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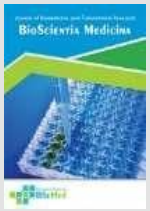

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A Rare Presentation of Choledochal Cyst in a Six-Month-Old Infant: Diagnostic and Surgical Challenges

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A B S T R A C T

Background: Choledochal cysts (CCs) are rare congenital anomalies of the biliary tree, characterized by cystic dilation of the intrahepatic and/or extrahepatic bile ducts. While the classic triad of abdominal pain, jaundice, and a palpable abdominal mass is well-described, the presentation in infants can be subtle and often involves diagnostic and surgical challenges. This case report describes a rare presentation of a Todani Type 1 choledochal cyst in a six-month-old infant who presented primarily with abdominal distension, highlighting the diagnostic pathway and surgical management. **Case presentation:** A six-month-old female infant presented with a two-month history of progressive abdominal distension. There was a history of pale stools at two months of age that lasted for one week. Physical examination revealed a well-nourished infant with icteric skin and sclera and a distended abdomen with a palpable, mobile mass measuring 7x5 cm. Laboratory investigations revealed hyperbilirubinemia and elevated liver enzymes. Abdominal ultrasound and subsequent CT scan with contrast confirmed the presence of a Todani Type 1 choledochal cyst. The infant underwent successful surgical excision of the cyst and Roux-en-Y hepaticojejunostomy. The postoperative period was uneventful, and the patient was discharged in stable condition. Follow-up at one month showed good recovery and no signs of complications. **Conclusion:** This case highlights an atypical presentation of a Todani Type 1 choledochal cyst in a young infant, where the primary symptom was abdominal distension rather than the classic triad. Early diagnosis through imaging modalities like ultrasound and CT scan, followed by complete surgical excision and Roux-en-Y reconstruction, resulted in a favorable outcome. This case underscores the importance of considering choledochal cysts in the differential diagnosis of abdominal distension in infants, even in the absence of jaundice or pain.

1. Introduction

Choledochal cysts (CCs) are rare congenital anomalies that affect the biliary tree. These anomalies are characterized by cystic dilatations, which can occur in the intrahepatic or extrahepatic bile ducts, or both. The incidence of choledochal cysts is estimated to be between 1 in 100,000 and 150,000 live births in Western populations. Notably, a higher prevalence has been reported in Asian countries. The classification system developed by Todani et al. is the most widely used for categorizing CCs. This system classifies choledochal cysts into five types, based on their anatomical location and morphology. Type I cysts are the most common, accounting for 80-90% of cases. These cysts involve fusiform or saccular dilatation of the extrahepatic bile duct.[1-3]

The underlying cause of choledochal cysts remains unclear, but the most widely accepted theory centers on an abnormal pancreaticobiliary duct junction (APBDJ). This theory, proposed by Babbitt, suggests

that APBDJ is a primary factor in the development of choledochal cysts. In APBDJ, the pancreatic duct joins the common bile duct outside the sphincter of Oddi, creating a long common channel. This anatomical variation allows for the reflux of pancreatic enzymes into the bile duct. The reflux of pancreatic enzymes can lead to inflammation and weakening of the bile duct wall, which can eventually result in cyst formation. The classic clinical presentation of choledochal cysts includes a triad of symptoms: abdominal pain, jaundice, and a palpable abdominal mass. However, this classic triad is more frequently observed in older children and adults. The presentation of choledochal cysts in infants can be more varied. Infants may present with symptoms such as jaundice, feeding difficulties, vomiting, and abdominal distension. The diagnosis of choledochal cysts in infants can be particularly challenging. This difficulty arises from the rarity of the condition and the non-specific nature of the symptoms that may manifest.[4-7]

Early diagnosis and surgical management are of paramount importance in cases of choledochal cysts. Prompt intervention is necessary to prevent the development of complications. Potential complications include cholangitis, pancreatitis, biliary lithiasis, and an increased long-term risk of cholangiocarcinoma. The preferred surgical treatment for Type I choledochal cysts involves complete excision of the cyst. Following cyst excision, a Roux-en-Y hepaticojejunostomy is performed to restore biliary-enteric continuity. This surgical approach aims to achieve complete removal of the cyst, thereby mitigating the risk of malignant transformation and preventing the recurrence of complications.[8-10] This case report describes an unusual presentation of a Todani Type 1 choledochal cyst in a six-month-old female infant. In this case, the infant presented primarily with progressive abdominal distension.

