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Cystic Enlargement of the Bile Duct: Case Report and Review of Literature

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Abstract

Congenital cystic dilatations of the bile duct are rare malformations representing 1 per 13,000 births with a female predominance. Diagnosis of this pathology is based on radiological imaging especially radiological investigations conducted on an ultrasound coupled with Computed tomography (CT) scan and especially a bili-MRI which allows the analysis of the bile duct malformations and the anomalies of the pancreaticobiliary junction. We will discuss in this clinical case the various radiological aspects of cystic dilatations of the biliary duct.

Keywords

Cystic Dilation, Bile Duct, Radiology, Malformation

1. Introduction

Congenital cystic dilations of the biliary duct are rare malformations that are grouped into several types, depending on the site, shape and distribution of the malformation. They represent 1 in 13,000 births [1] and are more frequent in Asia with a significant female predominance. Congenital cystic dilations are most often diagnosed in childhood as 75% of the cases diagnosed at the age of 10 years [1]. Radiological imaging plays a very important role in the diagnosis of these preoperative anomalies. Thus, through this clinical case, we will discuss the various radiological aspects of cystic dilatations of the biliary duct.

2. Case Report

A 24-year-old primigest with amenorrhea for 16 weeks was admitted in our hos-

pital for febrile jaundice. This illness had begun 45 days before her admission with the appearance of a cholestatic jaundice associated a right hypochondrium pain with no improvement after indulging in self medication. These symptoms aggravated 5 days before her admission when she presented fever and chills. This prompted the patient to consult our emergency department.

Physical examination found a conscious patient with a body temperature at 39.7˚C, tachycardia (120 beats/minute), polypnea (30 cycles/minute), an apparent yellowing of the sclerae and a right hypochondriac region pain.

Laboratory test revealed the following: elevated White blood cells (18,540/mm^3), CRP (240 mg/l), cholestasis with total bilirubin = 105 mg/l, and direct bilirubin = 86 mg/l, GGT = 838 UI/l, PAL = 1057 UI/l. Renal function was impaired with elevated serum creatinine = 18 mg/l and blood urea nitrogen = 0.9 g/l.

The abdominal ultrasound revealed a significant dilation of the common bile duct and the intrahepatic bile ducts without a visible obstacle.

An MRI showed malformed cystic dilation of the intrahepatic bile ducts, the right and left hepatic ducts and the common bile duct with its transverse axis measuring 10 cm (Figures 1-4).

The patient was hospitalized in our intensive care unit (ICU) for severe angiocholitis and was put on antibiotics.

A fetal expulsion occurred 24 hours later. Patient passed away afterwards in a setting of septic shock complicating her underlying angiocholitis.

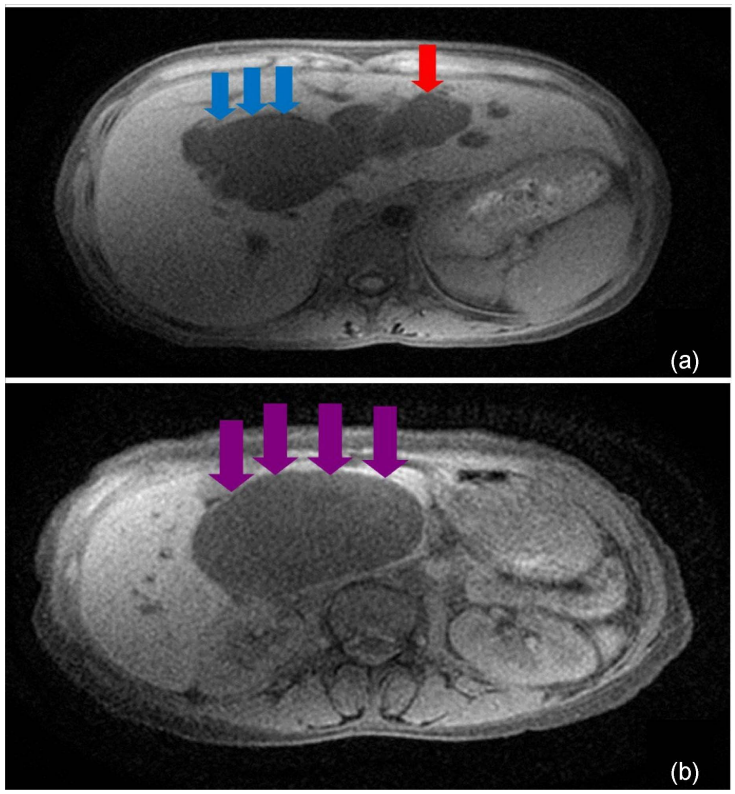

Figure 1. Axial section, T1 sequence showing massive dilation of liver bile ducts; (a) right (blue) and left (red) (b) common bile duct.

