Atypical IVA Hepatic Duct Cyst Mimicking Biliary Atresia: The Critical Role of Multimodal Imaging and its Diagnostic Pitfalls

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DISCLOSURES
The authors declare that they have no financial or non-financial competing interests to disclose.

CONSENT
Yes.

HUMAN AND ANIMAL RIGHTS
The procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation and with the Helsinki Declaration of 1975, as revised in 2000.

ABSTRACT

Biliary tree cysts are rare congenital anomalies, characterized by cystic dilatation of intrahepatic and/or extrahepatic bile ducts. Their incidence is approximately 1 case in 100,000-150,000 live births in western populations, notably more frequent in east Asia. Although most cases are diagnosed in the early childhood, up to 30% of the cases present in the neonatal period, often manifesting with prolonged jaundice as a nonspecific clinical feature. Their prompt differentiation from cystic biliary atresia is critical, due to its different management and prognosis.

We present the case of a 2-day old male neonate with conjugated hyperbilirubinemia, jaundice and abdominal distension. Ultrasonography revealed a cystic lesion distinct from the gallbladder, and further abdominal MRI and MRCP confirmed an atypical type IVA right hepatic duct cyst, without extrahepatic duct involvement. The case posed a diagnostic challenge due to overlapping clinical features with cystic biliary atresia. Early recognition of the accurate diagnosis, allowed for timely surgical planning, preventing complications as cholangitis, pancreatitis, and potential long-term malignant transformation.

This case accentuates the importance of maintaining broad differential diagnosis in neonatal cholestasis, and demonstrates how integrated imaging can accurately distinguish hepatic duct cysts from cystic biliary atresia. Moreover, early surgical referral is essential to mitigate long-term hepatobiliary morbidity.

CASE REPORT

BACKGROUND

Biliary tree cysts are rare congenital malformations, occurring in approximately 1 in 100,000–150,000 live births in Western populations and more frequently in East Asia. Although typically diagnosed in infancy or early childhood, up to 30% present in the neonatal period, often with nonspecific jaundice. Differentiating these cysts from cystic biliary atresia is critical, given distinct surgical timelines and prognoses. We report a rare atypical Todani type IVA right hepatic duct cyst in a 2-day-old male neonate, successfully distinguished from cystic biliary atresia through integrated ultrasonography and MRCP, enabling timely surgical management and prevention of long-term hepatobiliary morbidity.

CASE REPORT

Biliary tree cysts pose a rare congenital malformation, characterized by abnormal cystic dilatation of the intrahepatic and/or extrahepatic bile ducts [1]. Their incidence is estimated to be of 1 in 100,000-150,000 live births in western countries, yet are significantly more prevalent in east Asia, particularly in Japan, where their incidence may reach 1 in 1,000 live births [2,3]. Biliary duct cysts have a striking female predominance with a 4:1 ratio [4]. Although biliary duct cysts commonly present during infancy and early childhood, up to 20–30% manifest in the neonatal period, often with conjugated hyperbilirubinemia, jaundice, abdominal distension, or a palpable right upper quadrant mass [5]. The classical triad of jaundice, abdominal pain, and palpable mass is well described in literature, but rarely encountered in real clinical practice [6]. From the pathophysiological view, the most widely accepted mechanism involves an anomalous pancreaticobiliary ductal junction (APBDJ), permitting reflux of pancreatic enzymes into the bile ducts, leading to chronic inflammation of biliary tree, wall weakness with progressive cystic dilatation, and cyst formation [7-9].

The bile duct cysts are classified according to the Todani modification of the Alonso-Lej system, which to date delineates five principal types [10]. Type I biliary cysts are fusiform or cystic dilatation of the extrahepatic bile duct, representing 80–90% of cases, which by far are the most common type. Less frequently encountered bile duct cysts are: type II manifested as diverticula or outpouching of the extrahepatic bile duct, type III known as choledochoceles confined to the intraduodenal portion of the common bile duct, type IV characterized with multiple intrahepatic and extrahepatic cysts, and type V or Caroli disease that presents with multiple, saccular dilatations of the intrahepatic bile ducts [11]. Early differentiation of bile duct cysts from other causes of neonatal cholestasis, particularly biliary atresia is critical, given vastly different management pathways and prognoses [12].

