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Management and Outcomes of Wilson Disease in Pregnancy: A Case Report

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ABSTRACT

Pregnancy in a woman with Wilson disease (WD) can cause pre-eclampsia, miscarriage, and preterm labor and causes hepatic, neurologic, and hematologic complications aside from pregnancy-related difficulties. A 30-year-old female was brought to the emergency room (ER) in her 38th gestational week (GW) with the chief complaint of left foot swelling and weight gain. Aside from a platelet count of about 73000, the rest of the assessment and findings were normal. Regarding the reassurance of the platelet count and holding the medical treatment, throughout the vaginal delivery, a child with Apgar of 9-10 was delivered. WD in pregnancy tends to have complications, thus patient management and how the patient is treated are critical.

Introduction

Wilson disease (WD) is defined as a genetic disorder resulting in an inability to metabolize copper in the human body and consequently concentrating the copper in organs such as the brain and liver (1-3). Pregnancy in a woman with WD can cause problems during pregnancy, including pre-eclampsia, abortion, and preterm delivery (4); meanwhile, WD, aside from pregnancy effects, also causes hepatic, neurologic, and hematologic injuries in a pregnant woman. To date, few cases have been reported, and, here, we aim to report a case of WD in a pregnant woman.

Case presentation

A 30-year-old female was brought to the emergency room (ER) in her 38th gestational week (GW) with the chief complaint of left foot swelling and weight gain of about 1 kg in 5 days. The patient was a case of WD and documented liver cirrhosis from 5 years

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ago. It was her 4th gravidity, including two lives and one abortion. The abortion was one year ago at 16 weeks of pregnancy, and additionally, she expresses a history of severe admission to the hospital during her current pregnancy with a chief complaint of thrombocytopenia. The patient was under treatment (Penicillamine TDS) since the diagnosis; before the pregnancy, she reduced the dosage to BID and took QD since her pregnancy was confirmed. The patient was admitted to the gynecology ward. On initial physical examination, the patient was alert and responsive, vital signs were normal, blood pressure was 80/120 mmHg, and heart rate was 84 p / min. Moreover, neurological and ocular examinations were normal, and the Kayser-Fleischer ring was not visible. Fundus height was equal to 38-week, and no organomegaly was detected. Eventually, examination of lower extremities showed a 1.5 cm difference in leg circumference between the right and left leg.

Preliminary examination of the patient's blood test showed a reduction in platelet count to about 73,000, while the rest of the tests, including liver function test (LFT), coagulopathy tests, BUN, creatinine, and urine protein, were within the normal range. In addition, abdominal ultrasound showed normal results, and no liver cirrhosis was detected. Doppler ultrasound of the right leg was also normal. In fetal evaluation, fetal ultrasound showed an anterior placenta with an amniotic fluid index of 55 mm, and a biophysical examination of the fetus showed a normal result. Following consultation with gastrointestinal colleagues, they offered normal vaginal delivery in the case of platelets above 50,000 and no indication of obstetrics and gynecology for cesarean section. They also suggested performing prenatal endoscopy to detect the presence of esophageal varices and using peripheral blood smear (PBS) to diagnose any abnormalities such as schistocytes. Before delivery, they offered to hold the medical therapy and resume one week after the delivery.

Eventually, the endoscopy showed no signs of esophageal varices, and PBS revealed no abnormalities. Regarding the reassurance of the platelet count and the medical treatment, throughout the vaginal delivery, a child with Apgar of 9-10 was delivered. Subsequent to the delivery, because of heavy bleeding, 100-unit oxytocin and 1000 Mg misoprostol were prescribed, and the patient was monitored. Post-partum laboratory data showed 57000 platelet counts alongside normal results of LFT, BUN, creatinine, urine protein, and coagulation tests. Finally, in consultation with a gastrointestinal partner, they recommended 6 units of platelet transfer and initiation of medical treatment 1 week after delivery with penicillamine and zinc. The patient stayed in the ward for three days, and she was discharged after the patient's condition had been confirmed.

