acta medica

CASE REPORT

Pseudomonas-driven amyloid storm: a tale of two cases with rapidly progressive kidney injury

Saliha Yıldırım[1]
ORCID: 0000-0002-2693-1167

Betül Öğüt[2]
ORCID: 0000-0002-1385-7324

İpek Işık Gönül[2]
ORCID: 0000-0003-0058-5136

Özant Helvacı[1]
ORCID: 0000-0002-1382-2439

Yasemin Erten[1]
ORCID: 0000-0002-9741-6779

[1]Gazi University Faculty of Medicine, Department of Nephrology, Ankara, Türkiye

[2]Gazi University Faculty of Medicine, Department of Pathology, Ankara, Türkiye

Corresponding Author: Saliha Yıldırım
E-mail: dr.saliha@gmail.com

Received: 2 April 2025, Accepted: 20 June 2025, Published online: 30 September 2025

ABSTRACT

AA amyloidosis is a significant cause of chronic kidney disease and, if untreated, often leads to progressive kidney damage. This study aims to illustrate how AA amyloidosis can manifest as both slowly progressing chronic kidney injury and rapidly progressive acute kidney injury triggered by secondary conditions.

We describe two patients experiencing an amyloid storm related to Pseudomonas aeruginosa infection: one with bronchiectasis-related amyloidosis and another with paraplegia and decubitus ulcer-related amyloidosis. Clinical, laboratory, and kidney biopsy findings were analyzed.

Both cases demonstrated accelerated kidney failure within two weeks. The first case involved chronic decubitus ulcers and osteomyelitis, while the second had bronchiectasis with pneumonia. Despite infection control and colchicine therapy, both patients experienced irreversible renal damage.

These cases underscore the importance of aggressive infection control in preventing amyloid storm and highlight the interplay between chronic inflammation and acute infections in AA amyloidosis.

Keywords: amyloidosis, amyloid storm, bronchiectasis, decubitus ulcer, pseudomonas aeroginosa

INTRODUCTION

AA amyloidosis, also known as reactive or secondary amyloidosis, is a systemic disease resulting from elevated hepatic production of serum amyloid A (SAA), triggered by cytokine release—especially interleukin-1 (IL-1)—from activated macrophages [1,2]. It often arises secondary to genetic disorders (e.g., familial Mediterranean fever), rheumatoid arthritis, inflammatory bowel disease, connective tissue disorders, or chronic infections (e.g., osteomyelitis, decubitus ulcers, bronchiectasis) [3]. The kidney is a common target organ, with clinical presentations often including nephrotic syndrome [4]. While AA amyloidosis typically progresses to chronic kidney disease unless the underlying cause is adequately addressed, certain conditions can precipitate rapid renal failure [4]. An amyloid storm is a rare complication seen in patients with amyloidosis. It describes a sudden and overwhelming organ involvement by AA amyloid, presumably driven by an acute inflammatory trigger such as infection [5].

This report highlights two cases of AA amyloidosis complicated by accelerated kidney failure within two weeks. Both cases showed hallmark findings of AA amyloidosis on kidney biopsy and Pseudomonas aeruginosa infection. This study was conducted by the Declaration of Helsinki and approved by the Ethical Committee of Gazi University Faculty of Medicine (approval number 2024-1115, 12 July 2024). Written informed consent was obtained from all participants.

CASE PRESENTATION

Case 1: Paraplegic male with decubitis ulcer

A 45-year-old male was admitted to the emergency department with a one-month history of progressively worsening pretibial edema and a recurrent pressure ulcer over the right gluteal region. His medical history included paraplegia following a childhood traffic accident and surgical intervention for the same decubitus ulcer ten years ago. The ulcer was grade four with dark yellow discharge and a seven-centimeter diameter wound. The patient appeared pallor and cachectic and had grade three pretibial edema (BP: 110/60 mmHg).

