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Goodpasture's Disease: A Case of Rapidly Progressive Kidney Injury in an Elderly Patient

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ABSTRACT

Goodpasture's disease, a rare and aggressive autoimmune pathology, manifests with renal and/or pulmonary involvement due to the production of anti-glomerular basement membrane autoantibodies. We report the case of an 88-year-old patient presenting with rapidly progressive acute kidney injury associated with a history of chronic rhinitis, bronchitis, and an episode of hemoptysis, in whom the diagnosis of Goodpasture's disease was confirmed. Therapeutic management was limited by the unavailability of plasmapheresis in our department, highlighting the diagnostic and therapeutic challenges of this pathology in elderly patients. The clinical course was marked by the need for chronic hemodialysis.

KEYWORDS

Goodpasture's Disease, Rapidly Progressive Glomerulonephritis (RPGN), Geriatric Nephrology, Plasmapheresis.

MAIN ARTICLE

Introduction

Anti-glomerular basement membrane (anti-GBM) antibody disease, formerly known as Goodpasture's disease, is a rare, typically monophasic, and severe autoimmune disease characterized by the production of autoantibodies directed against the non-collagenous domain (NC1) of the α3 chain of type IV collagen (α3(IV)NC1), an essential component of the glomerular and alveolar basement membranes [1]. It can manifest with renal involvement, pulmonary involvement, or a combination of both leading to a pulmonary-renal syndrome, with a high risk of rapid progression to end-stage kidney disease and acute respiratory failure. Although anti-GBM disease is more common in young adults, it can also occur in elderly patients, often with atypical clinical presentations and a more severe course [2].Pulmonary involvement, particularly alveolar hemorrhage, is a feared complication. The break in immune tolerance underlying this disease may involve a modification of the autoantigen's quaternary structure, promoting the exposure of hidden epitopes, especially following exposure to respiratory toxins such as tobacco smoke [1]. In this context, early diagnosis and treatment are crucial for improving prognosis. This article presents the case of an 88-year-old patient with rapidly progressive glomerulonephritis (RPGN) due to anti-GBM antibodies, highlighting the diagnostic and management challenges.

Case Report

An 88-year-old man, a former smoker with chronic bronchitis, was admitted to the emergency department for incidentally discovered acute kidney injury (AKI). He presented with anuria, epigastralgia, nausea, and vomiting that had been progressing for several days. He also reported an episode of hemoptysis approximately one month prior to admission, which had resolved spontaneously. Clinical examination revealed no pulmonary manifestations.

Laboratory investigations showed severe AKI with urea at 1.48 g/L (24.6 mmol/L) (normal range: 0.17-0.43 g/L), creatinine at 109 mg/L (963 µmol/L) (normal range: 6-12 mg/L), and a marked systemic inflammatory response with C-reactive protein (CRP) at 230 mg/L (normal: < 5 mg/L) and procalcitonin at 2.17 ng/mL (normal: < 0.1 ng/mL). Renal ultrasound was normal, showing no hydronephrosis.

Anti-GBM antibody testing in the patient's serum was positive at 675 IU/mL (normal: < 20 IU/mL), and anti-neutrophil cytoplasmic antibody (ANCA) testing was negative. Given the rapidly progressive nature of the kidney injury, a renal biopsy was performed. Histological analysis revealed diffuse and severe proliferative extracapillary glomerulonephritis, affecting 90% of glomeruli with crescent formation. Direct immunofluorescence study showed intense, diffuse linear deposition of IgG along the glomerular basement membrane, a pathognomonic finding for anti-GBM disease (Figure 1).

The diagnosis of anti-GBM disease with rapidly progressive glomerulonephritis was formally established based on clinical, serological, and histological data.

The patient commenced emergency hemodialysis due to uremic symptoms and anuria. Induction therapy was rapidly initiated, combining high-dose corticosteroids (3 pulses of Methylprednisolone followed by oral Prednisone) and Cyclophosphamide. However, plasma exchange (PE), considered an essential treatment for the rapid removal of circulating autoantibodies, could not be performed due to its unavailability at our center.

No recovery of renal function was observed. The patient remained dependent on chronic hemodialysis, which is ongoing. No overt alveolar hemorrhage occurred during treatment.

