CASE REPORT

Atypical cyst of the right ductus hepaticus

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Abstract

Cystic dilatation is a rare disease of extra- or intra-hepatic bile ducts. According to the literature it is an disorder accompanying other diseases rather than an independent diagnosis. In the case of smaller dimensions it is often found by coincidence because of its asymptomatic course of development. Mostly it manifests itself through abdominal pain or icterus. The authors show a case report of 14-months old patient with the finding of a cyst in the right ductus hepaticus with extraordinary dimensions *(Fig. 2, Ref. 11)*. Full Text (Free, PDF) *www.bmj.sk*
Key words: congenital choledochal cyst, anastomosis Roux-en-Y.

As a rarity the extra-hepatic and intra-hepatic bile ducts are affected by larger or smaller localized dilatations. Mostly these are asymptomatic and their detection is made by coincidence. The number of findings of cystic dilations in bile ducts has increased as a result of the development of imaging techniques, namely USG (ultrasonography). The number of cysts varies from one solitary cyst up to polycystic changes in the bile ducts – Caroli syndrome. The incidence is described in different ways, from 1 case per 200,000 up to 1 per 2,000,000. They are more often detected in female patients, the ratio female/male is 4:1. Approximately 2/3 of cases are diagnosed till the 10th year of the patients• age (1). All cases require surgical treatment.

Case report

A 14-months old boy was hospitalized at the Pediatric Department of the Faculty Hospital in Nitra due to pathologically asymmetrically enlarged abdomen. Patient did not vomit, had no pain, had no febrile states (fevers) or icterus in the past. After the clinical examination laboratory examinations, USG, CT, MRI were performed. Imaging techniques proved large cystic formation under the whole liver which was significantly compressed and deformed (Fig. 1). Gall-bladder was also shown. There was a surgery indicated after surgical consultations.

Laboratory pre-operation examinations: Hb, Ht, RC, WC, PLT and hemo-coagulation examinations were within normal ranges. Albumin 36.0; total bilirubin 12.0; conj. 5.5; AST 1.23; ALT 0.67; GMT 7.78; ALP 18.8; AMS 0.4.

Surgery finding: After opening the abdomen a localized subhepatic cystic formation was detected with the volume of 1500 ml. There was a dark content visible through the wall. Cystic formation, respectively dilatation was localized within the entire right ductus hepaticus. This type of dilatation is not typical and it does not occur even in the Todani's classification of 1977 (Fig. 2) (11).

Left ductus hepaticus was approx. 15 cm long, deformed and stretched on the wall of the cyst. Ductus hepatocholedochus had appropriate width of approx. 3–4 mm and length of approx. 30 mm. Gall-bladder and ductus cysticus were without changes. Perioperative cholangio-scopy showed an anomalous branching in intra-hepatic bile ducts in the right hepatic lobe. There was normal branching of the bile ducts in the left lobe. Lumens of the bile ducts openings were approx. 1 mm; whereby the distance between both openings from the liver was 12 cm. The right bile duct was extremely dilated from the liver-mouth. The left bile duct was changed due to a chronic compression caused by the large cyst – it was significantly elongated (see above) and flattened, with a deficient wall. After an extirpation of the cyst a sufficient quality and nutrition of its wall was no guaranteed for preserving it. Since the anatomic ratios were changed by this finding and be-