Laboratory tests revealed severe anemia (hemoglobin: 7.8 g/dL), leukocytosis (13,700/µL with 88.6% neutrophils), and markedly elevated inflammatory markers (CRP: 242 mg/L, ESR: 130 mm/h). Renal function tests indicated acute kidney injury with a serum creatinine of 5.2 mg/dL and BUN of 38.9 mg/dL (Serum creatinine level three months before admission was 1.0 mg/dL). Hypokalemia (serum potassium: 2.7 mmol/L) and hypoalbuminemia (serum albumin: 1.1 g/dL) were also noted. Urinalysis showed nephrotic-range proteinuria (+3 protein dipstick, urine protein-to-creatinine ratio of 76,121 mg/g).

Leukocyte scintigraphy indicated osteomyelitis near the pressure wound, and wound cultures grew

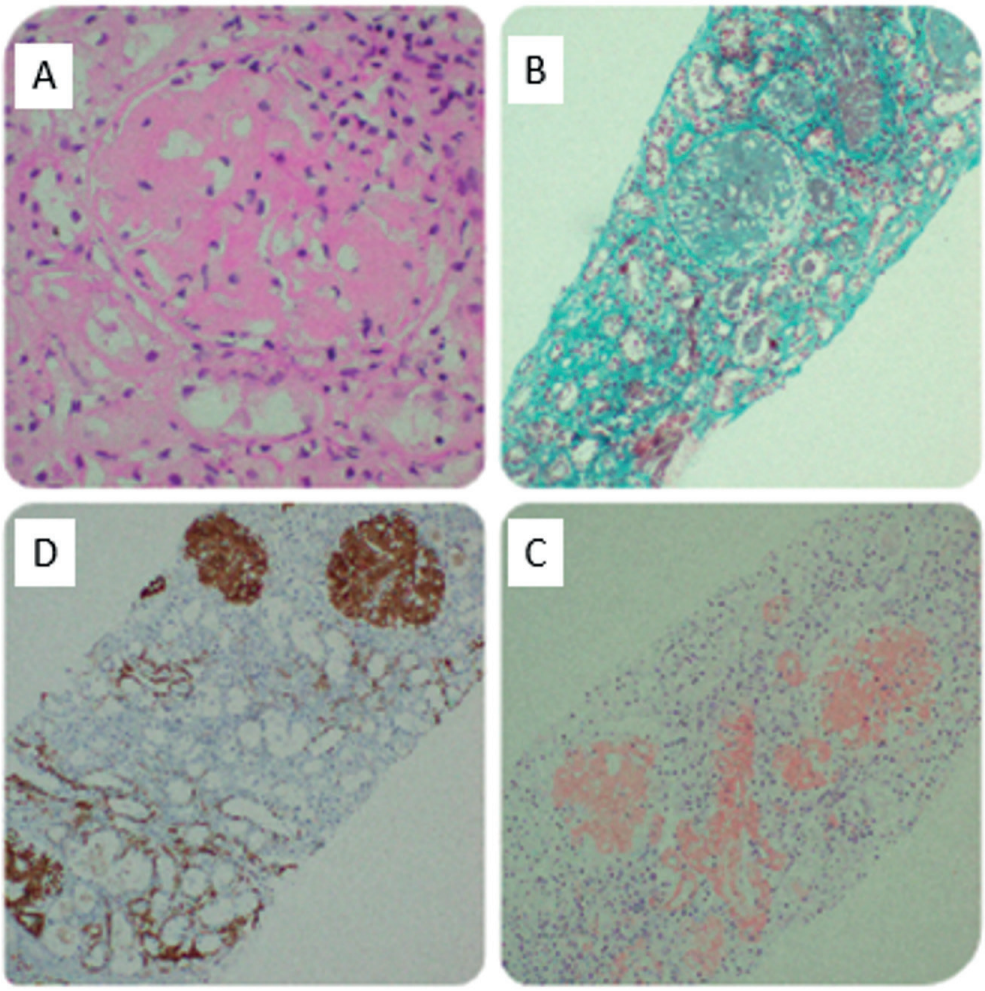

Figure 1. Kidney biopsy findings of case 1. A) Hematoxylin and Eosin (H&E) staining demonstrating amorphous eosinophilic material in the glomerulus. Glomerular tufts are expanded by acellular, eosinophilic, amorphous material suggestive of amyloid. There is associated interstitial inflammation and tubular atrophy. B) Masson's Trichrome staining of renal cortex reveals prominent mesangial expansion and sclerosis. There is moderate interstitial fibrosis and tubular atrophy (IFTA) with collagen deposition (green staining). Glomeruli demonstrate mesangial matrix expansion. C) Congo Red stain under bright-field microscopy showing amyloid deposits. Homogeneous, amorphous deposits are seen in glomerular mesangium and vessel walls, consistent