Case Report

Pregnancy in Biliary Atresia after Kasai Operation Complicated by Portal Hypertension

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Hepatic portoenterostomy or Kasai operation has been widely accepted as the standard therapy for biliary atresia. Recently, more female patients have grown up and reached adulthood; therefore, pregnancy in women with biliary atresia is sometimes inevitable. The authors report a 17-year-old woman with biliary atresia post Kasai operation at 3 months of age. After the operation, she became jaundice-free but developed portal hypertension with abnormal liver function. She had several episodes of esophageal variceal bleeding and was treated by beta-blocker and endoscopic sclerotherapy. Since then, she was lost to follow up for nearly 2 years. She came back again with 12 weeks of gestation with no symptoms of gastrointestinal bleeding for antenatal care. At 32 weeks of gestation, she presented with severe hematemesis from variceal bleeding and had thrombocytopenia from hypersplenism. She was treated with somatostatin analogue, fluid and blood component replacement and other supportive treatments. Cesarean section was performed when she was stable at 33 weeks of gestation. After the operation, her clinical status was improved and had no other complications. Her baby experienced complications of prematurity but improved after treatment. Pregnancy may affect the natural course of portal hypertension and worsen the clinical outcome. Pregnancy should be avoided in patients with portal hypertension, however it is not contraindicated. Pregnancy in biliary atresia patients needs intensive prenatal care.

Keywords: *Biliary atresia, Hepatic portoenterostomy, Pregnancy, Portal hypertension*

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Biliary atresia is characterized by complete obstruction of bile flow as a result of the destruction or absence of all or a portion of the extra-hepatic bile ducts. The disorder occurs in 1 in 10,000 live births and accounts for one-third of cases of neonatal cholestatic jaundice[1]. If it is unrecognized or uncorrected, cirrhosis inevitably develops within 3 to 6 months after birth[2]. The standard treatment for biliary atresia is Kasai operation[3]. In addition, the age of the patient at the time of surgery is critical. The results of surgery are dramatically better before 60 days of age[4]. After the operation, 50% of the cases experienced portal hypertension and hypersplenism[5]. Liver transplantation is essential in which portoenterostomy has failed. Without any treatment, death usually occurs within 2 years[1]. Because of the effectiveness of the operation and good medical management, a number of patients reach adulthood, become pregnant and experience birth giving. For instance, Shimaoka et al reported the results of a nationwide study on pregnancy of biliary atresia patients in Japan and described 16 successful pregnancies[6].

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Case Report

A biliary atresia patient had undergone Kasai operation at 3 months of age. After the operation, she became jaundice-free but later portal hypertension developed with hypersplenism, abnormal liver function and thrombocytopenia. She had multiple admissions because of hematemesis from esophageal variceal bleeding. She was treated with beta-blocker and twice endoscopic sclerotherapy. Since then, she was lost to follow up for nearly 2 years. At the age of 17, she came back again with 12 weeks of gestation with no symptoms of gastrointestinal bleeding during her antenatal care. Ultrasonography, however, showed liver cirrhosis and signs of portal hypertension with suspected pseudoaneurysm adjacent to the splenic artery. The magnetic resonance angiography confirmed an aneurysmal dilatation of splenic vein. Because of her thrombocytopenic condition, splenectomy was offered as the option of treatment but the patient declined. Gastroduodenoscopy at 22 weeks of gestation demonstrated obliterated esophageal varices at esophagus, obliterated gastric varices and diffuse mosaic pattern of the mucosa at the stomach. At 32 weeks of gestation, she was admitted due to severe hematemesis. She had a history of melena for 3 days before hematemesis about 300 ml with fainting. In addition, her fetal movement decreased.

