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Microscopic Polyangiitis Masquerading as Recurrent Acute Heart Failure in Advanced Chronic Kidney Disease: A Case Report

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Abstract

Background: Microscopic polyangiitis (MPA) is an antineutrophil cytoplasmic antibody (ANCA)-associated necrotizing small-vessel vasculitis. Its initial manifestations are frequently nonspecific, so diagnosis is often delayed, particularly in elderly patients with multiple comorbidities. In advanced chronic kidney disease (CKD), superimposed pauci-immune glomerulonephritis may be overlooked because renal dysfunction is attributed to pre-existing disease. Early recognition is essential, as untreated disease may rapidly become fatal. **Case presentation:** An 80-year-old man with stage 4 hypertensive CKD, permanent atrial fibrillation on apixaban, heart failure with preserved ejection fraction (HFpEF) presented with recurrent acute pulmonary edema, hemoptysis, and progressive anemia. Initial work-up suggested decompensated heart failure and pneumonia, with only transient improvement on diuretic and antibiotics. Repeated hospitalizations revealed persistent inflammation, worsening renal function, new proteinuria, microscopic hematuria, and anemia requiring transfusions. Despite extensive evaluation, the diagnosis remained unclear until immunological testing revealed strongly positive myeloperoxidase antineutrophil cytoplasmic antibody MPO-ANCA (>134 IU/mL) with negative anti-glomerular basement membrane antibody (anti-GBM). Severe renal atrophy and bleeding risk precluded biopsy, so a clinico-serological diagnosis of microscopic polyangiitis with pulmonary-renal syndrome was made. Bron-

choscopy confirmed diffuse alveolar hemorrhage. Rituximab, glucocorticoids, and avacopan were initiated, but the patient developed recurrent diffuse alveolar hemorrhage and fatal mesenteric infarction. **Conclusion:** This case shows how microscopic polyangiitis may masquerade as recurrent heart failure in patients with advanced CKD, causing diagnostic delay. Recurrent hemoptysis, unexplained anemia, persistent inflammation, and new proteinuria or hematuria should prompt evaluation for ANCA-associated vasculitis even when comorbidities offer an alternative explanation.

Keywords

Microscopic Polyangiitis, ANCA-Associated Vasculitis, MPO-ANCA, Pulmonary-Renal Syndrome, Diffuse Alveolar Hemorrhage, Chronic Kidney Disease, Heart Failure Mimic, Case Report

1. Introduction

Microscopic polyangiitis (MPA) is a systemic necrotizing small-vessel vasculitis within the spectrum of antineutrophil cytoplasmic antibody (ANCA)-associated vasculitides (AAV). MPA is strongly associated with myeloperoxidase-specific ANCA (MPO-ANCA) [1]. ANCA-mediated neutrophil activation, endothelial injury, and alternative-pathway complement activation drive the necrotizing vasculitis [2]. MPA has an annual incidence of 10 - 20 cases per million and peak onset in the sixth to eighth decades [3] [4]. Renal involvement occurs in up to 90% of patients while pulmonary involvement affects one-third. Their coexistence constitutes the pulmonary-renal syndrome, one of the most severe AAV presentations [5].

Clinical presentation is heterogeneous. Constitutional symptoms often precede organ-specific disease from weeks to months. Pulmonary involvement ranges from asymptomatic infiltrates to diffuse alveolar hemorrhage (DAH). Renal disease may begin with microscopic hematuria and proteinuria before progressing to irreversible failure [6]. These features mimic heart failure, pneumonia, embolism, infection, or malignancy, delaying diagnosis, especially in elderly patients with alternative diagnoses. Kidney biopsy remains the diagnostic gold standard, but may be contraindicated by renal atrophy, anticoagulation, advanced CKD, or bleeding risk [7]. Current guidance accepts a diagnosis, based on a characteristic pulmonary-renal syndrome and strongly positive MPO-ANCA, when biopsy cannot safely be obtained and delaying treatment would risk irreversible organ damage [8] [9].

We report a patient with advanced CKD and established heart disease in whom recurrent pulmonary edema, elevated natriuretic peptides, and transient diuretic response initially suggested decompensated heart failure. Progressive hemoptysis, transfusion-dependent anemia, inflammatory syndrome, de novo proteinuria, and hematuria lead to MPO-ANCA-associated vasculitis. Despite remission-in-

duction therapy, the disease progressed to diffuse alveolar hemorrhage and fatal mesenteric infarction. This case highlights the need for a high index of suspicion for AAV when pulmonary and renal manifestations evolve despite conventional heart-failure therapy.