Discussion

Wilson's disease is defined as an inherited disorder that results in impaired copper transfer in the liver, resulting in elevated copper concentrations in the liver and prompt hepatic cirrhosis, chronic hepatitis, and dysfunction of the nervous system field [5]. A large number of patients with WD disease are women of childbearing age, so it is essential to manage and control WD [6, 7]. Complications of WD in pregnancy involve difficult delivery and a higher risk of spontaneous abortion. In one study, the abortion rate was estimated at 40% higher than normal pregnancies [4]. As mentioned earlier, WD also causes neurological symptoms in pregnant women, and studies show that neurological damage is more significant than spontaneous abortion or even bone and liver damage [4, 8, 9].

It should be noted that in those studies, not all women with spontaneous abortion had a previous history of anti-copper therapy. Other complications in pregnant women with WD include limb edema, which occurs more often in these patients than in normal pregnant women, and also studies demonstrated no change in the Apgar score and the type of delivery amongst women with WD healthy patients [6]. In our case, a 30-years-old female the patient had previously been diagnosed with Wilson's disease and was being treated and discontinued treatment before pregnancy. Furthermore, the patient presented to the emergency department with limb swelling, consistent with studies. Early symptoms of WD often include liver dysfunction and neurological abnormalities[10]. In our study, because the onset of the disease was years ago and she was under medication, no signs of neurological symptoms and liver disorders were seen at the time of referral.

In pregnancy, due to predictable changes in liver biomarkers in the conditions such as HELLP syndrome, pre-eclampsia, or acute fatty liver pregnancy, differentiating the cause is challenging since the laboratory abnormalities remain unspecific; However, in WD, chronic hepatic impairment appears and triggers maternal liver complications. Studies evaluating changes in liver tests and pregnancy outcomes indicate that changes in liver tests are directly related to patient mortality and the difficulty of delivery and delivery outcome[11]. During the hospitalization, no changes were observed in the liver tests, abdominal ultrasound showed normal

results, and no liver cirrhosis was detected. The only problem we found in the patient's experiments was platelet depletion, which did not limit us according to the studies performed. Neurological manifestations vary in patients and mostly include tremors, dystonia, and ataxia. The patient may have a Kayser-Fleischer ring[12]. In our patient, there were no signs of neurological problems.

Studies have reported congenital anomalies in pregnant women with WD treated with D-penicillamine; the studies reported congenital abnormalities such as agenesis of the corpus callosum, micrognathia, and limbs contractures the postnatal course, chronic lung disease and developmental delays were reported[13]. None of the mentioned complications occurred in our patient, while the patient had stopped her treatment when she was informed of the pregnancy.

Pregnant women with WD need ongoing assessment and should have tests such as BUN, creatinine, liver function tests, and coagulation tests to find out their baseline amount. An ultrasound should be done to diagnose liver cirrhosis, and upper gastrointestinal endoscopy is essential for screening for esophageal or gastric varicose veins, especially in patients with portal hypertension and patients should undergo a physical and neurological examination to diagnose KF, tremor, dysarthria, and neurological defects [14]. The obstetrical indications determine the route of delivery in the WD patients and depend on laboratory tests and presence of liver cirrhosis and coagulation impairments, and the presence of high-grade esophageal varices[15]. Consulting with colleagues, checking vital signs, and preparing for antepartum or post-partum bleeding are necessary.

To the best of our knowledge, few cases of WD in pregnancy have been published to date, and most studies evaluate pregnancy complications and management in pregnant women with WD. In our case, the patient was under treatment since the diagnosis and decreased medication dosage before the pregnancy. In the assessment of the patient, ultrasonography was normal, and no liver cirrhosis was detected; in the endoscopy, no sign of esophageal varices was seen, and laboratory analysis revealed normal, excluding the decrease in the decrease platelet count, which was because of the disease. The patient was stable, and no contraindication for vaginal delivery was detected; hence vaginal delivery was performed. The patient's order was Penicillamine and Zinc for medication.

Conclusion

Although WD in pregnancy is rare, this problem should always be considered, and diagnostic tests related to complications should be performed. It is recommended that the treatment behold and careful patient monitoring be considered.

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this article.

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Conflict of Interests

The authors declare no conflict of interest.

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