This case is particularly noteworthy, while it highlights a neonatal right hepatic duct cyst with clinical features overlapping with biliary atresia, creating a significant diagnostic challenge. In neonates, distinguishing these two entities is vital, while biliary atresia demands urgent hepatoportoenterostomy in order to prevent irreversible cirrhosis [13], whereas hepatic duct cysts require complete excision and biliary reconstruction, typically via a Roux-en-Y hepaticojejunostomy to avoid long-term risks [14]. Nonetheless, complications of untreated hepatic duct cysts are debilitating, including recurrent cholangitis, pancreatitis, spontaneous rupture of the cyst with biliary peritonitis, ductal strictures leading to secondary biliary cirrhosis, portal hypertension, and an estimated 10–30% lifetime risk of malignant transformation into cholangiocarcinoma [15,16], underscoring the importance of prompt and accurate diagnosis.

Reporting this case adds significant value to the existing literature, by emphasizing the critical role of a multimodal diagnostic approaches in neonates, where high-resolution ultrasonography remains the first-line modality, capable of visualizing cystic dilatations separate from the gallbladder [17,18], while magnetic resonance imaging and magnetic resonance cholangiopancreatography offer superior anatomical delineation without ionizing radiation, obviating the potential need for invasive cholangiography [19,20]. This case also emphasizes the necessity of correlating radiological findings with clinical and laboratory parameters, contributing to accurate diagnosis and surgical management.

Therefore, this case report aims to illustrate the importance of maintaining a broad differential diagnoses in cases of neonatal obstructive jaundice, leveraging complementary imaging modalities to accurately distinguish hepatic duct cysts from biliary atresia, and underscores the imperative for early surgical intervention, to mitigate the risk of progressive hepatobiliary morbidity and malignant transformation.

The diagnosis and management of the neonatal biliary cyst in this case were guided by a comprehensive, multimodal imaging approach. Initial high-resolution ultrasonography was performed, which revealed a well-circumscribed anechoic cystic structure at the porta hepatis. Subsequent magnetic resonance imaging (MRI) and magnetic resonance cholangiopancreatography (MRCP) were conducted to delineate biliary anatomy and evaluate ductal continuity. Imaging confirmed a cystic dilation involving the right hepatic duct without extrahepatic duct involvement, consistent with an atypical Todani type IVA biliary cyst. Surgical excision of the cyst and a Roux-en-Y hepaticojejunostomy were performed at two weeks of age. Histopathological evaluation of the cyst wall confirmed the diagnosis. Due to the descriptive nature of the case, no statistical analyses were performed.

Ethical approval for publication of this single-patient case report was waived by the Institutional Review Board of our institution, as it does not involve experimental research or generalizable data collection. The procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation and with the Helsinki Declaration of 1975, as revised in 2000. Written informed consent for publication, including clinical data and accompanying images, was obtained from the patient's legal guardian. All patient identifiers have been removed to ensure anonymity.

We present the case of a full-term male neonate, born via spontaneous vaginal delivery, with an uneventful perinatal history, that started manifesting jaundice on the second day of life. The baby exhibited progressive yellow discoloration of the skin and sclera, accompanied by mild abdominal distension, but there was no vomiting, feeding intolerance, or signs of inflammation. There was no prior family history of liver disease, congenital anomalies, or consanguinity. On physical examination, the infant was icteric, mildly distended but non-

tender abdomen, with no palpable abdominal mass. There was no hepatosplenomegaly encountered, and other systemic examinations were unremarkable.

