CATASTROPHIC ANTIPHOSPHOLIPID SYNDROME DUE TO INADEQUATE ANTICOAGULATION LEADING TO END STAGE RENAL DISEASE.

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A 30 year old African American female with a history of systemic lupus erythematosus (SLE), hypertension, anti-phospholipid syndrome (APS) and autoimmune hemolytic anemia (AIHA) presented with nausea, vomiting and 6 lb weight loss. On admission, serum creatinine was 2.2mg/dL, hemoglobin 7.7g/dL (baseline 0.9 mg/dl and 10.8g/dl respectively) and INR of 2.5. Pt was diagnosed with recurring AIHA and started on steroids. Renal biopsy showed acute glomerular thrombotic microangiopathy (TMA) with ISN-RPS class II lupus nephritis. Serum creatinine improved to 1.7mg/dL. Patient was discharged on warfarin and tapering prednisone dose. INR was 1.1 on discharge.

Pt returned 8 days later with chest pain, shortness of breath and hemoptysis. Pt was intubated for respiratory failure. Blood Urea Nitrogen (BUN) was 190mg/dl and serum creatinine 14.5mg/dL. Chest CT showed alveolar hemorrhage. A diagnosis of possible CAPS was made. Anticoagulation and steroid therapy was continued. She received 4 days of plasmapheresis and IVIg therapy. Hemodialysis was initiated. Repeat renal biopsy showed severe, acute TMA involving arterial vessels, with ISN-RPS class III lupus nephritis and lupus vasculopathy, all worse compared to the prior biopsy. Patient was started on mycophenolate mofetil for the immune complex mediated disease. Despite overall improvement, renal function did not recover.

CAPS is a rapidly progressive disease, with multi organ involvement, seen in <1% of APS and carries a mortality of 50%. We hypothesize that CAPS was precipitated by lack of adequate anticoagulation on discharge. Inadequate anticoagulation for only a few days seems to be enough to incite CAPS. Our patient demonstrated a good clinical response overall, despite the presence of SLE which is a poor prognostic indicator. There is paucity of data on progression of renal disease in CAPS. The renal disease progressed to end stage in our patient necessitating continuation of renal replacement therapy eight months after presentation.