Rapidly Progressive Glomerulonephritis

Rapidly progressive glomerulonephritis (RPGN) is a type of nephritic syndrome associated with severe glomerular injury with rapid and progressive loss of renal function. This form of glomerulonephritis typically presents with severe oliguria and other signs of nephritic syndrome, including hematuria, hypertension, and mild proteinuria. If untreated, this form can cause death from renal failure within several weeks to months. This disease is commonly called crescentic glomerulonephritis due to the most common histologic picture with the presence of crescents in most glomeruli, caused by the proliferation of the parietal epithelial cells that line Bowman's capsule along with the infiltration of fibrin and plasma proteins like C3b. Monocytes and macrophages are also known to infiltrate, causing crescent formation. In cases of rapidly progressive glomerulonephritis, immunofluorescence helps differentiate between various causes based on the deposition pattern of those complexes in the glomeruli. These patterns include linear, negative, and granular immunofluorescence. RPGN can be caused by several different diseases, including Goodpasture syndrome, granulomatosis with polyangiitis (formerly Wegener's), and microscopic polyangiitis. While some forms respond to plasmapheresis, steroids, and cytotoxic agents, some patients eventually require chronic dialysis or transplantation despite therapy.

PLAY PICMONIC

Pathogenesis

Nephritic Syndrome

Nerd-cricket

Nephritic syndrome is characterized by inflammation of the glomeruli and is a set of symptoms that includes hematuria, hypertension, oliguria, and less than 3.5 grams per day of proteinuria. Patients with nephritic syndrome also commonly present with red cell casts in the urine and azotemia. Nephritic syndrome can be caused by several diseases, including Berger's disease, poststreptococcal glomerulonephritis, and rapidly progressive glomerulonephritis.

Crescent Shape

Crescent

Crescents are formed by proliferation of parietal cells and by migration of monocytes and macrophages. Fibrin strands are frequently prominent.

Crescents Consist of Fibrin and C3b

Crescent Consists of Fiber and Cat (3) Tree (b) bee

Crescents consist of fibrin and plasma proteins, including C3b. The fibrin is a product of blood clotting and hemorrhage, while the plasma protein C3b is a component of the complement cascade responsible for the inflammatory response. Crescents are formed by proliferation of parietal cells and by migration of monocytes and macrophages.

With Parietal Cells, Monocytes, and Macrophages

Pirate Cell, Monkey-monocyte, and Mac-men

The disease is commonly called crescentic glomerulonephritis due to the most common histologic picture with the presence of crescents in most glomeruli, caused by proliferation of the parietal epithelial cells that line Bowman's capsule along with the infiltration of fibrin and plasma proteins like C3b. Monocytes and macrophages are also known to infiltrate, causing crescent formation.

IMMUNOFLUORESCENCE

Linear Immunofluorescence

Linear In-moon-fluorescent

A linear pattern on immunofluorescence is due to antibodies attacking the glomerular basement membrane (GBM), as seen in Goodpasture syndrome. It is a type II hypersensitivity reaction involving antibodies against both the GBM and alveolar basement membrane, leading to hematuria and hemoptysis.

Negative Immunofluorescence

Negative In-moon-fluorescent

A pauci-immune subtype is identified by negative immunofluorescence. This negative immunofluorescence is due to the absence of immunoglobulin or C3 (IgC3) deposition. This subtype is associated with vasculitides such as granulomatosis with polyangiitis and microscopic polyangiitis.

Granular Immunofluorescence

Grain In-moon-fluorescent

This immunofluorescence pattern is commonly observed in post-streptococcal glomerulonephritis and diffuse proliferative glomerulonephritis.

Etiology

Goodpasture Syndrome

Gold-pastor

Goodpasture syndrome is a cause of rapidly progressive crescentic glomerulonephritis. Goodpasture syndrome is characterized by the presence of anti-glomerular basement membrane antibodies (GBM) that cross-react with pulmonary alveolar basement membranes to produce a clinical picture of pulmonary hemorrhage associated with renal failure. The Goodpasture antigen is a peptide within type IV collagen.

Granulomatosis with Polyangiitis (Formerly Wegener's)

Granny-llama with Polly-angel

Granulomatosis with polyangiitis is a small and medium vessel necrotizing vasculitis that typically affects the nose, lungs, and kidneys. Kidney involvement can cause a rapidly progressive glomerulonephritic pattern with crescent formation.

Microscopic Polyangiitis

Microscope Polly-angel

Microscopic Polyangiitis is an autoimmune small vessel vasculitis affecting multiple organs in the body. The small vessel inflammation leads to disease manifestation in the brain, lungs, intestines, and kidneys. Kidney involvement can lead to a rapidly progressive glomerulonephritic pattern with crescent formation.

Prognosis

Poor Prognosis

Gravestone

This form of glomerulonephritis typically presents with severe oliguria and other signs of nephritic syndrome, including hematuria, hypertension, and mild proteinuria. If untreated, this form can cause death from renal failure within several weeks to months. While some forms of this disease respond to plasmapheresis, steroids, and cytotoxic agents, some patients eventually require chronic dialysis or transplantation despite therapy.