

**IDIOPATHIC LIGHT CHAIN DEPOSITION DISEASE (LCDD)  
WITH LIGHT CHAIN CAST NEPHROPATHY (LCCN)  
TREATED WITH BORTEZOMIB. A CASE REPORT AND  
REVIEW OF TREATMENT/PROGNOSTIC FACTORS**

Manish Gera<sup>1</sup>, R. Jeevan<sup>1</sup>, S. Dhar<sup>1</sup>, M Shaheen<sup>2</sup> C.L. Phillips<sup>3</sup>. Internal Medicine Nephrology Inc. Terre Haute, IN<sup>1</sup>. Providence Med. Ctr. Terre Haute, IN<sup>2</sup> I. U. Pathology, Indianapolis, IN<sup>3</sup>

LCDD is a systemic disorder characterized by deposition of mono-typical light chains in various organs. Presence of LCCN with LCDD in absence of multiple myeloma (MM), when light chain load is less, is rare and is not well characterized. Moreover, treatment remains anecdotal.

We report a case of a 40 yr old lady who presented with nephrotic syndrome. Glomerulonephritis work up was unremarkable. Free kappa light chains were borderline high and could not be interpreted in view of high creatinine. Renal biopsy on light microscopy showed nodular expansion of matrix with laminations. Tubules showed protein casts with cellular response, including giant cell reaction. Immunofluorescence showed kappa antibodies labeling the mesangium. Electron microscopy showed sub endothelial electron dense deposits suggesting LCDD with LCCN. The patient was started on dialysis with peak creatinine around 5 mg/dl. Simultaneous treatment with bortezomib was started. Patient's renal function improved and patient remains off dialysis with creatinine stabilizing at 2 mg/dl at 1-year follow-up. A systematic review of literature was done.

a) This is the first reported case of LCDD with LCCN, treated with bortezomib with good response. As in MM, bortezomib can be the first line of treatment, given its favorable side effect profile. b) Acute renal failure due to LCCN in idiopathic LCDD may have better chance of renal recovery as apposed to MM, because of less light chain burden and less fibrogenic effect. c) LCCN may also augur better prognosis especially in LCDD with isolated kidney involvement as these patients may get detected and treated early.