FACTA UNIVERSITATIS
Series: **Medicine and Biology** Vol.13, No 2, 2006, pp. 90 - 93 UC 616.361:618.2

COMPLICATED CHOLEDOCHAL CYSTS IN PREGNANCY

Dragojlo Gmijović[1], Miroslav Stojanović[1], Milan Radojković[1], Ljiljana Jeremić[1], Zlatko Širić[2]

[1]*Surgical Clinic, Clinical Center, Niš, Serbia*
[2]*Radiology Institute, Clinical Center, Niš, Serbia*

Summary. *Choledochal cyst is congenital dilatation of the biliary tract, present primarily in infants. This condition in pregnancy is very rare, and the clinical manifestations are nonspecific and variable. In late pregnancy, the symptoms may exacerbate due to hormonal effects and the compression of the common bile duct by the enlarging uterus. The need for urgent therapy of the complications associated with danger of exposing the mother and fetus to surgical operation and ionizing radiation, always represent a therapeutic challenge to surgeons. Step-by-step treatment of complicated choledochal cyst in pregnancy must be carefully planned, once the diagnosis is established, in order to avoid complication during pregnancy. Definitive surgical management of the choledochal cyst should be delayed until the patient's general physiological condition becomes normal after elective cesarean section. We reported our experience in managing a case of choledochal cyst in pregnant patients, complicated with cholangitis and review the literature regarding choledochal cyst in pregnancy.*

Key words: *Choledochal cyst, pregnancy, cholangitis*

Introduction

A choledochal cyst in pregnancy is rare, and the clinical manifestations are nonspecific and variable (1). In late pregnancy, the symptoms may exacerbate due to hormonal effects and the compression of the common bile duct by the enlarging uterus (2,3). Diagnosis is often delayed until patients are presented with life-threatening complications like cholangitis, jaundice, pancreatitis, cystic rupture. The need for urgent therapy of the complications associated with danger of exposing the mother and fetus to surgical operation and ionizing radiation, always represent a therapeutic challenge to surgeons (3,4). Because of that, step-by-step treatment of complicated choledochal cyst in pregnancy must be carefully planned, once the diagnosis is established.

We reported our experience in managing a case of choledochal cyst in pregnant patients, complicated with cholangitis.

Case Report

A 28-year-old primigravida was in 32nd week gestation with classic Charcot's triade of cholangitis (upper abdominal pain, fever and jaundice). At examination, she had tenderness in the upper abdomen. Abdominal ultrasonography (US) revealed a 16 cm cystic mass in the hepatic hilum area. Serum total bilirubin was 220 mcm/L, and alcaline phosphatase and gama –GT were four times greater than normal values. Aspartate aminotransferase and alanine aminotransferase were significantly higher. Leukocyte count was 14.000 G/L, and CRP value 110 IU. Magnetic resonance imaging (MRI) showed a large type I choledochal cyst, presented like well-defined cystic mass in the liver hilum.

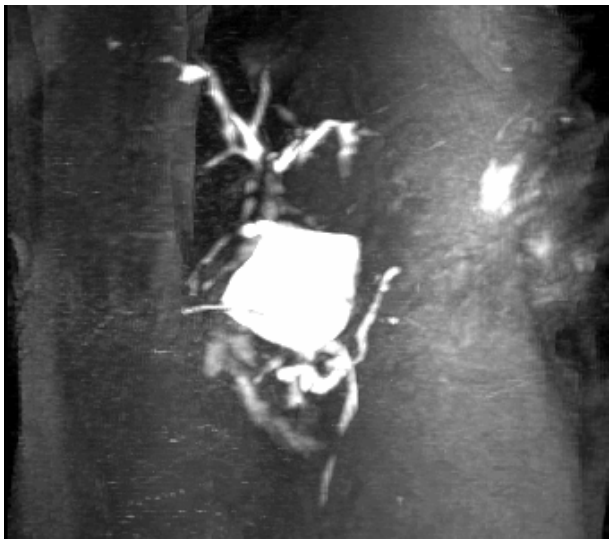


Fig. 1. MRCP –US guided percutaneous drainage of the choledochal cyst

Percutaneous cystic decompression was done under US guidance (Fig. 1), and. patient's condition stabilized after prolonged external drainage. An elective cesarean section was performed seven weeks later.

