Open Access Library Journal
2026, Volume 13, e16181
ISSN Online: 2333-9721
ISSN Print: 2333-9705

Dialysis-Requiring Pulmonary-Renal Syndrome with Renal Recovery in Granulomatosis with Polyangiitis: A Case Report

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How to cite this paper: Maaroufi, M., Harraz, H., Laklaai, Z., Jalloul, W., Chaanoun, K., Benjelloun, H. and Zaghba, N. (2026) Dialysis-Requiring Pulmonary-Renal Syndrome with Renal Recovery in Granulomatosis with Polyangiitis: A Case Report. *Open Access Library Journal*, **13**: e16181.
https://doi.org/10.4236/oalib.1116181

Received: October 20, 2026
Accepted: November 22, 2026
Published: November 25, 2026

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Abstract

Granulomatosis with polyangiitis (GPA) is an antineutrophil cytoplasmic antibody-associated vasculitis that can cause life-threatening pulmonary-renal syndrome. We report a 57-year-old man who presented with hemoptysis after six weeks of otalgia, purulent otorrhea, tympanic membrane perforation, and recurrent epistaxis. Chest computed tomography revealed bilateral pulmonary consolidations and multiple cavitary lesions. Bronchoscopy showed diffuse mucosal inflammation and hemorrhagic bronchoalveolar lavage fluid, supporting suspected diffuse alveolar hemorrhage in the clinical context. Urinalysis demonstrated marked hematuria and proteinuria, while serum creatinine rose rapidly from 3.21 to 10.62 mg/dL. Kidney biopsy revealed necrotizing crescentic glomerulonephritis, anti-proteinase 3 antineutrophil cytoplasmic antibodies were strongly positive, and sinus imaging demonstrated destructive sinonasal changes. The findings supported a diagnosis of GPA with severe pulmonary and kidney involvement. The patient received intravenous methylprednisolone followed by oral glucocorticoids and intravenous cyclophosphamide. Hemodialysis was temporarily required and was subsequently discontinued. At one year, the patient remained dialysis-independent, with a creatinine level of 1.10 mg/dL and no hematuria or proteinuria. This case emphasizes the value of early urinalysis in suspected pulmonary-renal syndrome and illustrates that dialysis-requiring kidney failure in GPA may be reversible with timely treatment.

Subject Areas

Respiratory Medicine

Keywords

Granulomatosis with Polyangiitis, Pulmonary-Renal Syndrome, Diffuse Alveolar Hemorrhage, Rapidly Progressive Glomerulonephritis, Cyclophosphamide

1. Introduction

Granulomatosis with polyangiitis (GPA) is a necrotizing vasculitis of small and medium vessels, frequently associated with proteinase 3-antineutrophil cytoplasmic antibodies (PR3-ANCA). Ear, nose, and throat (ENT) manifestations may precede pulmonary and renal involvement. Pulmonary nodules, cavitation, and alveolar hemorrhage may resemble infection or malignancy [1] [2]. The coexistence of suspected diffuse alveolar hemorrhage and rapidly progressive glomerulonephritis represents a potentially life-threatening pulmonary-renal syndrome. We report severe PR3-ANCA-positive GPA with destructive ENT disease and dialysis-requiring renal failure followed by marked recovery after induction therapy.

2. Case Presentation

A 57-year-old man without known chronic kidney disease was admitted with low-volume hemoptysis. Six weeks earlier, he had developed left-sided otalgia, purulent otorrhea with tympanic membrane perforation, and subsequently two episodes of epistaxis. The initial symptoms had been treated as an ear infection. He reported progressive deterioration in general condition without fever. He had never smoked and reported neither previous tuberculosis treatment nor known recent exposure. On admission, temperature was 36.5˚C, respiratory rate was 16 breaths/min, and oxygen saturation was 98% on room air. He was hemodynamically stable but pale, with bilateral painless pitting edema, ulcerations of the lingual frenulum and tongue, and a lower-lip ecchymosis. Urine dipstick testing revealed 4+ blood and 2+ protein.

Chest radiography showed bilateral perihilar and basal alveolar opacities. Thoracic computed tomography (CT) demonstrated bilateral consolidations and multiple nodular lesions, several with cavitation (**Figures 1(A)-(C)**). Hemoglobin was 10.6 g/dL, leukocytes 19.12 × 10^9/L, platelets 596 × 10^9/L, C-reactive protein 234.5 mg/L, and serum albumin 25 g/L. Initial serum creatinine was 3.21 mg/dL (estimated glomerular filtration rate [eGFR], 20 mL/min/1.73m^2). Microscopic hematuria was marked, 24-hour proteinuria was 1.2 g, and urine culture was negative. Kidney ultrasonography showed kidneys of normal size with preserved corticomedullary differentiation.