2. Case Presentation

An 80-year-old man was admitted in April 2024 with acute dyspnea, hemoptysis, and rapidly conducted atrial fibrillation. The overall clinical course, including key diagnostic and therapeutic milestones, is summarized in Figure 1. His history included long-standing hypertension with stage 4 hypertensive-related chronic kidney disease (CKD), permanent atrial fibrillation on apixaban since 2021, pulmonary embolism in 2020, pacemaker implantation for atrioventricular block in 2022, and HFpEF. Regular medications were apixaban, furosemide, ramipril, bisoprolol, weekly darbepoetin alfa, and sodium polystyrene sulfonate. He had completed a course of amoxicillin-clavulanic acid seven days earlier for a presumed pneumonia.

On admission, he reported progressive dyspnea, repeated low-volume hemoptysis, but no constitutional symptoms. Physical examination revealed bilateral mid-lung crackles on auscultation, oxygen saturation at 86% on room air, blood pressure of 170/100 mmHg, and rapid atrial fibrillation at 120 bpm. There was no limb edema, rash, purpura, digital ischemia, or neuropathy. Body temperature was 37.4˚C.

Biology showed normocytic normochromic anemia (hemoglobin 8.5 g/dL), C-reactive protein 44 mg/L, serum creatinin 284 µmol/L (eGFR 17 mL/min/1.73 m^2, CKD-EPI), blood urea nitrogen 21 mmol/L, and NT-proBNP 22,658 ng/L. Leukocytes count, platelets, haptoglobin, LDH, aminotransferases, and bilirubin were normal. Blood cultures were negative. Routine laboratory investigations performed before admission (January 15, 2024) showed no urinary sediment abnormalities (no dysmorphic red blood cells, red blood cell casts, leukocytes, or granular casts). Hemoglobin was 11.0 g/dL, serum creatinine 202 µmol/L, estimated glomerular filtration rate (eGFR, CKD-EPI) 26 mL/min/1.73 m^2, proteinuria 0.05 g/L, and the urine protein-to-creatinine ratio was 11 mg/mmol.

Chest computed tomography (CT) demonstrated bilateral pleural effusions, bilateral interstitial and alveolar infiltrates predominantly in the lower lung fields, interpreted as pulmonary edema with possible superimposed pneumonia (Figure 2, panels 1 - 2). Transthoracic echocardiography showed left ventricle (LV) ejection fraction 60%, elevated LV filling pressures, pulmonary hypertension, and a dilated inferior vena cava. There was no valve disease. Right ventricle was not dilated and its function was normal. These findings were consistent with acute decompensated HFpEF.

Patient was managed for acute cardiogenic pulmonary edema with rapid atrial fibrillation and presumed community-acquired pneumonia. Apixaban was temporarily discontinued because of hemoptysis. Intravenous furosemide, supple-

mental oxygen, non-invasive ventilation, tranexamic acid, and ceftriaxone were administered. There was initial clinical improvement following important diuresis, blood pressure, and heart rate control. Renal function deteriorated; patient became hypotensive prompting discontinuation of ramipril. There was no urinary tract obstruction.

Days after, the patient developed persistent fever (38˚C - 39˚C). Infective endocarditis was considered, but multiple blood cultures remained sterile. Repeated echocardiogram showed no vegetation or pacemaker device-related infection. Culture-negative endocarditis assessment was unremarkable. Complement levels (C3, C4), CH50, rheumatoid factor, and urinalysis were unremarkable. Urinary Legionella and pneumococcal antigens, sputum cultures, and Interferon-Gamma Release Assay (QuantiFERON) were negative. Nevertheless, ceftriaxone and spiramycin were given empirically for seven days for pneumonia. A chest radiograph obtained during a subsequent hospitalization again showed diffuse bilateral alveolar infiltrates with a butterfly-wing appearance and an enlarged cardiac silhouette, again suggestive of pulmonary edema (**Figure 2**, panel 3).

Further investigations excluded an infectious, malignant, or granulomatous process. Whole-body FDG PET-CT showed no inflammatory foci.

