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Case Report: Nursing Interventions on the Patient with Goodpasteur Syndrome

[1]Ms Tenzin Wosser, [2]Ms Lovely Negi

[1]Nursing Tutor, Medical Surgical Nursing, Holy Family College of Nursing, Okhla, India
[2]Nursing Tutor, Medical Surgical Nursing, Holy Family College of Nursing, Okhla, India

Email - [1]wlower949@gmail.com, [2]lovelynegi021193@gmail.com

Abstract: *Goodpasteur Syndrome also referred as anti-glomerular (anti-GBM) basement membrane disease is an autoimmune disease in which antibodies attach to collagen-fiber networks in the membrane, which triggers the membrane's destruction, causing alveolar hemorrhage, resulting in anemia, respiratory failure due to impaired gas exchange and acute or rapidly progressive Glomerulonephritis. The disease confirmed with pulmonary radiography, Kidney biopsy, and with the presence of increased anti-GBM antibodies. Imminent treatment should be possible combination of Plasmapheresis, immunosuppressive therapy and Haemodialysis.*
Case report: A 15 years old boy presented with symptoms of cough, haemoptysis, burning micturition and nausea, admitted in hospital and initially diagnosed with Anaemia and Renal dysfunction. Later shifted to Intensive care unit due to severe anaemia, continuous renal shut down and hypoxia. Diagnosis was confirmed with kidney biopsy which showed positive anti-GBM crescentic glomerulonephritis. He was kept on invasive ventilator (Volume Control) and treated with corticosteroids, cytostatic drug, multiple plasmapheresis and hemodialysis with anticoagulant therapy.
Discussion: The case report has enhanced the knowledge and skills in providing priority based comprehensive nursing care. The knowledge of Goodpasteur Syndrome is essential to act promptly in identifying and treating the disease. Through this case study, early diagnosis, prompt and aggressive treatment is associated with reduction of renal and pulmonary complications.

Keywords: Key: *Anti-glomerular (anti-GBM) basement membrane disease, Goodpasteur Syndrome, Cytostatic drug, Pasmapheresis.*

1. INTRODUCTION:

Goodpasteur Syndrome also referred as anti-glomerular (anti-GBM) basement membrane disease is an autoimmune disease that describes the clinical entity of diffuse pulmonary hemorrhage and acute or rapidly progressive glomerulonephritis(1).

The circulating autoantibodies in Goodpasteur Syndrome form antigen-antibody complexes in the glomerular basement membrane and causes crescentic glomerulonephritis. However, due to increase permeability of alveolar capillaries, autoantibodies also trespass the alveolar basement membrane and results in pulmonary hemorrhage. (2)
The disorder is named after Ernest Goodpasteur, an American pathologist who discovered it for the first time in 1919. However in 1967, Lerner Glassock and Dixon discovered anti-GBM antibodies which led to the understanding of the pathogenesis of Goodpasteur Syndrome. (3)

It is a rare syndrome. Its incidence is 0.5-1.8 cases per million per year in both European white and Asian populations. It commonly occurs in 20-30 years and 60-70 years of age group and more prevalent in younger men and older women. (4) It occurs more commonly in men and almost exclusively in smokers. (5)

Goodpasture syndrome appears to result from environmental factors with genetic predisposition. It has been found that mostly HLA-DR15 have been implicated in conferring increased genetic susceptibility to Goodpasture syndrome. The evrionmental factors include drugs (such as Alemtuzumab), Inhalation of Cocaine, infections such as influenza A2, smoking, exposure to metal dust, organic solvents or hydrocarbons and extracorporeal shock wave Lithotripsy then damage the alveoli, causing increased permeability and increased access to the basement membrane for

the autoantibodies. (6), (7) Circulating anti-glomerular basement membrane (GBM) antibodies attach to Alpha 3 chain of type 4 collagen-fiber networks in the membrane, which triggers the membrane's destruction, causing alveolar haemorrhage, resulting in anaemia and respiratory failure due to impaired gas exchange. (5) Disease in the younger age group is usually explosive, with haemoptysis, a sudden fall in haemoglobin, fever, dyspnoea, and hematuria.(9) Percutaneous kidney biopsy is the gold standard procedure to establish the diagnosis of Goodpasture syndrome. But lung biopsy may be performed where renal biopsy is contraindicated for any reason. (10)

This case report emphasizes on the nursing care of a 15 years old male child in India diagnosed with the Goodpasteur Syndrome.