with amyloid deposition. D) Immunohistochemistry for Serum Amyloid A (SAA) showing strong positive staining in glomerular and vascular deposits. There is diffuse, intense SAA positivity in glomerular deposits, confirming the diagnosis of AA-type amyloidosis.

Morganella morganii, Pseudomonas aeruginosa, and Escherichia coli. Intravenous meropenem was initiated.

The biopsy revealed positive staining for AA amyloid, with diffuse polymorphonuclear leukocyte-predominant tubulointerstitial inflammation. Moderate tubular atrophy and interstitial fibrosis were observed, along with 2/30 global glomerulosclerosis. These findings were consistent with AA amyloidosis and ongoing acute kidney injury (Figure 1).

Despite broad-spectrum antibiotics and colchicine therapy, serum creatinine rose from 5.2 mg/dL to 7.8 mg/dL over five days, accompanied by worsening edema and refractory hyperkalemia. Hemodialysis was initiated on day six. The patient remains on thrice-weekly hemodialysis six months post-discharge.

Case 2: Female with bronchiectasis

A 54-year-old female with bronchiectasis and immotile cilia syndrome presented to the emergency department with altered mental status and worsening respiratory symptoms. Her medical history included chronic bronchiectasis, occasional NSAID use, and recurrent lower respiratory tract infections.

Initial tests revealed mild renal impairment (serum creatinine: 0.85 mg/dL, BUN: 22 mg/dL) and elevated inflammatory markers (CRP: 197.2

