significant anatomical variant. When associated with distal cholangiocarcinoma, it complicates both diagnosis and surgical management. Awareness, careful preoperative imaging, and meticulous surgical technique are paramount to optimize outcomes and prevent complications.

Case presentation

We report a case of periampullary mass in a 84-year-old male patient. Patient presented with a 2-month history of progressive jaundice, right hypochondrial pain, anorexia, and significant weight loss (20 kg). The abdominal pain was intermittent, dull aching, aggravated by food intake, and associated with vomiting, early satiety, and generalized weakness.

There was no history of fever, hematemesis, melena, or prior jaundice. Past medical history was unremarkable for diabetes, hypertension, tuberculosis, pancreatitis, or prioabdominal surgery. The patient reported a 30-year history of alcohol consumption, discontinued 1 year prior. He denied smoking or drug abuse. Physical examination revealed distended abdomen with a centrally located umbilicus and a non-tender lump measuring 4 × 4 cm in the right hypochondrium and epigastrium. Liver span was 9 cm, with no ascites. In vital signs: pulse 88 beats per minute, blood pressure 130/80 mmHg, SpO_2 98% on room air, temperature 38°C. Other systemic findings were unremarkable. Laboratory data revealed a hemoglobin concentration of 10.3 gm/dl, platelet: 263 ($10^3/\mu l$), white blood cell counts: 7.37($10^3/\mu l$), PT/INR 15.1/1.24. Blood sugar levels 95 and 82 mg/dl, HbA1c:4.6%. Liver func-

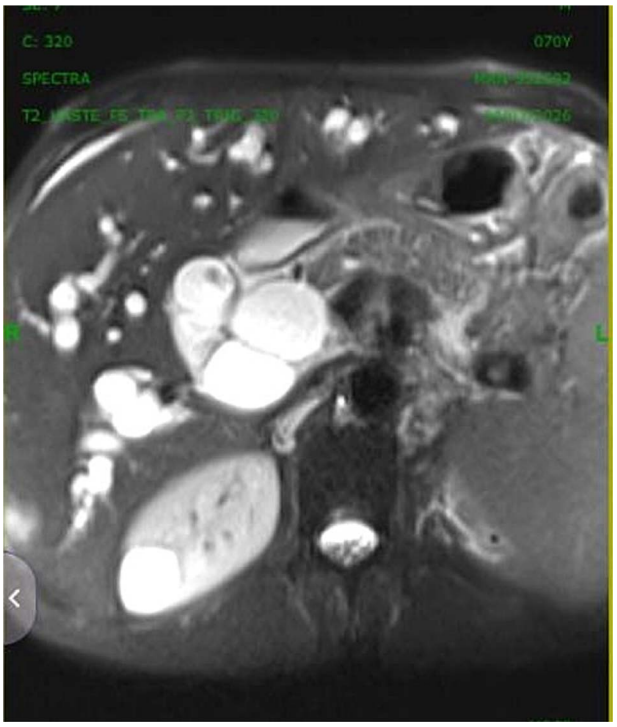

Figure 2 MRCP (axial view)-dilated CBD with septum.

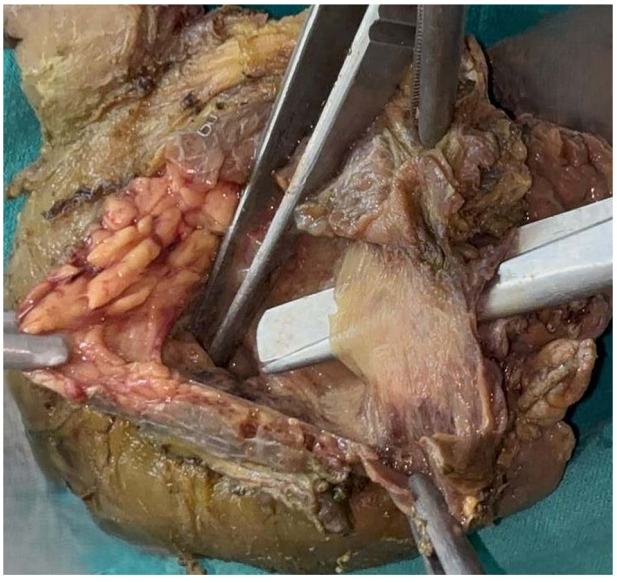

Figure 3 CBD septum.