2. Case Presentation

The patient, a six-month-old female infant, presented to the medical facility with a primary complaint of progressive abdominal distension. This distension had been observed for a duration of two months, indicating a gradual and evolving nature of the symptom. In addition to the primary complaint of abdominal distension, the patient's medical history revealed an episode of pale stools occurring approximately one week at two months of age. This particular symptom had resolved spontaneously. Furthermore, there was a reported decrease in appetite noted in the two weeks leading up to the patient's admission. A recent onset of slight yellow discoloration of the skin and eyes was also documented, suggesting the development of jaundice, albeit not as the primary presenting symptom. It is important to highlight the absence of certain symptoms in this patient. There was no reported occurrence of fever, vomiting, poor feeding, or significant jaundice initially. The absence of these symptoms is noteworthy as they are often associated with gastrointestinal or hepatobiliary disorders in infants. The lack of significant initial jaundice is particularly relevant, given the eventual diagnosis of a choledochal cyst, a condition frequently characterized by jaundice. Regarding the patient's past medical history, the birth history was significant for a term birth via spontaneous vaginal delivery. There were no reported perinatal complications associated with the birth, indicating an uncomplicated delivery process. The patient's growth and development were reported to be appropriate for her age until the onset of the presenting symptoms. This suggests that the patient had been developing normally prior to the emergence of the abdominal distension and associated symptoms. The family history was notable for the absence of any known family history of choledochal cysts or other congenital anomalies. This absence of a positive family history for similar conditions is relevant in considering the potential etiology of the patient's condition, although the etiology of choledochal cysts is often multifactorial and not strictly hereditary. Upon physical examination, the patient's general appearance was described as alert and interactive. The patient was also assessed as well-nourished,

indicating adequate nutritional status despite the reported decrease in appetite. The patient's weight was recorded as 7.5 kg, which was reported to be within the normal range for the patient's age. The patient's length was measured at 65 cm, also considered within the normal range for the patient's age. These anthropometric measurements suggest that the patient's overall growth was progressing within expected parameters. A more focused examination revealed icteric skin and sclera. Icterus, or jaundice, is the yellow discoloration of the skin and sclera (the white part of the eye) caused by elevated levels of bilirubin in the blood. The presence of icterus in this patient, although described as a recent onset and not initially significant, is a crucial clinical finding that suggests an underlying hepatobiliary dysfunction. Abdominal examination revealed a distended, soft, and non-tender abdomen. Abdominal distension refers to an abnormal enlargement of the abdomen, which can be caused by various factors, including fluid accumulation, organomegaly, or, as in this case, a cystic mass. The abdomen's softness indicates a lack of rigidity, while the non-tender nature suggests that the patient did not exhibit pain or discomfort upon palpation of the abdomen. However, a palpable mass was detected in the upper abdomen. This mass was described as smooth, mobile, and approximately 7x5 cm in size. The palpation of a mobile mass in the abdomen is a significant finding that necessitates further investigation to determine its origin and nature. The cardiovascular, respiratory, and neurological systems were reported as unremarkable. This indicates that the patient's heart function, lung function, and neurological status were within normal limits, suggesting that the patient's presenting symptoms were likely localized to the hepatobiliary system or gastrointestinal tract. Laboratory investigations were conducted to further evaluate the patient's condition. These investigations revealed abnormalities in the patient's bilirubin levels and liver enzymes. The total bilirubin level was measured at 6.5 mg/dL, which is significantly elevated compared to the normal range of 0.3-1.2 mg/dL. Bilirubin is a yellow pigment formed during the breakdown of red blood cells, and elevated levels in the blood can lead to jaundice. The direct bilirubin level was 4.8 mg/dL, also elevated above the normal range of 0-0.2 mg/dL. Direct bilirubin, also known as conjugated bilirubin, is a form of bilirubin that has been processed by the liver, and its elevation suggests a problem with bilirubin excretion. The liver enzymes, specifically SGOT (AST) and SGPT (ALT), were also elevated. SGOT (AST) was measured at 85 U/L, above the normal range of less than 40 U/L. SGPT (ALT) was measured at 92 U/L, also above the normal range of less than 40 U/L. AST and ALT are enzymes primarily found in the liver, and their elevation in the blood indicates liver cell damage or inflammation. The elevated bilirubin and liver enzyme levels collectively point towards a hepatobiliary disorder, suggesting an issue with the liver, gallbladder, or bile ducts. Hematological parameters, including hemoglobin and white blood cell count, were reported to be within normal limits. This indicates that the patient's red blood cell count and white blood cell count were within the expected ranges, suggesting that there was no evidence of anemia or infection based on these parameters. Urinalysis revealed the presence of bilirubin in the urine. Bilirubinuria, the presence of bilirubin in the urine, is another indicator of hepatobiliary dysfunction, as bilirubin is not normally excreted in the urine. Imaging findings played a crucial role in establishing the diagnosis. Abdominal ultrasound revealed a large cystic structure in the region of the common bile duct, consistent with a choledochal cyst. The gallbladder appeared normal, while intrahepatic biliary ducts were not significantly dilated. A choledochal cyst is a congenital anomaly involving cystic dilatation of the bile ducts. The ultrasound findings suggested the presence of such a cyst in the patient. Abdominal CT scan with contrast further detailed the findings. The CT scan demonstrated fusiform dilatation of the extrahepatic bile duct, approximately 6x4 cm in dimension, consistent with a Todani Type 1 choledochal cyst. There was no evidence of intrahepatic biliary dilatation or other associated