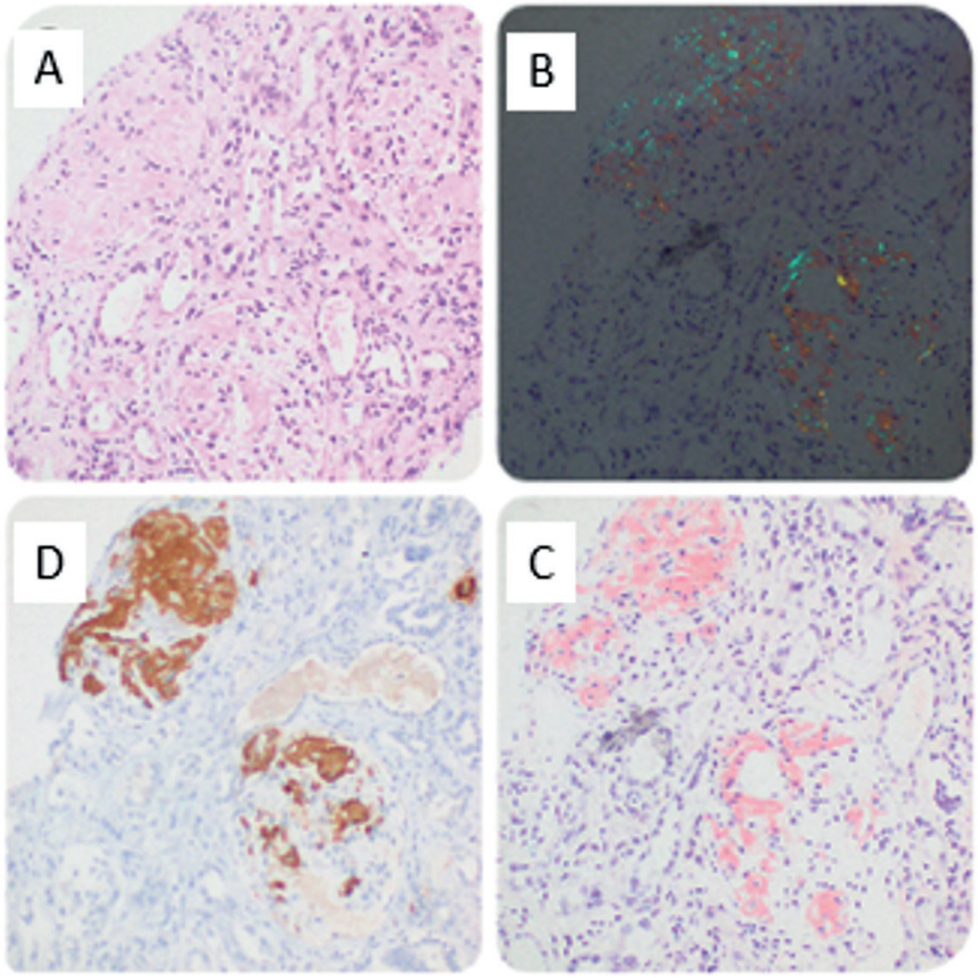

Figure 2. Kidney biopsy findings of case 2. A) Hematoxylin and Eosin (H&E) staining showing glomerular involvement by amyloid. Glomeruli appear expanded by acellular, pale eosinophilic material, with associated interstitial inflammation and tubular atrophy. B) Congo Red staining under polarized light showing apple-green birefringence of amyloid deposits. Glomerular and vascular deposits demonstrate characteristic apple-green birefringence under polarized light, confirming the presence of amyloid. C) Congo Red staining under bright-field microscopy highlights amorphous amyloid deposit in the glomeruli and vessel walls. D) Immunohistochemical staining for Serum Amyloid A (SAA) protein demonstrates positive staining in glomerular amyloid deposits. Strong SAA immunoreactivity in glomerular areas confirms the diagnosis of AA amyloidosis.

mg/L, procalcitonin: 4.6 ng/mL). Hemoglobin was 13.9 g/dL, and leukocytosis was noted (15,590/μL). Urinalysis demonstrated subnephrotic-range proteinuria (2358 mg/day) and microscopic hematuria (8 erythrocytes per high-power field). Sputum cultures isolated Pseudomonas aeruginosa. Chest CT revealed diffuse bronchiectasis with pneumonic infiltrations in the right lower lobe. Intravenous meropenem was initiated.

The biopsy showed positive staining for AA amyloid, tubulointerstitial mononuclear inflammation, moderate tubular atrophy, and interstitial fibrosis supporting the diagnosis of AA amyloidosis with acute kidney injury. Additionally, 2/29 global glomerulosclerosis was observed (Figure 2).

During hospitalization, serum creatinine rose to 2.17 mg/dL. Proteinuria at presentation was subnephrotic (2.4 g/day), which later progressed into the nephrotic range during hospitalization. Two months post-discharge, renal function declined (serum creatinine: 3.14 mg/dL) despite infection control and colchicine therapy.

DISCUSSION

AA amyloidosis arises from systemic deposition of SAA protein, which increases dramatically during acute inflammatory episodes [2]. Bronchiectasis, hemiplegia, and decubitus ulcers may be associated with amyloidosis through different mechanisms [6,7]. Infectious triggers, particularly Pseudomonas aeruginosa, are crucial in exacerbating this process. Pseudomonas aeruginosa infection is a potent inflammatory stimulus associated with functional amyloid biogenesis, a process by which bacterial amyloids can accelerate systemic amyloid deposition [8]. This dual mechanism likely amplifies the risk of amyloid storm, characterized by rapid and severe kidney injury.

In these cases, the presence of Pseudomonas aeruginosa coincided with significant elevations in inflammatory markers such as CRP and procalcitonin, driving an environment conducive to accelerated SAA production and tissue deposition. Furthermore, its involvement in chronic infections such as bronchiectasis and osteomyelitis provides a persistent source of inflammatory signaling, increasing susceptibility to amyloidogenic triggers [9,10]. Experimental studies have highlighted the role of Pseudomonas aeruginosa in secreting curli-like proteins that may directly interact with human amyloid precursors, potentially accelerating the aggregation process [8,10].