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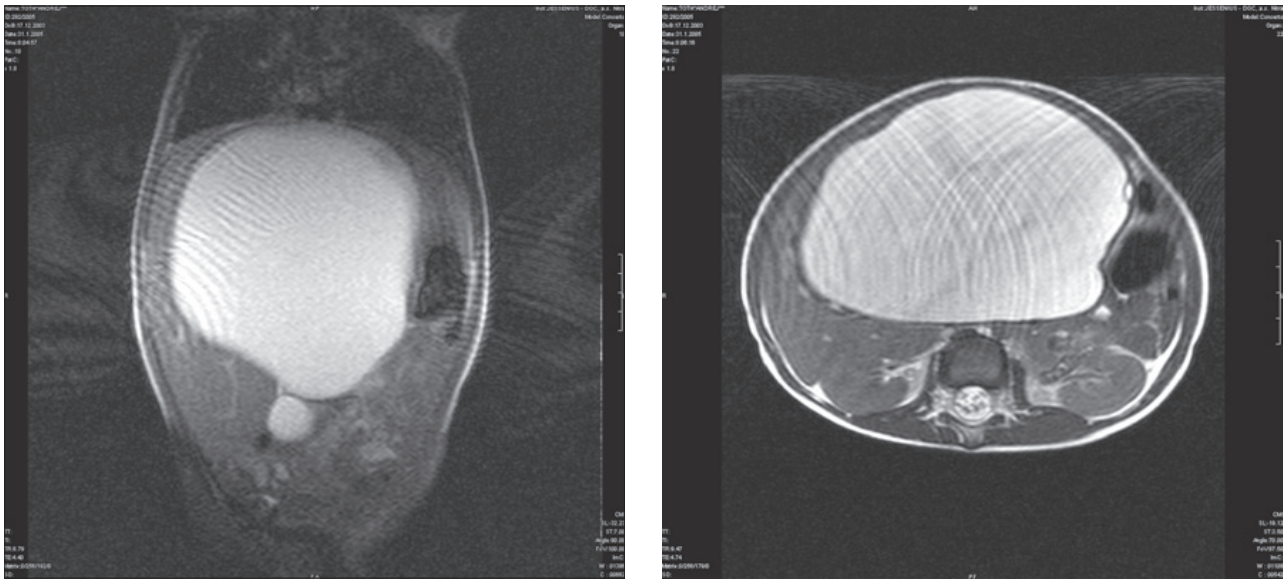

Fig. 1. MRI image made before the surgery.

cause of being afraid how the anastomosis will be healed we decided to make atypical reconstruction by the means of omega-loop – hepatico-jejuno anastomosis end-to-side with Völker's drainage with sewing the hepatics separately. Left anastomosis was drained through the inflow side and right one through the outflow side with the purpose to rework the stated intestinal loop for Roux-en-Y as soon as possible, should the healing be without complications. We had left drains for 3, respectively for 4 weeks. Post-operative course was complicated by a subphrenic abscess on the right side which was treated conservatively.

Laboratory examinations after the first surgery: Hb, Ht, RC, WC, PLT and hemo-coagulation examinations were within normal ranges, album. 39.0; total bilirubin 5.0; conj. 1.5; AST 0.69; ALT 0.5; GMT 5.24; ALP 12.1; AMS 0.49.

Histological examination: image of the wall consisting of a dense fibrotic material with intendances, part of its inner area was littered with a cylindrical epithelium with the indication of a soft papillarity. Larger part of this area is, however, with a terrain of erosions even ulcerations, bottom of which comprises of pre-dominantly chronic inflammatory cellulization and variously thick zones constituting a variously mature non-specific granulation tissue as an evidence of the proliferative inflammation processes. Clinically determined cyst in the ductus hepaticus is, from the formal pathogenesis point of view, deemed to be more fusiform eventually spherical dilatation of the bile duct; the right-side cysts, firstly of congenital origin caused by unknown causes and described spectrum of regressive changes in the mucous are deemed to be a phenomenon of a bile stagnation, respectively an abrasive effect of pancreatic enzymes in possible reflux. Part of the image comprises also of the glandular structures in the intra-mural localization.

Once the drainage was removed, a second surgery was performed within 2 months – this surgery was made without any complications and the Roux-en-Y hepatico-entero-anastomosis was kept. Immediate post-operative course was without any complications.

Later there were 2 frust cholangoitis detected during the post-operation period, these were healed conservatively.

Laboratory examinations after the 2nd surgery: Hb 9.7; RC 3.93;•Ht 28.8; WC 10.7; PLT 766; GMT 0.74; ALP 11.11; AST 0.37; ALT 0.43; total bilirubin 6.0; conj. 1.4; albumin 37.6; Fe 1.2; cholesterol 2.34; IgG 7.59; IgA 0.69; IgM 0.79; C3 1.77; C4 0.52.

With the follow-up CT we have found cystic formation of 4 cm located in the right lobe of the liver heading up to the center with the slight pressure on the bile ducts and also a compensation hypertrophy of the left lobe and atrophy of the right lobe. Stated formation was resolved on the specialized workplace by the means of percutaneous drainage with repeated lavages and alcoholization. However, the right-sided hemihepatectomy is not excluded during the ongoing course.

