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Rare cause of chronic kidney disease in young adult: A case report

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Monoclonal gammopathy of undetermined significance (MGUS) is consistent with immunoglobulin precipitation or deposition diseases occurring with B cell proliferation. Diagnostic features: proteinuria (>60% nephrotic range), microscopic hematuria (>65%), hypertension (>50%), renal insufficiency, antinuclear antibody (ANA) positive-speckled pattern (19%). Electron microscopy reveals 10-30 nm micro fibrils with random orientation in mesangium and glomerular capillary wall. A 36 years old male, presented with history of generalized weakness, nausea, reduced appetite and reduced urine output since 2 months. Hemodialysis was initiated in view of progressive renal dysfunction and underwent kidney biopsy. Autoimmune work up was not significant and diagnosis of dense deposit disease was made. Steroids started at 40 mg OD and tapered by 5 mg weekly. Hemodialysis (HD) sessions advised thrice a week through left femoral uncuffed non-tunnelled HD catheter. Left brachiocephalic arteriovenous fistula was created. Patient came for kidney transplant work up to our center with wife as a prospective donor. He was admitted for further work up and his biopsy was reviewed. Free kappa/lambda ratio of 28.38 (0.26–1.65). Biopsy impression was: widespread effacement/loss of visceral epithelial foot processes; massive mesangial accumulation of intermediate electron dense deposits with fine granular/short fibrillary appearance. He was diagnosed as a case of chronic kidney disease/stage VD/fibrillary glomerulonephritis. At present, patient is asymptomatic and on thrice a week HD at our center. Patient underwent six cycles of bortezomib and dexamethasone (once a week regimen). His initial serum creatinine was 10.8 mg/dl and now it had come down to 4.5 mg/dl. Repeat kappa/lambda ratio is 7.78.

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