NEPHROLOGY/IMMUNOLOGY

INDEX COPERNICUS INTERNATIONAL

ICV value for 2024 = 82.51

Case Report | International Journal of Medical Case Reports

Microscopic Polyangiitis Presenting With Diffuse Alveolar Hemorrhage In A Patient With Chronic Kidney Disease.

Arpita Palit[1], **Jaideep Chaudhary**[2]

[1]Medanta Hospital, Gurugram, Haryana, India.
[2]Max Hospital, Dwarka, Delhi, India.

ISSN 2455-0574

ABSTRACT

Background:

Diffuse alveolar hemorrhage (DAH) is a rare, life-threatening manifestation of pulmonary–renal syndrome and may occur in microscopic polyangiitis (MPA), a myeloperoxidase-antineutrophil cytoplasmic antibody (MPO-ANCA)-associated small-vessel vasculitis. Early diagnosis is particularly challenging in patients with pre-existing chronic kidney disease (CKD), in whom worsening renal function and pulmonary infiltrates may be misattributed to progression of CKD, infection, or fluid overload.

Case Report:

A 60-year-old man with type 2 diabetes mellitus, hypertension, and stage 4 CKD presented with fever, haemoptysis, weakness, oliguria, and hypoxemia. Investigations revealed severe anemia (hemoglobin 5.5 g/dL), leukocytosis, and acute-on-chronic kidney injury with serum creatinine rising from a baseline of 3.8 mg/dL to 6.4 mg/dL. Chest radiograph showed bilateral patchy consolidations. High-resolution computed tomography (HRCT) chest demonstrated diffuse ground-glass opacities with septal thickening, suggestive of DAH. Despite empiric broad-spectrum antibiotics, oxygen therapy, blood transfusions, and renal replacement therapy, hypoxemia and haemoptysis persisted. Flexible bronchoscopy on day 3 showed progressively bloodier bronchoalveolar lavage aliquots, confirming DAH. Serology revealed MPO-ANCA positivity, and a diagnosis of MPA presenting as pulmonary–renal syndrome was established. The patient received high-dose intravenous methylprednisolone, six sessions of plasma exchange, and two doses of rituximab. He showed progressive clinical improvement with resolution of haemoptysis, stabilization of hemoglobin, improved oxygenation, and recovery of renal parameters, and was discharged in stable condition on day 24.

Conclusion:

MPA should be considered in patients with CKD who develop haemoptysis, anemia, bilateral pulmonary infiltrates, and acute worsening of renal function. Early recognition of DAH using imaging, bronchoscopy, and serology, followed by prompt immunosuppressive therapy, can be lifesaving.

Keywords: Alveolar Hemorrhage, Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis, Chronic Kidney Disease, Microscopic Polyangiitis, Rituximab

INTRODUCTION

Diffuse alveolar hemorrhage (DAH) is a rare but fulminant clinicopathologic syndrome which is caused by disruption of the alveolar–capillary basement membrane. This leads to bleeding into the alveolar spaces and rapidly progressive respiratory compromise. Clinically, it usually presents with haemoptysis, anemia, diffuse pulmonary infiltrates and hypoxemic respiratory failure. DAH most often affects middle-aged to older adults and frequently requires intensive care support, especially when associated with concurrent renal involvement. In such patients, the coexistence of pulmonary hemorrhage and kidney dysfunction should immediately raise concern for a pulmonary–renal syndrome, a medical emergency that warrants urgent etiologic evaluation and prompt immunosuppressive therapy.[1]

Microscopic polyangiitis (MPA) is a necrotizing pauci-immune small-vessel vasculitis and one of the principal clinicopathologic forms of ANCA-associated vasculitis. It is strongly associated with myeloperoxidase (MPO)-ANCA, with anti-MPO specificity identified in the majority of patients, and most commonly involves the kidneys and lungs.