Six weeks after delivery, ERCP showed definitive mapping of the biliary tract and pancreato-biliary junction, with type I choledochal cyst and good position of the drainage catheter.

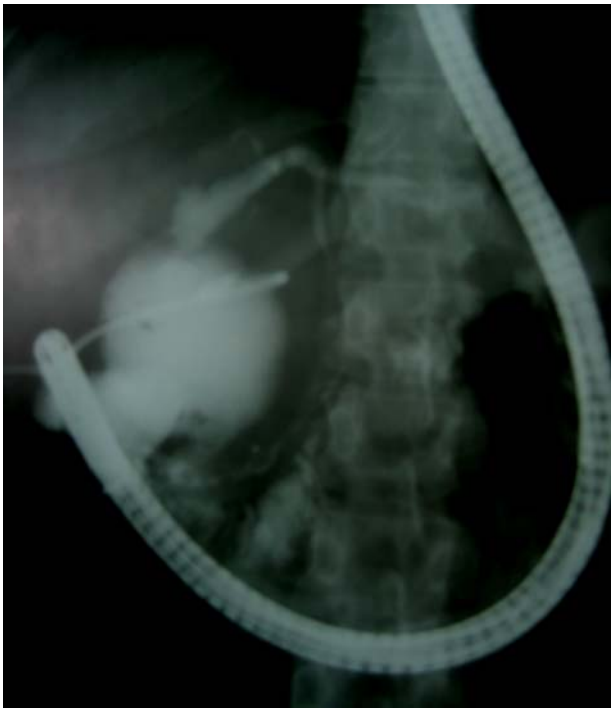


Fig. 2. ERCP after caesarean section

Eight weeks after delivery, patient received definitive near-total surgical excision of the cyst and hepaticojejunostomy with Roux-en Y loop. Her postoperative course was uneventful. Histology specimen showed well-defined cystic wall formed by collagenous fibers covered with one-layer cylindric biliary epithel (Fig. 3).