Flexible bronchoscopy showed severe diffuse bronchial mucosal inflammation without a visible endobronchial tumor (**Figure 1(D)**). Bronchoalveolar lavage (BAL) fluid was grossly hemorrhagic (**Figure 1(E)**), and BAL cytology identified no malignant cells. Microbiological testing of bronchial aspirate included routine bacteriological investigations, direct acid-fast bacilli (AFB) smear, Xpert MTB/RIF,

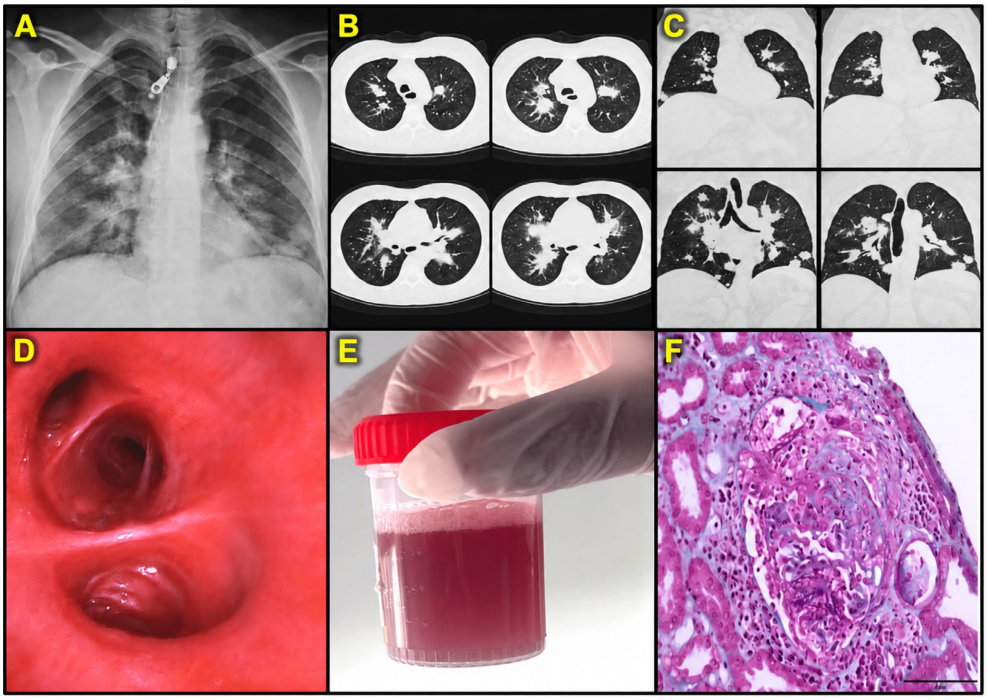


Figure 1. Radiologic, bronchoscopic, and histopathologic findings at presentation. (A) Chest radiograph with bilateral alveolar opacities. (B) Axial chest CT showing bilateral pulmonary consolidations, some cavitary. (C) Coronal chest CT reconstructions confirming multifocal bilateral lesions. (D) Bronchoscopic view showing diffuse bronchial inflammation. (E) Grossly hemorrhagic bronchoalveolar lavage fluid. (F) Renal biopsy showing necrotizing crescentic glomerulonephritis.

mycobacterial culture, and fungal culture on Sabouraud agar. Routine bacteriological investigations, AFB smear, and Xpert MTB/RIF were negative during the initial work-up. Mycobacterial culture and fungal culture were reported negative approximately one month after sampling, thus after the initial decision to initiate immunosuppression. Hemorrhagic BAL, associated with hemoptysis, anemia, and bilateral opacities, supported clinically suspected diffuse alveolar hemorrhage. Bronchial biopsies showed nonspecific fibro-inflammatory changes.

Creatinine increased to 4.12, 4.96, and 6.20 mg/dL over the first four hospital days. Kidney biopsy revealed necrotizing crescentic glomerulonephritis. Of seven glomeruli examined, three showed cellular extracapillary proliferation (cellular crescents) with fibrinoid necrosis. Glomerular capillary basement membrane rupture with fibrinoid necrosis was also documented, along with one globally sclerosed (obsolescent) glomerulus. The interstitium exhibited edema and a moderate inflammatory infiltrate composed of lymphocytes, plasma cells, neutrophils, and rare eosinophils, with small interstitial granulomas and multinucleated giant cells. Tubular epithelial regeneration and intratubular red blood cell casts were observed. Arteriolar vasculitis with fibrinoid wall necrosis and inflammatory infiltration was present, accompanied by minimal arteriolar hyalinosis (**Figure 1(F)**). These findings were consistent with renal involvement in granulomatosis with polyangiitis.

