ISSN: 2576-2842

Research Article

Journal of Gynecology & Reproductive Medicine

A Case Report About Polycythemia Vera and A Pregnant Woman

Shuxin Ma and Xia Luo*

Center for Reproductive Medicine, Cheeloo College of Medicine, Shandong University, Jinan, Shandong, 250012, China

***Corresponding Author**
Xia Luo, Center for Reproductive Medicine, Cheeloo College of Medicine, Shandong University, Jinan, Shandong, 250012, China.

Submitted: 2023, July 31; **Accepted:** 2023, Aug 29; **Published:** 2023, Sep 21

Citation: Luo, X., Ma, S. (2023). A Case Report About Polycythemia Vera and A Pregnant Woman. *J Gynecol Reprod Med, 7*(3), 126-128.

Abstract

Background: *Polycythemia Vera (PV), belonging to the myeloproliferative disorder, refers to the apparent overproduction of red blood cells. This pathological change culminates in blood hyperviscosity, which causing pregnant women with high risk of thrombosis. In this study, we introduced a rare case about a pregnant woman with PV in China.*

Case summary: *In this case report, we summarized the process of diagnosis and treatment about the pregnant woman with previously diagnosed PV during her gestation time. The first onset of this disease was in her twenty-second. She was supplied with hydroxyurea and interferon and got symptoms relieve. But she didn't undergo regular treatment after that. She was hospitalized during gestation in her thirty-first, treated with regular administration of phlebotomy, aspirin (100mg), and low molecular weight heparin (LMWH). Finally, the patient delivered a healthy newborn after the cesarean.*

Conclusion: *Early diagnosis and intervention can effectively prevent pregnancy complications. Flexible control of hematocrit levels combined with anticoagulation contributes to improving pregnancy outcomes.*

Keywords: Polycythemia Vera, Pregnancy, Phlebotomy

Abbreviations

HGB: Hemoglobin
HCT: Hematocrit
IFN: Interferon
ICU: Intensive Care Unit
LBR: Live Birth Rate
LMWH: Low Molecular Weight Heparin
MRI: Magnet Resonance Imaging
NT: Nuchal Translucency
NICU: Neonatal Intensive Care Unit
OGTT: Oral Glucose Tolerance Test
PV: Polycythemia Vera
PCR: Polymerase Chain Reaction
RCM: Red Cell Mass
WBC: White Blood Cell Count.

Introduction

Polycythemia vera (PV) is a relatively indolent myeloproliferative neoplasm, characterized by the clonal proliferation of myeloid cells, especially the trilineage expansion of morphologically normal red cells, white blood cells and platelets. The reported incidence in women younger than 40 years was 0.3 per 100,000. Pregnancy complicated with PV is an exceedingly rare disorder. Less than 200 cases have been reported in the worldwide literature and this is the first Chinese report.

Case Presentation

The hematological journey started when the patient was 22 years old. She was hospitalized at the local hospital because of dizziness and numbness in hands and tongue, with the appearance of flushed face and purple gums. Routine blood test showed hemoglobin (HGB) of 213g/l, erythrocytes of 7.99*1012/L and platelets of 492*109/L. Cytological examination of bone marrow suggested 'PV'. Detection of JAK2 (V617F) mutation was positive. Abdominal ultrasound examination showed splenomegaly. Then she got phlebotomy treatment three times and oral hydroxycarbamide. After that she became relived and repeated test showed HGB of 183g/L, erythrocytes of 6.73*1012/L, and platelets of 492*109/L. For further treatment, she came to the department of hematology in our hospital. Our hospital further completed the relevant examinations. Blood route test showed erythrocytes of 5.69×1012/L, HGB of 167g/L, hematocrit (HCT) of 50.5%, and platelet of

316×109/L. Bone marrow biopsy showed hypercellularity with prominent erythroid, granulocytic and megakaryocytic proliferation and a normal karyotype (46, XX). Quantitative polymerase chain reaction (PCR) of JAK2 (V617F) demonstrated the status of coexistence of mutant type and wild type. So, the diagnosis of polycythemia vera was conformed according to the WHO criteria.

Then she got phlebotomy treatment and administration with hydroxycarbamide and interferon (IFN) to control HCT. After the symptomatic relief, she left the hospital and kept the daily IFN administration for two years.

