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# Paroxysmal Nocturnal Hemoglobinuria in Pregnancy (A Case Report)

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## Abstract

Paroxysmal nocturnal hemoglobinuria (PNH) is a rare cause of hemolytic anemia that can be exacerbated by pregnancy. This case report reflects on this condition. A pregnant 27-year-old female, a known case of PNH, presented with a history of liquor drainage. Upon investigation, we found that her platelet levels were $75 \times 10^9$. The patient was following up with the hematology department, where they started her on eculizumab. PNH can induce thrombocytopenia in pregnancy, which can lead to thromboembolic events; the risk reaches 10% in pregnancy. Moreover, it can lead to an increased risk of infection, which also happened to our patient, as she presented with a labial abscess 5 months post-delivery. Conclusion: Pregnant women with PNH must have their platelet levels closely monitored to prevent thromboembolic events.

## Keywords

Paroxysmal Nocturnal Hemoglobinuria, Thrombocytopenia in Pregnancy, Eculizumab in Pregnancy

## 1. Introduction

Paroxysmal Nocturnal Hemoglobinuria is an acquired somatic mutation affecting Hematopoietic Stem Cells (HSCs) [1]. Red Blood Cells (RBCs) become sensitive to complement-induced destruction due to a deficiency in surface anchoring proteins responsible for complement regulation [1]. It is characterized by paroxysmal attacks of intravascular hemolysis, anemia, and thrombocytopenia [2].

Stress, for example, can exacerbate hemolytic crises [3]. In pregnancy, thromboembolism risk rises to 10% [4], which is the main cause of mortality during the gestational period [1] [2].

Safe anticoagulation is used prophylactically during gestation in 60% of pregnancies [5] and is usually prescribed once pregnancy is diagnosed [3] until 6 weeks

postpartum [5].

Another PNH-related complication that is exacerbated during pregnancy is bleeding, which can vary in its presentation from recurrent epistaxis to severe postpartum hemorrhage [6].

This case report presents a case of PNH that was exacerbated during pregnancy.

## 2. Case Report

A 27-year-old Pakistani female patient with a known case of paroxysmal nocturnal hemoglobinuria presented to the accident and emergency department as a primigravida at 35 weeks of gestation with liquor drainage.

The patient was previously following up with the hematology department, where they started her on eculizumab and a prophylactic dose of enoxaparin (30 mg) once daily. As recommended by the Royal College of Obstetrics and Gynecology (RCOG Green-top Guideline No. 37a) [7], low-dose prophylactic LMWH is given to women with low to moderate VTE risk, adjusting the dose based on weight and risk factors. Given the patient's weight (62 kg), 30 mg enoxaparin once daily was appropriate. Anti-Xa monitoring was not performed, as the testing was not available at the treating center.

She was also counseled regarding eculizumab during pregnancy, weighing the risks and benefits, which she agreed to. The patient did not receive meningococcal vaccination or antibiotic prophylaxis before starting eculizumab.

Upon presentation, the patient was conscious, alert, and oriented, with no history of bleeding, bruising, or dark-colored urine.

She was started on eculizumab injection for PNH to manage hemolysis. Initially, she was on 600 mg weekly for 1 month, then was changed to 900 mg every 2 weeks.

The patient's laboratory investigations were as follows:

Baseline hemoglobin is 7 - 8 g/dL, and platelets are $75 \times 10^9$ (similar to baseline).

As the fetal presentation was breech, the patient was counseled for a cesarean section.

The hematology department was consulted before proceeding with the procedure, and they advised:

- The patient can go for a cesarean section.
- To reserve 6 units of platelets for the operation, to be transfused, if needed, during the operation.

The patient underwent an emergency lower-segment cesarean section for a breech presentation in labor. The outcome was a single live male baby, birth weight of 2.840 kg, Apgar scores of 9 and 10 at 1 and 5 minutes, with a total blood loss of 600 mL.