Cytoplasmic ANCA with anti-PR3 specificity was strongly positive; anti-myeloperoxidase ANCA, anti-glomerular basement membrane antibodies, and antinuclear antibodies were negative. Sinus CT showed erosion and demineralization involving the nasal septum and turbinates, with nodular mucosal thickening. Nasofibroscopy found severe crusting rhinitis, but inferior turbinate biopsy was nonspecific. These findings established GPA after consideration of infectious and other immune-mediated mimics. The 2022 ACR/EULAR GPA classification threshold was also fulfilled [3].

Given organ-threatening disease, intravenous methylprednisolone was initiated on hospital day five at 15 mg/kg/day for three days, followed by oral prednisone 1 mg/kg/day for six weeks. Prednisone was subsequently reduced by 5 mg every 15 days; calcium and vitamin D supplementation was provided. Intravenous cyclophosphamide was administered at a dose of 0.6 g/m^2 per pulse, with mesna and hydration, for a total of six pulses. The first three pulses were administered at two-week intervals, followed by three additional pulses at three-week intervals.

Serum creatinine rose to a peak of 10.62 mg/dL in the setting of severe, rapidly progressive acute kidney injury, for which intermittent hemodialysis was initiated. Sessions were initially performed three times weekly and were progressively spaced as renal function improved. During the dialysis-dependent phase, follow-up serum creatinine measurements were generally taken on the day after dialysis sessions, and therefore reflected the effects of renal replacement therapy as well as any endogenous renal recovery. Serum creatinine values of 1.96 mg/dL after the second cyclophosphamide pulse and 1.38 mg/dL after the third were reported during the recovery phase but cannot by themselves establish dialysis-independent renal function. Serial clinical assessment and improving renal biochemistry guided the reduction and eventual cessation of dialysis; the internal jugular dialysis catheter was subsequently removed (**Table 1**).

Hemoptysis, oral lesions, and ENT manifestations resolved during induction. Serial chest radiographs documented progressive clearing of pulmonary infiltrates (**Figure 2**). Pneumocystis jirovecii prophylaxis was given with trimethoprim-sulfamethoxazole (TMP-SMX, double-strength 160/800 mg tablet); the reported daily regimen was one tablet, with renal-function-based adaptation during severe kidney impairment. Treatment was well tolerated, with no reported infectious or hematologic complications during induction or maintenance. Following six cyclophosphamide pulses, maintenance azathioprine was initiated at 150 mg/day. At one-year follow-up, the patient remained dialysis-independent and free of recurrent hemoptysis, with creatinine 1.10 mg/dL (eGFR, 69 mL/min/1.73 m^2) and no hematuria or proteinuria. Ongoing maintenance therapy comprised azathioprine 150 mg/day, prednisone 5 mg/day, and continued TMP-SMX prophylaxis, with parallel follow-up by the internal medicine team.

3. Discussion

This case demonstrates how ENT manifestations may obscure evolving systemic

Table 1. Evolution of renal biochemistry and hemodialysis during induction and follow-up.

Time point	Creatinine (mg/dL)	eGFR (mL/min/1.73m²)	Clinical course/dialysis context
Admission	3.21	20	Hematuria 4+; proteinuria 2+
Hospital day 2	4.12	15	Rapidly changing renal function
Hospital day 3	4.96	12	Rapidly changing renal function
Hospital day 4	6.20	9	Rapidly changing renal function
Peak	10.62	5	Severe progressive AKI; HD initiated (3 sessions/week initially)
After second CYC pulse	1.96	35	Recovery phase; creatinine usually checked the day after HD
After third CYC pulse	1.38	53	Recovery phase; HD progressively spaced then discontinued
One-year follow-up	1.10	69	No HD; normal urinalysis; on maintenance treatment

eGFR, estimated glomerular filtration rate; CYC, cyclophosphamide; HD, hemodialysis. eGFR estimates in acute kidney injury or during dialysis are not reliable measures of intrinsic kidney filtration. During HD, surveillance creatinine was usually obtained on the day after a session. Exact dialysis start/end dates, total session count, and pulse-related sampling dates were not available for this report.

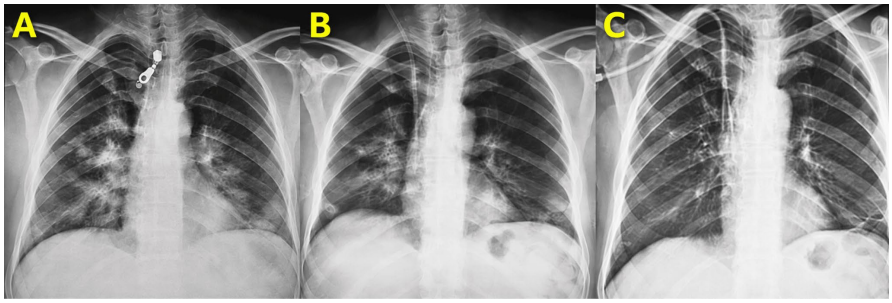


Figure 2. Serial chest radiographs following the first (A), second (B), and third (C) cyclophosphamide pulses, showing progressive resolution of bilateral pulmonary infiltrates.