Patient clinically transiently improved. Fever resolved; inflammatory markers normalized, but hemoglobin progressively declined to 8.0 g/dL. There was no other bleeding or hemolysis. The patient had courses of transfusion. Repeated urinalysis now showed persistent microscopic hematuria and newly developed proteinuria (protein concentration 0.40 g/L; protein-to-creatinine ratio 108 mg/mmol), both absent three months earlier.

The coexistence of recurrent hemoptysis, progressive anemia, inflammatory syndrome, worsening renal function, new hematuria and proteinuria prompted consideration of an underlying systemic vasculitis. An extended immunological work-up demonstrated strongly positive myeloperoxidase-specific ANCA (MPO-ANCA > 134 IU/mL; reference < 3.5 IU/mL), negative anti-glomerular basement membrane antibodies. ANCA testing was performed using a stepwise approach. Indirect immunofluorescence (IIF) demonstrated a perinuclear ANCA (P-ANCA) pattern. Anti-myeloperoxidase (MPO) antibodies were strongly positive by both multiplex immunoassay (BioPlex® 2200, Bio-Rad Laboratories; antibody index > 8, reference < 1) and fluorescence enzyme immunoassay (EliA® on the ImmunoCAP/Phadia 250 platform, Thermo Fisher Scientific; index > 134 IU/mL, negative if <3.5 IU/mL). In contrast, anti-proteinase 3 (PR3) antibodies were negative by multiplex immunoassay (BioPlex® 2200, Bio-Rad Laboratories; antibody index < 0.2, reference < 1). Overall, these findings were consistent with P-ANCA of anti-MPO specificity, supporting the diagnosis of MPO-ANCA-associated vasculitis. Antinuclear antibodies, antiphospholipid antibodies, anti-CCP antibodies, extractable nuclear antigen antibodies (anti-Sm, RNP, SSA, SSB, Scl-70, Jo-1) were all negative.

The patient was transferred to a tertiary center. A follow-up chest CT scan

performed on June 13 demonstrated marked resolution of the bilateral alveolar consolidations, marked regression of the bilateral pleural effusions, and persistence of only subtle residual ground-glass opacities. Bronchoscopy demonstrated hemorrhagic bronchoalveolar lavage fluid. The sequential lavage aliquots became progressively bloodier. Broncho alveolar lavage cultures remained sterile. These findings were considered consistent with diffuse alveolar hemorrhage.

Given renal atrophy, advanced CKD, chronic anticoagulation, and a high anticipated bleeding risk, kidney biopsy was judged unlikely to alter management and was not performed after multidisciplinary discussion. Based on the characteristic pulmonary-renal syndrome and strongly positive MPO-ANCA serology, with alternative diagnoses excluded, a diagnosis of microscopic polyangiitis was established.

Remission-induction therapy was initiated with high-dose glucocorticoids, rituximab, and avacopan.

The patient received intravenous methylprednisolone 500 mg/day from June 18 to June 20, followed by oral prednisone at 1 mg/kg/day with gradual tapering (60 mg/day until June 30, 30 mg/day until July 7, 20 mg/day until July 14, 15 mg/day until July 20). Rituximab was administered as four weekly infusions of 600 mg (June 21, June 27, July 4, and July 11), together with avacopan 30 mg twice daily. Infection prophylaxis included vaccination with the 20-valent pneumococcal conjugate vaccine (PCV20; Prevnar 20®) on June 23, trimethoprim-sulfamethoxazole (80/400 mg) three times weekly, and valacyclovir 500 mg once daily. Serial changes in hemoglobin, C-reactive protein, proteinuria, creatinine, and eGFR over following-up, relative to the timing of immunosuppressive treatment, are shown in **Figure 3**.

Despite treatment, the patient developed recurrent diffuse alveolar hemorrhage with respiratory distress and hypoxic cardiac arrest. Following resuscitation, invasive mechanical ventilation, norepinephrine, and intermittent renal replacement therapy were started. Apixaban was discontinued and was not resumed thereafter due to the persistent high bleeding risk. Empirical piperacillin-tazobactam therapy was initiated because of the severity of the patient's condition; however, repeated microbiological investigations were subsequently negative.

