

Spontaneous Rupture of Choledochal Cyst during Pregnancy: A Case Report:

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Abstract: Choledochal cyst is a rare congenital surgical entity, first reported by Weber in 1934, present as lump, jaundice and abdominal pain in young children and adolescents. Its complications include cholangitis, pancreatitis, liver abscess, biliary cirrhosis and portal hypertension. Rarely it can have spontaneous rupture in 2% of cases causing biliary peritonitis. We are presenting our experience of spontaneous rupture of choledochal cyst during pregnancy with brief review of literature.

Keywords: Choledochal cyst, peritonitis, pregnancy.

I. Case Report:

A 23 years old female in her first trimester, was admitted with upper abdominal pain and mild jaundice of 12 days duration. She was having recurrent attacks of such pain since childhood. Pain was colicky in nature and often associated with fever. She noticed mild jaundice 5 days ago not associated with itching. On examination she was jaundiced, afebrile and anaemic. Abdominal examination revealed a tender mass in right hypochondrium, firm, mobile. Mild hepatomegaly. No other organomegaly. Gynaecological examination proved 13 weeks viable pregnancy. Investigations; Hb: 9.5 gm%, TLC 12600/cmm with polymorphonuclear count 74%, liver enzymes raised: SGOT 54 and SGPT 62 IU, S. bilirubin 2.6 mg% direct type being greater. Alk. Phosphatase was 463 IU. Ultrasound of abdomen was suggestive of choledochal cyst type 1 with diameter of 18mm and few calculi in it. Gall bladder was distended but no calculi. Liver was mildly enlarged and no IHBR dilatation. Rest of abdomen was normal. With a diagnosis of cholangitis, she was managed with antibiotics and improved. In view of pregnancy, she was discharged after 7 days and advised to come for elective surgery after delivery.

She later on developed severe abdominal pain and collapsed at 7th month of pregnancy at her village and was operated at some peripheral hospital. Biliary peritonitis was found but no perforation seen. A drain tube was put in and she was sent to our hospital.

On arrival at our hospital, she was stable, afebrile and mildly jaundiced. Abdominal examination was unremarkable except a drain tube draining about 240ml bile per day. Since the previous details were known to us, she was evaluated further by ERCP and confirmed to have ruptured choledochal cyst. Biliary stenting was done and she improved gradually and drainage of bile eventually stopped. She was again discharged with advice to come after delivery for elective surgery.

Later on it was brought to our notice that she expired in post partum period of some unknown cause.

II. Discussions

Choledochal cysts represent an uncommon but important cause of jaundice in infants and of abdominal pain in children. More than 60% of cases are reported from Asia, Japan being the highest. More common in females (75%)¹ Yamaguchi et al² reported analysis of 1433 cases of congenital choledochal cysts in Japanese literature.

The classical triad of jaundice, right upper quadrant mass with abdominal pain is seen in less than 17% of the cases presenting to any tertiary care centre. 6 Two-thirds the cases are diagnosed before the age of 10 years and 85% cases before puberty. Cystic dilatation of the extrahepatic tree can be diagnosed using ultrasound as early as 15 weeks of gestation and a half of these would continue to be asymptomatic in early infancy with the other half presenting with jaundice and a palpable mass. The most important differential diagnosis is biliary atresia.³

It is more common in female sex and can present with following complications:

1. Choledocholithiasis (Intra and extrahepatic)
2. Cholangitis.
3. Acute pancreatitis (often recurrent).
4. Cholangiocarcinoma.
5. GB carcinoma.
6. Liver abscess.