abnormalities. The Todani classification is a widely used system for classifying choledochal cysts based on their anatomical location and morphology. Type 1 cysts, as seen in this case, involve dilatation of the common bile duct. The CT scan findings corroborated the ultrasound findings and provided a more detailed anatomical characterization of the choledochal cyst. Histopathological findings were obtained from tissue samples. Macroscopic examination revealed two pieces of tissue. One piece was described as a cyst that had been cut open, and the other piece appeared to be a lumenous part, suspected to be the ductus choledochus (common bile duct). The cyst tissue measured 7 x 4.5 x 4 cm, and after being cut open, it measured 7 x 5 x 2 cm. It contained a lumen or cavity with a diameter of 6 cm, thin thick walls, and had prints of 5 cups and 2 cassettes. The lumenous section measured 5.5 x 4.5 x 1.8 cm and had a brownish-white appearance with blackish parts, also with prints of 5 cups and 2 cassettes. These macroscopic findings describe the physical characteristics of the cyst and the adjacent tissue. Microscopic examination of the cyst tissue revealed pieces of tissue consisting of a cyst wall. The cyst wall showed connective tissue stroma with reactive spindle cells/fibroblasts containing a few lymphocytes, plasma cells, and polymorphonuclear leukocytes (PMN). Hyperemic capillaries and bleeding foci were also seen. The lumenous part showed a piece of tissue covered with cellular cells that could be seen from one layer to several layers. There was connective tissue and muscle tissue, hyperemic blood vessels, and areas of bleeding containing a light amount of lymphocytic cells, plasma cells, and PMN leukocytes. In other parts, tissue was visible in the form of sheets consisting of connective tissue stroma containing areas of bleeding and inflammatory cells. These microscopic findings provide details of the cellular and tissue composition of the cyst and the adjacent duct. The presence of inflammatory cells indicates an inflammatory process within the cyst and the surrounding tissue. Based on the culmination of the clinical presentation, laboratory investigations, imaging findings, and histopathological findings, the final diagnosis was established as a Todani Type 1 Choledochal Cyst (Table 1).

