

Patient Diagnosed of Budd-Chiari Willing to Get Pregnant: Presentation of a New Case of Full-Term Pregnancy

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Abstract

Budd-Chiari syndrome is an infrequent pathology characterized by the obstruction of the suprahepatic venous outflow. It can manifest in several ways and may become very severe. So far, there is great controversy on the issue of whether these patients should become pregnant, because of the complications this implies in this disease. Nowadays, there are very few cases of full-term pregnancy with good outcome reported. We present a case of a 36-year-old patient with subseptate uterus diagnosed of Budd-Chiari with Polycythemia Vera (jack2 gene mutation and heterozygosity for factor V Leiden). After being treated and the disease controlled, the patient got pregnant. This is one of the few cases that exist with good outcome. Strict follow-up of the patient was carried out at our hospital and no complications occurred throughout pregnancy and postpartum. The patient was treated prior to pregnancy with low molecular weight heparin and antiplatelet drugs until 24 hours prior to labor. Delivery occurred by cesarean section (CS) at 37 weeks without complications. The healthy newborn was discharged with the mother after one week of hospitalization. The proper control of this disease prior to pregnancy, together with strict monitoring conducted during it seems one of the keys to make possible pregnancy in young patients willing to get pregnant. In conclusion, this case could not support the contraindication of pregnancy in certain cases of this pathology. Moreover, the patient should be always informed of all the existing risks and complications.

Keywords: Budd-Chiari; Pregnant; Full-term pregnancy; Polycythemia Vera

Introduction

The Budd-Chiari syndrome is considered a rare disease (prevalence: 1-9/100,000) characterized by the obstruction

of the suprahepatic venous outflow, almost always due to thrombosis. Thus, a hepatic sinusoidal congestion is produced, creating portal hypertension [1]. The most frequent causes are those diseases that produce hypercoagulability such as myeloproliferative syndromes, hereditary coagulation factor deficiencies or antiphospholipid syndrome. The disease is most frequently diagnosed in young women with increased estrogens stimuli such as oral contraceptives or pregnancy itself [1, 2].

We present a case of a full-term pregnancy with subseptate uterus and Budd-Chiari syndrome, diagnosed with Polycythemia Vera (PV) with mutation of Jack2 gene and heterozygosity for factor V Leiden.

Case Report

A 36-year-old patient with Budd-Chiari syndrome and known Polycythemia Vera came to our center for control of her first pregnancy. Her medical history revealed a subseptate uterus without previous surgery. The patient was diagnosed of Budd-Chiari Syndrome at the age of 29 in a context of Polycythemia Vera (positive jack2 gene mutation and heterozygosity for factor V Leiden). The disease was diagnosed by symptoms such as ascites, hepatosplenomegaly, and venous ectasia in suprahepatic veins observed by Nuclear Magnetic Resonance. These symptoms were treated and controlled. During the following years, the patient was controlled in our hospital and the treatment received was oral anticoagulation and antiplatelet drugs. Subsequently, the patient expressed her desire to become pregnant and 6 months prior to pregnancy oral anticoagulant treatment was replaced by low molecular weight heparin.

Throughout pregnancy, no complications occurred; the patient was strictly followed at our hospital as high-risk pregnancy and by the Departments of Hematology and Digestive diseases with satisfactory outcome. The patient had appointments in Obstetrics every 2 - 3 weeks and tests on fetal welfare and growth were performed by means of ultrasound and Doppler. The fetal growth curve remained at around P40 throughout gestation and Doppler studies were always normal. No complications related with her pathology

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occurred. The treatment prescribed during pregnancy was low molecular weight heparin 80 mg and acetylsalicylic acid 100 mg daily. Blood test remained within normal values.

The patient came to the hospital at 36 weeks of pregnancy for uterine contractions without bleeding or other symptoms. She was admitted to the hospital and it was decided to perform a labor induction by cervical ripening at 37 weeks. Heparin was removed 24 hours prior to ripening. Finally, a cesarean section (CS) was performed for failed labor induction. After CS a healthy female newborn of 2880gr., Apgar Index 9/10 was born. After surgery the patient was controlled in the Intensive Care Unit without complications. Heparin therapy was resumed 6 hours after the CS. The patient was discharged 5 days after and subsequent analytical and clinical controls in consultations were all correct. Both, the patient and the neonate remain stable.