International Journal Of Medical Case Reports (ISSN 2455-0574) is an indexed medical journal indexed in Index Copernicus

Access this Journal Online

Quick Response Code

Website: www.ijomcr.net

Email:ijomcr@gmail.com

Dr Arpita Palit

Medanta Hospital, Gurugram, Haryana, India.
Email: arpitapalit13@gmail.com

Renal disease in MPA usually takes the form of pauci-immune necrotizing crescentic glomerulonephritis, whereas pulmonary disease may present as DAH or, less commonly, chronic interstitial fibrosis. Because MPA often lacks the granulomatous upper airway manifestations that may suggest granulomatosis with polyangiitis, its initial presentation can be more subtle and easily overlooked, particularly in patients with multiple comorbidities. Epidemiologically, MPA is more frequently recognized in older adults, and renal involvement is one of its dominant determinants of long-term outcome. Importantly, pulmonary manifestations may occur simultaneously with renal injury or may become clinically apparent only after kidney disease has already evolved. This overlap between pulmonary and renal injury makes MPA a prototypical cause of pulmonary–renal syndrome and emphasizes the need for clinicians to recognize DAH not merely as an isolated pulmonary catastrophe, but as a manifestation of systemic small-vessel vasculitis. [2]

The diagnosis of MPA presenting with DAH is especially challenging in patients who already have chronic kidney disease (CKD). In such individuals, a rising creatinine level may be incorrectly attributed to progression of pre-existing renal dysfunction, volume depletion, sepsis, or nephrotoxic exposure rather than superimposed active vasculitic glomerulonephritis. Likewise, pulmonary infiltrates in a patient with CKD may initially be misinterpreted as pulmonary edema, infection, or uremic lung injury. This issue is clinically important because DAH in ANCA-associated vasculitis is a severe manifestation associated with substantial early morbidity, and its outcome depends heavily on rapid recognition of the underlying autoimmune process. [3]

Therapeutically, ANCA-associated vasculitis with life-threatening organ involvement requires immediate induction treatment. High-dose glucocorticoids remain the cornerstone of initial therapy, while rituximab has emerged as an effective remission-induction agent that is noninferior to cyclophosphamide for severe ANCA-associated vasculitis and is now widely used in contemporary practice. Adjunctive plasma exchange has historically been considered in patients with severe renal impairment or diffuse pulmonary hemorrhage; however, evidence from large randomized trials has tempered expectations regarding its effect on the composite outcome of death or end-stage kidney disease, even though selected critically ill patients may still receive it on an individualized basis. In real-world settings, especially when DAH threatens ventilation and oxygenation, clinicians often continue to use a multimodal approach that combines pulse corticosteroids with either rituximab or cyclophosphamide, with plasma exchange considered in carefully selected cases. These therapeutic decisions are particularly complex in patients with CKD, in whom infection risk, dialysis dependence, drug toxicity, and the difficulty of assessing renal recovery all influence management. Accordingly, case-based descriptions remain valuable in illustrating how diagnostic reasoning and treatment sequencing are applied in acutely unstable patients. [4]

Despite advances in the understanding of ANCA-associated vasculitis, important knowledge gaps remain regarding the early recognition of MPA when it presents as DAH in patients with pre-existing CKD. Published literature has largely focused on DAH as part of pulmonary–renal syndrome in previously undiagnosed vasculitis, whereas fewer reports emphasize the diagnostic pitfalls created by chronic baseline renal impairment, where acute vasculitic injury may be underestimated and pulmonary hemorrhage may initially mimic infection or fluid overload. Similarly, although rituximab-based induction is now well established, the role and timing of adjunctive plasma exchange in individual cases with severe DAH and acute-on-chronic kidney injury remain matters of clinical judgment rather than uniform consensus. In this context, the present case is noteworthy because it highlights MPA presenting with DAH in a patient with advanced CKD, demonstrates the importance of integrating radiologic, bronchoscopic, and serologic findings for timely diagnosis, and illustrates favorable clinical recovery following aggressive immunomodulatory therapy. By documenting this presentation, our study seeks to reinforce awareness of vasculitic DAH in CKD, underline the need for early suspicion of pulmonary–renal syndrome despite pre-existing renal disease, and contribute practical insight into the management of this uncommon but life-threatening clinical scenario. [5]

CASE REPORT

A 60-year-old male was brought to the emergency department with complaints of fever, cough with blood-tinged sputum and reduced urine output since three days. He was a known history of type 2 diabetes mellitus (T2DM), essential hypertension and stage 4 chronic kidney disease (CKD). On presentation, the patient appeared sick and was having dyspnea. He was found to have tachycardia (HR=122 bpm), blood pressure of 150/77 mm of Hg and respiratory rate of 22 breaths per minute. Oxygen saturation at room air was found to be 82%. There was no fever at the time of admission. Physical examination showed presence of pallor and signs of respiratory ditress in the form of nasal flaring. On auscultation bilateral diffuse crepitations with occasional wheeze was present.