The surgical intervention for the six-month-old female infant diagnosed with a Todani Type 1 choledochal cyst was a critical component of her overall management. The date of the surgery was recorded as November 2023. This timing is significant as it reflects the point at which the diagnostic process had been completed, and the decision to proceed with surgical management was finalized. The promptness of surgical intervention following diagnosis is often a key factor in achieving favorable outcomes in cases of choledochal cysts, as it aims to mitigate the risk of complications such as cholangitis, pancreatitis, and the potential for malignant transformation in the long term. The procedure performed was a surgical excision of the choledochal cyst and Roux-en-Y hepaticojejunostomy. This is the standard surgical treatment of choice for Todani Type 1 choledochal cysts and represents a definitive approach to addressing the anatomical anomaly. Surgical excision refers to the complete removal of the cyst. This is a crucial step in the procedure. By excising the cyst, the source of potential complications is eliminated. The cyst itself can be a site of inflammation, stone formation, and, most importantly, a risk factor for the development of cholangiocarcinoma, a type of bile duct cancer. Therefore, complete excision is aimed at preventing these long-term risks. Roux-en-Y hepaticojejunostomy is a reconstructive surgical procedure. It is performed after the cyst has been excised to restore the continuity of the biliary tract and allow for proper bile flow from the liver into the small intestine. The Roux-en-Y technique involves creating a loop of the jejunum (a part of the small intestine) and anastomosing (connecting) it to the hepatic duct (the duct through which bile exits the liver). This reconstruction creates a new pathway for bile to flow, bypassing the area where the cyst was located. The Roux-en-Y configuration is specifically chosen for several reasons. It helps to prevent reflux of intestinal contents into the biliary tree. Reflux can lead to cholangitis, which is an infection of the bile ducts. The

Roux-en-Y technique minimizes this risk by creating a segment of the jejunum that is not in direct continuity with the duodenum (the first part of the small intestine), reducing the likelihood of duodenal contents, including bacteria, from ascending into the biliary system. This is achieved by the creation of a "Roux limb," which is a segment of jejunum that is divided and re-anastomosed in a way that creates a limb of jejunum used for the biliary anastomosis that is out of the direct flow of intestinal contents. The surgical approach employed was a right subcostal incision. This refers to the location and direction of the incision made in the patient's abdomen to access the surgical site. A subcostal incision is made below the rib cage (costal margin) on the right side of the abdomen. This approach is commonly used for surgeries involving the liver, gallbladder, and bile ducts, as it provides good access to these structures. The choice of a right subcostal incision in this case is consistent with the location of the choledochal cyst and the surgical procedures required for its excision and the subsequent reconstruction. The intraoperative findings provide a detailed account of what the surgeons observed and the steps they took during the operation. The findings confirmed the preoperative diagnosis of a Todani Type 1 choledochal cyst. The surgeons noted a large cystic dilatation of the common bile duct, which is the hallmark of this type of choledochal cyst. A crucial aspect of the surgery involved the careful dissection of the cyst from the surrounding structures. This is a meticulous and technically challenging part of the procedure. The common bile duct is in close proximity to vital structures, including the portal vein and the hepatic artery. The portal vein is a major blood vessel that carries blood from the gastrointestinal tract to the liver, while the hepatic artery supplies oxygenated blood to the liver. Injury to these structures during surgery can lead to significant complications, such as bleeding or impaired liver function. Therefore, the surgeons' careful dissection to separate the cyst from these vessels is of paramount importance. The next step, as indicated previously, was the complete excision of the cyst. This involved removing the entire cystic portion of the dilated bile duct. Following the excision, the Roux-en-Y reconstruction was performed. This involved creating a Roux-en-Y loop of the jejunum. The jejunum is the middle part of the small intestine. A segment of the jejunum was isolated and divided, and then a side-to-end anastomosis was created between the proximal jejunal loop and the common hepatic duct. This anastomosis established a new connection for bile flow from the liver. The distal jejunal loop was then anastomosed to the side of the jejunum at a point 30 cm distal to the hepaticojejunostomy. This step completes the Roux-en-Y reconstruction. The distance of 30 cm is a detail of surgical technique, often chosen to further minimize the risk of reflux and to ensure adequate drainage. During the procedure, the gallbladder was preserved. The gallbladder is a small organ that stores and concentrates bile. In some cases of biliary surgery, the gallbladder is removed. However, in this case, the surgeons opted to preserve the gallbladder. The decision to preserve or remove the gallbladder depends on various factors, including the condition of the gallbladder itself and the specific surgical circumstances. A nasogastric tube was placed intraoperatively. A nasogastric tube is a tube that is inserted through the nose into the stomach. It is often used after abdominal surgery to decompress the stomach, remove fluids or air, and allow the gastrointestinal tract to rest. It can also be used for administering medications or feedings. The placement of a nasogastric tube in this patient was a standard part of the surgical management to aid in the immediate postoperative period. The postoperative management of the patient is also critical for ensuring a smooth recovery. Following the surgery, the patient was monitored in the Pediatric Intensive Care Unit (PICU) for 24 hours. The PICU provides specialized care for critically ill patients, including close monitoring of vital signs, respiratory status, and other parameters. The initial 24-hour period in the PICU is a standard practice after major surgery, allowing for intensive observation and immediate intervention if