7. Biliary cirrhosis.
8. Portal hypertension.
9. choledochal cyst complicating pregnancy.
10. Choledochal cyst have been classified into 4 types depending upon nature of dilatation, generalized or localized. Todani's classification is commonly used and is as follows:

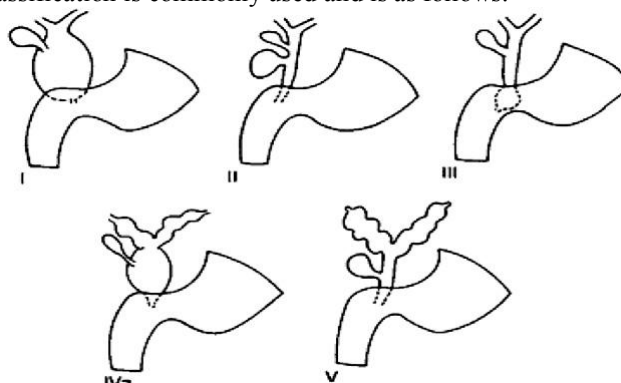


Fig. 1 Classification of choledochal cysts (Todani). Type I:

Cystic or fusiform dilatation of hepatic and common bile, duct (40%e85%). Type II: Diverticulum of the common bile, duct (2%e3%). Type III: Choledochocele:Intrahepatic CBD, dilatation (1.4%e5.6%). Type IVa: Intra- and extrahepaticbile duct dilatation (18%e20%). Type V: Intrathepatic bile duct dilatation

In favour of the literature nowadays it would be possible to systematize this malformation and make a division in two groups, depending on the cholangiographic findings and clinical presentation: 1. Cystic dilatations with a clinical neonatal presentation or beneath 2 years. 2. Fusiform dilatations with a later clinical predominance and associated frequently to pancreatitis and anomalous pancreatobiliary junction. Choledochocele is an entity that must be considered not only for its etiology but for its clinical presentation and treatment.⁴

Primary cyst excision and biliary Roux-en-Y reconstruction is the treatment of election. Regular long-term review of these patients is mandatory in the surveillance of sub-clinic cholangitis and the risk of possible long-term malignance of this entity.

Rupture in infancy and adulthood is common as reported by Jindal B and Bhatnagar V in 2008³. They have reported a rupture in two and a half months old infant.

Rupture of choledochal cyst especially during pregnancy is a major catastrophe. Usually it occurs during late stages of pregnancy as was seen in the present case. Many previously asymptomatic women present during pregnancy for a number of reasons, including obstruction of the cyst by the gravid uterus, further stasis of pancreaticobiliary secretions because of biliary hypomotility, and cyst rupture because of increased intra-abdominal pressure during pregnancy and labour. Presenting symptoms are usually abdominal pain, fever and vomiting, usually due to cholangitis or pancreatitis. Diagnosis by ultrasonography may be difficult because of obscuration and alteration of normal anatomy by the gravid uterus. Given that computed tomography scans expose the fetus to ionizing radiation, magnetic resonance imaging (MRI) has been recommended as the imaging modality of choice. Management of CC during pregnancy is difficult because of the surgical risk to both mother and fetus. Incidentally found CC should be followed with serial ultrasonography, and symptoms or rapid cyst enlargement should be treated conservatively as we had done. High intra-abdominal pressure during labour may cause cyst rupture, and many surgeons recommend elective caesarean section in the third trimester, definitive cyst excision and hepaticostomy should be performed after delivery. Cyst rupture may mandate emergent surgery for bile evacuation and washout, but this should be followed with external drainage, and definitive surgery should be performed during the postpartum period.⁶ This case was dealt at some peripheral centre without the knowledge of her past check up and operating surgeon has done rightly external drainage and sent to us for further evaluation.

T diamond et al⁷ first reported a case of ruptured choledochal cyst in pregnant female. In this patient during caesarean section obstetrician has noted biliary fluid in the abdomen and a surgical consultation was sought. Due to lower transverse incision surgeon could not explore much and a drain tube was put only. Post operatively condition deteriorated and was reexplored on forth day and ruptured cyst was found. Cystoduodenostomy was done.

The cause of rupture during pregnancy remains a dilemma but it seems to result from sluggish motility seen during pregnancy. Other factors may be pressure of gravid uterus on cyst wall, distal obstruction and calculus formed in cyst.³

Treatment of choledochal cyst, if detected in pregnancy, is conservative. N F Hopkins et al⁷ reported 7 cases of choledochal cysts out of which two were pregnant in late trimester and were treated after delivery by excision and hepaticojejunostomy.

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