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CLINICAL IMAGE

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A curious case of antiglomerular basement membrane antibody disease

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Key Clinical Message

Antiglomerular basement membrane (GBM) antibody disease is a vasculitis affecting glomerular capillaries, pulmonary capillaries, or both, with GBM deposition of autoantibodies. It can be both life- and organ-threatening and delayed diagnosis may be detrimental. Alveolar hemorrhage and microscopic hematuria should be excluded in anybody who is suspected of this disease.

KEYWORDS

acute kidney injury, alveolar hemorrhage, hematuria, renal biopsy

1 | CASE

A 21-year-old white man presented with gross hematuria for a week. Vital signs were stable, and laboratory data were significant for acute kidney injury with a serum creatinine of 1.6 mg/dL (baseline~0.9) and urine RBC count of >100/hpf. Abdominal CT scan excluded nephrolithiasis and pyelonephritis. Six months prior to presentation, he sought medical attention for hemoptysis and was treated with azithromycin for atypical pneumonia as the CT chest showed diffuse nodular

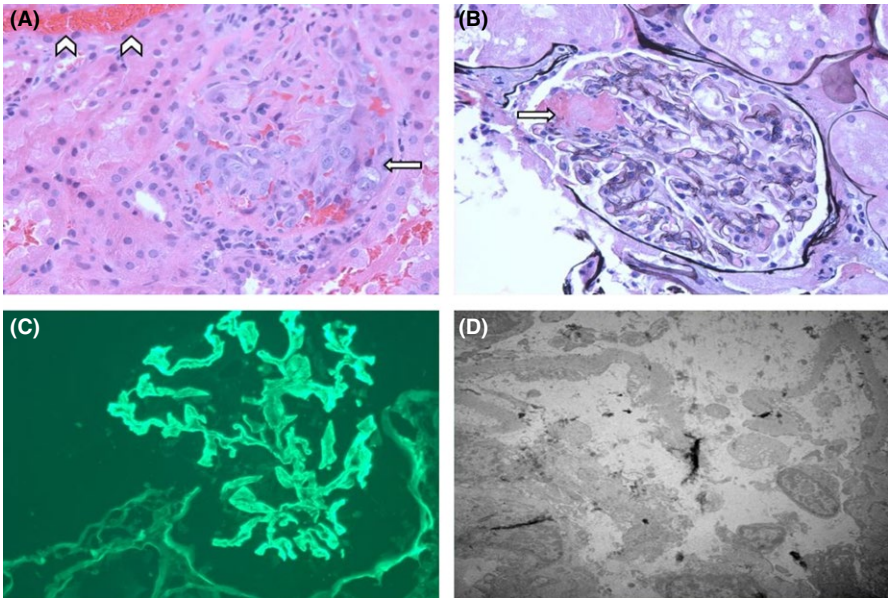

FIGURE 1 Renal pathology demonstrating anti-GBM antibody disease. A, Glomerulus containing cellular crescent [arrow], compressing glomerular capillary tuft. An intratubular RBC cast is also shown [chevrons] (H&E 40x10). B, Segmental fibrinoid necrosis in a glomerulus [arrow] (Silver stain 40x10). C, Immunofluorescence showing IgG linear staining along the glomerular capillary loops. D, Interruption of glomerular capillary basement membrane under electron microscopy, indicating severe glomerular injury

ground glass opacities. Anti-GBM antibodies were positive at that time but as the titer was reduced during the follow-up visit and his symptoms resolved, it was thought to be "false" positive. Urinalysis was not checked. This time, renal function worsened rapidly with increased antibody titer. He was started on pulse steroids and plasmapheresis. Renal biopsy confirmed anti-GBM disease with cellular crescents and segmental necrosis in 50% of the sampled glomeruli (Figure 1). No active vasculitis was identified. Plasmapheresis was continued for a total of 7 sessions with approximately 60 mL/kg volume replacement. He remained dialysis-dependent 2 months later, despite cyclophosphamide therapy.

False-positive results for anti-GBM do occur, especially in diabetics.[1] However, before suspecting this, it would be prudent to evaluate all symptomatic patients for diffuse-alveolar hemorrhage (DAH) and microscopic hematuria. Isolated pulmonary involvement is rare[2] and while serum creatinine may be preserved initially, especially in young patients with good renal functional reserve,[3] microscopic hematuria is a reliable clue to renal involvement. It is important to note that early diagnosis of anti-GBM disease and treatment with plasmapheresis is associated with better prognosis. For example, in a large, multicenter controlled trial[4] including 137 patients, Plasmapheresis followed by prednisone and cyclophosphamide therapy was associated with a significantly higher rate of renal recovery at 3 months compared with medical management alone (69% vs 49%, respectively). Similarly, plasmapheresis has been linked to reduction of mortality in patients with DAH.[5]

CONFLICT OF INTEREST

The authors have declared that no conflict of interest exists.

AUTHORSHIP

All authors: made substantial contribution to the preparation of this manuscript. AK: drafted the manuscript, performed literature search, and was the attending nephrologist for the case; ML: participated in patient care and assisted in drafting the manuscript; XZ: provided pathology images and pertinent input and was the attending renal pathologist for the case.

INFORMED CONSENT

Informed consent has been obtained for the publication of this clinical image.

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