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Nursing Care in Postpartum Atypical Hemolytic Uremic Syndrome: A Case Report

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ABSTRACT

Pregnancy-associated atypical hemolytic uremic syndrome (P-aHUS) is a systemic disease associated with high morbidity and mortality rates, caused by dysregulation of the alternative complement pathway, leading to uncontrolled complement activation resulting in thrombotic microangiopathy. In this case it's reported patient care management of a P-aHUS patient which developed in postpartum period with renal failure, anasarca edema, hypertension and important laboratory signs of the syndrome. Patient was effectively treated by hemodialysis and eculizumab therapy, which controls complement activation and discharged without signs of hemolysis. The patient was discharged to home from the hospital after 42 days. At follow-up 2 weeks after discharge, all signs and symptoms of renal failure had resolved. The patient has not had any inpatient readmissions to the hospital to date. There are no case report in P-aHUS nursing care. Discussion of this case contributes the literature describing nursing interventions associated with caring for patients with P-aHUS.

Keywords: Postpartum, Hemolytic uremic syndrome, Nursing

1.INTRODUCTION

Pregnancy associated atypical hemolytic uremic syndrome (P-aHUS) is a rare, life-threatening complication in pregnancy. It is seen in about one in 25.000 pregnancies and it's associated with a significant perinatal or maternal morbidity and mortality (1-3). P-aHUS is a severe, systemic disease, first described by Robson in 1968 (4), associated with uncontrolled complement alternative pathway activation, leading to hemolytic anemia, usually accompanied by thrombocytopenia, hypertension and acute renal failure (5). Here it's reported nursing care management of a severe P-aHUS case in the postpartum period, through timely diagnosis and treatment resulted in healing and remarkable renal recovery. The aim of this case report is; to describe a rare severe illness and to summarize the nursing care in these cases.

2. CASE PRESENTATION

A 29-years old female, free of previous medical history except tonsillectomy and migraine. She has 3.25 degrees myopia and astigmatism. She is non-smoker. Her elder sister was diagnosed with P-aHUS following by her first labor in March of 2017. Similarly P-aHUS symptoms was presented in postpartum period in our patient. Her pregnancy had been without complication and she followed routine antenatal appointments including light thrombocytopenia, iron deficiency and anemia (hemoglobin=8-9g/dL). She was transfused two units of blood just before the cesarean (Hgb=6.6g/dL) and four units more after the labor. She was admitted in March 2018 for a planned cesarean section 38 week and 5 days. One week after cesarean section, she presented hypertensive crisis, anasarca edema, oliguria, hematuria and she was hospitalized postpartum day 13. Diagnosis process and initiation of treatment were quickly because of her elder sister P-aHUS history.

Schistocytes were detected 3-4% on peripheral blood smear, while elevated lactate dehydrogenase (LDH)=1805 IU/L (90-240), total bilirubin (TBIL)=3.36mg/dL (0.3-1.5), direct bilirubin=0.62 (0-40), serum creatinine=1.88mg/dL (0.40-1.40), blood urea nitrogen (BUN)=33mg/dl (5-24), as well as decreased platelet count (PLT)= 41×10^9/L ($173\text{-}390\times10^9$/L), red blood cell (RBC)= $2.66\text{x}10^6$(3.92-5.08), hemoglobin=8.3g/dL(11.9-14.6) were recorded. Haptoglobulin levels were low

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at 0.315g/L (0.3-2), and direct Coombs testing was negative. ADAMTS-13 activity was normal %99.82 (40-130).

Glomerular filtration rate (GFR) was calculated as 15ml/dk/1.73m^2. Protein value in urine analysis was ≥300(0-25). She was treated with antibiotic, diuretic, beta blocker, antihypertensive, corticosteroid, antiemetic and antiasid. In the present case also occurred tonic-clonic seizures twice in first week her inpatient stay and antiepileptic therapy (epanutin) 2x100mg is added to treatment by neurologist. There was no seizure in the follow up. Daily plasma exchange was initiated on day 14 postpartum but there were no improvement in thrombocytopenia and signs of hemolysis. The case was immediately administered a meningococcal vaccine and prophylactic ciprofloxacin treatment before starting of anti-C5 eculizumab therapy. Renal function of the patient started to deteriorate gradually and on the 24th postpartum hemodialysis was performed. One week later eculizumab was started following an induction schedule at a dose of 900 mg intravenously (IV) per week for 4 weeks. The patient's need for hemodialysis gradually decreased within 22 days after eculizumab treatment and was discharged on the 42nd day with full recovery. After discharge eculizumab was followed by 1200 mg every second week for 7 months. And she is still going on receiving 1200 mg eculizumab monthly for 6 months and antihypertensive, beta blocker and antiepileptic treatment. In our case, symptoms and treatment options were similar to worldwide cases (6-11).

Our patient could not breastfeed and interact with her newborn baby because of her hospitalization and medication process. During our interview with her medical history she was upset sometimes. She did not want to remember that days. She has no more pregnancy plan because of morbidity risk.

