CRYOGLOBULINEMIC VASCULITIS PRESENTING AS PULMONARY RENAL SYNDROME IN A PATIENT WITH SCLERODERMA

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Pulmonary renal syndrome can manifest in scleroderma (Ssc) by a variety of mechanisms, including thrombotic microangiopathy, scleroderma-renal crisis, small vessel vasculitis, and anti-glomerular basement membrane (GBM) disease. We report the case of a 39 years old woman with a longstanding history of SSc presenting with diffuse alveolar hemorrhage and non-oliguric acute renal failure (peak serum creatinine 3.8 mg/dL from 0.7 mg/dL). Primary disease manifestations were limited skin involvement, Raynaud's phenomenon, interstitial lung disease and recurrent ulcerations affecting the lower extremities. She developed new onset pedal edema, acute renal failure with urinary red cell casts, and respiratory insufficiency requiring intubation.

Bronchoalveolar lavage revealed diffuse alveolar hemorrhage. Serum complement was low, rheumatoid factor elevated, and serum cryoglobulins were positive (monoclonal IgM and polyclonal IgG). Antinuclear and anti-Scl 70 antibodies were positive at high titers. Anti-GBM, -dsDNA, -Sm, -PR3 and -MPO antineutrophil cytoplasmic antibodies were negative as well as HIV and hepatitis C serologies. Renal biopsy revealed hyaline thrombi in the glomerular capillary loops and focal arterial fibrinoid necrosis. Immunofluorescence showed 2+ IgG and IgM staining of the capillary loops. Electron microscopy showed subendothelial deposits of approximately 29nm diameter. A diagnosis of cryoglobulinemic vasculitis was made. The patient was treated with high dose IV methylprednisolone, cyclophosphamide and plasmapheresis. The creatinine decreased to 1.9 mg/dL without dialysis and the alveolar hemorrahage subsided. We have not seen any other cases in the literature of cryoglobulinemic pulmonary renal syndrome in SSc.