Initial laboratory investigations revealed conjugated hyperbilirubinemia, with a total serum bilirubin of 8.4 mg/dL (direct fraction 5-5.9 mg/dL), mildly elevated transaminases, and normal synthetic liver function. Infectious and metabolic screens were unremarkable. Given the early onset of direct hyperbilirubinemia, an abdominal ultrasonography was performed, which demonstrated a well-circumscribed, anechoic cystic structure measuring approximately 4 cm in diameter, located at the porta hepatis and distinctly separate from the gallbladder. The gallbladder was typical on its size and shape, with a clear lumen, without any obstructions or filling defects.

To further delineate the anatomy and evaluate biliary continuity, magnetic resonance imaging (MRI) and magnetic resonance cholangiopancreatography (MRCP) were performed. Initially magnetic resonance imaging revealed a cystic dilation involving the right hepatic duct consistent with an atypical Todani type IVA biliary cyst, without extrahepatic ductal involvement, as documented in axial and coronal images presented under figures 1 and 2.

Further, with magnetic resonance cholangiopancreatography the cyst was shown to communicate with the common hepatic duct distally, ruling out cystic biliary atresia. And there were no pancreatic duct anomalies identified, as presented in figure 3.

Based on the clinical presentation, laboratory parameters, and imaging findings, a diagnosis of neonatal type IVA right hepatic duct cyst was established. The patient underwent elective surgical management with complete excision of the cyst and Roux-en-Y hepaticojejunostomy at two weeks of age. Intraoperative findings confirmed a large cystic dilation of the right hepatic duct with a normal-appearing gallbladder and no evidence of extrahepatic ductal involvement. Histopathology of the excised cyst wall showed fibrosis with inflammatory cell infiltration, consistent with right hepatic duct cyst. The postoperative course was uneventful. The infant's jaundice resolved progressively over the following weeks, with normalization of bilirubin levels and liver enzymes on follow-up. To date, the patient remains asymptomatic with appropriate growth and no evidence of recurrent cholestasis.

DISCUSSION

In this case report, we describe a 2-day old male neonate presenting with conjugated hyperbilirubinemia, jaundice and progressive abdominal distension, where ultrasonography imaging revealed a large cystic intrahepatic dilatation, which was further confirmed with MRCP as an atypical Todani type IVA right hepatic duct cyst. The diagnosis was established through a combination of high-resolution abdominal ultrasonography, which demonstrated a cystic mass distinct from the gallbladder, and MRCP, which confirmed the anatomical continuity with the biliary tree, and excluded extrahepatic ductal involvement. This early identification enabled prompt surgical referral and definitive management, reducing the risk of complications frequently associated with delayed diagnosis.

Etiology & demographics

Hepatic duct cysts pose a sparse congenital anomaly of the biliary tract, with an incidence ranging from 1 in 100,000-150,000 live births in western countries, though significantly higher in east Asian populations [22]. Various studies emphasize the predominance in females, with a female-to-male ratio of approximately 4:1 [23]. Our case involves a male neonate, adding to the subset of less commonly reported clinical presentations of biliary duct cysts in male infants.

Clinical & imaging findings

In terms of classification, the majority of biliary duct cysts are type I with a prevalence of 80–90%, manifesting as dilatation of the entire extrahepatic bile duct [24]. Yet, in our case we present a much rarer atypical Todani IVA hepatic duct cystic dilation, with no extrahepatic involvement [21,25]. Contrary, a typical IVA Todani classification would manifest with both intrahepatic and extrahepatic biliary duct dilation, which was not the case with our neonate. Therefore, this atypical Todani IVA variant observed in this case, poses significant and novel addition to the current research in this field. In addition, the age at presentation within 2-day of the neonatal period, places our case among a minority of significant clinical cases [26].