necessary. After the initial 24-hour period, the patient was transferred to the general pediatric ward. This indicates that the patient's condition had stabilized sufficiently to allow for a less intensive level of care. In the pediatric ward, the patient continued to receive postoperative care. Intravenous antibiotics were started postoperatively. Antibiotics are administered to prevent or treat infections. In surgical cases involving the biliary system, there is a risk of infection, including cholangitis. Intravenous administration ensures that the antibiotics reach therapeutic levels quickly. Later, the antibiotics were switched to oral administration. This transition usually occurs when the patient's condition improves, and they are able to tolerate oral medications. Pain management was an important aspect of the postoperative care. Analgesics were administered to control postoperative pain and ensure the patient's comfort. Effective pain management is crucial for promoting healing and facilitating early mobilization. Oral feeds were gradually introduced and well-tolerated. After abdominal surgery, there is typically a period where the patient is kept NPO (nothing by mouth) to allow the gastrointestinal tract to recover. Oral feeds are then gradually introduced, starting with clear liquids and progressing to more solid foods as tolerated. The fact that the oral feeds were well-tolerated indicates that the patient's gastrointestinal function was recovering appropriately. The patient's condition at the time of discharge was reported as stable. The patient was discharged one week after surgery. This timeframe suggests that the patient had made a good recovery and that there were no significant postoperative complications requiring further hospitalization. Follow-up care is essential after surgical intervention for choledochal cysts. At the one-month follow-up, the patient showed good recovery, and there were no signs of complications. This early follow-up is important for assessing the initial success of the surgery and identifying any potential early problems. Long-term follow-up is also crucial in these patients to monitor for potential late complications such as anastomotic strictures, cholangitis, or the development of hepatolithiasis (bile duct stones) (Table 2).

3. Discussion

This case report presents a six-month-old infant diagnosed with a Todani Type 1 choledochal cyst. The primary symptom that led to the patient's presentation was progressive abdominal distension. While the patient did exhibit jaundice, a common sign of biliary tract disorders, it was not the predominant symptom at the onset, nor was it the symptom that prompted the initial medical consultation. This particular presentation is noteworthy because it deviates from the classic clinical triad traditionally associated with choledochal cysts. The classic triad, typically observed in older children and adults, consists of abdominal pain, jaundice, and a palpable abdominal mass. In the case of infants, the clinical presentation of choledochal cysts can be highly variable and often non-specific. This variability poses a significant challenge in achieving early and accurate diagnoses. Infants may present with a range of symptoms, including jaundice, feeding difficulties, vomiting, and abdominal distension. The non-specific nature of these symptoms can overlap with those of other more common infantile conditions, which can lead to delays in considering and diagnosing choledochal cysts. The case presented here underscores the importance of recognizing that choledochal cysts can manifest in atypical ways, particularly in the infant population. Abdominal distension, as the primary presenting symptom, can be a significant indicator of underlying pathology in infants. In this specific case, the abdominal distension was likely attributable to the size of the choledochal cyst and its consequent effect on the displacement of other abdominal organs. The gradual accumulation of fluid within the cyst and the physical space occupied by the cyst can lead to a noticeable increase in abdominal girth, which was the primary concern in this patient. The atypical presentation in this case highlights the necessity for clinicians to maintain a high index of suspicion for choledochal cysts, even when the classic triad is not fully present.

Table 1. Summary of patients' clinical findings.