As there was no obvious discomfort so she didn't come for a follow-up visit until last year. She was hospitalized at the local hospital with the complain of left-sided numbness. Cranial magnet resonance imaging (MRI) demonstrated lacunar and softening lesions in left frontal lobe and bilateral corona radiate, suggestive of ischemic cerebral infarction. Accordingly, she got anti-coagulation treatment (aspirin and clopidogrel) for two months and gradually felt no numbness.

The patient was 31 years old when she became pregnant. At 6 weeks of gestation, she was treated with vitamin E and progesterone capsules because of a small amount of vaginal bleeding at the local hospital. At 8 weeks of gestation, she presented to our hospital for combination of obstetric department and hematological department. Blood route test showed erythrocytes of 6.11×1012/L, HGB of 144g/L, HCT of 46.4%, and platelet of 687×109/L. Treatment included phlebotomy and administration of aspirin (100mg), low molecular weight heparin (LMWH), progesterone and dydrogesterone. After she was released, she was always taking aspirin and maintained regular follow-up visits to the hematological department. Repeated routine blood tests showed a good control of HCT (< 48%). The noninvasive prenatal cfDNA testing suggested a high risk of aneuploidy of sexual chromosomes. However, the patient rejected the recommendation of amniocentesis. Other examinations during pregnancy including nuchal translucency (NT), fetal ultrasound, oral glucose tolerance test (OGTT), and fetal echocardiography were normal. At 37 weeks and 6 days of gestation, she presented at the obstetric department for an appropriate delivery method without any evident symptoms. Repeated tests showed erythrocytes of 4.00×1012/L, HGB of 121g/L, HCT of 36.6%, platelets of 436×109/L and roughly normal clotting, liver and renal function indicators for pregnancy.

The multidisciplinary team included obstetric, anesthetic, hematological, transfusion, intensive care unit (ICU), neonatal intensive care unit (NICU) and neurology departments. We agreed on a cesarean section under general anesthesia. After the cesarean a female newborn weighing 3200g with normal appearance was delivered. Apgar scores at first and fifth minute were both 10 points. The newborn was hospitalized at the NICU and released 7 days later as she was well. First postpartum blood routine test of the patient showed erythrocytes of 3.91×1012/L, HGB of 124g/L, HCT of 35.4%, platelets of 429×109/L. She stayed in ICU for 5 days, administrated with enoxaparin, oxytocin, and other supportive treatment. Then the patient was referred to the obstetrics department. Blood route test before discharge showed erythrocytes of 3.78×1012/L, HGB of 119g/L, HCT of 34.4%, and platelet of 446×109/L. Aspirin restarted in the postpartum period for 1 weeks.

Discussion

PV is a kind of hematologic malignancy, but pregnancies complicated with PV are relatively rare. The rough incidence in women during the reproductive period varies from 0.04 per 100,000 in those aged 20–34 years to 0.25 per 100,000 in those aged 35–39 years [1]. Pregnancies interfere with the prognosis of PV because of lower levels of HGB and HCT during gestation. PV also affects pregnancy and causes severe pregnancy complications, such as vaginal bleeding, early spontaneous abortions, premature delivery and preeclampsia. There were few reports on the management of pregnancies complicated with PV.

Some retrospective studies have reported the course and outcomes of PV pregnancies. A meta-analysis summarized 159 cases out of three studies and concluded that the overall live birth rate was only 66.7% of PV pregnancies [2]. Another study reported the live birth of 58.0% from 38 PV pregnancies [3]. A updated research reported 41 pregnancies in 20 women with PV. Bleeding and placenta pathology were the commonest complications (6/42, 14.6%), followed by bleeding at delivery (4/41, 9.8%). The incidence of preeclampsia and deep vein thrombosis was identical. During postpartum follow-up, one major hemorrhage (a spontaneous intrasplenic bleeding that required splenectomy) and four thrombotic events (one femoral vein thrombosis, one portal vein thrombosis and two cases of Budd-Chiari syndrome) occurred among 21 live birth pregnancies. PV-specific therapy, including administration with LMWH, lowdose aspirin, IFN and phlebotomy, significantly improved the obstetrical outcomes of 41 pregnancies in 20 women [4].