Furthermore, the classical triad of jaundice, palpable right upper quadrant mass, and abdominal pain, first described by Alonso-Lej et al., is rarely seen in full in neonates [11]. Indeed, as reported by Singham et al., most infants present with jaundice alone, while older children more frequently exhibit pain and intermittent cholangitis [24,27]. Thus, our case aligns with the more common neonatal presentation of isolated persistent jaundice without abdominal pain. Radiologically, the findings in our patient were of type IVA cysts with a well-circumscribed cystic mass in the porta hepatis, separated from the gallbladder, with no associated of extrahepatic ductal dilatation. Interestingly, we found no evidence of multifocal intrahepatic cysts involvement, effectively excluding Caroli disease [28].

Although biliary tree cysts are recognized as a cause of obstructive jaundice in infants, their presentation in the neonatal period is relatively uncommon, reported only in approximately 20–30% of cases [29], and their clinical presentation often overlaps clinically and biochemically with biliary atresia, an entity requiring urgent intervention to preserve hepatic function to prevent cirrhosis [30]. In the neonatal period, the principal differential diagnosis for persistent direct hyperbilirubinemia with bile duct dilatation is cystic biliary atresia, a progressive fibro-inflammatory obliteration of the extrahepatic biliary tree. Biliary atresia may present with a small, fibrotic gallbladder

and non-visualization of the common bile duct on ultrasound. However, an overlap can exist, particularly in early stages [31]. Some neonates with biliary atresia may even demonstrate transient cystic dilatation, termed "cystic biliary atresia," complicating the differentiation. Yet, critical imaging features that favored the diagnosis of a right hepatic duct cyst in our case included: a large cystic mass (>1.5 cm), clearly separated from the gallbladder, normal intrahepatic bile ducts (ruling against Caroli disease), and preservation of normal gallbladder morphology and contractility on feeding [32]. In contrast, in cystic biliary atresia we would have seen the triangular cord sign, an echogenic fibrotic cone anterior to the portal vein, along with absent or irregular gallbladder morphology [33]. Such findings were absent in our patient. Thus, this case underscores the pivotal role of imaging in the differential diagnosis of neonatal cholestasis. Further, MRCP provided non-invasive confirmation of the cyst's continuity with the biliary tree, solidifying the diagnosis and avoiding invasive intraoperative cholangiography as the sole diagnostic tool. Other less likely differentials would include cystic biliary hamartomas, mesenteric or duplication cysts, and cystic neuroblastoma, though these pathologies typically lack direct biliary communication [34].

This case highlights the complementary roles of ultrasonography and MRCP in the evaluation of neonatal jaundice and cholestasis. Ultrasonography is universally regarded as the first-line modality given its accessibility, lack of radiation, and excellent resolution for evaluating biliary anatomy. In our patient, ultrasound effectively demonstrated the cystic mass and its anatomical separation from the gallbladder, raising suspicion for a biliary tree cyst. MRCP then served as the gold standard non-invasive modality, providing high-resolution delineation of the biliary tree, confirming the cystic dilation of the right hepatic duct without multi-centric involvement, and demonstrating continuity with the common hepatic duct. MRCP avoids ionizing radiation and was instrumental in differentiating from cystic biliary atresia, which may require urgent porto-enterostomy. While hepatobiliary scintigraphy could have been considered as a diagnostic option to evaluate bile excretion into the intestine, it is generally less helpful in distinguishing between these structural anomalies, and would not have replaced the detailed anatomical mapping provided by MRCP [35].

Treatment & prognosis

The standard of care for hepatic duct cysts is complete cyst excision with biliary reconstruction, most commonly via Roux-en-Y hepatico-jejunostomy, which was performed in our patient [36]. Early surgical intervention is essential to mitigate the risks of recurrent cholangitis, secondary biliary cirrhosis, pancreatitis, cyst rupture, and ultimately malignant transformation [37]. The lifetime risk of cholangiocarcinoma in untreated hepatic duct cysts is estimated between 10–30%, increasing with age, further underscoring the importance of prompt diagnosis and treatment. Postoperatively, most patients have excellent outcomes, though lifelong surveillance is recommended due to the potential for anastomotic strictures or late neoplasia.