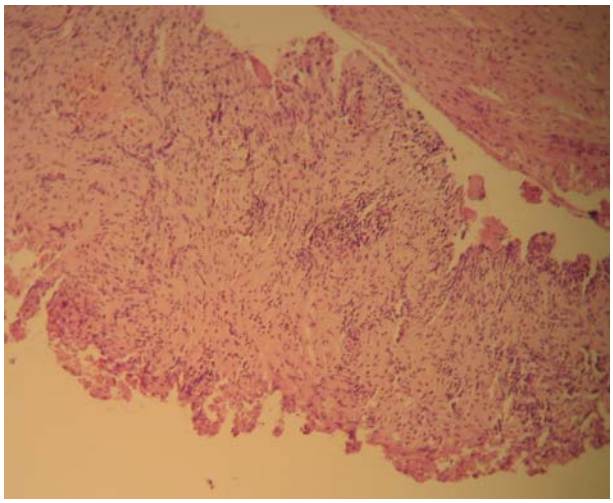


Fig. 3. Histology of the cystic wall. HEx 120

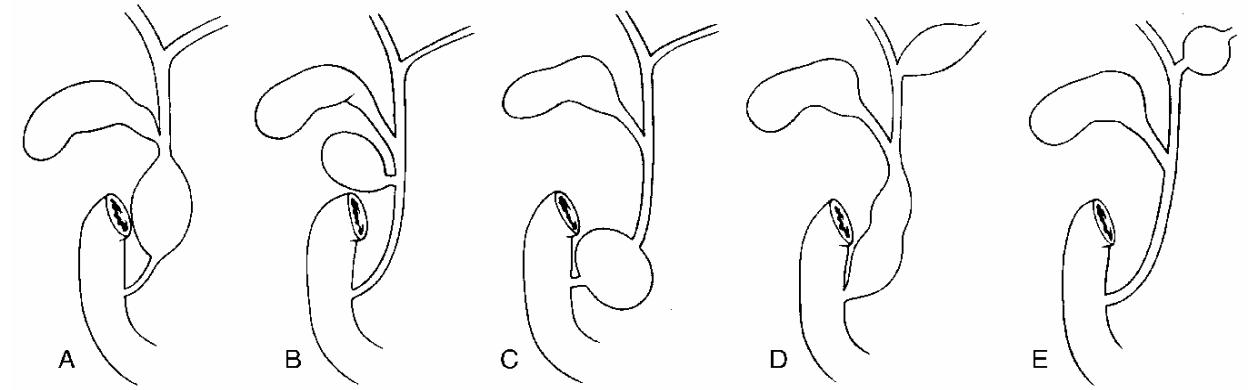
Discussion

Choledochal cyst is a rare congenital abnormality of the biliary tract (2,3), occurring in approximately one in 1-2,000,000 live birth (3,5), presented with the dilatation of either intra or extrahepatic biliary system or both. It affects females 4 times as often as males, and is present mainly in infancy and childhood (60%) (4). It was first described in 1852 by Douglas (4,5). The most accepted classification of the choledochal cysts is Todani's classification (6) into five types (table and graph 1).

The definitive etiology of choledochal cysts is not clear. There are two actual concepts: first concept suggests that choledochal cysts arise from inequality in the vacuolization of the biliary tract in early embryonic life (3). The second is common channel theory, proposed by Babbitt (2), based on an abnomality of the pancreaticobiliary junction and the formation of an abnormally long common channel (greater than 15 mm)

Table and graph 1. Todani's modification of the Alonso-Ley classification of choledochal cyst

Type I (a)	Type II (b)	Type III (c)	Type IVa (d)	Type V (e)
Fusiform dilation of the extrahepatic bile duct	Single saccular dilation or diverticulum of the extrahepatic bile duct	Dilation of the intraduodenal portion of the bile duct	Combined intra-and extrahepatic dilation of the bile duct	Isolated or diffuse intrahepatic biliary dilation (Caroli's disease when associated with hepatic fibrosis)
			Type IVb (d)	
			Multiple dilations of the extrahepatic bile duct	



outside the control of the sphincters of Boyden. Long common chanel permits reflux of pancreatic enzymes into the common bile duct, with persistent inflammation, epithelial denudation, thinning of the bile duct wall and distal obstruction, then eventually cyst formation.

The reported complications of choledochal cyst are cholangitis, jaundice, pancreatitis, cystic rupture, and malignancy (2). Incidence of biliary tract carcinoma in choledochal cysts varied from 2.5% to 17.5% (1-3). The age-related incidence of cyst-associated cancer has been shown to increase from 0.7 % in the first decade of life to 14.3 % after 20 years of age. Because of that, most euthors (1-3) advocated primary cyst excision. The classic characteristic triad (abdominal pain, jaundice and right hypochondrial mass) occurs mostly in childhood, and is seldom seen in adults (4).

Clinical presentation of the choledochal cysts during pregnancy is very uncommon, and it is very difficult to make the diagnosis clinically. The symptoms (pain and jaundice) caused by choledochal cyst in pregnancy may be due to hormonal effect, compression of the bile duct lumen and cyst by the gravid uterus and increase in intra-abdominal pressure during pregnancy (3). All of these symptoms exacerbated in late pregnancy.