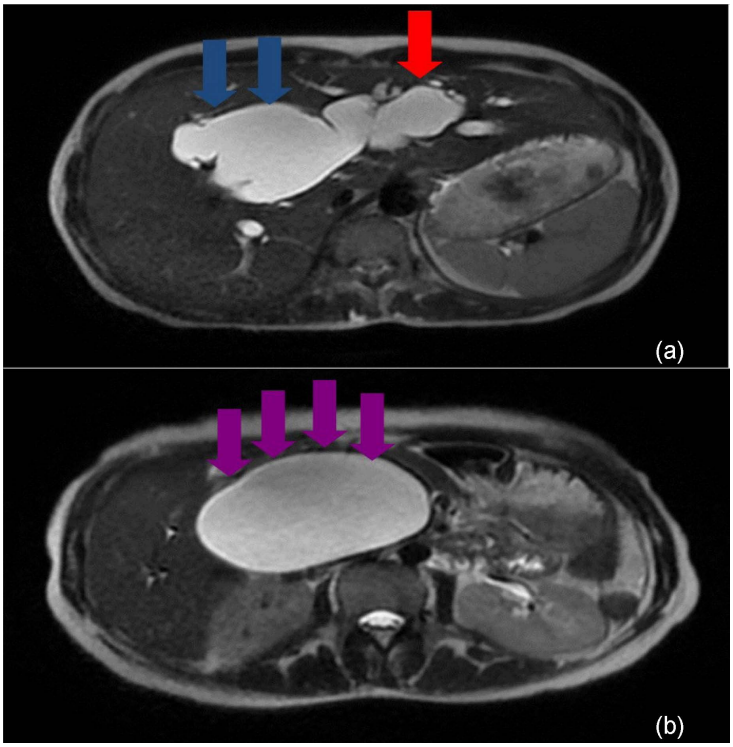

Figure 2. T2, axial section showing massive dilation of liver bile ducts; (a) right (blue) and left (red) (b) common bile duct.

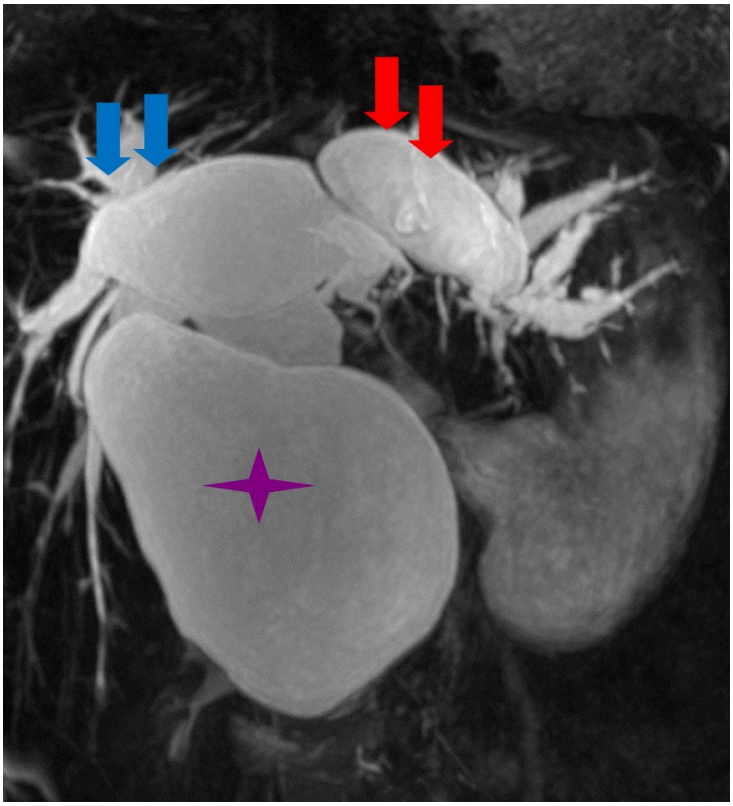

Figure 3. 3D MRCP showing important dilation of bile ducts, right (blue) left (red) and common bile duct (purple).