After the operation, the patient was kept in the postnatal ward for observation and to monitor her laboratory investigations. To avoid going into thrombosis post-op, the patient was maintained on prophylactic enoxaparin and the same pre-op Eculizumab dose.

Her investigations were as follows:

- 04 Feb: HB 10; PLLT 116
- 06/Feb: HB 8.1; PLT 46
- 07/Feb: HB 7.6; PLT 60
- 07/Feb: HB 7.7; PLT 44
- 08/Feb: HB 7.3; PLT 44
- 10/Feb: HB 7.7; PLT 55

Enoxaparin was withheld when platelet levels were <50, with continuation of Eculizumab. She did not require platelet transfusion pre- or post-op. She was then discharged from OB/GYN inpatient care in stable condition with a hematology follow-up appointment.

The patient presented again on postoperative day 11 with lower-segment cesarean section (LSCS) stitches, pain, and oozing.

Upon abdominal examination, the entirety of the wound was indurated with erythema, with a 1-cm gap in the middle that was oozing pus.

A CT scan of the abdomen was arranged, and the study showed a bulky uterus with a pre-uterine peritoneal fluid collection measuring 5.32.85 cm and a complication of dehiscence of the LSCS incision associated with mesenteric fat stranding and pelvic free fluid.

Since the patient was vitally stable and afebrile, conservative management was started with IV antibiotics: ceftriaxone and metronidazole, along with Clexane and Eculizumab.

At 5 months post-delivery, the patient was admitted again with complaints of labial swelling. She presented with a one-week history of pain and swelling in her labia. She had no history of discharge or fever.

On examination:

The patient was vitally stable and afebrile.

Per abdomen: soft, lax; no masses felt.

Per vagina: right labial swelling of around 4 x 4 cm, tender, no oozing.

Investigations: WBC 9.5; HB 10; PLT 185; coagulation profile within normal range.

Incision and drainage (I&D) was uneventful; a 6 × 6 cm right labial abscess was drained. The patient was finally discharged in stable condition with Eculizumab and oral antibiotics.

## 3. Discussion

The mainstay of PNH management is supportive care through packed RBC or platelet transfusions, whose requirements arise during the gestational period [6]. On the other hand, treatment is centered around the prevention of hemolytic anemia through a monoclonal antibody that antagonizes complement C5, namely Eculizumab [4], which carries a better prognosis by reducing the thrombosis rate [1].

The safety of eculizumab is still uncertain; some levels were found in cord blood samples; however, its levels were not detected in breast milk [6]. Discontinuation

of it during pregnancy resulted in thrombosis, which occurred during the postpartum period [6].

Infection rates in PNH are elevated due to leukocyte dysfunction or, probably, bone marrow dysfunction [8], which could explain the patient's pelvic collection and labial abscess after delivery.

Eculizumab is essential for the formation of the MAC complex, which is the mainstay of defense for Neisseria species. Patients receiving eculizumab should be vaccinated against Neisseria species. Receiving the vaccine can be done on the same day as receiving the eculizumab dose [9].

In a recent case study conducted in November 2024 on six pregnant PNH patients who were on a C5 inhibitor, all six patients gave birth to healthy children with normal development to date [10].

## 4. Conclusions

Eculizumab is started for pregnant patients with PNH to prevent hemolytic anemia. It is important for patients to have their CBC, LDH, reticulocyte count, and biochemical profile levels checked weekly for the first 4 weeks when starting on Eculizumab, and then monthly to prevent thromboembolic events.

Supportive management can be achieved through packed RBC or platelet transfusions. Once the patient is discharged postpartum, she should be followed up by both the obstetrics/gynecology and hematology teams in the outpatient clinic.

This report is limited by its single-case nature, its short follow-up, and a lack of available evidence, which highlights the need for multi-institutional cohorts to strengthen the evidence supporting the management of PNH in pregnancy.

## Disclosure

Informed consent was obtained from the patient.

## Conflicts of Interest

There is no conflict of interest regarding publishing the case report.

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