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Perspective

IgA vasculitis: A systemic form of IgA nephropathy

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ABOUT THE STUDY

IgA Nephropathy (IgAN), also known as Berger's disease or synpharyngitic glomerulonephritis, is a kidney and immune system disease. It is a type of glomerulonephritis, or inflammation of the kidney's glomeruli. Other major organs, such as the liver, skin, and heart, can be attacked by aggressive Berger's disease. The NORD list of rare diseases includes aggressive Berger's disease. Primary IgA nephropathy is distinguished by IgA antibody deposition in the glomerulus. Other diseases associated with glomerular IgA deposits include IgA vasculitis (formerly known as Henoch-Schönlein Purpura), which is widely regarded as a systemic form of IgA nephropathy. IgA vasculitis is more common in children and is characterised by a purpuric skin rash, arthritis, and abdominal pain. HSP nephropathy has a better prognosis than IgA nephropathy. Non-aggressive IgA nephropathy has traditionally progressed slowly to chronic kidney failure in 25-30% of cases over a 20-year period.

Pathophysiology

The disease is named after granular deposits of immunoglobulin A (IgA) in the mesangium (by immunofluorescence), a region of the renal glomerulus. Light microscopy reveals that the mesangium is hypercellular, with increased deposition of extracellular matrix proteins. Although the renal manifestation of Henoch-Schonlein purpura shares the same histological spectrum as IgA nephropathy, a higher frequency of severe lesions such as glomerular necrosis and crescents was observed. HSP nephritis has a higher frequency of glomerular fibrin staining than IgAN, but otherwise has a similar immunofluorescence profile. There is no clear explanation for the IgA accumulation. Exogenous IgA antigens have not been found in the kidney, but it is possible that this antigen is cleared before the disease manifests. It has also been suggested that IgA is the antigen. A recently advanced theory focuses on IgA1 molecule abnormalities. IgA1 is one of two immunoglobulin subclasses (the other is IgD) that are O-glycosylated on several serine and threonine residues in a proline-rich hinge region. IgA polymerisation appears to be caused by abnormal glycosylation of IgA molecules in tissues, particularly the glomerular mesangium. A similar mechanism has been proposed to explain Henoch-Schonlein purpura, a vasculitis that primarily affects children and can cause renal involvement that is nearly identical to IgA nephritis. Human studies, however, have discovered that degalactosylation of IgA1 occurs only in patients with IgA nephropathy in response to gut antigen exposures and to a lesser extent in healthy people. This strongly suggests that IgA1 degalactosylation is a byproduct of an underlying phenomenon (abnormal mucosal antigen handling) rather than the primary cause of IgA nephropathy. Human studies, on the other hand, have discovered that degalactosylation of IgA1 occurs only in IgA nephropathy patients in response to gut antigen exposures, and to a lesser extent in healthy people. This strongly suggests that IgA1 degalactosylation is a byproduct of a more fundamental phenomenon (abnormal mucosal antigen handling) rather than the primary cause of IgA nephropathy. Surprisingly, the IgA1 that accumulates in the kidney appears to come from the bone marrow rather than the Mucosa-Associated Lymphoid Tissue (MALT), which is the site of most upper respiratory tract infections. This, too, suggests an immune pathology rather than direct external interference.

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