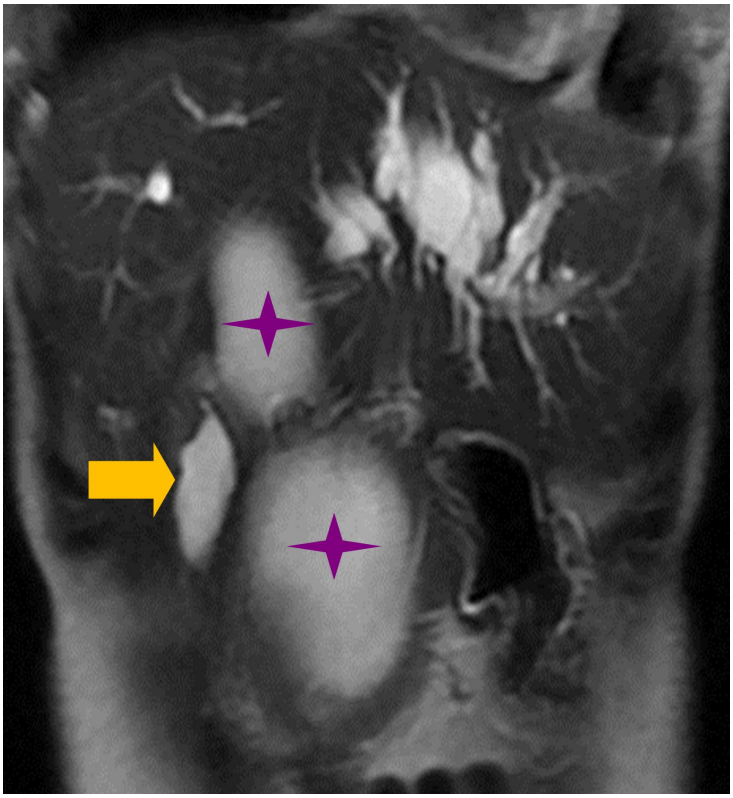

Figure 4. 2 weighted sequences, coronal section showing massive dilation of the common bile duct (purple) and normal gallbladder (yellow).

3. Discussion

Cystic dilatations of the bile duct rare congenital malformations. They are more common in Asia with a female predominance [1]. Its frequency is estimated between 1/100,000 and 1/175,000 births. Diagnosis of this congenital malformation is usually made before the age of 10 years. 20% - 50% of cases are discovered in adulthood in western countries.

Many theories have been proposed to explain its origin:

- In 1969, Babbitt [2] incriminated an anomaly of the pancreaticobiliary junction found in 57% - 96% of cases, characterized by three criteria:

- Long hepatopancreatic duct > 15 mm;
- Extra-duodenal union of the two distal canals which is distant from the sphincters;
- Connection angle > 30˚.

This anomaly of the-pancreaticobiliary junction is incriminated in:

- The reflux of pancreatic enzymes into the common bile duct [3] which results in irritation of its walls that would favor the formation of congenital cystic dilatations of the bile ducts,
- The degeneration of the dilated bile duct segments and the gallbladder.

Other possible causes have been mentioned: genetic, familial or immune factors, retroviral infection, functional anomalies of the sphincter of Oddi, congenital weakness of the bile duct wall, anomalies of autonomic innervation, com-

mon bile duct "hypoganglionosis", and any other factors responsible for obstruction of the main bile duct [3].

The clinical symptoms of cystic dilation of the bile tract are based on the classical triad: pain, jaundice and mass of the right hypochondriac region. This triad is more present in children below the age of 10 years and is found in only 10% of cases. Some cases are detected during antenatal care. Acute pancreatitis often reveals the choledochal cyst. Other revealing complications are lithiasis, which can block the common bile duct, cholangitis, Gallbladder perforation with choleperitoneum, biliary cirrhosis, and secondary degeneration of the cystic wall which has been reported from the age of 12 years [4].

In 1723, Vater described for the first time the dilatations of the biliary tree. In 1959, Alonso-Lej [5] proposed the first classification of these anomalies. Currently, the most widely used classification is that of Todani [6], which groups these malformations into five types according to their locations, their extent, and the type of dilation of the bile ducts.

