An exceedingly rare case of separated drainage of the cystic duct, the common bile duct and the main pancreatic duct documented on magnetic resonance cholangiopancreatography

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Abstract

Anatomical variations of the intra- and extra-hepatic biliary system are common, including those affecting the course and insertion point of the cystic duct. Adequate knowledge of such variations and an appropriate roadmap before any surgical, endoscopic or percutaneous procedure help in preventing associated iatrogenic complications. Magnetic resonance cholangiopancreatography (MRCP) can precisely delineate the anatomy of the biliary system preoperatively. We report the case of a 72-year-old female patient who presented with chronic right upper quadrant abdominal pain of 6 months duration, which had acutely worsened over the previous 2 weeks, and no other comorbidities. Blood tests were normal. Abdominal ultrasound revealed a slight dilatation of the common hepatic duct (10 mm) but no gallstones or sludge were demonstrated. MRCP excluded choledocholithiasis but revealed a cystic duct coursing medial to the common bile duct before anastomosing at the level of the ampulla of Vater. Trifurcation (Type 2 variant) of the intra-hepatic bile system was also observed. To the best of the Authors’ knowledge, this is the very first report of this exceptionally rare anatomic cystic duct anomaly on MRCP.

Main text

Anomalies of the biliary ductal system have been appreciated by physicians and anatomists for centuries and are common findings, possibly involving both the intra-hepatic biliary ducts (IHBD) and the extra-hepatic biliary ducts (EHBD).

Normally, the cystic duct (CD) joins the common hepatic duct (CHD) from a right lateral position approximately halfway between the porta hepatitis and the ampulla of Vater.

The significance of a low CD union was first described by Eisendrath in 1918 who noted that such anatomy might predispose to inadvertent bile duct injury during cholecystectomy [1]. Reports concerning the further clinical significance of this anatomic anomaly are relatively rare, but they observed an association with gallstone pancreatitis, Mirrizi syndrome, gallbladder cancer, and cystic dilatation of the biliary duct [2].

Despite a medial insertion of CD is reported in about 18% of patients, its low insertion has been observed more infrequent and has been described in only 6-11%; in addition, the combination of these two anatomical variations is even more uncommon, presenting merely in 4-5.5% of cases [3-4]. A medial and extremely low (intraduodenal) insertion of CD is an exceedingly rare finding, with percentages ranging from 1.66% in cadaveric studies [5] to 2% in cholangiopancreatographic studies [6], and only very few cases reported in the literature. Moreover, evidence of this singular configuration is supported only by endoscopic retrograde cholangiopancreatography (ERCP) images [1,4]. Here, we report a unique case of medial and extremely low (intraduodenal) insertion of CD and, to the best of the Authors’ knowledge, the very first report of this exceptionally rare anatomic anomaly on magnetic resonance cholangiopancreatography (MRCP).
A 72-year-old female reported to our Institution with chronic right upper quadrant abdominal pain of 6 months duration, which had acutely worsened over the previous 2 weeks. She had no other comorbidities. The patient had been empirically treated with ursodeoxycholic acid prescribed by her primary care physician without improvement. Blood tests were normal, with lipase levels within normal limits and no leukocytosis. In the suspicion of biliary stones, an abdominal ultrasound (US) was requested. US revealed a slight dilatation of the CHD (10 mm), but no gallstones or sludge were demonstrated; furthermore, the gallbladder and the intrahepatic biliary ducts did not reveal any obvious abnormality. To definitively exclude choledocholithiasis and gallstones, an MRCP was then performed. The MRCP excluded the presence of any stones in the biliary system and in the gallbladder, but revealed a very rare cystic duct anomaly, demonstrating a CD coursing parallel and medial to the common bile duct (CBD) before anastomosing at the level of the ampulla of Vater; therefore, three distinct bile draining ducts were noted separately arising from the ampulla: the CD, the CBD and the main pancreatic duct (Figure 1A-C). In addition, an anatomical variation of the IHBD was also observed, with a common confluence of the right posterior duct, the right anterior duct and the left hepatic duct (i.e., trifurcation or Type 2) (Figure 1D).

The CD normally joins the CHD approximately halfway between the porta hepatis and the ampulla of Vater from a right lateral position.

However, the course and pattern of entry of the CD into the CHD can be extremely variable, depending on both the timing of the process of separation of pars hepatica (which will become the liver, the IHBD system and the CHD) from pars cystica (which will form the CD, the CBD and the gallbladder) and the grade of rotation of the CD itself induced by the rotation of the duodenum during the fetal stages. Whether these anatomical variations are due to simple stochastic events or rather represent the outcome of delays and/or alterations in the “normal” rates of differential growth in the different embryonic duct structures is still unknown. Moreover, since the EHBD and the IHBD maintain luminal continuity from the very start of organogenesis, it cannot be excluded that variations in the configuration of one system may induce or, at least, contribute to a concomitant alteration in the other system, causing some sort of “ripple effect” on the following morphogenetic phases. Nonetheless, recent evidence seems to support well-distinct mechanisms regulating the development of both the EHBD and the IHBD systems, suggesting that the molecular pathways involved in the development of the EHBD are more closely related to the formation of the duodenum and the pancreas [7]. Therefore, further studies are warranted to elucidate the mechanisms responsible for variations in the remodeling process of the biliary system during embryogenesis.