Discussion

There is a controversy over Budd-Chiari syndrome and pregnancy because of the high risk involved [3]. Diagnosis often occurs in young women of childbearing age with pregnancy desire and there are different opinions regarding contraindication of pregnancy in this condition [4]. Up to now, there is a tendency towards contraindication of pregnancy in these patients due to the complications it implies: increased deterioration of hepatic function and, of both thrombotic and bleeding events or of ascites. In addition, there is an increased risk of recurrent postpartum thrombosis in patients with stable disease and, therefore, there is an increased need for surgical interventions such as angioplasty, Transjugular intrahepatic portosystemic shunt (TIPS) or portosystemic bypass [3-5].

There are few cases in the literature associated with pregnancy. In most cases, there are maternal complications or pregnancy loss. However, there are some cases of pregnancy with good outcome in terms of maternal and neonatal morbidity [6]. In 2009, a multicenter study was published, in which 16 patients diagnosed with this syndrome were analyzed and treated by surgery, interventional radiology and anticoagulation; subsequently, the patients got pregnant. From a total of 24 pregnancies, 16 newborns were delivered and only 4 of them were full-term pregnancies [7]. During pregnancy or postpartum complications such as thrombosis and hemorrhage were described. It has been proposed not to contraindicate pregnancy in women with Budd-Chiari syndrome already diagnosed and properly treated, either conservatively or by surgery [6, 7]. However, patients must be adequately informed of the potential risks, both maternal and fetal and strict control of pregnancy must be performed. One of the problems occurs because, in many cases, pregnancy is the triggering cause for the diagnosis of a completely unknown pathology to the patient, being in these cases of much

greater risk [6, 7].

Another issue is the cause of the syndrome. In most of the cases there is a hypercoagulability due to underlying conditions, as in our case [8]. These can be myeloproliferative disorders, factor V Leiden mutation, antithrombin deficiency or protein C and S deficiency. There are causes and conditions that have been associated with worse outcomes as in the case of factor II mutation, in the case of older subjects, or of previous transplant [9].

Regarding the treatment during pregnancy, there is not enough data to develop a protocol, but there are better treatment outcomes with anticoagulation with low molecular weight heparin throughout pregnancy and adding antiplatelet drugs if required according to the pathology [10]. For patients treated with oral anticoagulants, such as dicumarinics, it should be replaced by heparin prior to delivery, as in the case presented. However, on the other hand, it implies a risk; there is a high rate of bleeding and intrauterine hematomas during pregnancy and metrorrhagia during postpartum in these patients. Therefore, it is recommended to strictly control anticoagulation doses and stop the treatment in the moment previous to delivery [10].

The birth canal is another discussed point. CS has been related with postoperative increased risk of thromboembolism, with a higher blood loss and ascites. Thus, there is a trend towards vaginal delivery as long as there is no contraindication to it. In the series of Rautou et al. of the 16 pregnancies, only 4 were full-term and delivery was vaginal [7]. In our case a labor induction was performed to try vaginal delivery, but it was impossible due to dystocia; therefore, a CS was performed [11]. Furthermore, the end of pregnancy is also a controversial issue. There are very few cases of full-term pregnancy due to complications that are more frequent in this type of patients, such as intrauterine growth restriction (IUGR), placental pathology, and some cases of stillborn. Therefore, given the limited clinical experience, it is difficult to decide when to end pregnancy. In this case, it was decided to end pregnancy at 37 weeks due to good maternal and fetal evolution [11].

The newborn may also present complications due to the increased risk for preterm delivery and the possibility of having an underlying prothrombotic disorder. Therefore, a strict surveillance must be performed of the newborn [11].

Conclusion

There is still much controversy over pregnancy in patients with Budd-Chiari syndrome because of the small number of cases so far reported. Given the limited information, there are no protocols regarding the management of these patients.

The case here presented is one of the few examples of full-term pregnancy without maternal or fetal complications. This case could support the new proposal that aims not to contraindicate pregnancy in patients with this pathology

already diagnosed, treated and stable; always in a planned manner and with a very strict control throughout pregnancy. However, it is very important to inform the patient of all the potential risks and complications that assumes.

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