ZKTB

CASE REPORT

Successful Management of Pregnancy In Non-Cirrhotic Portal Hypertension: A Case Report

Non-Sirotik Portal Hipertansiyonda Gebeliğin Başarılı Yönetimi: Olgu Sunumu

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ABSTRACT

Introduction: *The co-existence of pregnancy and liver disease is considered to be a rare and complex clinical situation. Besides pregnancy has specific complications even in a healthy liver, the previous liver damage associated to portal hypertension develops additional risks as a result of marked hemodynamic disturbances.*

Case: *We report the management of a pregnancy of a 22-year-old patient without additional complications beside NCPH and a history of prophylactic endoscopic ligation of esophageal varices. The pregnancy continued until term and cesarean section was performed successfully.*

Conclusion: *In non-cirrhotic portal hypertension (NCPH) the liver function is usually preserved, and the increased risk of bleeding from esophageal varices in pregnancy is open to debate.*

Keywords: *Liver disease, pregnancy, portal hypertension*

ÖZET

Giriş: *Gebelik ve karaciğer hastalığı nadir ve karmaşık bir durum olarak ele alınır. Sağlıklı bir karaciğerde bile gebeliğe özgü komplikasyonların yanı sıra, önceki portal hipertansiyonuna bağlı karaciğer hasarı varlığı belirgin hemodinamik sorunlara yol açan ilave riskler geliştirir.*

Olgu: *22 yaşında non-sirotik portal hipertansiyonlu profilaktik endoskopik varis ligasyonu yapılmış bir hastanın ek komplikasyon olmayan gebeliği sunulmuştur. Gebelik terme kadar sorunsuz izlenmiş ve başarılı bir sezaryen gerçekleştirilmiştir.*

Sonuç: *Non-sirotik portal hipertansiyonda karaciğer fonksiyonları genellikle korunur ve gebeliğe bağlı özofagus varis kanaması riskindeki artış tartışmaya açıktır.*

Anahtar Kelimeler: *Karaciğer hastalıkları, gebelik, portal hipertansiyonı*

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Submitted: 30.09.2014
Accepted: 16.11.2014
DOI: http://dx.doi.org/10.16948/zktb.39119

INTRODUCTION

The co-existence of pregnancy and liver disease is considered to be a rare and complex clinical situation (1-4). Besides pregnancy has its specific complications with a healthy liver, the previous liver damage associated to portal hypertension develops additional risks as a result of marked hemodynamic disturbances (1-4). Portal hypertension is not a contraindication to pregnancy (5). The frequency and severity of complications during pregnancy are related to the severity of liver function (1, 6). In patients with cirrhosis and significant portal hypertension, 50% will develop maternal/fetal complications (1, 7). Pregnancy should be avoided in women with previous history of variceal bleeding and liver insufficiency (1, 2, 8). However, in those with well-compensated cirrhosis, pregnancy generally is considered to be safe (1, 6). Since the experiences on the issue are currently limited, no definite guideline regarding management of pregnancy and delivery exists. A meticulous follow-up and a proper management of complications with multidisciplinary approach in a tertiary care center are necessary. We report the management of an uncomplicated pregnancy of a young patient with NCPH in whom prophylactic endoscopic variceal band ligation and cesarean section were performed successfully.

CASE REPORT

A 22 year old patient admitted to our clinic with 6 weeks of pregnancy and ongoing comorbid liver disease. In past medical history, the patient was admitted to our clinic with intrauterine fetal death at 32 weeks of gestation in 2007. Since physical examination revealed splenomegaly, she was referred to a tertiary center where she made an uneventful vaginal delivery thereafter. In splenomegaly evaluation she had normal liver function tests, and negative viral hepatitis panel. As she underwent endoscopy, grade II-III esophageal varices were found. Immunological studies revealed all negative serology for anti-neutrophil cytoplasmic antibodies

(ANCA), smooth muscle antibodies (SMA), and anti-liver-kidney microsomal antibodies (Anti-KLM1), except for antinuclear antibodies (ANA). Finally, the patient was diagnosed as idiopathic portal hypertension according to the liver biopsy revealing hepatoportal sclerosis in 2009. As she was admitted to our clinic with 6 weeks' pregnancy in December 2009, she was informed about the complications, but the patient decided to continue her pregnancy.

An endoscopic examination of the upper gastrointestinal tract was performed that showed grade II-III varices of the lower esophagus. No fundal varices were seen. We referred the patient to a gastroenterology clinic for endoscopic variceal ligation. Three columns of esophageal varices were ligated (Speedband; Microinvasive, Boston Scientific Corporation, Watertown, Mass.). She was instructed to modify her activities to avoid straining and valsalva maneuver, and called for controls every 4 weeks until 28th week of gestation. Hemoglobin, hematocrit values and liver function tests were in normal range. Platelet count decreased to 60,000/mm^3 least. Maternal stool guaiac tests to screen occult bleeding were negative, and there were not any clinical symptoms during these follow-ups. Esophageal varices, reduced to grade I-II, were seen in the course of endoscopies done in the second and third trimester. After 28th week of gestation we continued the clinic visits every 2 weeks. Ultrasonography and fetal non-stress tests showed a normal growing fetus. Pregnancy reached term free of problems. When the uterine contractions started, the patient was admitted to delivery room. 4 U of packed red blood cells, 4 U of platelets, and 2 U of fresh-frozen plasma were reserved in case of variceal bleeding and postpartum hemorrhage. The indication of caesarean section had to be given by reason of late decelerations. A male baby, appropriate for his gestational age, with a 7/10 APGAR was delivered through a mid-line laparotomy incision under epidural anesthesia. The patient remained hemodynamically stable throughout the procedure, and systemic blood pressure was regulated by nitroglycerin perfusion at the post-operative period. After uneventful two days the mother was discharged and advised follow-up in the gastroenterology clinic.

