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"Caught In the Crossfire -A Case of Anti GBM Disease with Rapid Renal Decline"

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ABSTRACT

Background: Anti-glomerular basement membrane (anti-GBM) disease is a rare autoimmune disorder that primarily affects the kidneys and, less commonly, the lungs. The rapid progression of glomerulonephritis is a hallmark of this disease, leading to significant renal damage if not treated promptly. Early diagnosis and treatment are critical to preventing irreversible kidney injury.

Case Description: We report a case of a 45-year-old male who presented with 15 days of hematuria and 2 days of breathlessness. The patient had no history of hypertension or diabetes. Clinical examination revealed pallor, high blood pressure, and fine crepitations in bilateral lower lung fields. Laboratory investigations showed elevated serum creatinine, reduced hemoglobin, and significant proteinuria. Anti-GBM antibodies were markedly elevated, and a kidney biopsy confirmed the diagnosis of crescentic glomerulonephritis with a necrotizing pattern. The patient was treated with plasmapheresis, steroids, and cyclophosphamide, along with maintenance hemodialysis. Despite the aggressive nature of the disease, early intervention helped stabilize his renal function.

Conclusion: This case highlights the importance of rapid diagnosis and prompt initiation of aggressive treatment in patients with anti-GBM disease presenting as rapidly progressive glomerulonephritis. Early intervention is key to preserving renal function and improving patient outcomes.

Key Words: Anti-GBM disease, rapidly progressive glomerulonephritis, crescentic glomerulonephritis, plasmapheresis, autoimmune nephritis

Case Report

A 45-year-old male presented to the emergency department with complaints of red urine for 15 days and breathlessness for 2 days. The patient reported no associated fever, pain, or dysuria with the hematuria. His breathlessness was not accompanied by orthopnea, cough, or edema. He also experienced vomiting for 2 days. The patient had no prior history of hypertension, diabetes, or similar illness in his family.

Upon physical examination, the patient was pale, with a pulse rate of 100/min and blood pressure of 170/98 mmHg. Fine crepitations were heard in the lower lung fields bilaterally. Heart sounds were normal, and the abdomen was soft and non-tender. Neurologically, the patient was alert and oriented. Review of other systems was unyielding.

Laboratory findings were significant for severe anemia (hemoglobin: 7 g/dl), elevated serum urea (274 mg/dl), serum creatinine (14.8 mg/dl), and hyperkalemia (serum potassium: 7 mEq/l). Urinalysis showed 500 RBCs/HPF and proteinuria of 300 mg/dl. The urine protein-creatinine ratio was 650 mg/g of creatinine.

Further investigations revealed elevated anti-GBM antibodies (55 U/ml, normal <20 U/ml), and a kidney biopsy confirmed crescentic glomerulonephritis with a necrotizing pattern. Immunofluorescence showed linear staining for IgG along the glomerular basement membrane. pANCA and cANCA were negative.

The patient was diagnosed with renal-limited anti-GBM disease and treated with plasmapheresis, intravenous corticosteroids, cyclophosphamide, and maintenance hemodialysis.

Discussion

Anti-glomerular basement membrane disease, or anti-GBM, Goodpasture syndrome when the lungs are involved is an uncommon autoimmune disease. It is caused by the generation of circulating auto-antibodies that attack the non-collagen part of the alpha 3 chain of the type IV collagen. These antibodies are targetted against the

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glomerular and the alveolar basement membranes leading to a rapidly progressive glomerulonephritis and at times pulmonary hemorrhage.

Pathogenetic factors are related to both genetics and environmental influences. It has high correlation with particular HLA class II molecules such as HLA-DR15 and HLA-DR4 that are presumed to present collagen derived peptide to T cells leading to production of pathogenic antibodies. Several environmental factors such as smoking, viral infections and hydrocarbon exposure can also act as disease promoters, aggravating the immune response. Although, There was no clear environmental eliciting factor in this case, smoking is often associated with higher risk of pulmonary hemorrhages in anti-GBM patients.

