KSN SEOUL, KOREA 2023
April 27(Thu) - 30(Sun), 2023
Coex, Seoul, Korea

Submission No.: PG02-9035
Session Title: PG Education 2 (Glomerular Disease)
Date & Time, Place: April 27 (Thu), 10:30 - 12:00, Room 3

Membranous Nephropathy

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Membranous nephropathy (MN) is a major cause of nephrotic syndrome in adults, with a mean age of diagnosis at 50~60 years and a male predominance. Pathologically, it is characterized by thickening of the glomerular capillary walls on light microscopy, which results from immune complex deposition on the basement membrane of glomeruli. In 75~ 80% of patients, there is no underlying cause of MN (primary MN), and the remaining 20~25% of patients are associated with drugs, autoimmune diseases, infections, such as hepatitis B or hepatitis C, and malignancies (secondary MN).
The clinical course of the MN, commonly referred to as the rule of thirds, is variable; a third of the patients undergoing spontaneous remission, a third maintaining proteinuria and the remaining third progressing to loss of kidney function.
The understanding of the pathogenesis of primary MN has remarkably progressed since the identification of M-type phospholipase A2 (PLA2) receptor and thrombospondin type-1 domain-containing 7A (THSD7A) as target antigens. Over the past few years, several novel antigens have been identified using laser microdissection of glomeruli and mass spectrometry. It includes exostosin1 (EXT1) and exostosin2 (EXT2), neural epidermal growth factor-like 1 protein (NELL1) and semaphorin 3B (SEMA3B), protocadherin7 (PCDH)7, serine protease HTRA1 (HTRA1) and neural cell adhesion molecule 1 (NCAM1). EXT1 and EXT2 were detected in secondary MN associated with autoimmune disease, especially lupus nephritis, whereas NELL1, in some patients with malignancy, and SEMA3B mostly in children and young adults.
The management of MN depends on the risk for progressive loss of kidney function or complication of nephrotic syndrome. The clinical criteria were proposed to stratify the patients with MN into with low, intermediate, high and very high risk in an updated KDIGO guideline. The optimization of therapy in MN requires the accurate identification of patients according to the risk prediction and timely application of the effective therapies, especially immunosuppressive agents based on the risk-benefit balance. In patients with a high risk of kidney function decline, the combination of cyclophosphamide and corticosteroids has shown benefit on renal function. Calcineurin inhibitors (CNIs) and rituximab alone or in combination are considered to be less toxic, but there are limited data on renal survival. New CD20 antibodies, anti-plasma cell therapy, and anti-complement therapy might be considered in patients with refractory MN. In addition, monitoring the level of serum anti-PLA2R antibody would provide useful information for tailoring therapy in patients with PLA2R-associated primary MN.
In this lecture, recent advances in the pathophysiology of MN and updated guidelines in the diagnosis and management of patients will be presented.