RENAL MANIFESTATIONS OF POEMS SYNDROME

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POEMS syndrome is a rare disorder associated with Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal plasma cell disorder and Skin changes. Renal involvement in POEMS syndrome is rare with unique biopsy findings and a frequent response to steroid treatment. This is a case report of a biopsy proven pauciimmune membrano-proliferative glomerulonephritis in a Hispanic patient with POEMS.

41 y/o Hispanic female presented with dyspnea and generalized swelling. Exam revealed anasarca, extremity skin thickening with hyperpigmentation, hepatosplenomegaly and diffuse peripheral neuropathy. Labs: Hb/Hct 10/30; BUN/creatinine 30/1.5; total protein/albumin 6/2.1; UA: 2+protein, 5-10 red cells, no casts; 24 hour urine protein 338 mg; SPEP with M spike of 0.32g/dl; Urine/Serum IFE: IgG lambda chain. Lymph node biopsy revealed Castleman's disease (angiofollicular hyperplasia, a non-clonal disease of the lymph nodes). EMG revealed sensory motor polyneuropathy. Diagnosis: POEMS syndrome with Castleman's disease. Renal biopsy revealed lobulated glomeruli with mesangial hypercellularity and a "double contour" pattern; subendothelial electron dense deposits were present but no immune deposits were observed. Classic renal biopsy has been described as "pauciimmune membrano-proliferative" glomerulonephritis with absence of immune and light chain deposits, but with electron dense subendothelial deposits.

The mechanism of renal involvement in POEMS is unclear, but increased levels of IL-6 with subsequent increase of vascular endothelial growth factor (VEGF) have been implicated. Moreover, the same pathophysiology mechanism has been theorized for Castleman's disease. The increase of VEGF results in angiogenesis, capillary proliferation and endothelial hyperplasia in the glomerular vasculature, and it causes B-cell proliferation resulting in hyperplastic follicles and hence the enlarged lymph nodes. Interestingly, many patients have shown a dramatic improvement in renal function with steroid therapy along with decreasing IL-6 levels suggesting a central role of the proinflammatory state in the pathophysiology. Early recognition of renal involvement in POEMS syndrome is important to understand disease mechanisms and treatment options, including the role of steoids. The use of ACEI may play a role in controlling proteinuria in these patients.