Category	Detail
Patient demographics	
Age at presentation	6 months
Gender	Female
Presenting symptoms	
Primary complaint	Progressive abdominal distension (2-month history)
Other symptoms	History of pale stools (resolved after one week at 2 months of age), decreased appetite (2 weeks prior to admission), slight yellow discoloration of skin and eyes (recent onset)
Absence of symptoms	No reported fever, vomiting, poor feeding, significant jaundice initially
Past medical history	
Birth history	Born at term via spontaneous vaginal delivery, no perinatal complications
Growth and development	Appropriate for age until onset of symptoms
Family history	No family history of choledochal cysts or other congenital anomalies
Physical examination	
General appearance	Alert and interactive, well-nourished
Weight	7.5 kg (within the normal range for age)
Length	65 cm (within the normal range for age)
Skin and sclera	Icteric
Abdomen	Distended, soft, non-tender
Palpable mass	Upper abdomen, smooth, mobile, approximately 7x5 cm
Other systems	Cardiovascular, respiratory, and neurological systems unremarkable
Laboratory investigations	
Total bilirubin	6.5 mg/dL (normal range: 0.3-1.2 mg/dL)
Direct bilirubin	4.8 mg/dL (normal range: 0-0.2 mg/dL)
SGOT (AST)	85 U/L (normal range: <40 U/L)
SGPT (ALT)	92 U/L (normal range: <40 U/L)
Hematological parameters	Hemoglobin and white blood cell count within normal limits
Urinalysis	Presence of bilirubin
Imaging findings	
Abdominal ultrasound	Large cystic structure in the region of the common bile duct, consistent with a choledochal cyst; gallbladder appeared normal; intrahepatic biliary ducts not significantly dilated
Abdominal CT scan with contrast	Fusiform dilatation of the extrahepatic bile duct, approximately 6x4 cm, consistent with a Todani Type 1 choledochal cyst; no evidence of intrahepatic biliary dilatation or other associated abnormalities
Histopathological findings	**Macroscopic:** Two pieces of tissue, one of which is a cyst that has been cut open and the other piece appears to be a lumenous part (suspected of the ductus coledukus). Tissue was white, spongy, measuring 7 x 4.5 x 4 cm. From the cyst that has been cut, measuring 7 x 5 x 2 cm in cross section, 1 cavity with a diameter of 6 cm, thin thick walls, print 5 cups and 2 cassettes. From lumenous section, measuring 5.5 x 4.5 x 1.8 on cross section brownish white with blackish parts, print 5 cups 2 cassettes. **Microscopic:** From the cyst that has been cut microscopically, pieces of tissue in the form of a cyst wall consisting of connective tissue stroma with reactive spindle cells/fibroblasts containing a few lymphocytes, plasma cells, PMN leukocytes and histiocytes are visible. Hyperemic capillaries and bleeding foci were seen. From the microscopic lumen part, a piece of tissue in the form of a duct with a surface covered with columnar cells can be seen from one layer to several layers, there is connective tissue and muscle tissue, hyperemic blood vessels, the area of bleeding contains a light amount of lymphocyte cells, plasma cells, and PMN leukocyte cells. In other parts, tissue is visible in the form of sheets consisting of connective tissue stroma containing areas of bleeding and inflammatory cells.
Diagnosis	Todani Type 1 Choledochal Cyst

Table 2. Surgical management and follow-up.

Category	Detail
Date of surgery	November 2023
Procedure performed	Surgical excision of the choledochal cyst and Roux-en-Y hepaticojejunostomy
Surgical approach	Right subcostal incision
Intraoperative findings	Large cystic dilatation of the common bile duct, confirming Todani Type 1 choledochal cyst; cyst carefully dissected from surrounding structures (portal vein and hepatic artery); complete excision of the cyst performed; Roux-en-Y loop of jejunum created; side-to-end anastomosis between proximal jejunal loop and common hepatic duct; distal jejunal loop anastomosed to side of jejunum 30 cm distal to hepaticojejunostomy; gallbladder preserved; nasogastric tube placed intraoperatively.
Postoperative management	Monitored in PICU for 24 hours, then transferred to the general pediatric ward; intravenous antibiotics started postoperatively, later switched to oral; pain managed with analgesics; oral feeds gradually introduced and well-tolerated.
Discharge condition	Stable condition, one week after surgery
Follow-up (1 month)	Good recovery, no signs of complications

It is crucial to consider choledochal cysts in the differential diagnosis of infants presenting with abdominal distension, especially when other common causes have been excluded. Atypical presentations can easily lead to diagnostic delays, which in turn can increase the risk of complications.[11,12]