Variations in the course and insertion point of the CD are common in clinical practice and they typically remain asymptomatic throughout life. However, these anomalies still demand considerable attention during diagnostic investigations and interventional and surgical procedures since they might be the source of complications. For example, a CD with an extremely low and medial insertion has the tendency to overlie the CBD and thus be misidentified prior to interventional procedures, thus increasing the risk of inadvertent ERCP stent malposition during stone retrieval [8]. In addition, in the case of an extremely low union, the CD and the CBD may be joined by fibrous tissue, thus making clamping the CD difficult or
resulting in the inadvertent iatrogenic injury of the CD. Finally, the presence of an unusually long CD remnant after cholecystectomy or living donor liver transplantations has been associated with inflammatory changes and the formation of calculi [4].

Therefore, noninvasive techniques that can precisely delineate the anatomy of the biliary tract preoperatively could be of clinical value. In most cases, the normal-caliber CD is not seen on US or computed tomography (CT). Conversely, MRCP proved to be an accurate non-invasive imaging modality for mapping the CD anatomy and detecting aberrant ductal anatomy, relying on the high signal intensity of fluid-containing structures in T2-weighted images [3]. This technique allows to obtain high-resolution cross-sectional imaging, with both two- and three-dimensional projection, and its results are comparable to the more invasive ERCP and intraoperative cholangiograms. Moreover, MRCP is quick, is not associated with radiation exposure and does not require the administration of contrast media, needing only a simple glass of juice to evaluate the anatomy of both the intra- and extrahepatic biliary ducts [9]. Finally, it allows the synchronous evaluation of the IHBD, whose anatomical variations are often associated with variations of the downstream biliary system, as was demonstrated also in the present case [7].

Despite the potential surgical complications associated with these rare congenital malformations, CD aberrations have been demonstrated to predispose to several pathologic pancreaticobiliary conditions. For example, a low union of the CD has previously been associated with a higher risk of pancreatitis and even periampullary cancer and this association was ascribed to an anatomy-induced pressure process leading to chemical/bilious irritation  [2]; this association is probably supported also by the shorter and lesser exposure of the biliary epithelium to the protective or dilutional effect of peri-biliary mucus glands in the upper biliary tree which happens in the presence of a low union [1]. In addition, previous studies have found an association between low insertion of the CD and choledocholithiasis [10] which can be due to increased retrograde pressure in the CD leading to both bile stagnation and impaired gallbladder emptying. In the present case, however, no calculi were detected on both US and MRCP. Despite being contradictory to previous reports, the absence of biliary stones in the present case may be explained by differences in terms of biochemical and clinical contributors to stone formation. At the same time, the chronic abdominal pain reported by the patient could be attributed to unusual bile reflux into the CD or the pancreatic duct, likely favored by the relative proximity of these canals and the increased intra-luminal pressure at the level of the anastomosis. This hypothesis could also justify the evidence of the slightly wider CHD diameter reported on US and MRCP.

A CD coursing parallel and medial to the CBD before anastomosing at the level of the ampulla of Vater is an extremely rare anatomical variant of the biliary system that, if unrecognized, may complicate both surgical and interventional procedures. In an era of ever-increasing laparoscopic, endoscopic and percutaneous procedures of the hepatobiliary system, it is extremely important to have adequate knowledge of such variations before the actual procedure is performed. MRCP is pivotal to correctly identifying anatomic variants of both the IHBD and the EHBD, whose association is not unusual.
Abbreviations

IHBD: intra-hepatic biliary ducts; EHBD: extra-hepatic biliary ducts; CD: cystic duct; CHD: common hepatic duct; ERCP: endoscopic retrograde cholangiography; MRCP: magnetic resonance cholangiopancreatography; US: ultrasound; CBD: common bile duct; CT: computed tomography.

Declarations

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References


Figures
Figure 1

MRCP images demonstrating a cystic duct (CD, green outline, 1B) coursing parallel and medial to the common bile duct (CBD, red outline 1B) before anastomosing at the level of the ampulla of Vater with the main pancreatic duct (yellow outline, 1B); no stones were detected in both the gallbladder and the extra-hepatic biliary system. Oriented MRCP image of the same patient demonstrating the concomitant presence of Type 2 variant (trifurcation) of the intra-hepatic bile duct (red arrow, 1D).