In this case report, the patient was diagnosed with PV at 22 years. Hydroxyurea treatment is the first-line cytoreductive drug due to its high efficiency, convenient administration, minimal toxicity and low cost. IFN can be an alternative agent for young patients, pregnant women and patients tolerant or resistant to hydroxyurea treatment. Scheduled phlebotomies, in place to control hematocrit□45%, effectively attenuate clinical manifestations induced by hyperviscosity [5]. These major medical protocols prevent the PV progression on the patient's first episode.

At eight weeks of gestation, the blood routine test results were suggestive of the dominant erythrocyte proliferation stage. Thrombotic events are the main threat to the patient's health. Considered the cornerstone of treatment for down-regulating the red cell mass (RCM), the anti-thrombotic value of phlebotomies has been extensively elaborated [6]. A detailed analysis reported by the European LeukemiaNet multi-institutional series demonstrated that the live

birth rate (LBR) of pregnancies after PV diagnosis was 77%, while the LBR of pregnancies before PV diagnosis was 49%. Phlebotomies in the first trimester contributed to strict hematocrit control < 40% [7]. Since the patient had a history of ischemic cerebral infarction before pregnancy, LMWH was used for prophylactic anticoagulation. As the primary anti-thrombotic prophylaxis approach, administration with 200mg aspirin daily decreased the incidence of cardiovascular death or nonfatal cardio-embolic events (stroke, venous thrombosis, pulmonary embolism, myocardial infarction) [6]. Besides, fetal preservation therapy was necessary. Discharge medication was the regular administration of aspirin until parturiency.

At 37 weeks and 6 days of her gestation, the team of multidisciplinary collaboration set out strict protocols for this pregnancy with various and complicated associated diseases. Aspirin was prudently withdrawn 24 hours before the Cesarean at 38 weeks. Postoperatively, we administrated enoxaparin to reduce the thrombosis risk. The hematocrit level and platelet count were carefully kept below 45% and 450×109/L separately before and after surgery.

In conclusion, the precise diagnosis of PV before or during pregnancy contributes to timely clinical intervention. Besides, routine thrombophilia screening is recommended to prevent cardiac-cerebral vascular comorbidity. Effective multidisciplinary collaboration, a well-controlled hematocrit and administration with aspirin or LMWH can significantly improve pregnancy outcomes.

HGB	(g/L)	HCT (%)	RBC ($\times 10^{12}$/L)	Platelets ($\times 10^{9}$/L)
First episode	213	-	7.99	492
Reexamination at our hospital	183	-	6.73	213
Post-treatment review	167	-	5.69	316
8 weeks of gestation	144	46.4	6.11	687
37 weeks and 6 days of gestation	131	38.6	4.26	539
Before operation	121	36.6	4.00	436
After operation	124	35.4	3.91	429
Before discharge	119	34.4	3.78	446

HB hemoglobin, HCT hematocrit, WBC white blood cell count

Table 1: Repeated tests of HGB, HCT, RBC and Platelets.

References

1. Barbui, T., & Finazzi, G. (2006). Myeloproliferative disease in pregnancy and other management issues. ASH Education Program Book, 2006(1), 246-252.
2. McNally, R. J., Roman, E., & Cartwright, R. A. (1999). Leukemias and lymphomas: time trends in the UK, 1984–93. Cancer causes & control, 10, 35-42.
3. Harrison, C. (2005). Pregnancy and its management in the Philadelphia negative myeloproliferative diseases. British journal of haematology, 129(3), 293-306.
4. Wille, K., Bernhardt, J., Sadjadian, P., Becker, T., Kolatzki, V., Huenerbein, K., & Griesshammer, M. (2021). The management, outcome, and postpartum disease course of 41 pregnancies in 20 women with polycythemia vera. European Journal of Haematology, 107(1), 122-128.
5. Tefferi, A., Vannucchi, A. M., & Barbui, T. (2021). Polycythemia vera: historical oversights, diagnostic details, and therapeutic views. Leukemia, 35(12), 3339-3351.
6. Guglielmelli, P., & Vannucchi, A. M. (2020). Current management strategies for polycythemia vera and essential thrombocythemia. Blood Reviews, 42, 100714.
7. Gangat, N., & Tefferi, A. (2021). Myeloproliferative neoplasms and pregnancy: overview and practice recommendations. American journal of hematology, 96(3), 354-366.