On examination, the vital signs were stable. The conjunctivae were moderately pale. Examination of her heart and lungs revealed no abnormality. The abdomen was soft, no tenderness, no superficial vein dilatation; splenomegaly was detected but the liver could not be palpated. The fetal heart rate was normal and the uterus height was 2/4 above the umbilicus. The uterine contraction was noted. The vaginal examination found the cervical os closed. Laboratory analysis revealed hematocrit of 27.6%, hemoglobin of 9.5 g/dl, platelet of 69,000 cell/mm^3, aspartate aminotransferase of 33 IU/L (normal value: 0-38 IU/L), alanine aminotransferase of 42 IU/L (normal value: 0-38 IU/L), alkaline phosphatase of 164 IU/L (normal value: 39-117 IU/L), total protein of 4.8 g/dL (normal value: 6.6-8.7 g/dL), albumin of 2.6 g/dL (normal value: 3.4-5.5 g/dL), total bilirubin of 0.55 mg/dL (normal value: 0.00-1.00 mg/dL), prothombin time of 12.1s (normal value: 11.0-15.0 s). Ultrasonography showed breech presentation and an approximately 1,700 grams fetus. Biophysical provocative test was normal. Gastroduodenoscopy found one esophageal varices lesion without red color sign at lower esophagus, large amounts of clot blood at the gastric fundus. She was given Sandostatin IV drip, proton-pump inhibitor, ceftriazone, pack red cells, pooled platelet, vitamin K and fluid replacement.

Throughout her hospital course, the patient continued to have hematemesis and coffee ground fluid from the nasogastric tube off and on, but it was still controllable. On the fourth day of admission, there was watery discharge from her vagina. The obstetric team decided to perform a Cesarean section when her clinical condition was stable. On the eighth admission day at 33 weeks of gestation, Cesarean section was scheduled. Laboratory analysis, prior to the operation, revealed hemoglobin of 11.9 g/dl, hematocrit of 34% and platelet concentration of 36,000 cells/ mm^3. She got pooled platelet before the operation and internal jugular venous cut down was performed. Low transverse Cesarean section with tubal resection was carried out. Her blood loss was approximately 400 ml. No complication occurred following delivery. After delivery, she was fine. She had neither hematemesis nor melena. Her laboratory analysis revealed hemoglobin of 11.1 g/dl, hematocrit of 31.1% and platelet concentration of 44,000 cells/mm^3. She was discharged with improved clinical status.

Her baby was a boy, weight 1,960 grams. His Apgar scores were 5, 8 at 1 and 10 minutes after birth respectively. He later developed neonatal hypoglycemia, neonatal jaundice, and transient tachypnea of the newborn. After 7 days of treatment, his status was well and he was discharged.

About 3 months later, she came for follow up at the OPD. Her clinical condition was well. She had not had hematemesis since the delivery. Her baby was doing well.

Discussion

Kasai portoenterostomy was developed for 30 years ago; it has been accepted as the definitive therapy for biliary atresia. Earlier patients have grown and reached their adulthood. Apparently, pregnancy in women with biliary atresia has become more frequent[3]. The pregnancy may affect the natural course of the disease and even worsen the clinical outcome. The main risk for complication is variceal bleeding which may be life-threatening[3,6]. Some studies reported massive bleeding from esophageal varices that leaded to circulatory failure and sometimes abortion, hypoproteinemia, serious atopic dermatitis, placental abruption and fetal distress. Problems after delivery are deterioration of liver function, which might need liver transplantation, and ascending cholangitis[6]. For

babies, most of them were born at term with low birth weight. Neither biliary atresia nor any congenital anomaly was found. Sometimes it is necessary to terminate pregnancy as they had problems from prematurity such as respiratory distress syndrome, neonatal jaundice[3].

The route of delivery depends on the patients' clinical status. In stable with minimal esophageal varices, the vaginal delivery with a short second stage should be encouraged. However, vaginal delivery may carry the risk of rupture of the varices in unstable varices. On the other hand, during cesarean section, one should be aware of hemorrhage due to coagulation factors deficiency from liver impairment[7]. In the present report, a biliary atresia patient developed portal hypertension with gastroesophageal varices, hypersplenism, and thrombocytopenia, after Kasai operation. Pregnancy made her varices worsen. She underwent cesarean operation to prevent further bleeding from the varices. Her baby encountered some problems from prematurity and fetal distress. However, the newborn was well after proper treatment.