vasculitis. Otorrhea, tympanic perforation, and epistaxis preceded the pulmonary-renal syndrome by several weeks. The immediately available urine dipstick provided a crucial clue to glomerular injury and prompted urgent kidney assessment. Neither bronchial nor nasal biopsies were diagnostically specific, whereas necrotizing crescentic kidney lesions, PR3-ANCA positivity, cavitary pulmonary lesions, and destructive sinonasal changes strongly supported GPA. The 2022 ACR/EULAR criteria facilitate classification once vasculitis is established and mimics have been excluded; they should not be used as standalone diagnostic criteria [3].

Diffuse alveolar hemorrhage should be considered in patients with hemoptysis, anemia, and bilateral infiltrates even without hypoxemia. Hemorrhagic BAL sup-

ported this diagnosis in the present patient. In view of the pulmonary cavities, infection was specifically considered: initial routine bacteriological assessment, AFB smear, and Xpert MTB/RIF on bronchial aspirate were negative. Negative anti-glomerular basement membrane antibodies helped evaluate a further important pulmonary-renal mimic. The combined assessment and kidney biopsy supported prompt treatment.

For severe ANCA-associated vasculitis with organ-threatening manifestations, contemporary EULAR and KDIGO recommendations support induction with glucocorticoids plus either rituximab or cyclophosphamide [4] [5]. Intravenous pulse cyclophosphamide is an established option: the original CYCLOPS trial reported comparable induction efficacy between pulse and daily oral cyclophosphamide [6], whereas its long-term follow-up demonstrated a higher relapse rate after pulse therapy [7]. Rituximab is another guideline-supported option, although trial results should not be overgeneralized to rituximab alone in dialysis-dependent disease: RAVE excluded patients with serum creatinine > 4.0 mg/dL [8], and the RITUXVAS rituximab arm included two intravenous cyclophosphamide pulses [9]. PEXIVAS supported reduced-dose glucocorticoid regimens with fewer serious infections and no worse primary composite outcome of death or end-stage kidney disease [10]. KDIGO advises consideration of plasma exchange in patients with creatinine > 3.4 mg/dL, dialysis requirement or rapidly increasing creatinine, or hypoxemic alveolar hemorrhage [5]. In the present case, plasma exchange was discussed owing to severe renal involvement and dialysis requirement, despite preserved initial oxygenation, but was not performed because of practical and resource-related constraints. Glucocorticoid-cyclophosphamide induction, which was locally available and feasible, was initiated, with subsequent marked clinical and renal improvement. The favorable outcome cannot establish that adjunctive plasma exchange would have been unnecessary or ineffective.

The most important outcome was sustained renal recovery following initial dialysis dependence. Severe acute glomerulonephritis does not invariably result in irreversible loss of renal function; recovery following treatment of active vasculitis has been documented [11] [12]. In our patient, renal histopathology demonstrated prominent active lesions, including cellular crescents, fibrinoid necrosis, arteriolar vasculitis, and tubular epithelial regeneration, suggesting a potentially reversible component of kidney injury. At one year, a serum creatinine level of 1.10 mg/dL, sustained dialysis independence, and normal urinalysis provided stronger evidence of renal recovery than early measurements obtained following hemodialysis sessions. The combined clinical, serological, radiological, and histopathological findings strongly supported the diagnosis of GPA. Continued specialist follow-up is essential in PR3-ANCA-positive disease because of the risk of relapse and the need to monitor immunosuppressive treatment toxicity [4] [5].

4. Conclusion

GPA should be considered urgently when persistent ENT symptoms, hemoptysis,

pulmonary infiltrates, and urinary abnormalities coexist. Bedside urinalysis may reveal otherwise occult glomerulonephritis. Dialysis-requiring renal failure can still be reversible when organ-threatening vasculitis is recognized and treated promptly.

Ethical Approval and Informed Consent

Ethical approval was not required for this single-patient case report according to institutional policy. Written informed consent was obtained from the patient for publication of the case report and accompanying clinical images.

Funding

No external funding was received for this work.

Data Availability

The data supporting this case report are derived from a confidential patient medical record and cannot be made publicly available due to patient privacy and confidentiality considerations. Relevant de-identified clinical information is included in the manuscript.

Author Contributions

Conceptualization, methodology, investigation, data curation, formal analysis, visualization, and writing—original draft preparation: Mehdi Maaroufi. Writing—review and editing, supervision, and validation: Hanaa Harraz, Zakaria Laklaai, Wafaa Jalloul, Khadija Chaanoun, Hanane Benjelloun, and Nahid Zaghba. All authors have read and approved the final version of the manuscript.

Conflicts of Interest

The authors declare no conflicts of interest.

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