The diagnostic workup of this patient followed a systematic approach, beginning with initial laboratory investigations. The laboratory findings revealed conjugated hyperbilirubinemia and elevated liver enzymes. These findings are suggestive of biliary obstruction and/or hepatobiliary dysfunction. Hyperbilirubinemia, specifically the elevation of conjugated bilirubin, indicates that the liver is able to process bilirubin but is having difficulty excreting it. This can occur due to a blockage in the bile ducts, as seen in choledochal cysts. Elevated liver enzymes, such as SGOT (AST) and SGPT (ALT), are indicative of liver cell damage or inflammation. In the context of a choledochal cyst, the elevated enzymes may result from the backup of bile into the liver or from inflammation of the liver tissue itself. While these laboratory findings are not specific to choledochal cysts, they play a crucial role in raising clinical suspicion for a hepatobiliary disorder and prompting further investigation. Following the initial laboratory investigations, imaging modalities were employed to visualize the biliary tree and confirm the diagnosis. Abdominal ultrasound was utilized as the first-line imaging modality. Ultrasound is a non-invasive, readily available, and cost-effective tool that can provide valuable information about the abdominal organs. In this case, the ultrasound revealed a large cystic structure in the region of the common bile duct, which was consistent with a choledochal cyst. Ultrasound is particularly useful in pediatric patients because it does not involve ionizing radiation. It can effectively visualize fluid-filled structures like choledochal cysts and can also provide information about the size, location, and characteristics of the cyst. Additionally, ultrasound can be used to assess the gallbladder and intrahepatic biliary ducts, as was done in this case, to evaluate for any associated abnormalities. While ultrasound is a valuable screening tool, it may not always provide the detailed anatomical information necessary for surgical

planning. Therefore, in this case, a CT scan of the abdomen with contrast was performed. CT scans provide cross-sectional images of the body, offering a more detailed anatomical assessment. The CT scan confirmed the presence of a Todani Type 1 choledochal cyst and further delineated the extent of the cystic dilatation of the extrahepatic bile duct. The CT scan also played a crucial role in ruling out other associated anomalies. It provided a comprehensive view of the abdominal structures and helped to ensure that there were no other congenital abnormalities that might impact the surgical approach or the patient's overall management. In this particular case, the CT scan demonstrated fusiform dilatation of the extrahepatic bile duct, measuring approximately 6x4 cm, consistent with the diagnosis of a Todani Type 1 choledochal cyst. Another imaging modality that is often used in the evaluation of choledochal cysts is magnetic resonance cholangiopancreatography (MRCP). MRCP is a non-invasive imaging technique that uses magnetic resonance imaging to visualize the biliary and pancreatic ducts. It provides detailed images of the biliary anatomy and can be particularly useful in identifying anomalous pancreaticobiliary ductal junctions (APBDJ), which are frequently associated with choledochal cysts. In this case, while MRCP is a valuable tool, the clinical team determined that the findings from the ultrasound and CT scan were sufficient to establish the diagnosis and proceed with surgical planning. The decision to forgo MRCP may have been based on factors such as the clarity of the findings from the other imaging modalities, the need to expedite the surgical intervention, or the availability of MRCP. The diagnostic process in this case highlights the importance of a stepwise approach, starting with less invasive and readily available tests like ultrasound and progressing to more detailed imaging like CT scans when necessary. The combination of laboratory investigations and imaging studies is essential for accurately diagnosing choledochal cysts and for guiding appropriate management strategies.[13-15]

