

A RARE CASE OF CO-EXISTING SYSTEMIC LUPUS ERYTHEMATOSUS (SLE) AND MICROSCOPIC POLYARTERITIS (MPA) PRESENTING AS PULMONARY RENAL SYNDROME (PRS)

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PRS is characterized by co-existence of diffuse alveolar hemorrhage and glomerulonephritis and has been associated with SLE, Good pasture's syndrome and Anti Neutrophil Cytoplasmic Antibody (ANCA) associated vasculitides such as Wegener's Granulomatosis (WG) and MPA. We report a rare case of PRS in which both SLE and MPA were diagnosed at the same presentation.

A 29 year old female with history of still birth presented with acute respiratory distress and delirium after having flu like symptoms for few days. Physical exam revealed pallor, severe tachypnea, hypoxemia (Spo2 82%) and diminished breath sounds bilaterally. Initial labs were within normal limits except Hb 6.1 with BUN/Cr 12/1.1. CXR showed bilateral consolidations. CTPA ruled out PE and showed diffuse bilateral consolidations. She had massive hemoptysis and was emergently intubated. Bronchoscopy revealed diffuse alveolar hemorrhage. She remained intubated and hemoptysis started resolving but developed rapidly progressive anuric ARF in next 10 days, BUN/Cr 147/8.08 and was started on dialysis .Immunological workup showed positive ANA (titer 1:1280) with positive dsDNA and Smith AB suggestive of SLE. Anti cardiolipin and anti glomerular basement membrane (GBM) antibodies were negative. Interestingly p-ANCA titer (1:640) was also positive with positive myeloperoxidase Ab but negative c-ANCA & Proteinase Ab suggestive of microscopic polyarteritis. Renal biopsy showed fibrinoid necrotizing glomerulonephritis with crescent formation consistent with ANCA associated glomerulonephritis but without any SLE associated lesions. Meanwhile patient was already started on IV steroids and cyclophosphamide. She was eventually extubated and discharged on azathioprine but would require life long dialysis. This case illustrates the role of kidney biopsy in establishing cause of PRS in patients who had coexisting 2 different immunological disease processes.