ANCA-Associated Vasculitis and Systemic Manifestations with Volatile Solvent Exposure

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Introduction

ANCA-associated vasculitides (AAV) are a group of immune-mediated disorders in which systemic tissue injury results from the interaction of an initiating inflammatory event and a subsequent highly specific immune response. This can result in widespread vascular damage to nearly any microvascular system in the body, most commonly the glomeruli, lungs and sinuses.

History of Present Illness

A 54-year-old male with a history of nephrolithiasis and pericarditis complaining of prolonged headaches, dyspnea, and vomiting. He admits to using new volatile solvents at his job (Methyl-ethyl-ketone) to dissolve countertops without respirator or gloves immediately prior to symptoms. He was admitted for acute renal failure with metabolic acidosis.

Hospital Course

The patient was transferred to intensive care for emergent dialysis was initiated after finding he was in acute renal failure with a serum creatinine of 10.4 with new metabolic acidosis. Broad spectrum antibiotics were initiated for risk of community-acquired pneumonia due to hypoxia and chest X ray with suggestion of edema versus a multifocal pneumonia. Bronchoscopy was performed and did not show alveolar hemorrhage. Diagnosis of P-ANCA vasculitis was confirmed by renal biopsy and the patient started on high dose corticosteroids and rituximab. Echocardiogram was performed and showed an ejection fraction of 34% with apical and lateral left ventricular wall akinesis. Given profound P-ANCA cardiomyopathy, the patient was started on losartan, spironolactone, and carvedilol. Patient's condition stabilized and he was able to be discharged on steroids and PJP prophylaxis;he was instructed to follow-up with outpatient for dialysis and erythropoietin the next day. He had outpatient cardiac MRI scheduled as well.

Images

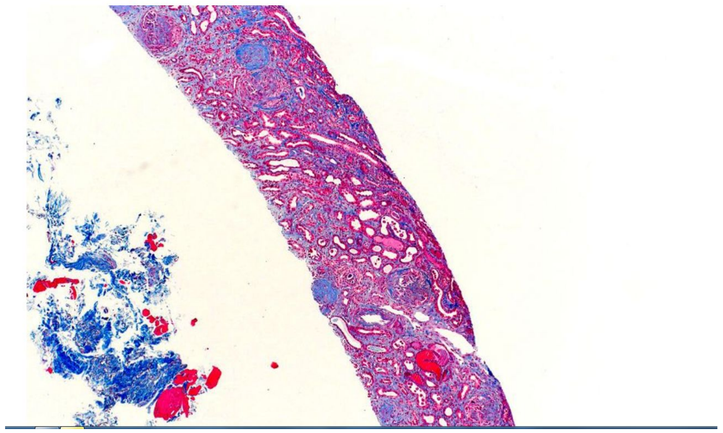

Figure 1: Low power: Glomerular biopsy with immunofluorescence on ethanol-fixed neutrophils, perinuclear/nuclear staining (P-ANCA) is observed.

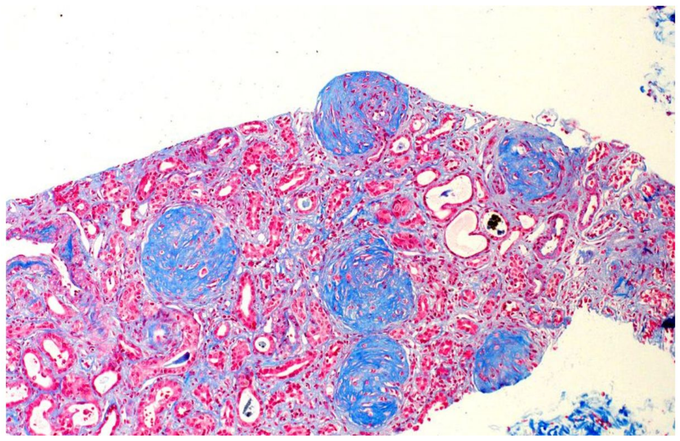

Figure 2: Global view of biopsy with immunofluorescence against P-ANCA.

Discussion

The systemic reach of vasculitides is not limited to renal and pulmonary, as seen with this patient's new-onset gastrointestinal and cardiac abnormalities. Furthermore, this case illustrates noninfectious inhalation exposure as a possible inciting event for the development of ANCAs. AAV incidence is 1.2 - 2.0 cases per 100,000 individuals. Roughly 89% of cases occur in white adults with equal male/female distribution. In one study of 155 similar-presenting patients at four months, 14 percent died, and 35 percent lived but required dialysis; 51 percent did not require dialysis and had no evidence of active vasculitis. Another study showed that remission was induced in 57% of 96 patients with eGFR of ≤10 . Treatment follows an algorithm developed based on BVAS score which relies on signs of end-organ damage to make the decision between rituximab or cyclophosphamide. Studies have shown that rituximab is an effective alternative to cyclophosphamide for new diagnosis of AAV. Plasma exchange is recommended based on three indications: the need for dialysis or a serum creatinine of >4.0 mg/dL, positive anti-GBM auto-antibody, or pulmonary hemorrhage. Rituximab regimen is 1g initially followed 14 days later by another 1g dose. Oral steroid therapy should occur concomitantly at specific doses.

References

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