DISCUSSION

Pregnancy alters systemic hemodynamics in response to developing maternal and fetal needs (3). Maternal plasma volume and cardiac output increased approximately 50% during pregnancy (3, 9). Portal hypertension also shares these alterations, but, unfortunately, compensatory mechanisms essential for the maintenance of blood pressure are insufficient. In consequent, patients develop hemodynamic changes known as hyperdynamic syndrome (3). In non-cirrhotic portal hypertension there is a pathologic increase in portal blood flow due to splanchnic vasodilatation in contrast to well-preserved liver function. These alterations are the underlying features of basic physiopathology of the increased portal pressure (3). NCPH patients have a spectrum of presentation involving massive splenomegaly, portal hypertension, well tolerated episodes of variceal bleeding and preserved liver function and fertility. In the management of pregnancy with NCPH, the obstetrician needs to decide on two features, both prominent and controversial: firstly, the prophylactic treatment of varices; and secondly, how to approach delivery.

Bleeding from esophageal varices is the main risk other than ascites, encephalopathy, hepatorenal syndrome, and splenic artery aneurysm rupture. Variceal bleed may not be as frequent and dreaded as previously supposed. The incidence of variceal bleeding in pregnant patients with known portal hypertension has been reported in up to 43% (10-13). İnside the pregnant NCPH group, mortality rate is between 2% to 6% (5). The least incidence and the good perinatal outcome in the series of Pajor and Lehoczky (11) are related to the diagnosis and the treatment preceding pregnancy. Variceal bleeding is expected to occur during the 2nd and 3rd trimesters when maternal blood volume and uterine compression of the inferior vena cava and collateral vasculature maximally increase (14). In contrast, the increased blood volume and cardiac output are thought to have compensated through the utero-placental circulation without affecting the portal venous pressure (15).

In our case, the patient bled neither at the pregnancy nor at the labor and delivery. This was achieved firstly by our strict follow-up for the maternal and fetal wellbeing, and also for the pregnant's lifestyle modifications. Furthermore, endoscopy played the main role in our management both for diagnosis and for prophylactic treatment. We performed three screening endoscopies at each three trimesters. Upper endoscopy during pregnancy appears to be safe for premature labor or fetal malformations, with the main risk being fetal hypoxia from sedative drugs or positioning (16). After the diagnosis of esophageal varices in the first course, prophylactic treatment was decided in order to prevent variceal bleeding in late pregnancy. There still remains controversy about the prop-

hylactic treatment options. Since Starkel et. Al (17). Reported the first EVL; it has largely been performed in prophylaxis and considered as an effective minimally invasive treatment during pregnancy. Banding acts locally without improving the physiopathology, and several trials showed that banding results are similar to beta-blocker therapy in re-bleeding prevention (18, 19). On the other hand, its effect to be local is the advantage of EVL against medical therapy that has the potential morbidities such as fetal growth retardation, neonatal hypoglycemia, and neonatal bradycardia. EVL has been reported to be superior to endoscopic sclerotherapy in re-bleeding (20). EVL is effective, but for a short time as portal pressure and flow are not modified. This makes endoscopic follow-up necessary (21). We also performed two screening endoscopies at controls of our case. Another and maybe the one that needs more to be illuminated area of controversy is the way of delivery. While some investigators (15) give no role to elective termination or caesarean section unless in case of obstetric indications, others (4) advocate performing elective caesarean section or operative vaginal delivery under extradural analgesia in order to decrease the risk of variceal bleeding due to the increased intra-abdominal pressure with repetitive Valsalva maneuver. Bleeding from pelvic and abdominal wall collaterals should be kept in mind in caesarean section. Nevertheless, all experts (5) share the point of ensuring blood products availability before labor. If caesarean section is performed, regional anesthesia might exclude the disadvantages of general anesthesia for variceal hemorrhage. The hypertensive response to intubation, straining during weaning, receiving a number of drugs and increased requirement of oxytocin could be avoided in regional anesthesia. The performance of regional anesthesia in portal hypertension is not without problems, either. The extradural veins engorge, as well. Thrombocytopenia may contraindicate any form of regional anesthesia. We decided to deliver the baby via vaginally under epidural anesthesia, but caesarean section indication was given because of fetal distress. During and after the operation, no complications occurred.

Finally, pregnancy in NCPH has good maternal and fetal outcomes. Although the physician should be aware of the specific risks and complications, there needs to be definite guidelines for appropriate management of pregnancy and delivery. We think endoscopic variceal ligation for prophylactic treatment of oesophageal varices during pregnancy and epidural anesthesia for delivery are effective and relatively safe in these patients.

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