Cyclophosphamide was withheld given the patient's advanced age, frailty, and rapidly evolving multi-organ failure. Nine days later, the patient developed progressive abdominal distension, diffuse abdominal pain, and feculent vomiting. Contrast-enhanced abdominal CT revealed extensive bowel wall pneumatosis, highly suggestive of transmural bowel necrosis involving the transverse colon, left colon, and part of the small intestine. The superior and inferior mesenteric arteries were opacified only in their proximal segments, consistent with extensive distal mesenteric arterial occlusion. Unfortunately, the CT images were not available for inclusion in the manuscript. The patient subsequently developed refractory shock and died of multiorgan failure despite maximal supportive care.

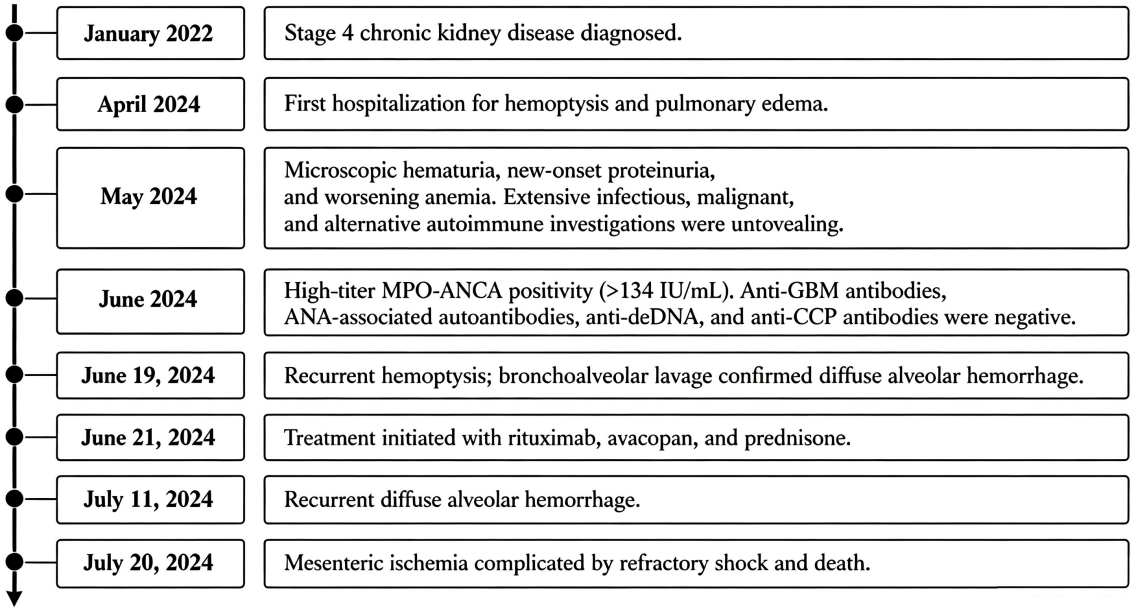

Figure 1. Clinical timeline summarizing key diagnostic milestones and therapeutic interventions from the initial diagnosis of stage 4 chronic kidney disease through the patient's death.

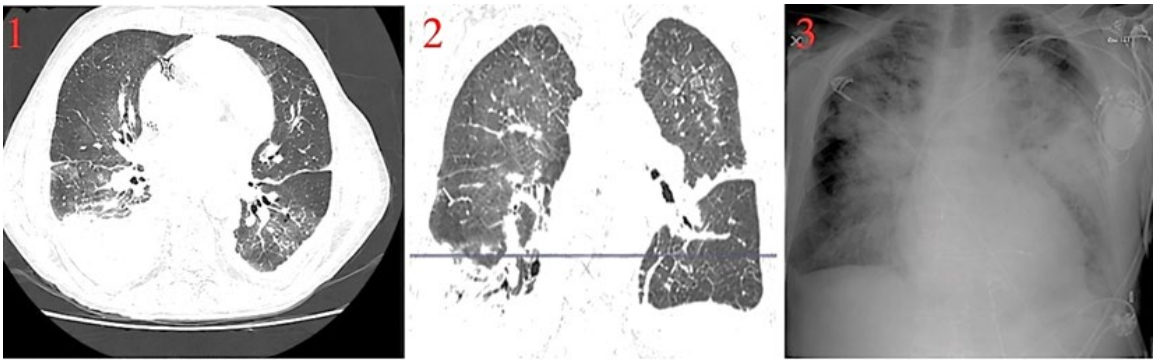

Figure 2. Thoracic imaging findings. (1, 2) Axial and coronal views of non-contrast chest computed tomography showing bilateral pleural effusions and diffuse alveolar-interstitial infiltrates, initially interpreted as pulmonary edema. 3) Chest radiograph showing diffuse bilateral alveolar infiltrates with a butterfly-wing appearance and an enlarged cardiac silhouette, suggestive of pulmonary edema at a subsequent admission.