Discussion

Presentation of the cystic dilatations of the bile ducts is, because of its rare occurrence, mostly in the casuistics. Differential diagnostics is complicated in spite of the progress achieved in the imaging techniques – USG, CT, MRI, ERCP. Most often the diagnosis takes into account a mesentrial cyst or cyst omentum majus (1, 2, 3). We have no experiences with the effectiveness of the PET examinations used by this type of disease. The occurrence of the cysts with smaller dimensions has been significantly increased. Almost every time these are asymptomatic cysts with the coincidental finding made by the examinations indicated due to other reasons. Etiology of the choledochal-cysts occurrence is unknown. Their occurrence is related with pathologic reflux of the pancreatic enzymes into the bile ducts (9).

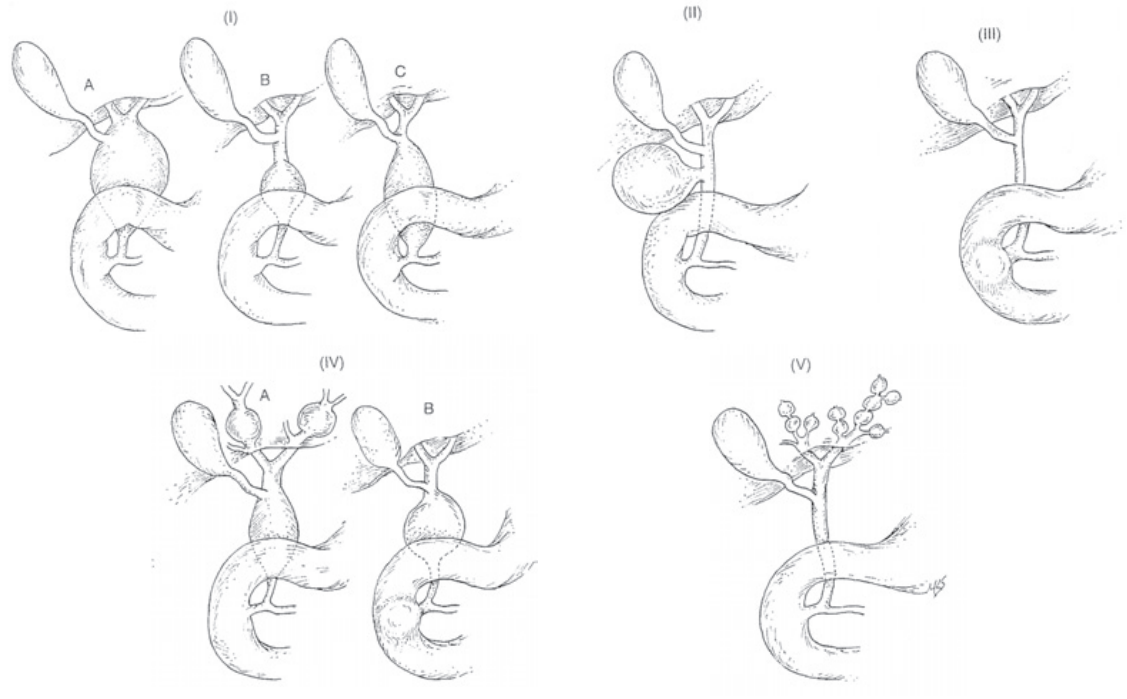

Fig. 2. Todani's classification of choledochal cysts.

Bile ducts cysts are mostly diagnosed in the child age. Clinical manifestations are various and they depend on the size and localization of the cyst – pain in abdomen, icterus, malfunctions in passage, resistance by palpation, cholangoitis, pancreatitis, but often they are asymptomatic. In the adult age the patients with choledochal-cyst have increased occurrence of cholangiocarcinoma in 2.5–28 % of patients (4). Treatment is also surgical and it comprises of a complete extirpation of the cysts. Drainage operations with keeping the cyst are not recommended due to a high risk of a cholangio-carcinoma occurrence (5, 6).

Conclusion

Cystic dilatation of the bile ducts is a rare disease which always requires a surgical solution (7, 8, 10). Among the hepato-entero, or hepatico-entero anastomosis is the Roux-en-Y anastomosis an proven type of anastomosis that we recommend also according to our experiences whereby this type of anastomosis can be constructed by the means of a modified method. There is an increased risk of cholangitis and hepatic cirrhosis occurrence in all patients. Often the patient becomes a suitable candidate for liver transplantation in the future.

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Received July 10, 2007,
Accepted September 20, 2007.