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Crescentic glomerulonephritis

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Extracapillary proliferative glomerulonephritis (GN) or crescentic GN (CGN) is not a specific disease, but a histologic manifestation of severe glomerular damage. CGN refers to a case of crescent formation (usually more than 50%) in a wide range of glomeruli and is often characterized by rapid progressive glomerulonephritis (RPGN), which rapidly decreases renal function. Crescent is defined as the presence of at least two layers of extra-capillary cell proliferation fill Bowman spaces completely or partially (25% or 10%). Crescent is classified into cellular crescent, fibrocellular crescent, and fibrous crescent depending on the ratio of cells and matrix, and it is known to develop from cellular crescent to fibrous crescent.
Crescent is composed of various proportions of epithelial cells proliferating along Bowman capsule and mononuclear cells infiltrating this site. In the early stages of crescent, the proportion of mononuclear cells is relatively high and epithelial cells predominate over time. In the fibroepithelial phases, various inflammatory cells including lymphocytes and fibroblasts are observed. In the crescent, mononuclear cells are usually observed more frequently in the case of segmental necrosis or Bowman capsule rupture and epithelial cells are more frequently observed in case of immune-complex mediated glomerulonephritis.

It is known that when the glomerular capillary basement membrane is ruptured, the various blood components and fibrinogen are introduced into the Bowman's space and start to be formed the crescent. As a result of the activation of the coagulation mechanism, fibrin is produced in the fibrinogen and inflammatory cells such as macrophages infiltrate to phagocyte the fibrin into the Bowman capsule and fibrin. Plasma proteins and cytokines stimulate epithelial cells to cause proliferation of epithelial cells and rupture of Bowman capsules occurs. Fibroblasts enter the Bowman's space in the interstitial tissue around the glomerulus through the gap of the ruptured Bowman capsule, and extracapillary cell proliferation occurs. In these cells, matrix proteins such as collagenous and non - collagenous proteins are increased and deposited in tissues. The crescent may evolve to fibrous crescent or be disappeared by apoptosis.
CGN can be primary or secondary. Primary crescentic glomerulonephritis is classified into five type. Type 1 is anti-GBM glomeruolonephritis, characterized by linear deposition of IgG along the glomerular basement membrane which is caused by the formation of autoantibodies against collagen in the glomerular basement membrane. Type 2 is immune-complex mediated glomerulonephritis and representative examples of type 2 CGN are lupus nephritis, Henoch-Schönlein purpura nephritis, IgA nephropathy and membranous proliferative glomerulonephritis, type 1. Immunofluorescence studies show granular immune-complex deposition in the glomerulus. Type 3 is kidney-limited or systemic small vessel vasculitis, which is deposition of immunoglobulin or complement is absent or few (pauci-immune) and antineutrophil cytoplasmic antibody (ANCA) is positive. ANCA-related glomerulonephritis, granulomatosis with polyangiitis or microscopic polyangitis are the representative examples. Type 4 is a combination of type 1 and type 3, and type 5 is ANCA-negative, pauci-immune renal vasculitis (5% to 10% of cases).
In this lecture, it will be briefly introduced the etiology, epidemiology and pathophysiology, and summarized the pathologic findings of CGN.