Initial laboratory investigations showed severe anemia (hemoglobin level of 5.5 g/dL), leukocytosis (TLC of 14,740 cells/mm^3) and a normal platelet count of 272,000 cells/mm^3. Renal function tests were significantly deranged. Serum creatinine level was found to be 6.4 mg/dL which were significantly elevated from a baseline of 3.8 mg/dL suggestive of acute-on-chronic kidney injury. Ultrasonography of the abdomen showed features consistent with early medical renal disease, along with chronic cystitis and prostatomegaly. Chest radiography revealed patchy bilateral consolidations. High-resolution computed tomography (HRCT) of the chest showed presence of diffuse ground-glass opacities and interlobular septal thickening and intralobular lines. These HRCT findings raised suspicion for possibility of diffuse alveolar hemorrhage (DAH) (Figure 1).

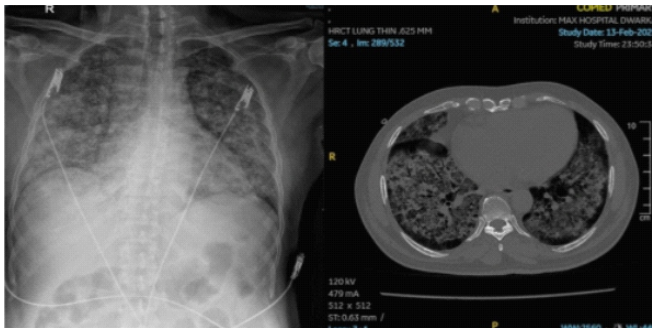

FIGURE 1: Chest X-ray showing bilateral diffuse patchy airspace opacities suggestive of alveolar consolidation (Left), HRCT chest showing diffuse ground-glass opacities with superimposed interlobular septal thickening and intralobular lines consistent with a "crazy paving" pattern (Right).

The patient was admitted to the intensive care unit. Broad-spectrum intravenous antibiotics (teicoplanin, meropenem and clindamycin) were started considering the possibility of severe pneumonia or sepsis. He also received supplemental oxygen therapy, packed red blood cell transfusions for severe

anemia and renal replacement therapy in view of worsening renal function and oliguria. Despite aggressive management the patient continued to have persistent hypoxemia and increasing haemoptysis. On the third day of hospitalization flexible bronchoscopy was performed, which showed presence of progressively bloodier aliquots on bronchoalveolar lavage. These findings confirmed the diagnosis of diffuse alveolar hemorrhage. In view of diffuse alveolar haemorrhage serological testing was done which revealed positivity for myeloperoxidase-antineutrophil cytoplasmic antibodies (MPO-ANCA). Based on the clinical presentation of pulmonary hemorrhage, renal dysfunction, and serological findings a diagnosis of microscopic polyangiitis (MPA) presenting as pulmonary–renal syndrome was established. Following confirmation of DAH, high-dose intravenous methylprednisolone (1 g daily) was initiated.

Given the severity of presentation plasmapheresis was started on the ninth day of hospitalization. The patient underwent total six cycles of plasma exchange. Following high dose methylprednisolone and plasmapheresis gradual improvement in oxygenation and renal parameters was observed. On the sixteenth day of hospitalization rituximab was administered in a dose of 500 mg with a second dose given subsequently as per protocol. Patient showed marked clinical improvement and haemoptysis resolved completely. Hemoglobin levels and renal function also improved. The patient received a total of six doses of intravenous methylprednisolone, two doses of rituximab, and six sessions of plasmapheresis during his hospital stay. He was successfully weaned off supplemental oxygen and did not require further respiratory support. on day 24 of hospitalization discharge from hospital was planned. At the time of discharge, the patient was clinically stable with improved respiratory status and renal parameters, He was advised close outpatient follow-up for ongoing management of ANCA-associated vasculitis.

