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Renal Amyloidosis: An update and focus on newly described entities.**Md. Shahrier Amin**

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Amyloidosis is a systemic protein folding disease where insoluble 7-12 nm fibrils having a β -pleated sheet architecture are deposited in the extracellular space in different tissues. Depending on the amyloid precursor protein, there are different types of systemic and organ specific amyloidosis. Differentiating between the different types is crucial, because subsequent management depends on the type and extent of the amyloidosis. When the kidneys are involved, patients often present with proteinuria. Amyloidosis is seen in up to 5% of adult patients with nephrotic syndrome. Proper interpretation of findings on a kidney biopsy is crucial. Pathologic diagnosis requires special stains, immunofluorescence and electron microscopy. Mass spectrometry is sometimes necessary for definite characterization of some rare types of amyloidosis and is an eligibility criterion for targeted therapy in some clinical trials. Immunoglobulin light chains (AL) and serum amyloid A protein (AA) form the basis of most common forms of amyloidosis, accounting for up to 90% of cases. While AL amyloidosis is often associated with lymphoproliferative disorders, AA amyloidosis is commonly seen with chronic inflammatory disorders including infections. Several other amyloidogenic proteins have recently been described and associated with particular histopathologic features. These include leukocyte chemotactic factor 2 (ALECT2), apolipoprotein A-IV and gelsolin. LECT2 amyloidosis is particularly seen in patients of specific ethnicities. Apolipoprotein A-IV amyloidosis shows a peculiar predilection to deposition in the medulla. This lecture will help to educate the audience about common forms of amyloidosis and gain further insight about newly described entities.

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