Discussion

This case report illustrates the diagnostic and therapeutic challenges posed by anti-GBM disease in elderly patients.

Firstly, the diagnosis requires a high index of clinical suspicion. Although rare, this disease should be considered in any patient presenting with RPGN, even in the elderly and in the absence of a complete pulmonary-renal syndrome [3].The clinical presentation can be atypical, with less specific symptoms and potentially longer diagnostic delays. In our patient, severe renal involvement was the predominant feature, while the reported hemoptysis was prior and resolved, with no active pulmonary signs at admission. Although hemoptysis can guide the diagnosis [4], its absence or non-concomitant nature should not rule out the disease.

Secondly, diagnostic confirmation relies on key investigations. Renal biopsy with immunofluorescence study is indispensable, revealing the pathognomonic linear IgG deposits (Figure 1) and assessing the severity of lesions (percentage of crescents) [5].Measurement of

circulating anti-GBM antibodies confirms the autoimmune nature [6].ANCA testing is also relevant, as co-positivity with anti-GBM antibodies is more frequent in older individuals [7], although it was negative in our case.

Thirdly, therapeutic management is limited by treatment availability. Standard therapy consists of a combination of immunosuppressants (corticosteroids and cyclophosphamide) and plasma exchange [8].PE plays a crucial role by rapidly removing pathogenic autoantibodies and significantly improving renal prognosis [9].The unavailability of PE at our center constituted a major therapeutic limitation. Although immunosuppressive therapy and dialysis prevented alveolar hemorrhage and ensured patient survival, the absence of PE likely contributed to the unfavorable progression to end-stage kidney disease. This prognosis was already burdened by the severity of the initial presentation (high creatinine, anuria, high percentage of crescents) and advanced age, which are recognized factors for poor renal and overall prognosis [10].

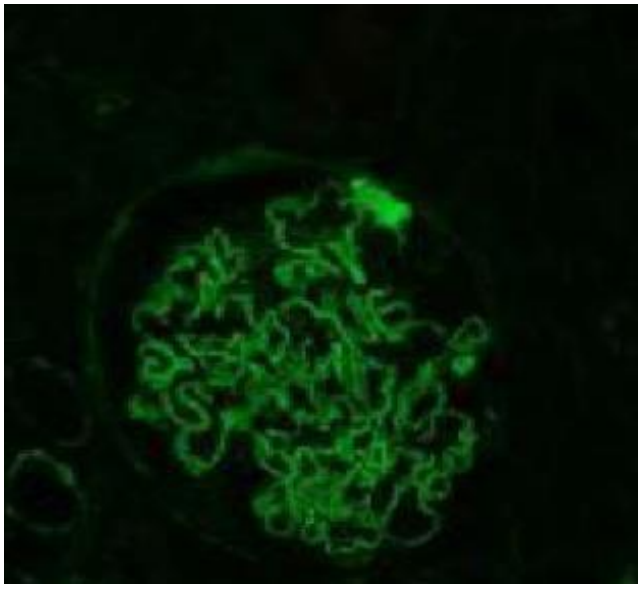
This case underscores the importance of rapid access to PE and the need to refer patients to specialized centers when possible. It also serves as a reminder that even without renal recovery, immunosuppressive therapy remains essential to control disease activity and prevent potentially fatal complications like alveolar hemorrhage. Management should be personalized, carefully weighing the benefit-risk ratio of immunosuppressants, particularly in elderly and frail patients.

Conclusion

Anti-GBM disease, although rare, should be systematically considered in the differential diagnosis of rapidly progressive kidney injury, even in elderly patients and in the absence of overt pulmonary involvement. This case report illustrates the importance of a rapid diagnostic approach, including renal biopsy and anti-GBM antibody testing. The unavailability of plasma exchange limited optimal management and potentially influenced the unfavorable renal outcome. Increased vigilance is essential when facing anti-GBM disease. Early diagnosis, confirmed by renal biopsy and anti-GBM antibody testing, allows for the prompt initiation of appropriate therapy and improves the prognosis of this severe condition.

FIGURES:

Figure 1: Renal biopsy, immunofluorescence. Linear IgG deposits along the glomerular basement membrane.



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The authors declare no conflict of interest.

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