Goodpasture's disease preceded by an upper respiratory infection without pulmonary complications: A case study

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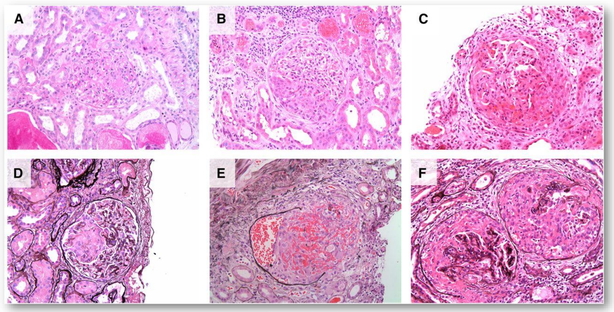
ABSTRACT

Goodpasture's disease is a rare autoimmune disease with anti-glomerular basement membrane antibodies that can damage the kidney and lungs. We report a case of a 59-year-old Caucasian female with Goodpasture's disease with rapidly progressive glomerulonephritis preceded by an upper respiratory infection with no other pulmonary manifestations.



Renal histopathology in anti–glomerular basement membrane (anti-GBM) GN.*(A–C) Hematoxylin and eosin–stained sections demonstrating (A) segmental fibrinoid necrotizing lesion in early anti-GBM GN; (B) small, circumscribed cellular crescent; and (C) large, circumferential cellular crescent. (D–E) Demonstrate the use of Jones methylamine silver stain to delineate glomerular and tubular basement membranes, clearly identifying a segmental area of extracapillary proliferation (D). (E) Demonstrates obliteration of the glomerular architecture and rupture of Bowman's capsule, with extravasation of red blood cells into the urinary space, and significant peri-glomerular inflammation. (F) Adjacent glomeruli with synchronous cellular crescent formation typical of anti-GBM disease.*

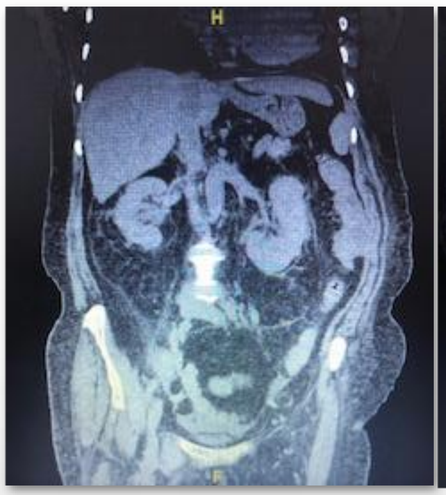
INTRODUCTION

Goodpasture's disease is a rare presentation of an autoimmune process that attacks both the lungs and the kidneys. Cases of Goodpasture's disease range from 0.5-1.8 cases per million per year, disease by its presentation of hemoptysis with proteinuria and hematuria. According to the National Institute of Health summary it is thought that a combination of genetic and environmental factors, such as cigarette smoke, inhaled hydrocarbons, and viruses play a role in the development of this autoimmune condition that attacks Alpha 3 chain of type IV collagen which is present in high concentration in the glomerular basement membrane of the kidneys and also present in the alveoli of the lungs. The syndrome presents as alveolar hemorrhage, proteinuria, and hematuria. Other presenting symptoms include bleeding from the nose or hemoptysis, which occur before the kidney disease in about two thirds of cases and is present in 82%-90% of the adults, pallor which is the most common clinical sign, cough that presents in approximately 40%-60% of the cases, dyspnea in approximately 57%-72% of cases, and occasional heart murmur in approximately 20%-25% of cases. Once the syndrome presents, is recognized clinically, urinalysis to look for blood and proteinuria is ordered along with blood test to look for anti-glomerular basement membrane (anti-GBM) antibodies. The gold standard for diagnosis is kidney biopsy looking for crescentic glomerulonephritis with linear staining of immunoglobulin G (IgG) on immunofluorescence. Clinical features all include kidney dysfunction but lung hemorrhage in only present half of them.

