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Case Report

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PR3-ANCA associated vasculitis with rapidly progressive glomerulonephritis (RPGN):

an uncommon cause of rapidly developing renal failure

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Abstract

PR3-ANCA-associated vasculitis is a rare autoimmune disease characterized by systemic inflammation, primarily affecting the respiratory tract but occasionally leading to rapidly progressive glomerulonephritis (RPGN). We present the case of a 64-year-old male with diabetes, hypertension, and COPD, who developed RPGN and pulmonary manifestation, initially manifesting as frothy urine, pedal edema, and dyspnoea. Diagnostic evaluation revealed positive PR3-ANCA, proteinuria, hematuria, and significant renal pathology, necessitating hemodialysis and immunosuppressive therapy with cyclophosphamide . This case underscores the need for early recognition and tailored management of PR3-ANCA vasculitis to improve patient outcomes.

Keywords: Antineutrophil Cytoplasmic Antibody-Associated Vasculitis, Proteinase 3, Glomerulonephritis Rapidly Progressive, Cyclophosphamide

INTRODUCTION

PR3-ANCA(+) associated vasculitis is a rare autoimmune condition characterized by systemic inflammation and predominantly small vessel involvement, often leading to target organ damage. PR3-ANCA vasculitis patients exhibit significantly more upper and lower respiratory tract manifestations, including destructive nasal/sinus lesions, granulomas, and pulmonary nodules or cavities.[1,2,3] Rapidly progressive glomerulonephritis (RPGN), developing over days to months, is a medical emergency, frequently resulting in acute kidney injury necessitating urgent management. Renal involvement is more commonly associated with MPO-ANCA.[4,5] However, PR3-ANCA is also implicated. Approximately 40% to 50% of adult RPGN cases are classified as pauci-immune, which includes both PR3-ANCA and MPO-ANCA types.[6]

We describe a case of PR3-ANCA associated vasculitis with rapidly progressive glomerulonephritis (RPGN) to raise awareness of the disease, since PR3-ANCA vasculitis present predominantly with granulomatous inflammation rather than renal involvement. In addition, an exact diagnosis will affect the treatment strategies like the use of steroids, rituximab or other immunosuppressive therapies.

CASE PRESENTATION

PR3-ANCA, A 64 year-old male from North 24 parganas, West Bengal, presented to our OPD with history of frothy urine and bilateral pedal edema for the last 1 month, which has been gradually increasing. He was also having significant cough and dyspnoea since the last 7 days. The patient had been diagnosed with type 2 diabetes mellitus, hypertension, and chronic obstructive pulmonary disease (COPD) for which he has been on treatment for the last 16 years. There was no history of fever, chest pain, or joint pain. The patient has history of frequent nasal congestion and discharge suggestive of upper respiratory tract involvement . There was history of burning sensation in bilateral distal lower limb without any weakness in the body.

On general survey the patient had no pallor, icterus, cyanosis, clubbing or lymphadenopathy. There was bipedal edema upto mid leg. Blood pressure was 138/82 mm Hg and pulse rate was 110/ min, palpable on all 4 limbs, regular with no radio-radial or radio-femoral delay. The respiratory rate was 30/ min. and SpO2 was 85% on room air. Examination of the respiratory system revealed bilateral rhonchi and coarse crepitations; cardiovascular and gastrointestinal system examinations were unremarkable. There was mild tenderness over the paranasal sinuses. Musculoskeletal system revealed normal tone bulk; no significant power loss in all 4 limbs, no joint swelling, tenderness or deformity. On neurological examination, pupils were bilaterally equal and reacting to light, plantar reflexes were flexor and all superficial reflexes were normal with decreased pain and touch sensation over the distal lower limb.