2. CASE REPORT :

A 15 years old Indian male presented to the hospital with a one week history of cough, haemoptysis, burning maturation and nausea. He was a non-smoker and had no significant past medical history.

On admission, his vital signs were stable. Initial Laboratory investigations showed Haemoglobin: 7gm/dl, Serum creatinine: 7.8 mg/dl, Sodium: 133mEq/L, Potassium: 4.4 mEq/L, Uric acid: 8.6 mg/dl, Total Leukocyte Count: 12.8 x 103cells/mm3, Serum albumin: 2.5 g/dl. Later on 5th day of hospitalization, he was shifted to Intensive care unit (ICU) in view of severe anaemia, renal dysfunction and hypoxia (SPO2: 85% @ 2l/min Oxygen).

In ICU, patient was kept in positive pressure room. Investigations including all ICU routine test, serum procalcitonin, Extractable Nuclear Antigen profile (ENA), Antinuclear Antibody profile (ANA), Anti- proteinase -3 (PR-3), Anti- Myeloperoxidase (MPO), Anti- Glomerular Basement Membrane (Anti-GBM), Complement component3 and 4(C3 and C4) and anti-neutrophil cytoplasmic antibodies (ANCA) was sent. The reports were non-significant except for ANCA and Anti-GBM report which was positive. Urine examination showed proteinuria (2+), and hematuria (RBCs: 70-80/hpf). Patient also underwent Kidney biopsy which revealed Anti- GBM crescentic glomerulonephritis.

Patient started on Injection Solumedrol 500 mg OD for 3 days followed by Tab Wysolone 10mg OD and underwent haemodialysis and plasma exchange regularly thereafter. Patient also received2 doses of Injection Rituximab 500 mg. He was improving clinically but on day 14, patient started developing fever, shortness of breath and massive hematemesis for which elective intubation was performed. On fifth day of ventilation, he was extubated and placed on 4L/min Oxygen via simple face mask. Later on, when patient's clinical condition improved, he was shifted to Semi-ICU and got discharge from medicine ward on day 21st of hospitalization.

During the course of hospitalization, patient has received 3 units of packed red blood cell (PRBC) in view of anaemia, underwent haemodialysis 15 times and had plasmapheresis 7 times.

3. NURSING INTERVENTIONS :

Ineffective airway clearance related to thick secretion from infectious process as evidenced by increased mucous production and altered respiratory rate, rhythm and depth.

Ineffective breathing pattern related to respiratory muscle fatigue as evidenced by abnormal breathing pattern. (11)

1) Assessing the patency of the airway.
2) Monitoring rate, rhythm, depth of respiration and SPO_2.
3) Auscultating lungs for breath sounds.
4) Performing analysis.
5) Providing semifowler's position to the patient.
6) Administering supplemental oxygen to the patient (FiO2: 40 % to 100 %).
7) Performing endotracheal suctioning using aseptic technique (if patient is intubated).
8) Assessing the sputum colour, consistency and amount.

Risk for infection related to compromised host defense secondary to immunosuppressive therapy.

1) Assessing the patient for sign and symptoms of infection including fever, redness, edema, pain, purulent discharge from catheters (Urinary catheter, central line catheter).
2) Monitoring colour of endotracheal secretions.
3) Monitoring white blood cell (WBC) counts.
4) Placed patient in a positive pressure room.
5) Ensuring any articles used should be properly disinfected or sterilized before use.
6) Maintaining proper hygiene before contacting the patient or his unit.
7) Maintaining strict asepsis for endotracheal suctioning, Foley's catheter care and central line care.

Impaired urinary elimination related to inflammation of the nephrons and glomerulus as evidenced by oliguria/anuria.