3. Discussion

Microscopic polyangiitis is a systemic necrotizing small-vessel vasculitis within the spectrum of ANCA-associated vasculitides, characterized histologically by pauci-immune necrotizing inflammation without granulomatous lesions. Circulating MPO-ANCA, detected in 60% - 80% of patients, play a central pathogenic role by activating primed neutrophils, promoting endothelial injury, and triggering the alternative complement pathway, particularly through C5a-mediated amplification of inflammation [1] [2].

This case illustrates diagnostic pitfalls of MPA in elderly patients with comorbidities. The initial presentation was strongly suggestive of acute decompensated heart failure. Established HFpEF, markedly elevated NT-proBNP, pulmonary hy-

pertension, bilateral pleural effusions, and diffuse infiltrates that improved transiently with diuresis provided a convincing alternative explanation and delayed consideration of vasculitis. Such delays are well recognized in AAV, since constitutional and pulmonary manifestations frequently mimic infection, heart failure, pulmonary embolism, and malignancy [1] [5].

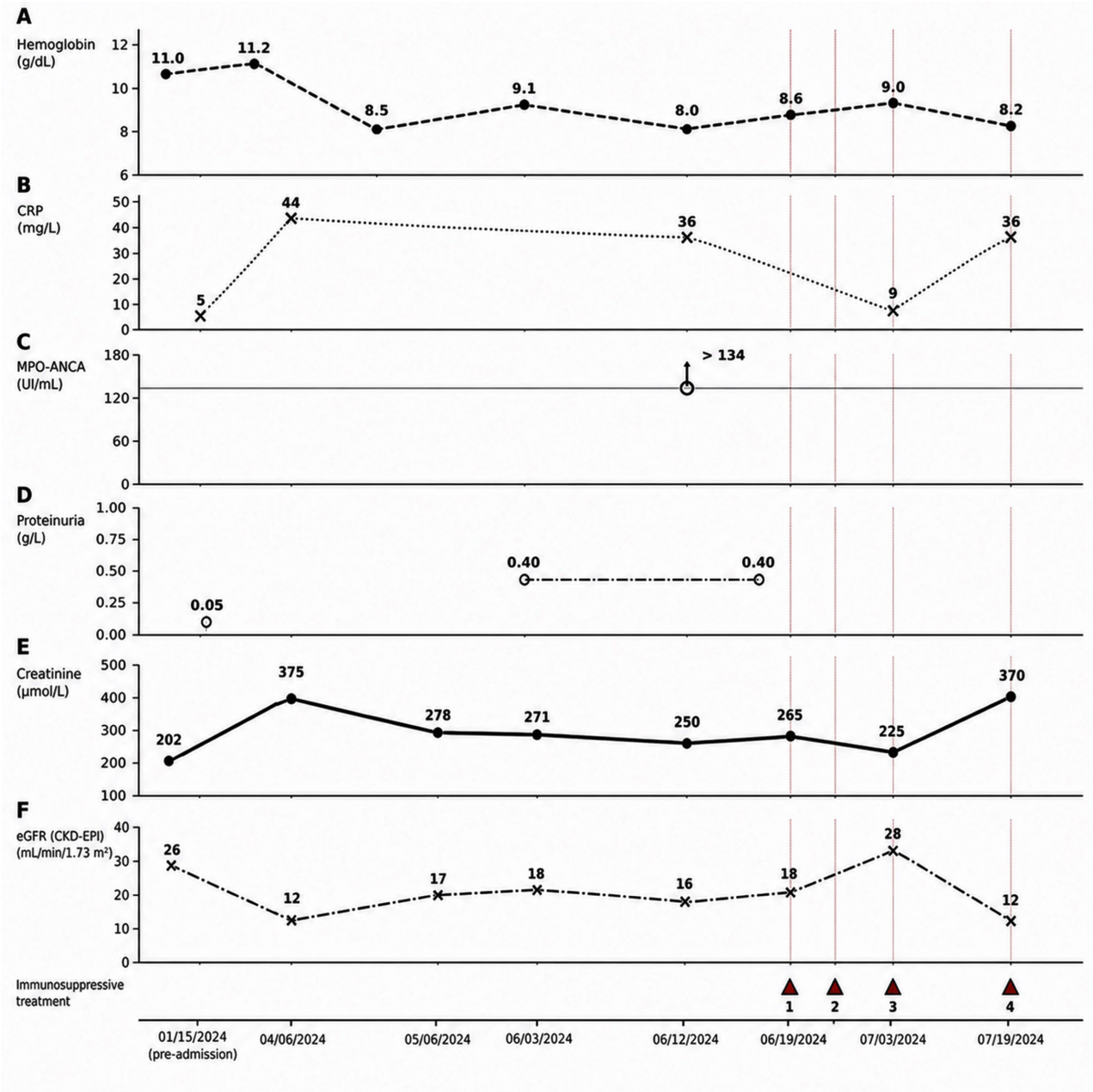

Figure 3. Serial trends in selected laboratory parameters — (A) hemoglobin, (B) C-reactive protein, (C) MPO-ANCA titer, (D) proteinuria, (E) serum creatinine, and (F) estimated glomerular filtration rate (CKD-EPI) — relative to the timing of immunosuppressive treatment (red triangles, numbered 1 - 4, denote the four weekly rituximab infusions).

One of the most important clinical clues was recurrent hemoptysis, initially attributed to pulmonary edema, anticoagulation, and presumed pneumonia. Alt-

hough hemoptysis is common with pulmonary congestion or anticoagulant therapy, recurrent episodes with progressive anemia should raise suspicion for diffuse alveolar hemorrhage (DAH), particularly without a focal pulmonary lesion. DAH is among the most severe manifestations of AAV, occurring in 12% - 55% of patients with MPA [5] [6]. Hemoptysis itself may nonetheless be absent in up to a third of patients, making diagnosis particularly challenging. In this case, bronchoscopy demonstrated hemorrhagic bronchoalveolar lavage, confirming DAH after several weeks of diagnostic uncertainty.