DISCUSSION

This case highlights a classic but diagnostically treacherous pulmonary–renal presentation of microscopic polyangiitis (MPA), made more difficult by the patient's pre-existing stage 4 chronic kidney disease. In the large retrospective series by Lauque et al[6] alveolar hemorrhage associated with MPA occurred in patients with a mean age in the mid-50s and was frequently accompanied by necrotizing glomerulonephritis, underscoring that simultaneous pulmonary and renal injury is a defining clinical pattern rather than an exceptional association. Similarly, Schirmer et al[7] in their monocentric cohort of 144 patients with MPA, confirmed that renal involvement is one of the dominant organ manifestations and strongly shapes long-term outcome. Our patient fits this phenotype closely: he presented with haemoptysis, profound anemia, bilateral pulmonary infiltrates, and acute worsening of baseline renal dysfunction. The notable teaching point, however, is that pre-existing CKD can obscure the vasculitic nature of renal decline. In many real-world settings, a rise in serum creatinine in a diabetic or hypertensive patient is initially attributed to progression of chronic nephropathy, sepsis-associated acute kidney injury, or drug-related nephrotoxicity. The present case demonstrates why that assumption can be dangerous. The abrupt increase in creatinine from 3.8 to 6.4 mg/dL, in parallel with pulmonary hemorrhage and hypoxemia, was much more consistent with an acute pulmonary–renal syndrome than with uncomplicated CKD progression. In that respect, our case extends the observations of Lauque[6] and Schirmer[7] by showing how chronic baseline renal impairment may delay recognition of superimposed pauci-immune vasculitic activity.

The case therefore reinforces an important clinical message: in older adults with CKD, haemoptysis plus unexplained anemia and new bilateral opacities should prompt immediate evaluation for ANCA-associated vasculitis even before renal biopsy is feasible.

A second important aspect of this case is the way the diagnosis of diffuse alveolar hemorrhage (DAH) was established through integration of imaging, clinical deterioration, and bronchoscopy. As emphasized by West et al[8] haemoptysis and dyspnea are common in ANCA-associated DAH but are nonspecific, and high-resolution computed tomography is more sensitive than plain radiography in identifying diffuse ground-glass change. Likewise, Park et al[9] stressed that early bronchoscopy with bronchoalveolar lavage is often required both to confirm DAH and to exclude infection. Our patient's course mirrored this diagnostic pathway precisely. Initial chest radiography showed bilateral patchy consolidations that could reasonably have been interpreted as severe pneumonia or pulmonary edema, especially in the context of CKD and oliguria. HRCT then demonstrated diffuse ground-glass opacities with septal thickening, raising suspicion for alveolar hemorrhage, but the definitive turning point came with bronchoscopy on day 3, which showed progressively bloodier lavage aliquots. That finding was decisive because it shifted the working diagnosis from predominantly infectious lung disease to pulmonary capillaritis-related hemorrhage. This sequence is clinically important: in unstable patients, DAH is often masked by empiric antibiotic treatment and competing explanations for infiltrates. Our case supports the approach advocated by West[8] and Park[9] that bronchoscopy should not be unduly delayed once DAH is suspected, particularly when severe anemia, persistent hypoxemia, and bilateral diffuse opacities coexist. The case also demonstrates the value of radiologic-bronchoscopic correlation in CKD patients, where volume overload and infection are common mimics.

The serologic profile and renal context in this patient are also highly consistent with MPA rather than other ANCA-associated vasculitic syndromes. Jennette et al[10] described MPA as a pauci-immune necrotizing small-vessel vasculitis in which crescentic glomerulonephritis and hemorrhagic pulmonary capillaritis are common and together constitute one of the classic forms of pulmonary–renal syndrome. More recently, Geetha et al[11] analyzing patients with renal involvement in the RAVE trial, confirmed that renal disease remains a central determinant of presentation and treatment response in severe ANCA-associated vasculitis. In our patient, MPO-ANCA positivity in the setting of DAH and acute-on-chronic kidney injury provided a compelling clinico-serologic diagnosis of MPA, particularly because no upper airway granulomatous features suggested granulomatosis with polyangiitis. Although renal biopsy would have added histopathologic confirmation, the patient's critical respiratory status, severe anemia, and urgent need for immunosuppressive treatment made immediate clinico-serologic decision-making appropriate. This reflects an important practical principle in vasculitis care: while tissue diagnosis remains desirable, life-threatening organ involvement often necessitates treatment before biopsy can be safely obtained. Our case further illustrates that in patients with advanced CKD, the threshold for suspecting active vasculitic nephritis must be lower, not higher. The improvement in renal parameters after steroids, plasma exchange, and rituximab retrospectively supports the interpretation that a substantial component of renal dysfunction was inflammatory and reversible rather than solely chronic structural kidney disease. Thus, this case aligns well with the disease construct outlined by Jennette[10] and the

renal-treatment observations reported by Geetha[11] while emphasizing the added diagnostic difficulty imposed by longstanding CKD.

