CASE REPORT

Ruptured choledochal cyst during early pregnancy with successful fetomaternal outcome

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SUMMARY

Choledochal cyst is a rare congenital malformation which is usually found in young children and adolescents but can rarely present in the adult age group. It can present with several complications like cholangitis, pancreatitis, biliary cirrhosis, portal hypertension, liver abscess, malignancy and rarely spontaneous rupture (in 2% of cases) causing biliary peritonitis. A case of spontaneous rupture of choledochal cyst during second trimester of pregnancy is reported where the patient was managed by minimal surgery, peritoneal washout, placement of T-tube and planned for definitive treatment in the postpartum period. The pregnancy continued till 35 weeks with favourable fetomaternal outcome.

BACKGROUND

Choledochal cyst is a congenital malformation where intrahepatic or extrahepatic bile ducts become dilated and thin walled. It is a rare malformation with the incidence of approximately 1 in 1000 live births in the Asian population as compared to 1 in 100 000–150 000 live births in western countries.¹ Rupture of choledochal cyst during pregnancy may pose a diagnostic challenge as clinical features of ruptured cysts are obscured by the physiological changes of pregnancy.² Many complications of choledochal cyst have been reported during pregnancy but spontaneous rupture of the cyst is rarely reported as most ruptures occur in childhood.² A rare case of spontaneous rupture of choledochal cyst during second trimester of pregnancy is being reported which was managed with conservative surgery along with continuation of pregnancy till 35 weeks of gestation.

CASE PRESENTATION

A 24-year-old woman, primigravida with 16 weeks of gestation presented with severe pain in upper abdomen and vomiting to the gynaecology casualty. There was history of anorexia, epigastric burning pain and fever for the past 1 week for which she took treatment from some other hospital near her residence in the form of pain killers, antiemetics and antacids. General physical examination revealed mild anaemia and jaundice, pulse rate of 120/min, blood pressure 77/36 mm Hg and respiratory rate was 34/min. Abdominal examination revealed enlarged liver with upper abdomen tenderness, distension and ascites. Uterus was palpable at 16 weeks corresponding with the period of gestation.

INVESTIGATIONS

Haematological and biochemical parameters revealed haemoglobin 10.4 g/dL (reference range: 11–13 mg/dL), total leucocyte count $11.4 \times 10^9/L$ (reference range: $4-11 \times 10^9/L$), platelet count $230 \times 10^9/L$ ($150-400 \times 10^9/L$), $1.5-4.5 \times 10^5/mm^3$), serum bilirubin 2.09 mg/dL (reference range: 0.2–1.2 mg/dL) with direct bilirubin 2.04 mg/dL (reference range: 0.1–0.4 mg/dL), serum glutamic oxaloacetic transaminase (SGOT) 65.1 IU/L (reference range: 8–48 U/L), serum glutamic pyruvic transaminase (SGPT) 121.9 IU/L (reference range: 7–56 U/L), alkaline phosphatase (ALP) 530 IU/L (reference range: 45–115 U/L), random blood sugar 138 mg/dL and serum amylase 41.1 U/L (reference range: 40–140 U/L). No abnormality was detected in her coagulation profile and kidney function tests. Viral markers (HIV, hepatitis B virus surface antigen (HBs Ag), hepatitis C virus, hepatitis A virus) were negative. Upper abdomen ultrasound (figure 1) revealed a mildly enlarged liver, moderate intrahepatic biliary dilatation, a large cystic lesion of approximately $9 \times 9 \times 4.6$ cm in the region of porta, communicating with the common hepatic duct and closely abutting gall bladder and pancreatic head. Multiple small calculi were seen within the lesion with posterior acoustic shadow. Mild splenomegaly along with moderate amount of fluid in abdominal cavity was present. Findings were suggestive of ruptured choledochal cyst. Obstetric ultrasound showed single live fetus of 18 weeks gestation. Magnetic resonance cholangiopancreatography (MRCP) was performed and a diagnosis of choledochal cyst Todani type 1a was made (figure 2). A final diagnosis of biliary peritonitis secondary to ruptured choledochal cyst was made.

TREATMENT

Patient underwent laparotomy after 12 hours of presentation followed by cholecystectomy and T-tube insertion for drainage of biliary fluid in the surgery department. Patient remained on ventilator support for 72 hours followed by improvement in clinical condition. The drainage tube remained throughout pregnancy draining 600–800 cc of biliary fluid every day. Her follow-up obstetric ultrasound revealed severe oligohydramnios and scaphocephaly at 26 weeks of gestation. At 28 weeks of gestation, patient developed breathlessness and right-sided pneumothorax. This was managed by right intercostal drainage and antibiotics. This

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Figure 1 Ultrasound image of epigastric region showed large anechoic cystic lesion (white arrow) with dependent sludge (black arrow) causing dilatation of intrahepatic biliary radicals.

could be a spontaneous primary pneumothorax, or in rare case, the possibility of air entering via some pre-existing defect in the diaphragm due to the presence of drain in the abdomen can be postulated. Successive obstetrics ultrasound revealed severe oligohydramnios till 30 weeks (figures 3 and 4), but this improved at 32 weeks of gestation spontaneously. The patient had multiple admissions in the surgery and gynaecology departments throughout the course of pregnancy but was managed conservatively. Her haematological and biochemical parameters were within normal limits in all admissions apart from mild jaundice (total bilirubin varying from 2.09 to 5.16 mg/dL). At 35 weeks of gestation, patient presented with leaking per vagina and preterm labour pains. Her haematological and biochemical parameters were within normal limit (haemoglobin 11.9 g/dL and total bilirubin 0.76 mg/dL). She delivered a healthy male baby of 2.16 kg vaginally.

