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MEMBRANOUS GLOMERULONEPHRITIS WITH LIGHT CHAIN RESTRICTED DEPOSITS: A REGISTRY DATA FROM SOUTH ASIA

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INTRODUCTION AND AIMS: Classically, the immune complex in primary membranous nephropathy (PMN) is polytypic. There have been very few reports of membranous nephropathy (MN) with light-chain deposits. However, the literature on their clinical presentation and outcome is very scanty. We report our MN registry data on the rare entity.

METHODS: The present report is a prospective study of patients with MN with a monoclonal light chain on the immunofluorescence (IF), the current research is a part of our MN registry. Patients were followed monthly with proteinuria, serum creatinine and albumin for six months, then every three months. Definition: Presence of glomerular basement membrane thickening with positive granular staining for IgG by IF with a single chain phenotype (kappa (k) or lambda (λ)) and presence of subepithelial granular deposits by electron microscopy. Clinical outcomes of complete/ partial remission (CR/PR) were as per the KDIGO guidelines.

RESULTS: A total of 5 (1.42%) patients had MN with light-chain deposits over the last seven years. Mean age of the patients was 33.2 ± 6.55 years. The mean proteinuria, serum albumin and serum creatinine was 5.73 ± 2.17 g/day, 2.86 ± 0.51 g/dL and 1.34 ± 1.19 mg/dL, respectively. None of the patients had a lymphoproliferative disorder. Only one (20%) patient had elevated free light chain ratio. Four (80%) patients were M-type phospholipase A2 receptor (PLA2R) negative (tissue and serum) and 1 (20%) was positive for PLA2R (both serum (135 RU/ml) and tissue). Three (60%) cases had monoclonal IgG3/k, one IgG3/λ, whereas one patient with PLA2R positivity had an IgG3/IgG4k subtype. Three were treated with cyclical cyclophosphamide and steroids; two (67%) patients treated achieved CR, and one patient (33%) with elevated baseline creatinine had a reduction in serum creatinine with persistent proteinuria at the end of 12th month of follow-up. One patient with PLA2R related MN was treated with rituximab and achieved CR. The patient with an elevated free light chain at baseline was treated with Bortezomib/Thalidomide/Dexamethasone, had complete remission at 12 months, however, had a progressive rise in creatinine over the next 40 months of follow-up. Details mentioned in Table 1.

CONCLUSIONS: MN with light-chain deposits is a rare clinical entity and usually not associated with a lymphoproliferative disorder. The disease responds favorably to the traditional drugs used to treat PMN.