Therapeutically, this case supports the contemporary strategy of immediate high-dose glucocorticoids followed by rituximab-based remission induction in organ-threatening ANCA-associated vasculitis. In the landmark RAVE trial, Stone et al[12] showed that rituximab was not inferior to cyclophosphamide for remission induction in severe ANCA-associated vasculitis. The RITUXVAS trial by Jones et al[13] similarly demonstrated that a rituximab-based strategy was comparable to cyclophosphamide-based treatment in severe renal vasculitis. These studies are highly relevant to the present patient, who had both life-threatening pulmonary hemorrhage and marked renal impairment. Once DAH was confirmed on bronchoscopy and MPO-ANCA returned positive, pulse methylprednisolone was appropriately initiated, followed by rituximab. The favorable course—resolution of haemoptysis, improved oxygenation, stabilization of hemoglobin, and recovery of renal parameters—supports the effectiveness of this approach in critically ill MPA. Of note, Cartin-Ceba et al[14] specifically evaluated DAH secondary to ANCA-associated vasculitis and found that rituximab treatment was associated with higher rates of complete remission at 6 months than cyclophosphamide in that cohort, including among patients with respiratory failure. Although caution is warranted in extrapolating cohort data to every individual case, our patient's response is directionally concordant with that observation. Another practical advantage of rituximab in a patient like ours is avoidance of cumulative cyclophosphamide toxicity, which is particularly relevant in older individuals with CKD, anemia, and infection vulnerability. Therefore, the present case not only aligns with randomized evidence from Stone and Jones but also adds bedside support to the growing preference for rituximab-based induction in selected patients with MPA-associated DAH and acute-on-chronic kidney injury.

Historically, plasma exchange was often added in patients with severe renal failure or pulmonary hemorrhage, and the MEPEX trial by Jayne et al[15] helped establish that adjunctive plasma exchange could improve short-term renal recovery in severe renal vasculitis. However, the much larger PEXIVAS trial by Walsh et al[16] later showed that plasma exchange did not reduce the composite outcome of death or end-stage kidney disease in severe ANCA-associated vasculitis overall, even though diffuse pulmonary hemorrhage was one of the inclusion criteria. More recently, the PEXIVAS DAH analysis reported by Fussner et al[17] examined outcomes specifically in the subgroup with alveolar hemorrhage and further refined the discussion around whether plasma exchange meaningfully changes respiratory or survival outcomes in this setting. In our patient, plasma exchange was started after the diagnosis had been clarified and was used as part of a multimodal rescue strategy because of severe DAH, marked hypoxemia, and significant renal dysfunction requiring dialysis support. It is impossible from a single case to isolate the independent contribution of plasma exchange from that of glucocorticoids, rituximab, transfusion support, and renal replacement therapy. Nevertheless, the patient improved after combined therapy without need for mechanical ventilation, suggesting that individualized use of plasma exchange may still be reasonable in carefully selected critically ill patients despite the neutral overall PEXIVAS result. Overall, our case emphasizes that in MPA presenting with DAH on a background of CKD, prompt recognition and rapid institution of combined immunomodulatory therapy can still lead to favorable short-term recovery, even when presentation is initially confounded by competing diagnoses such as infection, pulmonary edema, or progression of chronic kidney disease.

CONCLUSION:

This case underscores the complexity from diagnostic and management point of view in cases of microscopic polyangiitis presenting as diffuse alveolar hemorrhage in a patient who had pre-existing chronic kidney disease. In such as a case both pulmonary and renal manifestations may be initially misattributed to more common conditions. This case highlights the importance of early suspicion of pulmonary–renal syndrome, timely use of bronchoscopy and serology for timely diagnosis and prompt initiation of immunosuppressive therapy. A multidisciplinary as well as individualized treatment approach can lead to favorable outcomes even in critically ill patients with overlapping comorbidities.

Source Of Funding: None Declared.