2.1. Nursing Management

Our patient was treated in the intensive care unit with severe hemolytic uremia that developed suddenly after cesarean section. During this period, hypertension, diffuse edema, anuric renal failure, liver dysfunction, anemia, thrombocytopenia, convulsions and anxiety are among the main problems. In addition, the patient experienced deterioration and delay in all functions expected after cesarean section. The main nursing diagnoses (ND) related to childbirth in this process can be listed as inadequacy in mother-infant attachment, maternal role performance, breastfeeding, family relations, and ineffective coping. In the care plan, current and potential risk diagnoses (risk for /electrolyte imbalance, impaired skin integrity, bleeding related thrombocytopenia, infection related with eculizimab therapy and immunosupression, ineffective cerebral tissue perfusion, sensory and cognitive alterations and confusion related with convulsion and uremic toxins) some have been addressed according to NANDA (North American Nursing Diagnosis Association) and NIC (Nursing Intervention Classification) (12).

2.2. Nursing Care Plan

Nursing Diagnosis 1. Excess fluid volume related with acute renal failure

Expected Outcome: The fluid-electrolyte balance of the individual will be maintained.

Interventions:

- Follow the peripheral, sacral, pretibial and periorbital edema
- Follow fluid intake and output
- Daily weight tracking
- Measure abdomen or extremity circumference
- Check laboratory results related to fluid retention (urea, creatine, sodium, potassium)
- Follow fluid overload symptoms (dyspnea, hypertension, CVP increase, neck vein engorgement)
- Restrict salt and fluid in the diet
- Check skin integrity
- Observe the effects and side effects of drugs given for edema (12-15).

Nursing Diagnosis 2. Impaired physical activity/Activity intolerance related with anemia, uremic toxins and cesarean operation

Expected Outcome: Optimal values are reached in laboratory results related to anemia and uremia.

Interventions:

- Provide a bed rest
- Evaluate the patient's motivation and reactions (saturation, heart rate) to the activity
- Help self-care requirements. Patient participation is encouraged as much as possible.
- Provide sufficient energy source
- Follow the patient's sleep pattern
- Follow treatments and laboratory findings for anemia and acute renal failure (12-15).

Nursing Diagnosis 3. Imbalanced nutrition: less than body requirement related to anorexia and nausea

Expected Outcome: The patient will eat and maintain a balanced diet.

- Apply the desired antiemetic before the meal.
- Elevate the head of the bed or place the patient in a position to avoid aspiration.
- Keep patient and bedding clean when vomiting occurs
- Immediately remove odor-causing substances (eg bedding, food)

- Avoid procedures that cause pain and nausea near mealtimes
- Carry out oral care after vomiting
- Apply a cool, damp cloth to the patient's wrists, neck, and forehead.
- Recommend that cold foods and other foods be less fragrant (12-15).

Nursing Diagnosis 4. Anxiety related to lack of knowledge of diagnostic tests, disease process and therapeutic regimen

Expected Outcome: She will make necessary decisions to change the negative situation around her and to take appropriate actions in accordance with her decisions.

Interventions:

- Assess the level of anxiety, the factors that influence the onset of anxiety.
- Evaluate patient level of understanding of diagnosis.
- Provide actual information about diagnosis, action, and prognosis.
- Provide a comfortable relaxed environment to express their feelings, fear, anger and perception.
- Provide opportunity for questions and answer patient and family members honestly.
- Encourage the family to accompany the client
- Assess the client's expectations to treatment and care.
- Teach relaxation techniques to reduce anxiety (12-15).

Nursing Diagnosis 5. Risk for bleeding related with thrombocytopenia

Expected Outcome: Uterus is felt to be firm on postpartum palpation.

- Follow up bleeding for wound and incision areas
- Follow the wound healing process
- Follow up the bleeding of the dressings
- Vital signs, especially blood pressure and heart rate, as indicated at risk level.
- Check hemoglobin and hematocrit values
- Follow the coagulation factors
- Follow the uterine height and stiffness regularly
- Observe the lochia for bright red bleeding and clotting
- Avoid injection (12-15).

Nursing Diagnosis 6. Risk for impared skin integrity

Expected Outcome: Skin and mucous membranes be structurally complete and show normal physiological function.

- Use a risk assessment tool (eg. Braden, Norton Scale)
- Evaluate the skin at first admission and every day
- Assess skin for pallor or redness
- Assess the individual's ability to move around in a chair or bed
- Assess nutritional status
- Determine if there is urinary or stool incontinence
- Turn the patient every 1-2 hours as appropriate
- Turn the patient carefully to avoid injuries to sensitive skin (eg avoid shear forces)
- Teach in-bed exercises
- Consult a dietitian about increasing the intake of high-energy foods (12-15).

3. DISCUSSION

P-aHUS can occur during pregnancy, but as confirmed by the data from the French Cohort (6) most cases occur in the post-partum period (>75%), when inflammation, the release of fetal cells into the maternal circulation, infections, and hemorrhage may trigger systemic complement activation. Similarly our case and her elder sister were P-aHUS cases as in the majority. It's important to detect genetic predisposition for the family. We informed the patient and family about the treatment, prognosis, complications, emergency and social transmitted infections and a new pregnancy that can trigger the relaps of the disease. It's important of a conscious, true, thorough, complete, and holistic assessment made by the nurses in the postpartum period, as well as the true diagnosis, and true nursing interventions that reduce the risk of morbidity and mortality.

4. CONCLUSION

As in the present case, the possibility of syndrome should be considered in such symptoms occurring in the postpartum period. It is thought that discussing care and treatment of this rarely seen case report will contribute to the literature.

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