Differential Diagnoses

The differential diagnosis for a hepatic duct cyst includes choledocholithiasis, in which intraductal calculi cause biliary dilatation and obstructive jaundice; biliary atresia, particularly in infants, where progressive fibrosis and obliteration of the extrahepatic bile ducts may mimic cystic dilatation; and Caroli's disease, a congenital disorder characterized by segmental, saccular dilatation of the intrahepatic bile ducts. Other considerations include biliary stricture due to prior inflammation, surgery, or trauma; biliary cystadenoma and biliary cystadenocarcinoma, which present as cystic masses arising from the biliary epithelium; and pancreaticobiliary maljunction, a congenital anomaly that can lead to bile duct dilatation in the absence of a true cyst.

This case illustrates an atypical Todani type IVA right hepatic duct cyst, presenting in the neonatal period, underscoring important considerations in the differential diagnosis of neonatal cholestasis. It emphasizes the critical role of integrated imaging, initial ultrasonography complemented by MRCP, in accurately characterizing the biliary anatomy and distinguishing hepatic duct cysts from cystic biliary atresia, thereby guiding appropriate and timely surgical management.

This case highlights important learning points as: hepatic duct cysts, although rare, must be considered in neonates with persistent jaundice and a cystic porta hepatis mass. Also, differentiating biliary tree cysts from cystic biliary atresia is crucial, given divergent surgical timelines and prognoses. And finally, multimodal imaging, particularly the combination of US and MRCP, plays a pivotal role in diagnosis, surgical planning, and prevention of long-term complications.

TEACHING POINT

Hepatic duct cysts are rare congenital anomalies that may present in the neonatal period with features overlapping other causes of cholestasis, such as cystic biliary atresia. Multimodal imaging, particularly high-resolution ultrasonography and MRCP, is essential for accurately delineating biliary anatomy, confirming cyst–biliary communication, and guiding timely surgical management to prevent long-term hepatobiliary complications.

QUESTIONS

Question 1: Which of the following is NOT a common complication of untreated hepatic duct cysts?

a. Recurrent cholangitis
b. Secondary biliary cirrhosis
c. Spontaneous cyst rupture
d. Acute appendicitis (applies)
e. Malignant transformation into cholangiocarcinoma

Explanation:

Untreated hepatic duct cysts may lead to recurrent cholangitis, secondary biliary cirrhosis, pancreatitis,

spontaneous rupture with biliary peritonitis, and a lifetime risk of malignant transformation (10–30%). Acute appendicitis is unrelated to hepatic duct cyst pathology. ["Complications of untreated hepatic duct cysts are debilitating, including recurrent cholangitis, pancreatitis, spontaneous rupture of the cyst with biliary peritonitis, ductal strictures leading to secondary biliary cirrhosis, portal hypertension, and an estimated 10–30% lifetime risk of malignant transformation into cholangiocarcinoma"].

Question 2: Which imaging modality is considered the first-line investigation for suspected hepatic duct cysts in neonates?

a. MRCP
b. MRI
c. Ultrasound (applies)
d. CT scan
e. PET-CT

Explanation:

High-resolution ultrasonography is the first-line modality for suspected hepatic duct cysts in neonates due to its accessibility, lack of radiation, and ability to visualize cystic structures and their relationship to the gallbladder. ["High-resolution ultrasonography remains the first-line modality, capable of visualizing cystic dilatations separate from the gallbladder"].

Question 3: Which of the following findings favors the diagnosis of a hepatic duct cyst over cystic biliary atresia?

a. Large cystic mass >1.5 cm (applies)
b. Triangular cord sign
c. Small, fibrotic gallbladder
d. Absent gallbladder contractility
e. Echogenic fibrotic cone anterior to the portal vein

Explanation:

A large cystic mass (>1.5 cm), distinct from the gallbladder with preserved gallbladder morphology and contractility, favors the diagnosis of a hepatic duct cyst. Cystic biliary atresia often shows the triangular cord sign, a small fibrotic gallbladder, and absent contractility. ["Critical imaging features… included: a large cystic mass (>1.5 cm), clearly separated from the gallbladder… and preservation of normal gallbladder morphology and contractility"].

