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# Case report- A case of light chain nephropathy

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## Introduction

Light Chain Deposition Disease (LCDD) is a rare disease characterized by deposition of monoclonal non-amyloid light chains in multiple organs (1). It was first described about three decades ago. Due to varied clinical presentation and many differential diagnosis on morphology it is both under reported and under diagnosed (1, 2, 3). Here we report a case of 65 year old male patient who was diagnosed with light chain nephropathy and was a case worth presenting in view of very high quantity of these light chains with normal calcium and no lytic lesions which seldom exists in literature globally.

## Case report

A 65-year-old male presented with chief complaints of generalized body aches with significant history of NSAID intake followed by decreased urine output. Personal and family history was unremarkable. General physical examination revealed pallor, an emaciated patient and rest of the examination was normal. Investigations revealed normocytic normochromic anaemia with decreased haematocrit (24%); increase in blood urea nitrogen (110 mg/ dl); increased serum creatinine (5 mg/dl),normal serum calcium levels (8.7 mg/dl) and normal blood sugar levels (Fasting sugar 85mg/dl, HbA1c 5.8%). A 24-hour urine protein excretion was increased (4.1 g); although serum albumin levels were almost normal (4.0 mg/dl). Urine dipstick was suggestive of 2+ albumin. Ultrasound showed normal kidney size with increased echogenicity. Corticomedullary differentiation was maintained in both kidneys. There was no Hydronephrosis. No organomegaly was noted. Further laboratory work up showed normal C3, C4 complement component and C-ANCA ,P-ANCA were normal. A diagnostic renal biopsy was performed. Multiple sections stained with H & E,MT,PAS, Silver methanamine and Congo red included renal medulla and cortical parenchymal core containing up to 13 glomeruli three (23%) globally sclerosed.The glomeruli showed non proliferative morphology, significant capillary wall thickening or mottling splits were not seen. There was no evidence of crescent formation, tuftnecrosis, subendothelial / congophillic deposits or intracapillary thrombi in the visualized glomeruli. Tubular atrophy and interstitial fibrosis involved about 15 to 20 percent of sampled cortex. Tubules showed evidence of severe injury with inspissation of scattered hyaline granular and many atypical casts with waxy to granular appearance, brittle appearance, fracture planes also associated with inflammatory and epithelial cell reaction at places ,significant interstitial oedema ,multifocal chronic

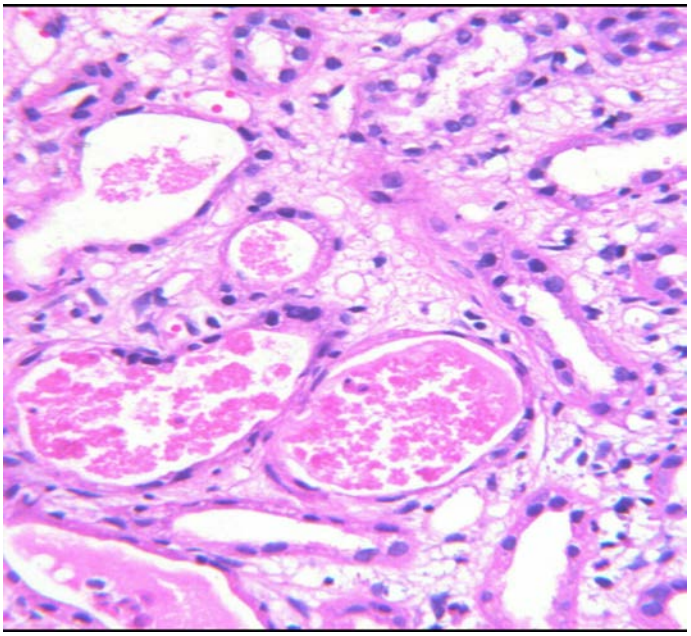
interstitial inflammation admixed with occasional neutrophils and eosinophils were noted.

Many atypical appearing tubular casts showing 3+ pattern less staining for kappa light chains and concurrent negativity for lambda light chains were also observed.

So a diagnosis of light chain nephropathy was made and the case was worth reporting keeping in view the number of light chains as they were significantly high. Such high levels of these light chains seldom exist in literature pertaining to light chain nephropathy.

**Morphological Features**

Severe tubular injury with inspissations of many atypical intratubular casts showing evidence of kappa light chain restriction in DIF studies. Glomeruli reveal non proliferative morphology and DIF studies reveal insignificant glomerular immune deposits.Multifocal chronic interstitial inflammation admixed with few neutrophils and eosinophils and mild increase in tubule interstitial chronicity are observed.



Further laboratory work up revealed 33% plasmacytosis in bone marrow and M spike in SPEP.Serum Free light chains increased, Kappa-32200 mg/l, Lambda-32.40 mg/l, Kappa/Lambda ratio-964.072.

**Discussion**

Monoclonal immunoglobulin deposition disease is a rare paraproteinemia characterized by the deposition of monoclonal immunoglobulins in the renal basement membrane , manifesting in $5^{th}$ -$6^{th}$ decade of life (4).MIDD is further grouped into light chain deposition disease, heavychain deposition disease and light and heavy chain deposition disease(1,4) .

Nephrotic range proteinuria is common but full blown nephrotic syndrome is seen in only one quarter patients . Clinical Presentation is in the form of proteinuria, microscopic hematuria, hypertension and variable degrees of renal insufficiency (2,3,5) Histopathological features predominantly include nodular sclerosing glomerulonephritis. The deposits are mainly made of alpha subunits and do not stain Congo red as they are non amyloid (6).

Bone marrow involvement is a Hallmark and the criteria for MM are met in 40 – 50 % cases . The clonal production of nephrotoxic FLCs is responsible for the kidney injury and gammopathies(7)

This patient had kappa chain nephropathy which is extremely rare . The treatment for this condition is mostly symptomatic and in research yet .

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