Conflict Of Interest : None

REFERENCES

1. Lara AR, Schwarz MI. Diffuse alveolar hemorrhage. Chest. 2010 May;137(5):1164-71.
2. Shi J, Shen Q, Chen XM, Du XG. Clinical characteristics and outcomes in microscopic polyangiitis patients with renal involvement: a study of 124 Chinese patients. BMC Nephrol. 2019 Sep 2;20(1):339. doi: 10.1186/s12882-019-1535-3. PMID: 31477033; PMCID: PMC6719371.
3. Karras A, Jayne D. Microscopic polyangiitis: new insights into pathogenesis, clinical features and therapy. Autoimmun Rev. 2018;17(12):1203-1209.
4. Stacey HL, Francis L, Smith RM, Jones RB. Practical Management of ANCA-Associated Vasculitis: A Clinician's Perspective. Glomerular Dis. 2024 Dec 18;5(1):84-102. doi: 10.1159/000543159. PMID: 39991192; PMCID: PMC11845170.
5. Wutzl AL, Foley RN, O'Driscoll BR, Reeve RS, Chisholm R, Herrick AL. Microscopic polyangiitis presenting as pulmonary-renal syndrome in a patient with long-standing diffuse cutaneous systemic sclerosis and antibodies to myeloperoxidase. Arthritis Rheum. 2001 Dec;45(6):533-6. doi: 10.1002/1529-0131(200112)45:6<533::aid-art379>3.0.co;2-d. PMID: 11762687.
6. Lauque D, Cadranel J, Lazor R, Pourrat J, Ronco P, Guillevin L, Cordier JF. Microscopic polyangiitis with alveolar hemorrhage. A study of 29 cases and review of the literature. Medicine (Baltimore). 2000;79(4):222-233. doi:10.1097/00005792-200007000-00003
7. Schirmer JH, Wright MN, Vonthein R, Herrmann K, Nölle B, Both M, et al. Clinical presentation and long-term outcome of 144 patients with microscopic polyangiitis in a monocentric German cohort. Rheumatology (Oxford). 2016;55(1):71-79. doi:10.1093/rheumatology/kev286
8. West S, Arulkumaran N, Ind PW, Pusey CD. Diffuse alveolar haemorrhage in ANCA-associated vasculitis. Intern Med. 2013;52(1):5-13. doi:10.2169/internalmedicine.52.8863
9. Park MS. Diffuse alveolar hemorrhage. Tuberc Respir Dis (Seoul). 2013;74(4):151-162. doi:10.4046/trd. 2013. 74.4.151
10. Jennette JC, Thomas DB, Falk RJ. Microscopic polyangiitis (microscopic polyarteritis). Semin Diagn Pathol. 2001;18(1):3-13.

11. Geetha D, Specks U, Stone JH, et al. Rituximab versus cyclophosphamide for ANCA-associated vasculitis with renal involvement. J Am Soc Nephrol. 2015;26(4):976-985. doi:10.1681/ASN.2014010046
12. Stone JH, Merkel PA, Spiera R, et al. Rituximab versus cyclophosphamide for ANCA-associated vasculitis. *N Engl J Med*. 2010;363(3):221-232. doi:10.1056/NEJMoa0909905
13. Jones RB, Tervaert JWC, Hauser T, et al. Rituximab versus cyclophosphamide in ANCA-associated renal vasculitis. N Engl J Med. 2010;363(3):211-220. doi:10.1056/NEJMoa0909169
14. Cartin-Ceba R, Diaz-Caballero L, Al-Qadi MO, et al. Diffuse alveolar hemorrhage secondary to antineutrophil cytoplasmic antibody-associated vasculitis: predictors of respiratory failure and clinical outcomes. *Arthritis Rheumatol*. 2016;68(6):1467-1476. doi:10.1002/art.39562
15. Jayne DRW, Gaskin G, Rasmussen N, et al. Randomized trial of plasma exchange or high-dosage methylprednisolone as adjunctive therapy for severe renal vasculitis. J Am Soc Nephrol. 2007;18(7):2180-2188. doi:10.1681/ASN.2007010090
16. Walsh M, Merkel PA, Peh CA, et al. Plasma exchange and glucocorticoids in severe ANCA-associated vasculitis. N Engl J Med. 2020;382(7):622-631. doi:10.1056/NEJMoa1803537
17. Fussner LA, King TE Jr, Rhee RL, et al. Alveolar hemorrhage in antineutrophil cytoplasmic antibody-associated vasculitis: results of an international randomized controlled trial (PEXIVAS). Am J Respir Crit Care Med. 2024;209(5):565-573. doi:10.1164/rccm.202308-1522OC

*****

Author Contribution :
AP-contributed to patient management, data collection, and manuscript drafting. **JC**-participated in data interpretation, literature review, and critical manuscript revision.

Received : 15-01-2026 Revised: 20-02-2026 Accepted : 10-03-2026