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CASE REPORT

An Unplanned Pregnancy with Life-Threatening Paroxysmal Nocturnal Hemoglobinuria: A Case Report

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Abstract

A 26-year-old Bahraini female with a past medical history significant for diabetes type 2 on metformin and paroxysmal nocturnal hemoglobinuria. An allogenic stem bone marrow transplantation was done from full match siblings approximately one year ago. The patient presented to the labor and delivery ward for possible normal vaginal delivery at 39 weeks gestational age. After hematology review the patient was taken to caesarian section due to failure of progress and the surgery was done successfully under neuraxial anesthesia (epidural top-up). The case illustrates the effectiveness of neuraxial anesthesia in such patient with such rare disease with possible coagulopathy.

Keywords: Bone marrow disorders; Epidural; Neuraxial anesthesia; Paroxysmal nocturnal hemoglobinuria; Unplanned pregnancy

Introduction

Paroxysmal nocturnal hemoglobinuria (PNH) is a rare, acquired, life-threatening blood disease. It is characterized by hemolytic anemia, hypercoagulable state, and impaired bone marrow function.[1] Its incidence rate is not clearly known due to its rarity and diagnostic difficulty.[2] PNH is primarily observed in younger adults with a slight female predominance, and the median age of diagnosis is in the early 30s.[3,4] It is closely related to aplastic anemia.[1] In fact, up to 30% of newly diagnosed cases of PNH evolve from aplastic anemia. The median survival after diagnosis is 10 years; however, some patients can survive for decades with only minor symptoms.[1]

PNH is associated with an increased risk of complications such as exacerbated hemolytic anemia and life-threatening thromboembolism during pregnancy.[5] These maternal complications are associated with fetal morbidity and mortality.[6] The most common cause of death is liver vein thrombosis (Budd-Chiari syndrome). Fetal wastage (30%) and premature birth rate (16%) are also high.[7] Therefore, female patients with PNH are discouraged from becoming pregnant.[5]

Here, we discuss the case of a 26-year-old pregnant woman with PNH who got pregnant against medical advice and presented for elective normal vaginal delivery.

Case Presentation

The patient was a 26-year-old Bahraini female with a past medical history significant for type 2 diabetes mellitus on metformin and classical PNH for which allogeneic stem bone marrow transplantation using fully matched bone marrow of her siblings had been done about a year prior to presentation. In March 2019, the patient was examined in an obstetrics clinic where 9 week pregnancy was confirmed, and she was advised to terminate the pregnancy due to PNH, immunosuppressants, and coagulopathy but she refused. She had a miscarriage the same month. The patient got pregnant again the following month, in April 2019, which was confirmed in the obstetric clinic. She was following up with the Hematology team during this period, and they kept her on vitamin D and checked her PNH panel regularly which was within normal limits.

The patient presented to the Labor and Delivery ward with labor pain, for possible normal vaginal delivery at 39 weeks of gestational age. On admission, she was generally well, afebrile (temperature 36.8°C), and vitally stable (heart rate 76 bpm, blood pressure 117/81 mmHg, and oxygen saturation 98%). Physical examination was unremarkable. Laboratory investigation on admission was notable for anemia 11.1 g/dL with a baseline of 11 g/dL; platelet count was 217 x 10^9 /L; coagulation profile was normal (INR 0.91, PT 10.9 sec, PTT 43%).

Hospital Course and Management

The Hematology team was consulted in view of patient's history of bone marrow transplantation. Complete blood count (CBC), iron study, liver function test (LFT), renal function test, PNH panel, vitamin B12, and vitamin-D were requested. The blood test result out and revived, all of them were within normal range.

The PNH panel analysis showed CD55- and CD59-deficient red cell population absent. Immunophenotyping was performed using gating antibodies CD45, CD15, CD64, CD235a, CD14 and CD24 as well as fluorescent aerolysin (FLAER), and all of them were within normal range.

The patient progressed into active labor and was shifted to the labor room where epidural was inserted at L4L5 level. Three hours later, the patient was taken for emergency cesarean section due to failure to progress to full dilation.

Cesarean section was done after topping up the epidural with 10 mL lidocaine 2% + 10 mL bupivacaine 0.5% for acceptable level of block according to the American society of anesthesiologists monitoring standards. Cefazolin 1 g and Midazolam 1 mg were given to the patient, and hemodynamic stability was achieved throughout the surgery with 1.5 L of crystalloids with no vasopressors on board.

The patient delivered a baby girl with APGAR 8 and 9, weight 3.382 kg, blood group O positive, with blood loss of 450 mL.

The patient was discharged on December 16, 2019, with an advice to follow up with Hematology every three months.

Discussion

PNH is associated with hemolytic anemia and coagulopathy (low platelet count), and the patient is prone to deep vein thrombosis.[1] Therefore, PNH has been considered as a contraindication for pregnancy.[10] The most common presentation of PNH is fatigue (80%) followed by dyspnea (64%) and hemoglobinuria (62%).[9] Thrombosis is seen in only 16% of the cases.[8]

PNH has three main subtypes: classical PNH in which the patients have evidence of PNH in the absence of another bone marrow failure disorder; PNH in the context of other primary bone marrow disorders, such as aplastic anemia or myelodysplastic syndrome; and subclinical PNH in which patients have small PNH clones but no clinical or laboratory evidence of hemolysis or thrombosis.[1]

The use of both general anesthesia and local anesthesia has been suggested for patients with PNH.[11,12] However, we chose to give our patient epidural labor analgesia due to the fact that she did not have absolute contraindication for its use.

The postoperative course of this patient was uneventful, without any complication with regard to the use of neuraxial anesthesia during surgery as the epidural catheter was removed one day postoperatively.

This is the first case documented in the Kingdom of Bahrain that was treated successfully after taking into consideration the patient's medical condition, choosing the right type of anesthesia, and monitoring her closely postoperatively. Such case reports of PNH with rare incidence are valuable resources for future research.

Conclusion

This case report demonstrated that neuraxial anesthesia was an effective method for managing cesarean section in pregnant women with PNH. It is important to bear in mind the role of bone marrow transplant and hematology management in optimizing the coagulation status of the patient.

Conflict of Interest

None

Acknowledgement

None

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