ejpmr, 2023,10(7), 416-418

SJIF Impact Factor 6.222

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Report

ISSN 2394-3211

EJPMR

A CASE REPORT ON PULMONARY RENAL SYNDROME IN ANCA ASSOCIATED VASCULITIS

Alwy Joseph[1]*, Divya Thomas[1] and Dr. Shamnad P.[2]

[1]Pharm D. Intern, Nazareth College of Pharmacy, Othera P.O, Thiruvalla, Kerala.
[1]Pharm D. Intern, Crescent College of Pharmaceutical Sciences, Payangadi, Kannur.
[2]Consultant, MBBS, MD, DrNB Nephrology, Department of Nephrology, Believers Church Medical College Hospital, Thiruvalla, Kerala.

***Corresponding Author: Alwy Joseph**

Pharm D. Intern, Nazareth College of Pharmacy, Othera P.O, Thiruvalla, Kerala.

Article Received on 10/05/2023 **Article Revised on 30/05/2023** **Article Accepted on 21/06/2023**

ABSTRACT

Pulmonary renal syndrome (PRS) is a life-threatening condition characterised by renal failure associated with respiratory failure and manifesting as rapidly progressive glomerulonephritis (RPGN) with diffuse alveolar hemorrhage (DAH). This condition is usually associated with ANCA vasculitis. So it is a key element that life-threatening complications can result from this condition. So timely diagnosis and management are crucial in this situation.

KEYWORDS: Pulmonary renal syndrome, Rapidly progressing glomerulonephritis, ANCA vasculitis, Anti phospholipid antibody syndrome, Pulmonary Hemorrhage.

INTRODUCTION

Pulmonary renal syndrome (PRS) is characterised by renal failure accompanied by respiratory impairment. Rapidly progressing glomerulonephritis with diffuse alveolar haemorrhage (DAH) is a symptom of pulmonary renal syndrome (PRS), a life-threatening condition.[1] The majority of cases of pulmonary-renal syndrome are associated with ANCAs, either c-ANCA or p-ANCA, due to autoantibodies against the target antigens proteinase-3 and myeloperoxidase, respectively. Treatment of generalised ANCA-associated vasculitis consists of corticosteroids and immunosuppressive agents such as cyclophosphamide (as induction therapy) or azathioprine (as maintenance therapy once remission has been achieved).[2]

CASE REPORT

A 77-year-old female with a past medical history of type 2 diabetes mellitus, dyslipidemia, and systemic hypertension. She presented with complaints of bilateral upper and lower limb weakness and rashes, difficulty swallowing, and slurring of speech for the past 10 days. On evaluation, the patient was hemodynamically stable, had purpuric rashes over the extremities, and had quadriplegia. Her investigations revealed anemia, thrombocytopenia, an increased creatinine level of 5.8 mg/dl, and active sediments in the URE. The possibility of RPGN with systemic vasculitis was considered. The results of the skin biopsy pointed to cutaneous vasculitis. Since there were numerous RBCs in the urine upon microscopic examination, a renal biopsy was deferred. The patient was initiated on IV Methyl Prednisone Pulse 500 mg in 3 doses. Later, P-ANCA was positive, and as a result, the patient was initiated on induction therapy with IV cyclophosphamide 500 mg.