- Type I (80% - 90%): Cystic dilatation of the common bile duct. This is the most frequent form. There are 3 sub-types:
  - Ia: Dilatation of a part or all of the extrahepatic bile duct with a cystic duct originating from the cyst.
  - Ib: Segmental dilation of the common bile duct.
  - Ic: Fusiform dilation of the common bile duct and the common hepatic duct.
- Type II (3%): Diverticula of the common bile duct. It is a diverticulum that comes from the common bile duct or the common hepatic duct.
- Type III (5%): Choledochocele. This is an intramural dilation of the distal part of the common bile duct that protrudes into the second part of the duodenum. According to the classification of Sarris and Tsang:
  - Type A: the ampulla opens into the top of the cyst, pours into the duodenum via a distinct orifice:
    - A1: common pancreaticobiliary canals,
    - A2: separate canals,
    - A3: small intra-mural choledochocele.
  - Type B: the ampulla directly flows into the duodenum.
- Type IV (10%): Multiple cystic dilatations. There are 2 types:
  - Type IVa: intrahepatic and extrahepatic dilations of the biliary tree.
  - Type IVb: multiple extrahepatic cysts.
- Type V (1%): Caroli disease is a rare congenital disease characterized by multiple sack like dilatations of the intrahepatic bile ducts. It can be bilateral sometimes. It can present early complications such as intrahepatic lithiasis, and recurrent bacterial cholangitis. It can also have delayed complcations such as biliary cirrhosis, portal hypertension and malignant degeneration. It may be associated with renal anomalies polycystic kidney disease, Cacchiricci disease, congenital hepatic fibrosis.

The initial diagnosis of cystic bile duct dilation is essentially based on ultra-

sound studies. It is a reliable tool in the evaluation of the importance of the dilation of the biliary tree. It remains the most sensitive and the most specific in the search for intracystic calculi. It shows a well-limited thin-walled cystic or fusiform formation, containing hyperechoic material corresponding to biliary sludge.

The CT scan allows good characterization of the lesions and shows an iso or hypodense thin walled mass unenhanced by contrast which contains a sludge. This mass is located just above the pancreas. The Bili-MRI [7] is the reference imaging modality allowing a more defined exploration of bilio-pancreatic tree. It shows all the malformations of the biliary tree allowing the differential diagnosis with an obstructive pathology and is very useful in the detection of bilio-pancreatic anomalies. It allows us to suspect a malignant degeneration by showing: a endoluminal polypoid tumor or an irregular thickened cystic wall. It shows lesions in the form of a regular, well-defined mass of variable size in continuity with the main bile duct with a hypo-intense liquid T1 signal, hyper intense T2 sgnal, and unenhanced after injecting a contrast agent.

The surgical treatment consists of a complete resection of the cyst with a Roux-en-Yhepatico-jejunal anastomosis [5].

Choledochocele may be treated with endoscopic sphincterotomy or surgical excision.

4. Conclusion

Cystic dilatations of the bile duct are rare malformations. Radiological imaging plays an essential role in diagnosing this affection, especially a Bili-MRI which allows an accurate biliary imaging. Surgery is the main treatment method for this affection, though endoscopy can sometimes be performed.

References

[1] Khmekhema, R., Zitounia, H., Ben Ahmeda, Y.S., *et al.* (2012) Surgery of Cystic Dilatation of the Bile Duct in Children. Results of 16 Observations. *Journal de pédiatrie et de puériculture*, **25**, 199-205.

[2] Babbitt, D.P. (1969) Congenital Choledochal Cysts: New Etiological Concept Based on Anomalous Relationships of the Common Bile Duct and Pancreatic Bulb. *Annals of Radiology*, **12**, 23.

[3] Ono, S., Fumino, S. and Iwai, N. (2011) Diagnosis and Treatment of Pancreaticobiliary Maljunction in Children. *Surgery Today*, **41**, 601-605. https://doi.org/10.1007/s00595-010-4492-9

[4] Iwai, N., Deguchi, E., Yanagihara, J., Iwai, M., Matsuo, H., Todo, S., *et al.* (1990) Cancer Arising in a Choledochal Cyst in a 12-Year-Old Girl. *Journal of Pediatric Surgery*, **25**, 1261-1263. https://doi.org/10.1016/0022-3468(90)90525-E

[5] Saluja, S.S., *et al.* (2010) Choledochal Cyst with Chronic Pancreatitis: Presentation and Management. *Journal of the Pancreas*, **11**, 601-603.

[6] Todani, T., Watanabe, Y., Narusue, M., *et al.* (1977) Congenital Bile Duct Cysts: Classification, Operative Procedures, and Review of Thirty-Seven Cases Including Cancer Arising from Choledochal Cyst. *American Journal of Surgery*, **134**, 263-269. https://doi.org/10.1016/0002-9610(77)90359-2

[7] Nakamura, H., Katayose, Y., Rikiyama, T., Onogawa, T., Yamamoto, K., Yoshida, H., *et al.* (2008) Advanced Bile Duct Carcinoma in a 15-Year-Old Patient with Pancreaticobiliary Maljunction and Congenital Biliary Cystic Disease. *Journal of Hepato-Biliary Pancreatic of Surgery*, **15**, 554-559. https://doi.org/10.1007/s00534-007-1310-x