Question 4: According to the Todani classification, type IVA cysts are typically characterized by:

a. Multiple intrahepatic cysts only
b. Extrahepatic fusiform dilatation only
c. Multiple intrahepatic and extrahepatic cysts (applies)
d. Diverticulum of the common bile duct
e. Choledochocele confined to the intraduodenal portion of the CBD

Explanation:

Type IVA cysts usually present with multiple intrahepatic and extrahepatic cystic dilatations. The presented case was atypical because it involved only the right hepatic duct without extrahepatic involvement. ["Type IV characterized with multiple intrahepatic and extrahepatic cysts… Contrary, a typical IVA Todani classification would manifest with both intrahepatic and extrahepatic biliary duct dilation"].

Question 5: What is the standard surgical treatment for hepatic duct cysts?

a. Percutaneous drainage
b. Liver transplantation
c. Endoscopic retrograde cholangiopancreatography (ERCP) with stenting
d. Complete cyst excision with biliary reconstruction (applies)
e. Observation and periodic ultrasound monitoring

Explanation:

The gold standard treatment for hepatic duct cysts is complete cyst excision with biliary reconstruction, most often via Roux-en-Y hepaticojejunostomy. This approach reduces the risk of complications and malignant transformation. ["The standard of care for hepatic duct cysts is complete cyst excision with biliary reconstruction, most commonly via Roux-en-Y hepatico-jejunostomy"].

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FIGURES

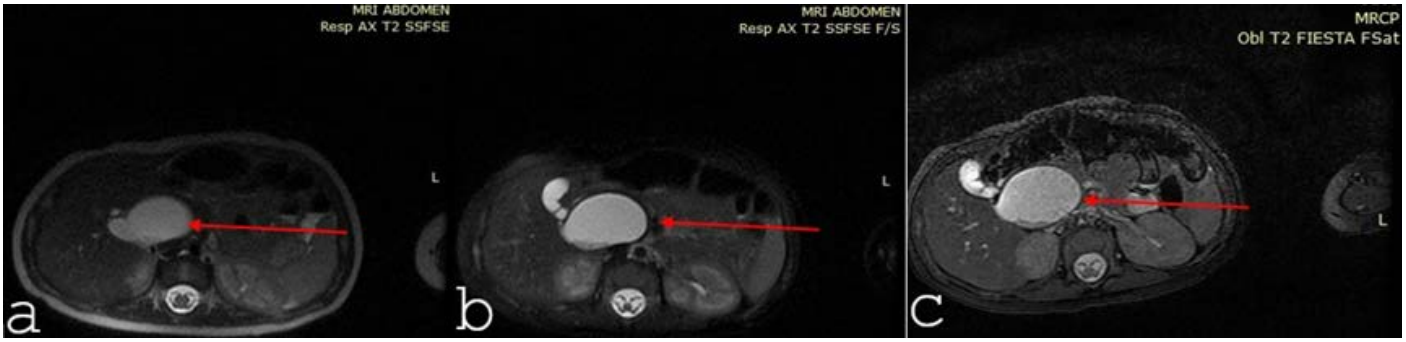

Figure 1: A 2-day old male neonate with atypical type IVA right hepatic duct cyst. Axial magnetic resonance imaging T2-weighted Single-Shot Fast Spin Echo (a) and fat saturation (b, c) images, demonstrating a 30 × 38 mm cystic lesion arising from the right hepatic duct, well differentiated from the adjacent gallbladder (red arrows).