Clinical presentation is characterized by rapidly progressive glomerulonephritis. The renal changes are usually the most pronounced, frequently resulting in hematuria, proteinuria and in severe renal failure. Histopathological examination reveals presence of crescentric glomerulonephritis, a hallmark feature of severe glomerular injury. Immunoflorescence studies show a picture of linear deposition of Ig G class of antibodies along the glomerular basement membrane. This finding is highly suggestive of anti GBM disease. These crescents compress the glomerular capillary tuft, leading to glomerular necrosis and irreversible renal damage if left untreated.

In this case, the patient presented with a classic feature of anti-GBM disease -hematuria and rapidly progressive renal failure. His elevated serum creatinine, significant proteinuria, and positive anti-GBM antibodies confirmed the diagnosis. Crescentic glomerulonephritis was identified on kidney biopsy, further supporting the diagnosis.

Pulmonary involvement although absent in this case is an important aspect of this disease. Clinical presentation of hemoptysis, dyspnea or diffuse pulmonary infiltrates on chest imaging should prompt a suspicion of pulmonary hemorrhage. The disease has a strong predilection for smokers. Younger male smokers appear to be at higher risk. This kind of pulmonary involvement was not present in this case and this spared the patient from more respiratory symptoms although renal damage was massive.

In anti-GBM disease urgent management is crucial, intensive and early treatment is required to prevent organ losses that are beyond salvageable. plasmapheresis is A proven method, useful in circulation of anti-GBM antibodies removal from the bloodstream. High-dose corticosteroids are given to the patient concomitantly to control the inflammatory response while cytotoxics like cyclophosphamide are used to prevent new antibodies from being generated.

Despite these available therapies that are provided, the outcome is still dismal for patients who present with severe renal failure. In this particular case, the patient's creatinine levels at time of presentation were markedly elevated and the patient was started on dialysis. For these patients with such severe kidney injury, recovery of kidney function to normal is often a far fetched dream, dialysis is inevitable and/or renal transplant is required emergently or later on. The most important determinant for predicting outcomes is the promptness of diagnosis and initiation of treatment. Several studies show that patients who receive early treatment, preferably before the dependence of dialysis sets in, tend to have better outcomes than those who do not. Time is Kidney! Patients who present later in the course of disease tend to perform worse with regards to both short term and long term outcomes. they frequently become dialysis dependent even if aggressive immunosupression is started. The rapidity and severity of this disease is attributable to formation of large crescents in the glomeruli, which, at first are cellular, but later ,can become fibrous and lead to scarring in kidneys and the disease process becomes irreversible at this stage. Cellular crescents are made of proliferating epithelial cells and macrophages which lead to destructiom of glomerular basement membrane and the classical clinical features.

Strangely, the existence of anti-GBM antibodies has also been linked to other autoimmune disorders such as ANCA-associated vasculitis. About 10% of persons diagnosed with anti-GBM disease are identified as 'double-positive' because those patients test both anti-GBM and ANCA antibodies. These patients are clinically inclined to be more vasculitis-like, making them more responsive to treatment than those who have anti-GBM disease only. However, this patient was also ANCA-negative and during the course of illness the patient had more typical isolated anti-GBM disease with severe renal involvement.

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As discussed earlier, the degree of renal failure at diagnosis is the most important determinant for predicting outcomes. Patients who present with high serum creatinine levels or require dialysis will have a lower rate of renal recovery. Patients with serum creatinine levels above 5.7 mg/dl or those requiring dialysis at presentation have a poorer prognosis, with a high likelihood of progressing to ESRD despite treatment. The presence of pulmonary hemorrhage, especially among the active smokers, is another important predictor of poor prognosis.

In a nutshell, anti-GBM disease remains a diagnostic and therapeutic challenge because of its rapid clinical onset and the high degree of potential of irreversible organ damage. Timely identification and treatment are necessary to achieve favorable outcome. Early intervention with plasmapheresis, immunosuppressive therapy, and supportive care remains the cornerstone of management in these cases, with the goal of preserving renal function and improving long-term outcomes. However, as exemplified by the case above, even with treatment, the prognosis remains poor in patients who present late or with significant renal involvement. Further research is much needed better management of this deadly disease.

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