tion tests: Total Bilirubin:11 mg/dl, Direct Bilirubin:10.5 mg/dl, Alkaline Phosphate:639 iu/l. Thyroid function tests: TSH of 1.786 μIU/ml, low T3 at 64.28 ng/dl, and normal T4. Screening for HIV, HBsAg, and HCV is negative. Renal function test:ure 8 mg/dl .reatinine 0.52 mg/dl. Electrolytes sodium 132 mmol/l and potassium 3.5 mmol/l.

Imaging findings

Ultrasonography revealed a 2.5 × 2.0 cm mass in the common bile duct and pancreatic head, causing obstruction with mild hepatomegaly. MRCP showed an enlarged liver, malignant CBD stricture, proximal and intrahepatic dilatation, pancreatic duct dilatation, and a distended gallbladder. Renal ultrasound demonstrated bilateral cysts: right 2 × 1.9 cm, left 1.7 × 1.5 cm.

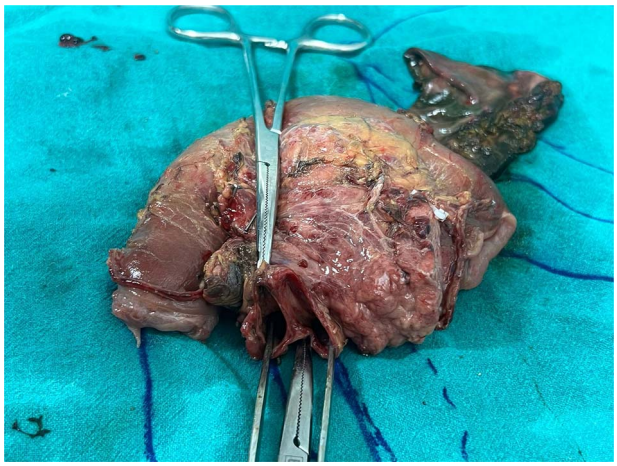

Figure 4 CBD septum seen after opening of CBD.

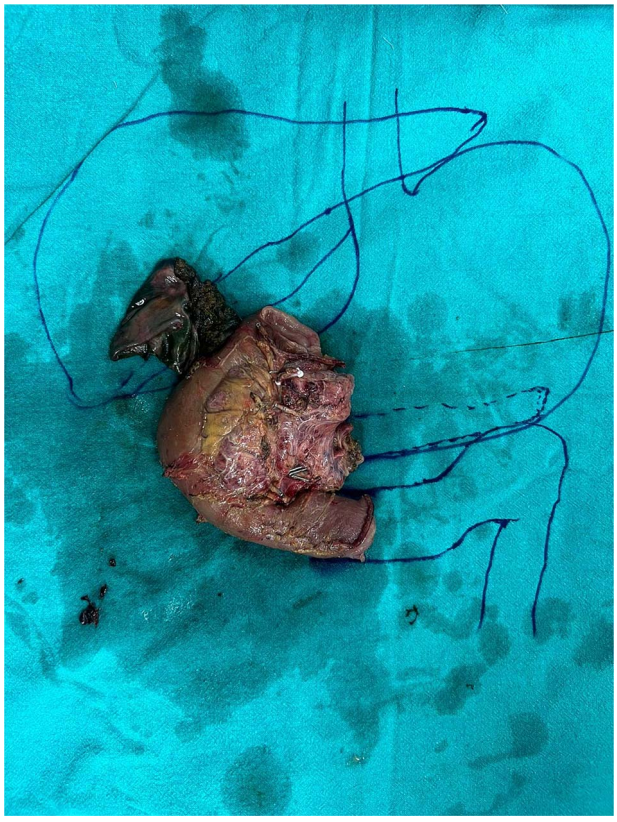

Figure 5 Distal CBD mass.

