Collagen Collisions: Infiltrative diseases of glomerular basement membranes

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Abstract

Although type IV collagen is a normal structural component of a healthy glomerular basement membrane, there are other collagen subtypes which are not normal constituents, specifically collagen type III which is present in the interstitium and vessel walls, but not in glomeruli. There are rare diseases characterized by abnormal glomerular deposition of type III collagen, some of which are associated with a known genetic mutation such as LMX1B in Nail Patella Syndrome, while other diseases remain elusive without evidence of familial or genetic correlation such as in Collagenofibrotic glomerulopathy. Although the two diseases both involve abnormal glomerular type III collagen deposition and may have overlapping histologic features, there are distinct utrastructural features which can aid in differential diagnosis