1) Monitoring strict urine output of the patient.
2) Monitoring Kidney Function Test (KFT), Urine culture and Urine R/M.

3) Catheterizing the patient as advised by doctor.
4) Administering diuretic as prescribed by doctor.
5) Assisted dialysis technician in performing haemodialysis as prescribed by doctor.

Fatigue related to decreased hemoglobin and decreased oxygen carrying capacity of the blood as evidenced by report of lack of energy.

1) Assessing the patient's ability to perform activities of daily living (ADLs) and the demands of daily living.
2) Monitoring hemoglobin, hematocrit, RBC counts, and reticulocyte counts.
3) Identifying the cause of fatigue.
4) Assisting patient in developing a schedule for daily activity and rest.
5) Providing supplemental oxygen therapy as prescribed by doctor.
6) Transfused Packed Red blood Cell (PRBC) as ordered by physician.

Deficient knowledge related to newly diagnosed disease condition as evidenced by questioning.

1) Assessing the patient's and his family abilities to learn about disease condition.
2) Educating patient and his family about disease condition, its cause, sign and symptoms and its treatment.
3) Educating the importance of compliance to the treatment.
4) Clarifying the doubts of patient and family members.

4. DISCUSSION:

Case report presented by Jagoda Stojkovikj et al showed that patient (49 years old) came with the pulmonary haemorrhage and glomerunephritis and diagnosed with Goodpasteur syndrome using renal biopsy. The p-ANCA levels were also positive (8.93 U/ml). In the current study, patient had similar complaints and diagnosed same while p-ANCA levels were negative. (2)

Study conducted by Rui Fernandes et al presented case report of 27 years old white man with Goodpasteur syndrome without absence of anti-Glomerular basement membrane antibodies (anti-GBM). The current case report showed 15 years old black man diagnosed with goodpasteur syndrome and positive anti-GBM antibodies.(1)

The similar case has been reported by Joseph McAllister, Pradeep Nagisetty and Kay Tyermanin 2022, of a 14-year-old boy was presented with acute kidney failure and severe pulmonary haemorrhage due to anti-GBM disease, confirmed on auto-antibody testing. Later diagnosed with thrombocytopenia and moderately low ADAMTS13 activity suggestive of Thrombocytopenic thrombotic purpura (TTP). Due to poor renal prognosis, the patient had undergone dialysis and kept on extracorporeal membrane oxygenation (ECMO) for pulmonary haemorrhages. Treated with corticosteroids, plasma exchange (PEX), Rituximab, and Cyclophosphamide. (4)

A case study conducted by Zhong et al, 2020 on 38 year old Chinese man who was presented with lung lesion with chief complaints of haemoptysis without fever, cough, chest pain and oedema. Laboratory testing revealed that the urinary protein level and urine erythrocyte count were 7.4 g/24 hours and 144/high-power field (HPF), respectively. Serological testing for anti-GBM antibodies was negative. Chest computed tomography revealed multiple exudative lesions in both lungs, indicating alveolar infiltration and hemorrhage. Kidney biopsy revealed cellular crescent formation and segmental necrosis of the globuli, with linear IgG and complement C3 deposition on the GBM. These findings were consistent with the diagnosis of anti-GBM antibody nephritis. The patient underwent 7 sessions of plasmapheresis and also administered with intravenous methylprednisolone and cyclophosphamide. After renal function stabilization, he was discharged under an immunosuppressive regimen comprising of glucocorticoids and cyclophosphamides. (12)

5. CONCLUSION:

Anti-GBM is rare in young children and adults. An aggressive treatment with methylprednisolone, PEX, Rituximab and cyclophosphamide need to be considered. (13) Despite prolonged periods of ventilation, a full pulmonary recovery is possible and disease remission achievable, allowing future kidney transplantation. (14)

The above case report has enhanced the knowledge and skills in providing priority based comprehensive nursing care. Thus, the knowledge of Goodpasteur Syndrome is essential to act promptly in identifying and treating the disease. (15)

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