Diagnosis and management plan

The patient was diagnosed with a primary periampullary mass, consistent with distal cholangiocarcinoma, and imaging suggested a possible septate common bile duct, a rare anatomical variant. The planned procedure is a Whipple procedure (pancreaticoduodenectomy), with high-risk consent obtained.

Discussion

Anomalies of the biliary tract are rare, and among them, a septate common bile duct (CBD) is an unusual congenital anomaly. This condition results from incomplete recanalization of the primitive biliary duct during embryonic development, leading to an intraluminal septum that partially or completely divides the duct. While often silent for decades, bile flow may be impaired, causing complications.

One important mechanism linking congenital anomalies to malignancy is chronic inflammation, which can progress to dysplasia and carcinoma. In a septate CBD, bile flow is abnormal and partially obstructed, especially when sludge or stones are present. Over years, this abnormal flow causes chronic irritation of the duct lining and subclinical cholangitis. Persistent irritation promotes cell proliferation and increases the risk of genetic mutations, creating conditions favorable for malignant transformation.

In elderly patients, such as the 84-year-old male in this case, the bile duct lining may have been exposed to these changes for decades, predisposing to distal cholangiocarcinoma. The coexistence of a congenital septum and malignancy complicates diagnosis. Distal cholangiocarcinoma typically presents as focal stricture or narrowing of the distal CBD, often with proximal biliary dilatation. In this patient, ultrasonography and MRCP revealed a mass in the distal CBD and pancreatic head region, with proximal and mild intrahepatic biliary dilatation, consistent with a periampullary lesion. MRCP also suggested irregularities in the CBD, possibly due to a septate duct, which can be better assessed with high-resolution imaging.

Normally, the adult CBD measures about 5 mm in diameter and 60–80 mm in length, while the common hepatic duct has a similar diameter but variable length. A septum divides the lumen into two narrower channels, reducing effective diameter. In benign cases without malignancy, endoscopic papillary large balloon dilation (EPLBD) has been used successfully to dilate the duct and remove stones or sludge. Compared to endoscopic sphincterotomy, EPLBD carries a lower risk of complications such as bleeding.

This case underscores the importance of recognizing rare congenital biliary anomalies in patients presenting with obstructive jaundice and suspected malignancy. A septate CBD may remain asymptomatic for many years but can predispose to malignant change and complicate diagnosis. Advanced imaging modalities like MRCP are invaluable for identifying these anomalies preoperatively, allowing surgeons to anticipate and manage potential complications during surgery.

Conclusion

This case highlights the rare coexistence of a Choi Type I septate common bile duct and distal cholangiocarcinoma. The intraluminal septum, demonstrated by MRCP and confirmed pathologically, likely promoted malignant transformation through chronic biliary stasis and epithelial irritation. In patients with distal biliary obstruction and periampullary masses, high-resolution imaging is essential to differentiate congenital anomalies from tumor spread. Accurate preoperative recognition and intraoperative confirmation are critical to achieving R0 resection margins and safe biliary reconstruction, reducing risks of ductal injury or residual stasis during radical pancreaticoduodenectomy.

Author contributions

Dr. Santosh Thorat contributed to surgical management, study conceptualization, manuscript drafting, and final approval of the manuscript. Dr. Mayur Baviskar contributed to data interpretation, supervision of study design, and critical revision of the manuscript. Dr. Dattatray Toradmal contributed to patient management, and draft review and editing. Dr.Aniket Kuldipak contributed to data collection, literature search, and draft preparation support. Dr. Nikita Gaikwad contributed to data collection, literature search, and draft preparation support. Dr. Nikhil Gavhane contributed to data collection, literature search, and draft preparation support.

Conflicts of interest

The authors declare no conflict of interest.

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