The precise etiology of choledochal cysts remains a subject of ongoing investigation, but the most widely accepted theory revolves around the concept of an abnormal pancreaticobiliary ductal junction (APBDJ). This theory, initially proposed by Babbitt, posits that APBDJ plays a crucial role in the development of choledochal cysts. APBDJ is an anatomical variation where the pancreatic duct and the common bile duct join outside the normal location of the sphincter of Oddi. The sphincter of Oddi is a muscular valve that controls the flow of bile and pancreatic juice into the duodenum. In individuals with APBDJ, this anatomical abnormality results in a long common channel where the pancreatic and biliary secretions mix. The significance of this long common channel lies in the fact that it allows for the reflux of pancreatic enzymes into the bile duct. Pancreatic enzymes, such as amylase and lipase, are highly potent and can cause significant damage to the bile duct wall. The reflux of these enzymes leads to inflammation, weakening, and eventual dilatation of the bile duct, ultimately resulting in the formation of a choledochal cyst. The chronic inflammation and weakening of the bile duct wall can lead to a variety of pathological changes. The bile duct epithelium may undergo metaplasia, which is a change in the type of cells lining the duct. There may also be proliferation of the smooth muscle and connective tissue in the duct wall. These changes contribute to the progressive dilatation and cystic formation characteristic of choledochal cysts. While the APBDJ theory is widely accepted, it does not fully explain all cases of choledochal cysts. There is evidence to suggest that other factors, such as genetic predisposition, may also play a role. Some studies have reported familial clustering of choledochal cysts, which suggests a possible genetic component. However, the specific genes involved in the development of choledochal cysts have not yet been definitively identified. Furthermore, not all individuals with APBDJ develop choledochal cysts, and some patients with choledochal cysts do not have an identifiable APBDJ. This suggests that the pathogenesis of choledochal cysts is likely

multifactorial, involving a complex interplay of anatomical, genetic, and environmental factors. The understanding of the etiology and pathogenesis of choledochal cysts is crucial for developing strategies for prevention and early detection. Further research is needed to fully elucidate the mechanisms underlying the development of these cysts and to identify individuals who may be at increased risk.[16-18]

The most widely used classification system for choledochal cysts is the one proposed by Todani et al. This classification system categorizes choledochal cysts into five main types, based on their anatomical location and morphology. The Todani classification is essential for standardizing the description and management of choledochal cysts. Type I cysts are the most common, accounting for the majority of cases. These cysts involve fusiform or saccular dilatation of the extrahepatic bile duct. The dilatation can affect the entire extrahepatic bile duct or a segment of it. In this case report, the patient was diagnosed with a Todani Type 1 choledochal cyst, which is consistent with the typical presentation of this condition. Type II cysts are less common and involve a diverticulum, which is a pouch-like protrusion, arising from the extrahepatic bile duct. These cysts are typically smaller than Type I cysts and may be located in various positions along the extrahepatic bile duct. Type III cysts, also known as choledochoceles, involve dilatation of the intraduodenal portion of the common bile duct. These cysts protrude into the lumen of the duodenum and may cause obstruction of the bile flow. Type IV cysts are characterized by multiple dilatations involving both the intrahepatic and extrahepatic bile ducts. This type is further subdivided into Type IVA, which involves multiple dilatations of both intrahepatic and extrahepatic ducts, and Type IVB, which involves multiple dilatations of the extrahepatic bile ducts alone. Type V cysts, also known as Caroli disease, involve multiple segmental dilatations of the intrahepatic bile ducts. This type is often associated with other congenital anomalies, such as renal tubular ectasia. The accurate classification of choledochal cysts is important for several reasons. It helps to guide surgical planning, as the surgical approach may vary depending on the type and location of the cyst. It also provides prognostic information, as certain types of cysts may be associated with a higher risk of complications. Additionally, the Todani classification facilitates communication and research among healthcare professionals involved in the management of choledochal cysts.[19,20]

4. Conclusion

This case report highlights a distinctive presentation of a Todani Type 1 choledochal cyst in a six-month-old infant, where the primary clinical manifestation was progressive abdominal distension, deviating from the classic triad of symptoms typically associated with this condition. The case underscores the importance of considering choledochal cysts in the differential diagnosis of abdominal distension in infants, even in the absence of jaundice or pain. Early diagnosis, facilitated by imaging modalities such as ultrasound and CT scans, is crucial for prompt and effective management. Surgical intervention, involving complete excision of the cyst and Roux-en-Y hepaticojejunostomy, remains the cornerstone of treatment, aiming to prevent complications and ensure a favorable outcome. This case also emphasizes the challenges in diagnosing choledochal cysts in infants due to the variability and non-specificity of symptoms. Clinicians must maintain a high index of suspicion and adopt a systematic approach to investigation to avoid delays in diagnosis and potential complications. Furthermore, this report contributes to the existing body of knowledge by illustrating an atypical presentation, thereby expanding the understanding of the clinical spectrum of choledochal cysts in the pediatric population. Long-term follow-up is essential to monitor for potential complications and ensure the patient's continued well-being.

5. References

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