Initial screening examination in the evaluation of complicated choledochal cyst is abdominal ultrasound. Difficulties may arise due to distortion of the normal abdominal anatomy and gravid uterus during pregnancy. The cyst may be misdiagnosed as an ovarian tumor or mucocele (8). ERCP or computed tomography (CT) may provide more accurate information, but ionizing radiation should probably be avoided in pregnancy (9). However, MRI can provide diagnostic information without exposing the mother and fetus to ionizing radiation, and the relationship between the choledochal cyst and the biliary tree can be clearly visualized (10-12). Therefore, MRI may be the investigation of choice when a large cystic structure is seen in the expected region of the common bile duct in a pregnant woman (1-3).

Surgical treatment of choledochal cyst in late pregnancy requires critical evaluation not only due to the physiological changes that occur during pregnancy, but also due to the risk of fetal mortality and maternal morbidity. Step-by-step treatment must be carefully planned, once the diagnosis is established. A more conservative approach may be adopted to minimize the risks of surgery (3). In our case, obstructive cholangitis was solved by US guide percutaneous cystic decompression at admission. Elective cesarean section was performed to avoid the stresses that might be imposed on the cyst by labor and vaginal delivery (2,3).

Definitive surgical treatment consists of total or near-total excision of types I, II and IV choledochal cysts with subsequent hepaticojejunostomy with Roux-Y loop. Cysto-jejunostomy is abandoned because of the higher incidence of postoperative complications and danger of malignant potential. Complete excision of the extrahepatic bile duct from the hepatic hilum to the pancreaticobiliary duct junction is the most radical slution, in some cases combined with pancreaticoduodenectomy or hepatic resection. Endoscopic sphincterotomy of type III cysts is the method of choice in the treatment of this condition. Treatment for type IVa cysts (combined extra and intrahepatic biliary dilatation) is still controversial. Total cyst excision including hepatectomy (12,13) has been recommended. Concerning type V cysts, some authors recommended hepatic resection for unilobar Caroli's disease (3).

In conclusion, although choledochal cysts occurs rarely in pregnancy, a high index of suspicion is essential to avoid a delay in diagnosis, particularly when a large cystic structure is seen in the region of the common bile duct. MRI should be used as the definitive diagnostic examination due to high resolution of the biliary tree, without the problems associated with exposing the mother and fetus to ionizing radiation. Treatment of complicated cysts must be carefully planned in order to avoid complications during pregnancy. Definitive surgical management of the choledochal cyst should be delayed until the patient's general physiological condition become normal after elective cesarean section.

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KOMPLIKOVANA CISTA DUCTUS CHOLEDOCHUS-A U TRUDNOĆI

Dragojlo Gmijović[1], Miroslav Stojanović[1], Milan Radojković[1], Ljiljana Jeremić[1], Zlatko Širić[2]

[1]Hirurška klinika, Klinički centar, Niš
[2]Institut za radiologiju, Klinički centar, Niš

Kratak sadržaj: *Holedohalna cista predstavlja kongenitalnu dilataciju bilijarnog stabla koja se uglavnom javlja kod novorođenčadi. Ovo stanje u trudnoći je veoma retko sa nespecifičnim i promenljivim kliničkim manifestacijama. U kasnijoj trudnoći može doći do egzacerbacije tegoba usled hormonskih efekata i kompresije glavnog žučnog voda uvećanim uterusom. Potreba za urgentnom hirurškom terapijom komplikacija povezana sa opasnošću od izlaganja majke i fetusa hirurškoj intervenciji i jonizujućem zračenju, uvek predstavlja terapijski izazov za hirurga. "Step by step" lečenju komplikovane holedohalne ciste u trudnoći mora biti pažljivo isplanirano čim se pojavi dijagnoza, da bi se izbegle komplikacije tokom trudnoća. Definitivno hirurško lečenje holedohalne ciste treba odložiti dok opšte fiziološko stanje pacijenta ne postane normalno nakon elektivnog carskog reza. Saoštili smo naše iskustvo u tretmanu slučaja holedohalne ciste kod trudnica, komplikovanih olangitisom i dali pregled literature u vezi holedohalne ciste u trudnoći.*

Ključne reči: *holedohalna cista, trudnoća, holangitis*