

GOODPASTURE'S SYNDROME IN A 40 YEAR OLD MALE WITH MICROSCOPIC HEMATURIA

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Anti-glomerular basement membrane (anti-GBM) antibody disease, a rare autoimmune disorder, involves circulating antibodies directed against α -3 chain of type IV collagen in glomerular and alveolar basement membranes. It can clinically present with mild renal insufficiency to RPGN. We present an interesting case of a previously healthy 40 year old male who presented with fatigue and was found subsequently to have microscopic hematuria. A urologic work-up was negative. However, he progressed rapidly to oliguric acute renal failure with a BUN/Creatinine of 67/9.1 requiring hemodialysis.

He underwent an extensive immunologic work-up and was found to have elevated titers of anti-GBM antibodies (62 units). A renal biopsy showed crescentic glomerulonephritis with linear pattern of IgG staining. Plasmapheresis and immunosuppressive therapy was initiated. After 9 days of plasmapheresis, he developed flank pain and was found to have a retroperitoneal hematoma secondary to bleeding from the biopsy, after which his plasmapheresis was discontinued. Patient is currently on chronic hemodialysis.

This case illustrates the difficulty of diagnosis of anti-GBM disease with only renal involvement. Most patients present with a combination of RPGN and lung hemorrhage, although 30% to 40% present with isolated renal involvement. Renal disease can present with hematuria, proteinuria or features of acute renal failure. A creatinine of more than 5.6 mg/dl with oliguria/ anuria, more than 50% crescents, pulmonary involvement before renal, and dialysis dependent patients portray a worse prognosis. However, even in patients with high serum creatinine, early diagnosis and prompt intensive treatment with plasma exchange, steroids and immunosuppressive therapy (cyclophosphamide) can recover renal function and improve mortality.