Clinically, the rapid renal decline observed in both cases underscores the importance of early identification and aggressive management of Pseudomonas aeruginosa infections. Standard antimicrobial treatments, while crucial, may not fully address the downstream amyloidogenic effects, suggesting a need for adjunct therapies targeting SAA production or amyloid deposition. Emerging research into anti-inflammatory agents, SAA-targeted therapies, and interventions aimed at bacterial amyloid pathways could offer new avenues for mitigating amyloid storm in similar clinical scenarios. These findings suggest that Pseudomonas aeruginosa is more than an inflammatory trigger; it may play a direct pathogenic role in the amyloidogenic cascade. As a result, infection prevention strategies and the need for multidisciplinary approaches to managing high-risk patients with AA amyloidosis are important.

In conclsion these cases highlight the critical role of Pseudomonas aeruginosa in precipitating amyloid storm in patients with AA amyloidosis. Its association with functional amyloid biogenesis raises the possibility of directly contributing to amyloidogenic processes beyond serving as an inflammatory trigger. Preventing and promptly treating Pseudomonas aeruginosa infections in high-risk patients is essential to mitigate the risk of catastrophic renal outcomes. Future research should explore targeted therapies addressing the infection and the amyloidogenic cascade.

Author contribution

Study conception and design: SY, BÖ; data collection: SY, BÖ, and ÖH; analysis and interpretation of results: SY, BÖ; draft manuscript preparation: SY, BÖ, YE. All authors reviewed the results and approved the final version of the manuscript.

Ethical approval

The study was approved by the Ethical Committee of Gazi University Faculty of Medicine (Protocol no. 2024-1115/12.07.2024).

Funding

The authors declare that the study received no funding.

Conflict of interest

The authors declare that there is no conflict of interest.

REFERENCES

[1] Gillmore JD, Lovat LB, Persey MR, Pepys MB, Hawkins PN. Amyloid load and clinical outcome in AA amyloidosis in relation to circulating concentration of serum amyloid A protein. Lancet 2001;358(9275):24-9. https://doi.org/10.1016/S0140-6736(00)05252-1

[2] Lachmann HJ, Goodman HJB, Gilbertson JA, et al. Natural history and outcome in systemic AA amyloidosis. N Engl J Med 2007;356(23):2361-71. https://doi.org/10.1056/NEJMoa070265

[3] Mirioglu S, Uludag O, Hurdogan O, et al. AA amyloidosis: A contemporary view. Curr Rheumatol Rep 2024;26(7):248-59. https://doi.org/10.1007/s11926-024-01147-8

[4] Feitosa VA, Neves PDMM, Jorge LB, Noronha IL, Onuchic LF. Renal amyloidosis: A new time for a complete diagnosis. Braz J Med Biol Res 2022;55:e12284. https://doi.org/10.1590/1414-431X2022e12284

[5] Kukuy OL, Beckerman P, Dinour D, Ben-Zvi I, Livneh A. Amyloid storm: Acute kidney injury and massive proteinuria, rapidly progressing to end-stage kidney disease in AA amyloidosis of familial Mediterranean fever. Rheumatology (Oxford) 2021;60(7):3235-42. https://doi.org/10.1093/rheumatology/keaa772

[6] Akçay S, Akman B, Ozdemir H, Eyüboğlu FO, Karacan O, Ozdemir N. Bronchiectasis-related amyloidosis as a cause of chronic renal failure. Ren Fail 2002;24(6):815-23. https://doi.org/10.1081/jdi-120015683

[7] Comarr AE. Amyloidosis in paraplegia correlation with decubitus ulcers. Calif Med 1955;82(4):332-4.

[8] Dueholm MS, Petersen SV, Sønderkær M, et al. Functional amyloid in Pseudomonas. Mol Microbiol 2010;77(4):1009-20. https://doi.org/10.1111/j.1365-2958.2010.07269.x

[9] Legger GE, Dermer CWE, Brunger AF, van Daele PLA, Nienhuis HLA. The relation between C-reactive protein and serum amyloid A in patients with autoinflammatory diseases. Pediatr Rheumatol Online J 2022;20(1):106. https://doi.org/10.1186/s12969-022-00757-9

[10] Rouse SL, Matthews SJ, Dueholm MS. Ecology and biogenesis of functional amyloids in pseudomonas. J Mol Biol 2018;430(20):3685-95. https://doi.org/10.1016/j.jmb.2018.05.004