Congenital anomalies of the biliary system: choledochal cysts

Case reports

Introduction
Bile duct cysts are rare congenital anomalies. The incidence of these anomalies is 1:100,000–150,000 and more frequently reported between female (F:M=3:1) [1-2]. Biliary tree cystic dilatations can affect intra- and extrahepatic biliary ducts, or both. Most bile duct cysts are diagnosed during infancy or childhood, but also it can be discovered at any age. The etiology of this condition is unknown, but the anomalous junction between the common bile duct and the pancreatic duct allowing pancreatic juice reflux into biliary tree is the most accepted etiopathogenic concept [1]. Choledochal cyst is related with biliary tree that causes two most frequent complications: stone formation and malignant transformation [1-3].

Table 1. Todani classification

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
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<tbody>
<tr>
<td>I</td>
<td>A fusiform dilation of the extrahepatic bile duct.</td>
</tr>
<tr>
<td>Ia</td>
<td>Dilatation of extrahepatic bile duct (entire).</td>
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<tr>
<td>Ib</td>
<td>Dilatation of extrahepatic bile duct (focal segment).</td>
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<tr>
<td>Ic</td>
<td>Dilatation of the common bile duct portion of extrahepatic bile duct.</td>
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<td>II</td>
<td>An extra- or intrabiliary duct diverticulum.</td>
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<tr>
<td>III</td>
<td>A choledocholecyst.</td>
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<tr>
<td>IVa</td>
<td>Multiple communicating intra- and extrahepatic duct cysts.</td>
</tr>
<tr>
<td>IVb</td>
<td>Multiple cystic dilatations involving only the extrahepatic bile duct.</td>
</tr>
<tr>
<td>V</td>
<td>Caroli disease.</td>
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</tbody>
</table>

The incidence of biliary tract cancer in patients with choledochal cysts increases with age. Moreover, the diagnosis of adult choledochal cysts is frequently delayed due to nonspecific clinical symptoms which classically presents right upper quadrant abdominal pain, jaundice and palpable mass. These symptoms are more common in children than in adults, while they can present with abdominal pain, cholangitis, pancreatitis. Furthermore, choledochal cyst can be asymptomatic for many years and diagnosed incidentally.

According Todani classification, these cysts are divided into five main types with several subtypes. Magnetic resonance cholangiopancreatography (MRCP) is useful, noninvasive tool with good overall accuracy in detection and classification of choledochal cysts. The treatment of choice is surgery, typically Roux-en-Y hepatoenteroanastomosis is performed [4]. Prognosis after this operation usually is perfect, nevertheless patient needs further follow-up because of the increased risk of cholangiocarcinoma.

Results
The „gold standard” for detecting and staging choledochal cyst is MRCP, which is a non-invasive imaging technique to visualize intra and extrahepatic biliary tree and pancreatic ductal system. MRCP can detect and evaluate choledochal cyst anatomy, size, site, and shape of bile duct dilatation. It also avoids the risk of pancreatitis and cholangitis associated with invasive procedures such as ERCP and percutaneous cholangiography. The limitations of MRCP are the presence of intra-abdominal clips from previous surgery, patient movement, or claustrophobia.

Todani classification of bile duct cysts divides choledochal cyst into five main types (Table 1):

- Type I
  - A fusiform dilation of the extrahepatic bile duct.
  - Type Ib: Dilatation of extrahepatic bile duct (focal segment).
  - Type Ic: Dilatation of the common bile duct portion of extrahepatic bile duct.
- Type II
  - An extra- or intrabiliary duct diverticulum.
- Type III
  - A choledocholecyst.
- Type IV
  - Multiple communicating intra- and extrahepatic duct cysts.
  - Type IVa: Fusiform dilatation of the entire extrahepatic bile duct with extension of dilatation of the intrahepatic bile duct.
  - Type IVb: Multiple cystic dilatations involving only the extrahepatic bile duct.
- Type V
  - Caroli disease.

Four patients were examined, three of them were women (the average age 49.7 years) and one man (39 years old). The predominant symptom was abdominal pain presented in all four patients.

The structure of cysts were:
- 1 case of type I
- 2 cases of type II
- 1 case of type V

Treatment in all these cases was surgery. In none of these patients malignancy was detected.

Conclusions
Main bile duct cysts are rare pathology with non-specific clinical manifestation. It is important for the radiologist to be acquainted with this problem. Correct diagnosis can be made after excluding other diseases, such as cholangiocarcinoma or bile duct stones. Usually prognosis after surgery is perfect, patients need long-term follow-up because of the increased risk of cholangiocarcinoma.

Methods
The pictorial review demonstrates the spectrum of congenital biliary duct system anomalies. Presentation provides basic morphological classification of congenital biliary duct anomalies, the diagnostic value of MRCP, as well as its advantages and disadvantages. The type of cyst was classified according Todani classification. Four clinical cases with congenital biliary system anomalies are analysed.

Bibliography