Progressive deterioration of kidney function was initially attributed to pre-existing stage 4 CKD and recurrent heart failure, but the appearance of de novo proteinuria and microscopic hematuria represented a genuine change from baseline and is typical of pauci-immune necrotizing glomerulonephritis. Such findings should never be dismissed as simple progression of hypertensive nephrosclerosis. In advanced CKD, superimposed glomerulonephritis is easily overlooked, as worsening renal function is often attributed to cardiorenal syndrome, dehydration, or nephrotoxicity. Current international recommendations emphasize that new-onset active urinary sediment, particularly with systemic inflammation or pulmonary manifestations, should prompt urgent investigation for AAV [8] [9].

The decline in hemoglobin despite the absence of overt bleeding or hemolysis was another important feature. Although anemia is common in advanced CKD, the combination of worsening anemia, recurrent hemoptysis, elevated inflammatory markers suggested chronic pulmonary blood loss rather than isolated erythropoietin deficiency; retrospectively, this represented another manifestation of ongoing diffuse alveolar hemorrhage.

The diagnosis was established based on pulmonary–renal syndrome with strongly positive MPO-ANCA serology. Kidney biopsy remains the diagnostic gold standard, confirming pauci-immune necrotizing crescentic glomerulonephritis and providing prognostic information regarding chronicity and reversibility of renal injury, but is not mandatory in every patient [7]. Current guidance holds that treatment should not be delayed when clinical suspicion is high, particularly in life-threatening disease, and that biopsy may reasonably be deferred when it carries unacceptable risk or is unlikely to alter management [2] [8]. In our patient, renal atrophy, advanced CKD, anticoagulation, and bleeding risk made biopsy impractical, so pulmonary hemorrhage, active urinary sediment, and high-titer MPO-ANCA were considered sufficient for a diagnosis.

The presentation also fulfills the 2022 ACR/EULAR classification criteria for microscopic polyangiitis, which assign weighted scores to clinical and laboratory features after exclusion of alternative diagnoses. Positive MPO-ANCA serology contributes the highest positive score, while the absence of sinonasal disease and eosinophilia further supports classification as MPA; although such criteria should not replace clinical judgment, they provide additional support for the diagnosis here [10].