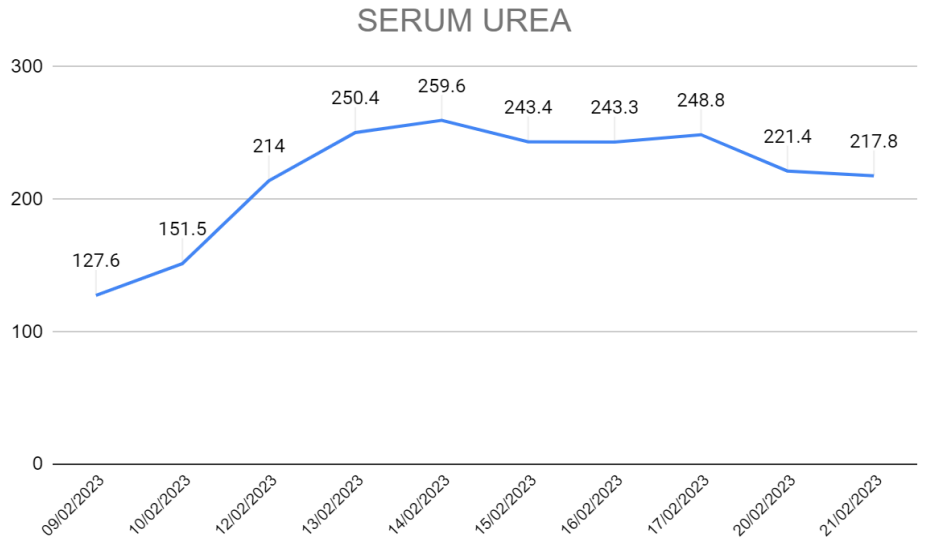
The patient developed breathing difficulty and started showing new-onset chest signs. The Chest X-ray showed bilateral fluffy infiltrates. An HRCT was performed to rule out infection or Diffuse alveolar hemorrhage (DAH), and the results were suggestive of DAH. The patient was initiated on plasmapheresis due to overall multisystem involvement; however, on the second day, the patient's health condition deteriorated, and she succumbed to death due to pulmonary hemorrhage.

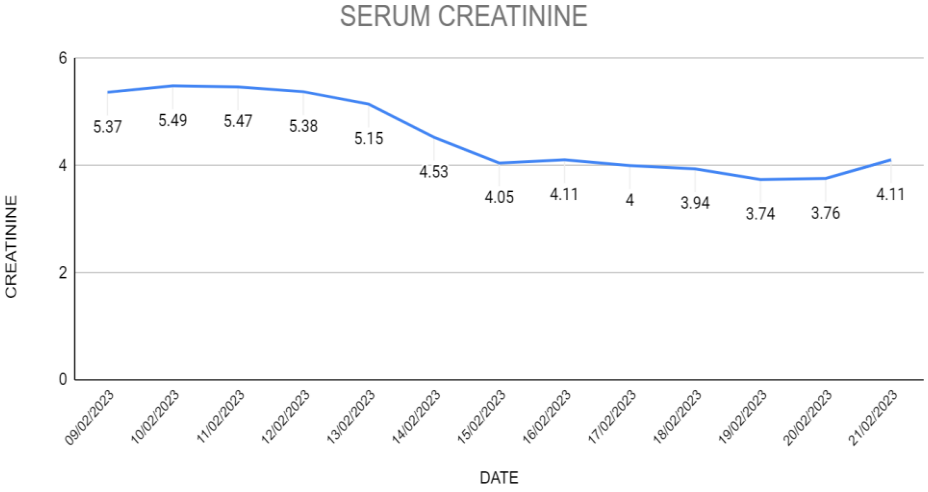
DISCUSSION

Pulmonary renal syndrome (PRS) is a term most commonly used to describe the combination of glomerulonephritis and pulmonary hemorrhage as a manifestation of multisystem autoimmune disease. It is often associated with ANCA vasculitis and Anti-GBM disease.[1] It usually affects the lungs and kidneys. The upper respiratory tract and lower respiratory tract are also involved, which causes symptoms such as rhinitis, sinusitis, deafness, or epistaxis, and alveolar hemorrhage that usually manifests as cough, hemoptysis, shortness of breath, and/or hypoxemia. It commonly affects the kidneys (sometimes only one organ is involved, called "renal-limited vasculitis"), causing hematuria, proteinuria, and elevated serum creatinine levels.[3] The diagnosis of ANCA is very important because it involves