In conclusion, pregnancy should be avoided in patients with portal hypertension. Nevertheless, they are not contraindicated. Management of the complications arising during pregnancy is similar to management in the non-pregnant patients. Provision of proper care for mother and fetus requires the skills of multiple specialists such as specialists in maternal-fetal medicine, gastroenterologist, nutritionist, and surgeon[8].

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การตั้งครรภ์ในผู้ป่วยโรคท่อน้ำดีตีบตันที่มีภาวะความดันสูงในเส้นเลือดพอร์ตัลภายหลังการผ่าตัดโดยวิธีของคาซาย

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การผ่าตัดต่อระหว่างท่อน้ำดีของตับกับลำไส้หรือที่เรียกกันว่าการผ่าตัดของ Kasai นั้นถือว่าเป็นการผ่าตัดที่เป็นมาตรฐานในการรักษาโรคท่อน้ำดีตีบตัน ซึ่งให้ผลภายหลังการผ่าตัดที่ดีทำให้มีผู้ป่วยสามารถเจริญเติบโตเข้าสู่วัยผู้ใหญ่ ซึ่งบางครั้งในเพศหญิงก็ไม่สามารถหลีกเลี่ยงการตั้งครรภ์ได้ ผู้ศึกษาได้รายงานผู้ป่วยโรคท่อน้ำดีตีบตันเพศหญิง อายุ 17 ปี ได้รับการผ่าตัด Kasai ตั้งแต่อายุ 3 เดือน ภายหลังการผ่าตัด ผู้ป่วยไม่มีภาวะดีซ่าน แต่มีภาวะความดันเลือดพอร์ตัลสูงและการทำงานของตับบกพร่อง ผู้ป่วยมีเลือดออกจากเส้นเลือดขอดที่หลอดอาหารหลายครั้งและได้ทำการรักษาโดยได้ยา beta blocker และการทำ endoscopic sclerotherapy ผู้ป่วยไม่มาตรวจติดตามประมาณ 2 ปี ภายหลังได้กลับมาเมื่อตั้งครรภ์ได้ 12 สัปดาห์เพื่อมาฝากครรภ์ ซึ่งขณะนั้นยังไม่มีภาวะเลือดออกในทางเดินอาหาร เมื่ออายุครรภ์ได้ 32 สัปดาห์ ผู้ป่วยมาโรงพยาบาลด้วยอาเจียนเป็นเลือดมากจากเส้นเลือดขอดและมีเกล็ดเลือดต่ำจากที่มีม้ามโต ได้รับการรักษาด้วย somatostatin analogue เลือดและสารประกอบของเลือดรวมทั้งการรักษาตามอาการอื่น ๆ ได้ทำการผ่าท้องคลอดเมื่ออาการดีขึ้นขณะอายุครรภ์ 33 สัปดาห์ หลังการผ่าตัดไม่มีภาวะแทรกซ้อนและอาการก็ดีขึ้น บุตรของผู้ป่วยมีภาวะแทรกซ้อนที่เกิดจากการคลอดก่อนกำหนดและอาการดีขึ้นภายหลังการรักษา การตั้งครรภ์นั้นส่งผลให้การดำเนินโรคของภาวะความดันเลือดพอร์ตัลสูงรุนแรงมากขึ้น ดังนั้นควรหลีกเลี่ยงการตั้งครรภ์ในผู้ป่วยที่มีภาวะนี้ อย่างไรก็ดีก็ไม่ได้เป็นข้อบ่งห้ามที่จะตั้งครรภ์ ดังนั้นในผู้ป่วยภาวะความดันเลือดพอร์ตัลสูงที่ตั้งครรภ์นั้นต้องการการดูแลทั้งก่อนตั้งครรภ์ และขณะตั้งครรภ์อย่างใกล้ชิด