

VENOUS THROMBOEMBOLISM AS THE INITIAL PRESENTATION OF PRIMARY RENAL AMYLOIDOSIS

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Amyloidosis is a generic term that refers to the extracellular tissue deposition of fibrils composed of low molecular weight subunits of a variety of proteins. Primary or AL amyloid is due to deposition of protein derived from immunoglobulin light chain fragments.

Hematologic manifestations associated with Amyloidosis, include anemia, thrombocytopenia and bleeding diathesis which may be seen in up to 44% of cases. Venous Thrombo Embolism (VTE) is a less well recognized complication.

We report a 65 year old Hispanic female who presented with history of two episodes of idiopathic VTE and hypoalbuminemia. Her initial presentation was 2 years ago, with bilateral lower extremity (LE) swelling and pain which was due to deep venous thrombosis (DVT). Evaluation for a hypercoagulable state was negative. She was treated with Coumadin for 6 months. She presented again 6 months later, with right LE DVT and was restarted on Coumadin. At this time, she was noted to have a serum albumin of 2.5 gm/dl which was persistently low for 2 years duration and proteinuria of 5gm/day with a normal complete blood count, renal panel and echocardiogram. Serum protein electrophoresis showed a monoclonal spike in the alpha region. Renal biopsy revealed primary amyloidosis. Kappa: Lambda ratio was normal and urine light chain assay was negative. Bone marrow biopsy shows only 1% plasma cells.

In a review of medical records of 2,132 patients with biopsy proven AL amyloidosis from Mayo clinic, they found that thromboembolic phenomena can antedate the diagnosis of amyloidosis. In a study published from Cleveland clinic the incidence of VTE in amyloidosis was up to 11% at 10 years, which was comparable to the incidence of VTE in multiple myeloma. Impairment of thrombin - antithrombin pathway, in association with low antithrombin biological activity has been recognized to have possible pathogenic role in hypercoagulability of amyloidosis. This case illustrates that primary amyloidosis can be an underdiagnosed cause of VTE and Nephrotic syndrome should be considered in the differential diagnosis of unexplained VTE.