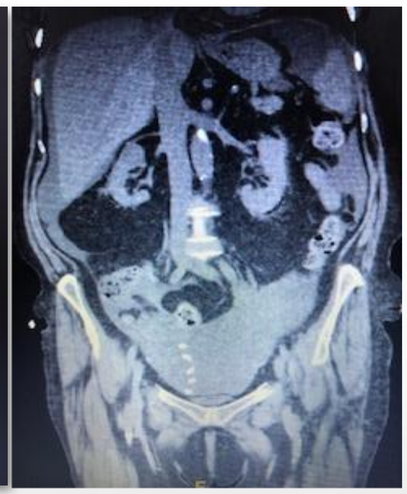
CASE PRESENTATION

This is a case of a 59-year-old Caucasian female who initially presented to the clinic with gross hematuria, subjective fevers, non-productive cough, chills and body aches who was initially diagnosed with subacute sinusitis in the outpatient setting. Her gross hematuria had been attributed to a recent procedure of lithotripsy with right ureteral stent placement due to an obstructing right distal ureteral calculus. She eventually went to the emergency room a week after her presentation due to progressively worsening nausea and vomiting. Her urinalysis in the emergency room showed proteinuria, large hematuria, trace leukocyte esterases, red blood cell count >100/HPF, white blood count of 21-50/high powered field (HPF), and rare bacteria. She was diagnosed with a urinary tract infection and sent home on sulfamethoxazole and trimethoprim (Bactrim DS) 800mg-160 mg one tablet twice daily for seven days, and ondansetron ODT 8 mg sublingually three times daily as needed for nausea for 4 days. She returned back to the emergency room four days later due to persistent nausea, vomiting, and persistent non-productive cough. Initial labs in the emergency room showed a white blood cell count of 12.2 x 103, blood urea nitrogen of 119 mg/dL, creatinine of 14 mg/dL and similar urinalysis findings as before, with proteinuria, large hematuria, trace leukocyte esterase, red blood cell count >100/HPF, white blood cells 51-100/HPF, rare bacteria.

Patient CT Abdomen/Pelvis



4/2/2019: *Nonspecific bilateral perinephric stranding is seen which may be related to chronic medical renal disease. Recommend correlation with urinalysis to exclude GU tract infection. Previous noted obstructing right proximal ureteral stone is no longer seen. 3 mm nonobstructing calcification in the lower pole the right kidney is noted. Left kidney appear grossly unremarkable. Small incompletely characterized hypodensity in the upper pole the right kidney is again noted measuring simple fluid by density measurement and could represent small cyst*



8/21/2020: *Substantial interval involutional of the kidneys relative to 4/2/2019 suggesting rapid progression to end-stage renal disease. Indwelling peritoneal dialysis catheter*

PATIENT LEFT KIDNEY BIOPSY 04/05/19: IMMUNE COMPLEX MEDIATED, CRESCENTIC AND NECROTIZING GLOMERULONEPHRITIS, WITH FEATURES MOST CONSISTENT WITH ANTI-GLOMERULAR BASEMENT MEMBRANE GLOMERULONEPHRITIS.

CASE PRESENTATION CON'T

Computed Tomography (CT) abdomen and pelvis without intravenous (IV) contrast showed mild nonspecific bilateral perinephric stranding of uncertain etiology. The previous noted obstructing right ureteral stone was no longer seen. The patient was fluid resuscitated, given IV ceftriaxone 1 gram once in the emergency room, had blood cultures and a urine culture drawn. She was admitted for pyelonephritis. Nephrology and cardiology were consulted. On initial consultation, nephrology suspected acute tubular necrosis due to recent sulfamethoxazole and trimethoprim treatment or other causes of acute kidney injury. An erythrocyte sedimentation rate, ANCA profile and placement of a right internal jugular tesio catheter for dialysis access were ordered. Chest x-ray showed mild cardiomegaly and no other acute cardiopulmonary abnormalities. The following day, the patient was started on high dose IV prednisone 80 mg daily, auto-immune work up was initiated, and a renal biopsy was ordered. Two sets of blood cultures from two different sites showed no growth in 5 days, and urine culture showed gram positive cocci <10,000 colony forming units/HPF. Biopsy was obtained three days after her second emergency room presentation and hemodialysis was started. The patient's creatinine level continually improved, and she was eventually discharged home in stable condition with pending biopsy results. Upon outpatient follow up, her pathology showed immune complex mediated, crescentic and necrotizing glomerulonephritis, with features most consistent with anti-GBM glomerulonephritis. Nephrology tapered off her corticosteroid treatment and the patient was found to have a high titer of anti-GBM antibodies. She was eventually started on immunotherapy until anti-GBM titers were sufficiently low for planned renal transplant.

DISCUSSION

Our patient had a preceding upper respiratory infection with no further pulmonary complications, but did have proteinuria and hematuria. She was diagnosed with Goodpasture's disease after inpatient admission for acute kidney failure and pyelonephritis without hemoptysis which clouded the clinical picture of classic Goodpasture's disease when the clinical presentation is more obvious prior to confirmatory diagnosis. The patient's lack of classic symptoms and possible subclinical symptoms of traditional Goodpasture's disease and complications from recent urological intervention likely resulted in her acute decline and delay of diagnosis. Her initial presentation was complicated by presumed acute pyelonephritis from recent stent placement for nephrolithiasis. The acute renal failure was treated appropriately while inpatient with close nephrology follow up with kidney biopsy obtained confirming anti-GBM disease. In this case, her rapidly declining kidney function should have warranted earlier dialysis treatment with or without renal biopsy regardless of any confounding factors which might have led to better clinical outcomes for this patient.

CONCLUSION

Awareness of classic symptoms and possible subclinical symptoms of traditional Goodpasture's disease can be helpful in making the correct diagnosis and subsequent management of this rare condition.

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