

Apla2r- significance and management in Primary Membranous Nephropathy

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This case is about the road map and management approach of a typical nephrotic syndrome. An individual who presented with a complicated nephrotic picture with thrombosis of pulmonary artery, edema and pleural effusion associated with constitutional symptoms. Base line workup revealed heavy proteinuria. Keeping in view the fact that nephrotic syndrome is not a diagnosis but a manifestation of a serious underlying condition merit evaluation further both non invasive and invasively as per need which need to be treated in its own right while managing complications and symptoms. Exhaustive efforts were put in to hunt for the culprit cause. Widespread workup in this particular case lead to the presence of anti phospholipase receptor anti bodies (aPLA2R). Age appropriate cancer screening was unremarkable. This was quite reassuring that aPLA2R is consistent with the diagnosis of primary membranous nephropathy. Detection of this antibody has diagnostic and prognostic value precluding kidney biopsy. Titers of this antibody are considered a guiding tool to monitor treatment adequacy and hence disease remission.

Land mark trials and research work reviewed extensively during the management of this complex case. After going through historical treatment modalities of primary membranous nephropathy, based on critical appraisal of relevant literature, rituximab has been opted as a standard of care for this particular renal condition over cyclophosphamide and cyclosporin. We confidently diagnosed idiopathic membranous nephropathy based on the antibody titer without the kidney biopsy and managed it with rituximab successfully and uneventfully by periodic monitoring and swift decline in aPLARtiter. Aim of sharing experience through this particular clinical case is to highlight two important points. First, patients can be saved from the need of kidney biopsy which is an invasive procedure having morbidity risks.

Second rituximab for induction of remission and maintenance as a first choice drug which is highly evidence based. It is advantageous in terms of tolerability, ease of administration, side effect profile and significantly lesser relapse rates. These characteristics promote cost effectiveness which is the most favorable factor towards health economics. Management strategies for refractory and resistant cases have also been highlighted by comparatively discussing role of cyclophosphamide, cyclosporine and steroid use. Rare treatment has also been summarized.

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