OUTCOME AND FOLLOW-UP

Repeat MRCP was done in the postpartum period, which revealed similar choledochal cyst as in the previous MRCP (figure 5). Patient did well in the postpartum period and was planned for elective surgery for choledochal cyst resection after 6 weeks.

Figure 2 Maximum intensity projection coronal images show a large choledochal cyst (big arrow) and moderate dilatation of intrahepatic biliary radicals (small arrow).

Figure 3 Choledochal cyst which showed reduced amount of liquor on 28 weeks scan .

DISCUSSION

Focal dilations of biliary tract are known as biliary cyst and lesions affecting only the common bile duct are known as choledochal cyst. The most accepted classification for these cyst has been proposed by Todani.³ According to this classification, there are five types, of which type 1 is the most common.

- ▶ Type 1: focal or complete dilatation of the extrahepatic bile duct.
- ▶ Type 2: biliary diverticulum arising from supra-duodenal part of the bile duct.
- ▶ Type 3: focal dilatation of the intramural portion of distal bile duct.
- ▶ Type 4: multiple communicating intrahepatic and extrahepatic biliary cysts.
- ▶ Type 5: also known as Caroli's disease and these are cystic dilations of intrahepatic bile ducts.

These cysts are usually seen in childhood and diagnosis during pregnancy is rare. Congenital choledochal cyst can be diagnosed

Figure 4 Choledochal cyst which showed reduced amount of liquor on 28 weeks scan.

Figure 5 T2 weighted axial image of abdomen shows large choledochal cyst (big arrow) with large number of dependent small calculi (small arrow).

in the fetus as early as 15 weeks of gestation by ultrasonography and is commonly seen in female fetuses. On the basis of review of the literature, this malformation can be divided into two types, cystic dilatation with neonatal presentation and fusiform dilatation with later presentation or adult presentation. Latter one usually presents with complications. Treatment of choice for choledochal cyst is primary cyst excision and biliary Roux-en-Y reconstruction.⁴ Long-term follow-up is usually required due to risk of developing malignancy. Choledochal cyst in an asymptomatic female can complicate during pregnancy leading to increased fetomaternal morbidity and mortality. Complications can be in the form of cyst obstruction by gravid uterus, stasis of pancreaticobiliary secretions, preterm labour pains, cholangitis, abortions and spontaneous rupture.² The risk of spontaneous rupture of cyst ranges between 1.8% and 2.8%.² Spontaneous rupture of the cyst is more commonly seen during childhood and adulthood and occurrence during pregnancy is very rare. Choledochal cyst rupture during pregnancy is a major catastrophe.⁵ Many theories have been suggested for spontaneous choledochal cyst rupture and the most often reported theories are weakness of the cyst wall with raised pressure within the cyst due to the obstruction of bile duct by strictures or stones, injury caused by numerous interventions, that is, amniocentesis, back flow of amylase rich fluid in biliary duct, the presence of abnormal pancreaticobiliary junction and increased pressure within cyst wall during labour.² Cyst rupture is usually seen in the late stages of pregnancy⁴ but in this case, it was seen in early pregnancy. Patients commonly present with abdominal pain, vomiting and fever due to pancreatitis and cholangitis. Diagnosis of choledochal cyst is slightly difficult during pregnancy because of changes in the normal anatomy and masking by the gravid uterus. MRI is the modality of choice for diagnosis as CT is associated with radiation exposure to the fetus. Various management options have been reported, but generally it is recommended that the intervention be as least invasive as possible given the risks to mother and fetus.

Incidental diagnosis of cyst during pregnancy should be monitored with ultrasonography and any cyst rupture or rapid enlargement in the cyst should be managed with conservative surgery. Spontaneous cyst rupture requires urgent bile evacuation in an

emergency; external drainage should be done either with percutaneous transhepatic drain or T-tube drainage as done in our case and definitive surgery should be performed preferably in the postpartum period.² Ultrasound-guided percutaneous transhepatic choledochal cyst drainage had also been used by few authors.⁶ The first case of choledochal cyst rupture during pregnancy was reported by Diamond *et al* in 1986⁷; since then many similar cases were reported but spontaneous rupture of cyst during pregnancy with continuation of pregnancy and successful outcome is still rare. As we have already discussed, there is a very high risk of spontaneous cyst rupture during labour because of the raised intra-abdominal pressure and it was recommended by some studies to conduct an elective caesarean section in these patients and final/complete surgery should be conducted in the postpartum period preferably after 6 weeks of delivery. This case was operated at our tertiary care centre for choledochal cyst rupture in the second trimester and external drainage was done with T-tube. The patient delivered a healthy baby spontaneously at 35 weeks of gestation. In review of the literature, we have not found any case of choledochal cyst rupture during early trimester of pregnancy followed by successful outcome. This case report is rare and explains a good fetal prognosis in pregnant patients with choledochal cyst rupture managed with minimal surgery.

Learning points

- ▶ Choledochal cyst presents commonly in infancy and adulthood but very rarely they can be found during pregnancy, so clinicians need to be aware of this condition.
- ▶ Delayed diagnosis and treatment may be catastrophic for the child and the mother. Patients should be managed in a tertiary care specialised centre with all the facilities available where delivery can be planned carefully.
- ▶ Such cases can be managed as reported with least invasive options to minimise risks and definitive treatment deferred until postpartum, if possible.

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