THE KOREAN SOCIETY OF NEPHROLOGY
APCN & KSN 2024
June 13(Thu) - 16(Sun), 2024 Coex, Seoul, Korea
Promoting Sustainable Kidney Health: The Asia-Pacific and Beyond

Submission No. : GN03-0003
Session Title : Glomerulonephritis 3 (Pathology)
Session Topic : Renal Pathology
Date & Time, Place : June 15 (Sat) / 10:30-12:00 / Room 1 (GBR 101 - 102)

Glomerular Diseases with Nonamyloid Fibrillar Deposits

Kelly Smith
University Of Washington, United States

Electron microscopic examination of glomerular diseases has helped define morphologically distinct forms of glomerular diseases. This lecture will focus on the characterization of non-amyloid glomerular diseases with deposits that are organized into fibrils, the salient morphologic features that are helpful to distinguish these entities, the pathophysiology of these diseases, and the clinical implications for accurately defining the nature of non-amyloid fibrillar glomerular diseases. The most common form of kidney disease caused by non-amyloid fibrillar deposits is fibrillary glomerulonephritis. Laser capture microdissection and tandem mass spectrometry have demonstrated that DNA J homolog subfamily B member 9 (DNAJB9) is nearly universally present in fibrillary glomerulonephritis. The concordance of positive DNAJB9 staining with strict morphologic criteria for fibrillary glomerulonephritis is close to 100%, with anti-DNAJB9 immunohistochemical tests reporting at least 98% sensitivity and 98% specificity for the diagnosis of fibrillary glomerulonephritis, making DNAJB9 a highly specific and sensitive marker for this disease. Other kidney diseases with organized fibrillar deposits include immunotactoid glomerulopathy, cryoglobulinemic glomerulonephritis, monotypic fibrillary glomerulonephritis, fibronectin glomerulopathy, cryofibrinogen-associated glomerulonephrits, collagenofibrotic glomerulopathy, and LMX1B-associated nephropathy. Understanding the differences between these diseases is critical for accurate diagnosis and guiding appropriate therapy.

Keywords: DNAJB9, immunotactoid, monotypic, glomerulopathy, fibrillary