

RARE ASSOCIATION OF IgA NEPHROPATHY AND LYMPHEDEMA DISTICHIASIS SYNDROME: FIRST CASE REPORT

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A 45 year old female presented with complaints of intermittent hematuria for 30 years. The first episode was along with an episode of acute pharyngitis 30 years ago and had resolved spontaneously. After this, she started noticing progressively increasing swelling of her lower extremities, left more than right and multiple layers of eye lashes. She was diagnosed with lymphedema-distichiasis syndrome. Her mother and two maternal aunts had similar disease presentation. She continued to have intermittent hematuria for many years generally along with minor infections but had never sought medical attention. Physical examination revealed significant edema of both lower extremity left greater than right and multiple layers of eye lashes. Laboratory tests revealed blood urea nitrogen of 66 mg/dL, serum creatinine of 8.5 mg/dL, red turbid urine with more than 50 RBCs/hpf with RBC casts. Daily protein excretion was 3.0 grams. IgA serum level was 526 mg/dl, C3, C4 were within normal limits. Antibodies to HIV, hepatitis, ASO were negative. In a renal biopsy specimen with total of 8 glomeruli, all had increase in mesangial matrix and 2 had fibrinous crescents, effacement of podocytes on electron microscopy. Immunofluorescence staining showed mesangial granular deposits of IgA and C3. The patient was diagnosed with IgA nephropathy and started on steroids and cyclophosphamide. Her renal function improved and she was discharged with a BUN of 30 mg/dL and serum creatinine of 2.4 mg/dl.

We have noted several possible associations between lymphedema distichiasis syndrome and IgA nephropathy. Both diseases are known to have familial forms. Recurrent cellulitis in a patient with lymphedema can lead to progression of IgA nephropathy. Lymphatic obstruction has also been known to increase IgA levels and their subsequent deposition in glomeruli can lead to IgA nephropathy. Also increased IgA production in IgA nephropathy can lead to vasculitis and lymphangitis which may worsen lymphedema.