Several differential diagnoses were considered and excluded. Infective endocar-

ditis was repeatedly investigated given persistent fever and pacemaker, but blood cultures remained sterile, serial echocardiograms showed no vegetation, and FDG PET-CT showed no cardiac uptake. Sarcoidosis, tuberculosis, pulmonary embolism, and occult malignancy were also excluded through appropriate investigations.

Treatment was initiated with glucocorticoids, rituximab, and avacopan. Rituximab has become an accepted first-line remission-induction therapy for severe AAV following the RAVE and RITUXVAS trials, which demonstrated efficacy comparable to cyclophosphamide with a more favorable toxicity profile in selected populations, particularly elderly patients [11] [12]. Avacopan is an oral C5a receptor antagonist targeting the alternative complement pathway [13]. Current recommendations therefore support combining avacopan with rituximab or cyclophosphamide in patients with severe disease at substantial risk of glucocorticoid-related adverse effects [8] [9].

Despite immunosuppressive therapy, our patient developed rapidly progressive DAH requiring mechanical ventilation, vasopressor support, and renal replacement therapy. DAH remains one of the strongest predictors of mortality in MPA: several observational studies report mortality rates exceeding 40% among patients requiring intensive care, particularly elderly individuals with advanced renal impairment [14] [15]. An early landmark cohort study found that pulmonary hemorrhage at presentation was independently associated with a more than eightfold higher risk of death, a prognostic weight that has persisted even in the era of modern therapy [16].

The decision not to perform plasma exchange deserves comment. Historically favored for severe pulmonary hemorrhage or rapidly progressive glomerulonephritis, plasma exchange was shown in the PEXIVAS trial not to significantly reduce death or end-stage kidney disease in severe AAV [17]. Current guidance reserves plasma exchange for selected patients, particularly those with concomitant anti-GBM disease or refractory life-threatening DAH. As our patient had negative anti-GBM antibodies and deteriorated despite appropriate induction therapy, plasma exchange was not considered likely to alter the outcome.

An unusual feature was acute mesenteric infarction. Gastrointestinal involvement is less common than renal or pulmonary disease in MPA. Involvement of mesenteric vessels may cause intestinal ischemia, perforation, or hemorrhage. It carries an extremely poor prognosis, usually reflecting widespread systemic vascular injury [18]. The bowel infarction may have reflected progression of systemic necrotizing vasculitis or thromboembolic disease, as anticoagulation had been withheld; however, neither mechanism can be definitively established.

4. Conclusion

This case illustrates the diagnostic challenge of microscopic polyangiitis in an elderly patient with advanced CKD and cardiovascular disease, in whom recurrent pulmonary edema and transient diuretic response masked an underlying

ANCA-associated vasculitis. Recurrent hemoptysis, transfusion-dependent anemia, inflammatory syndrome, de novo proteinuria, and hematuria revealed a pulmonary–renal syndrome secondary to MPO-ANCA-positive MPA. Despite remission-induction therapy, the disease progressed to diffuse alveolar hemorrhage and fatal mesenteric infarction. Maintaining a high index of suspicion for AAV in recurrent or atypical heart failure, particularly when pulmonary and renal abnormalities coexist, together with early ANCA testing and multidisciplinary assessment, is essential to avoid diagnostic delay and irreversible multiorgan damage.

5. Learning Points

- MPA should be considered in elderly patients presenting with recurrent "heart failure" that responds only transiently to conventional therapy, and/or accompanied by unexplained pulmonary infiltrates or hemoptysis.
- New-onset proteinuria, microscopic hematuria, or active urinary sediment in patients with pre-existing CKD should prompt evaluation for superimposed glomerulonephritis rather than being attributed solely to CKD progression.
- Diffuse alveolar hemorrhage may initially mimic pulmonary edema or pneumonia, especially in patients on anticoagulants, and should be suspected with recurrent hemoptysis and unexplained anemia.
- Early ANCA testing in an evolving pulmonary-renal syndrome may shorten diagnostic delay and allow earlier immunosuppression before irreversible multiorgan damage develops.
- A clinicoserological diagnosis of MPO-ANCA-associated vasculitis is acceptable when renal biopsy is contraindicated and the clinical presentation strongly suggests pulmonary-renal syndrome.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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