On admission, hemoglobin was 10.6 (MCV 95.4, MCH29.2, MCHC 30.6 – normocytic, normochromic), the total leucocyte count (TLC) was 21500 (N89 L05 M 06)/cumm, platelets count was 2.6 X 105/ L. Serum urea was 169 mg/dl, creatinine 6.5mg/dl, sodium 133meq/L, potassium 4.7meq/L. Routine urine examination showed proteinuria (2+) and RBC (2+); urine culture showed no growth, and 24 hr urine protein excretion was 1263 mg/day. Serum inflammatory markers procalcitonin was 7.70 (<0.5)ng/ml and CRP was 118.7mg/dl . Sputum CBNAAT was negative and a 12 lead ECG was showed normal sinus rhythm. USG abdomen showed right kidney 8.3 cm, left kidney 9.3 cm in longitudinal diameter. No significant change in Cortico-medullary differentiation . Chest X-ray was done which showed bilateral (Right> Left) reticular infiltrates, HRCT thorax showed bronchiectatic changes in both lower lobe with no features suggestive of ILD, nodule, cavity. 2D Echo study was within normal limits. Serum Anti Proteinase 3-Anti -Neutrophil Cytoplasmic Antibody (PR3-ANCA) reports were positive. A kidney biopsy was done which showed over 50% of the glomeruli showing extensive crescent formation.

Based on the history and investigations, the diagnosis of PR3-ANCA associated vasculitis with rapidly progressive glomerulonephritis (RPGN) along with Lower respiratory tract infection was done. In view of the elevated urea and creatinine reports and oliguria , patient was started on hemodialysis (HD) on alternate days. The patient was also treated with moist oxygen, empirical antibiotics, nebulization, insulin . The antidiabetic and antihypertensive medication doses were titrated.

SpO2 increased to 96% with oxygen flow of 10L/min through face mask. With treatment, patient improved significantly with decreased cough, no dyspnea at rest without O2 supplementation. However, patient remained oliguric although not showing any sign/ symptoms of fluid overload with alternate day HD. Leukocytosis decreased with total count 8600/cu.mm and serum urea and creatine were 90mg/dl and 4.6mg/dl respectively. After controlling infection, Inj. Cyclophosphamide pulse was given .

DISCUSSION

The presented case underscores the complexity of diagnosing PR3-ANCA-associated vasculitis, particularly in patients with overlapping comorbidities such as diabetes mellitus, hypertension, and COPD. The patient's clinical presentation of increased Frothiness of Urine, pedal edema, dyspnea, and cough coupled with laboratory findings of proteinuria, hematuria, elevated inflammatory markers, positive PR3-ANCA and biopsy findings strongly pointed towards RPGN secondary to PR3-ANCA vasculitis.

PR3-ANCA vasculitis differs from MPO-ANCA vasculitis in terms of genetic predisposition and pathogenesis. Genetic variants such as HLA-DPB1 have been linked to PR3-ANCA positivity.[7] Pathophysiologically, PR3 antibodies activate neutrophils, leading to endothelial damage and granulomatous inflammation. Pulmonary involvement is common due to neutrophil-mediated injury in alveolar capillaries.[8] Although typical pulmonary involvement in AAV is in the form of diffuse alveolar hemorrhage (DAH), pulmonary nodules/cavities or ILD, structural airway abnormalities like bronchiectasis has been reported in 19% of AAVs overall and 13% in patients with proteinase 3-ANCA.[9]

Given the patient's oliguric state, hemodialysis alongside supportive measures such as oxygen therapy and antibiotics were initiated.[10,11,12] The role of steroids was limited because of diabetes. So, the patent was started on Intravenous pulse administration of cyclophosphamide (CYC). Despite improvements in systemic symptoms and inflammatory markers, presence of persistent oliguria, these patients may develop irreversible renal damage if diagnosis is delayed. Non responders may be tried with Inj. Rituximab. This case emphasizes the need for heightened clinical suspicion and early intervention to prevent long-term complications.

CONCLUSION

This case report presents a 64-year-old male diagnosed with PR3-ANCA-associated vasculitis manifesting as RPGN. The patient's complex presentation required a multidisciplinary approach involving hemodialysis, antibiotics, cyclophosphamide and in some cases, rituximab. This case highlights the importance of prompt diagnosis and tailored management in PR3-ANCA vasculitis to improve outcomes and mitigate organ damage

CONFLICT OF INTEREST

None Declared

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