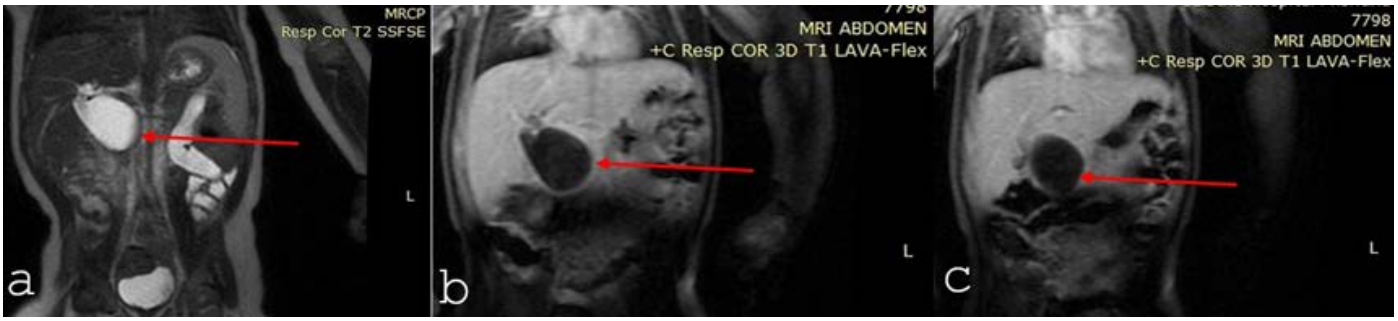

Figure 2: A 2-day-old male neonate with an atypical type IVA right hepatic duct cyst. Coronal magnetic resonance imaging T2-weighted Single-Shot Fast Spin Echo (a) and contrast enhanced images (b, c) demonstrate a 30 × 38 mm cystic lesion arising from the right hepatic duct, clearly differentiated from the adjacent gallbladder (red arrows).

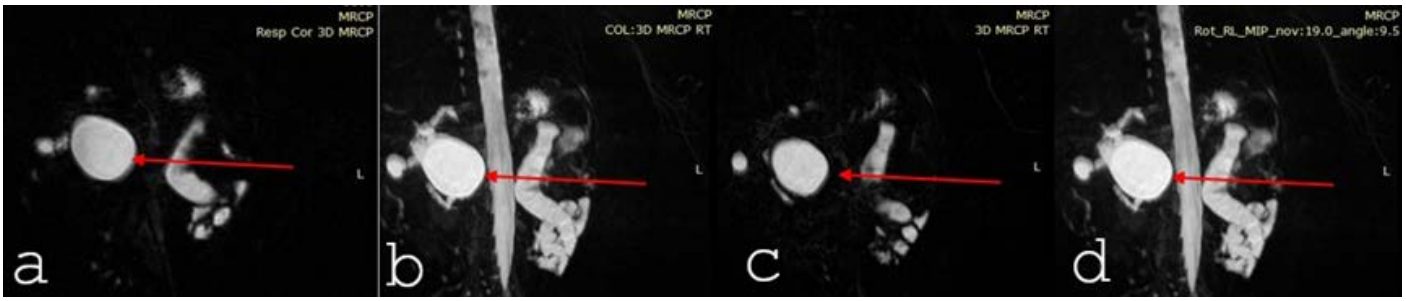

Figure 3: A 2-day-old male neonate with an atypical type IVA right hepatic duct cyst. Coronal magnetic resonance cholangiopancreatography images (a-d), demonstrate a 40 × 35 mm cystic dilation of the right hepatic duct, clearly distinct from the adjacent gallbladder and continuous with the intrahepatic biliary tree (red arrows).

KEYWORDS

Hepatic duct cyst; cystic biliary atresia; Todani IVA; magnetic resonance cholangiopancreatography; magnetic resonance imaging; ultrasonography.

ABBREVIATIONS

APBDJ = Anomalous Pancreaticobiliary Ductal Junction
BA = Biliary Atresia
CBD = Common Bile Duct
CHD = Common Hepatic Duct
CRP = C-Reactive Protein
CT = Computed Tomography
HD = Hepatic Duct
IV = Intra Venous
MRCP = Magnetic Resonance Cholangiopancreatography
MRI = Magnetic Resonance Imaging
RUQ = Right Upper Quadrant
US = Ultra Sonography

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