many systems. A detailed history and examination are required. Laboratory tests should include assessment of inflammatory markers, renal function, and serologic tests, including ANCA, antinuclear antibodies, and anti-glomerular basement membrane antibodies. A chest X-ray, CT scan, and MRI can be used to evaluate the structure of the chest, brain, eyes, ears, and throat in more detail. Diagnosis may be confirmed by biopsy, which shows perinuclear cytoplasmic fluorescence on ethanol-fixed neutrophils. ANCA vasculitis should be differentiated from other conditions because it mimics many other conditions, and thus most diagnosis are still confused with other conditions. Therefore, conditions such as infection, antiphospholipid antibody syndrome, thrombocytopenic purpura, sickle cell disease, atrial mucinous embolism, and atheroma cholesterol embolism are some of them.[4] Treatment of ANCA vasculitis primarily focus on remission induction therapy initiated at the initial stage or or on the recurrent diagnosis of Granulomatosis with polyangitis (GPA) or Microscopic polyangitis (MPA). Induction lasts 3 to 6 months. The first-line agents to induce remission in severe GPA and MPA include cyclophosphamide or rituximab, as well as glucocorticoids. Although recent evidence suggest that plasmapheresis is beneficial for severe ANCA patients, it is not routinely recommended for most patients. Plasma exchange (PLEX) can be considered in selected patients in conjunction with expert vasculitis opinions. Remission can be maintained using rituximab, azathioprine, and methotrexate. Leflunomide and Mycophenolate mofetil can be used when rituximab, azathioprine, and methotrexate are contraindicated or intolerable. The role of low-dose prednisone in maintaining remission has not been definitively demonstrated or established.[5]

In this case, our patient had a poor prognosis due to RPGN, age, and the late treatment due to the delayed diagnosis. Her renal parameters, such as creatinine and urea, were deranged. These all contribute to the burden of the disease. Despite receiving methylprednisolone and cyclophosphamide therapy, she could not be saved. Therefore, prompt recognition is required, as respiratory failure and end-stage renal failure can develop rapidly.[6]





CONCLUSION

This case highlights the poorer outcomes in patients with ANCA vasculitis when there is a delayed diagnosis and also highlights the potential complications that may arise. The importance of paying attention to the symptoms and signs that this disease presents cannot be overstated.

ACKNOWLEDGEMENT

The authors would like to thank the Department of Nephrology, Believers Church Medical College Hospital, Department of Neurology, Believers Church Medical College Hospital, Department of Pharmacy Practice Nazareth College of Pharmacy, Othera, Thiruvalla, Pathanamthitta and Department of Pharmacy Practice Crescent College of Pharmaceutical Sciences, Payangadi, Kannur for their extreme support and guidance

CONFLICTS OF INTEREST

There are no conflicts of Interest.

ABBREVIATIONS

PRS: Pulmonary Renal Syndome
RPGN : Rapidly progressing Glomerulo Nephritis
DAH : Diffuse Alveolar Hemorrhage
C- ANCA : Antineutrophil Cytoplasmic Autoantibody, Cytoplasmic
P- ANCA: Perinuclear Antineutrophil Cytoplasmic Autoantibody anti
URE : Urine Routine Examination
RBCs : Red Blood Cells
HRCT : High Resolution Computed Tomography
Anti GBM Disease: Anti Glomerular Basement Membrane Disease
MRI: Magnetic Resonance Imaging
GPA : Granulomatosis with Polyangitis
MPA: Microscopic Polyangitis
PLEX : Plasma Exchange

REFERENCE

1. Saladi L, Shaikh D, Saad M, Cancio-Rodriguez E, D'Agati VD, Medvedovsky B, Uday KA, Adrish M. Pulmonary renal syndrome: a case report of diffuse alveolar hemorrhage in association with ANCA negative pauci-immune glomerulonephritis. Medicine, Jun, 2018; 97(23).
2. Brusselle GG. Pulmonary-renal syndromes. Acta clinica Belgica, Apr 1, 2007; 62(2): 88-96.
3. Almaani S, Fussner LA, Brodsky S, Meara AS, Jayne D. ANCA-associated vasculitis: an update. Journal of clinical medicine, Apr 1, 2021; 10(7): 1446.
4. Yates M, Watts R. ANCA-associated vasculitis. Clinical Medicine, Feb, 2017; 17(1): 60.
5. Samman KN, Ross C, Pagnoux C, Makhzoum JP. Update in the management of ANCA-associated vasculitis: recent developments and future perspectives. International Journal of Rheumatology, Apr 8, 2021; 2021: 1-4.
6. Aggarwal N, Rai AK, Weiss R, Kupfer Y. ANCA Vasculitis: Fatal Pulmonary Hemorrhage. Chest, Oct 1, 2016; 150(4): 504A.