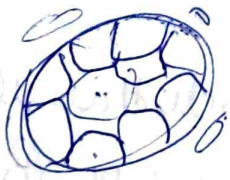
NEPHRITIC SYNDROME

Manifestations

1. Hematuria - red blood cells & red cell casts in urine.
2. oliguria.
3. Hypertension
4. Azotemia - elevated BUN & creatinine levels
5. Proteinuria - mild <1g /24 hour sample.

→ Acute proliferative glomerulo nephritis.



→ PSGN.
→ RPGN
→ goodpasture.
→ Non streptococcal GN.

Post streptococcal Glomerulo nephritis.

age: 6-10.

Etiology

Streptococcal infection of —1-4 week (latent period)→ PSGN.
- pharyngitis
- impetigo (skin)

- infection by →
→ Streptococcus pyogene Group A β Hemolytic strain 12, 4, 1
- Incubation – 1-4 weeks

CF

sore throat —10-14 days→
- cola coloured urine.
- malaise, fever, oliguria.
- periorbital edema.

Pathogenesis

Infection of pharynx or skin by nephritogenic strains by β hemolytic S.C grp A. 12, 4, 1 of

↓

planting of streptococcal antigen from circulation in Subendothelial region of Glomerular Basement membrane

(Spe B. Napi) → MC

Specific antibodies react with Streptococcal antigens in the Subendothelial region and forms in situ Immune Complexes.

↓

Immune complexes initiate an inflammatory response by activating complement and Other mediators of Inflammation.

Inf. mediators – attract neutrophils and monocytes and stimulate proliferation of mesangial & endothelial cells.

↓

IC detaches and migrate across GBM. rebuild to form IC on subepithelial side of the GBM.

↓

Glomerular damage with glomerulo nephritis.

↓

Nephritic syndrome.

if u have time, draw figure S.C

Morphology

figure

GROSS

- enlarged kidney, pale capsular surface & cortex.
- Flea Bitten appearance – petechial haemorrhage

Microscopy → LM, IF, EM.

figure

LM: enlarged hypercellular Glomeruli (G)

FIGURE SC

- hypercellularity → ① proliferation & swelling of endothelial cells, mesangial cells.
② neutrophils, monocytes – infiltration
→ severe case → crescent formation.
→ global & diffuse involvement.

Tubules: red cell (casts) in lumen, degenerative changes in tubular epith cells.

Interstitium: edema & inflammatory cells

Electron microscopy

✓ figure

electron dense subepithelial deposits on the epithelial side of GBM.

↓

Subepithelial Humps.

Immuno fluorescence microscopy.

- Granular deposits of IgG, C3, IgM in mesangium and along GBM.

↓

Granular fluorescence.

"starry sky appearance".

Non-streptococcal Acute GN (post infectious GN).

Bacteria - staphylococcal endocarditis. pneumococcal pneumonia

virus - hep B, C, HIV, mumps.

parasite - malaria.

Lab findings of PSGN ★

- ↑ ASO antibody
- hypocomplementemia ★.
- ↑ blood urea & creatinine.

Rapid Progressive (Crescentic) glomerulonephritis

RPGN is a clinical syndrome characterized by (acute renal failure).

1. rapid & progressive loss of renal function,
2. features of nephritic syndrome,
3. severe oliguria.

↓

If not treated → death in weeks-months due to renal failure.

- Crescentic GN.

Pathogenesis

- caused by primary glomerular diseases as well as systemic diseases.

→ most are Immunologically mediated.

Classification of RPGN

Type I - Anti GBM antibody

→ Goodpasture syndrome.

Type II - Immune complex deposition.

→ PSGN.
→ lupus nephritis.
→ Henoch-Schönlein purpura.
→ IgA nephropathy.

Type III - Pauci immune.

1. ANCA associated
2. Idiopathic
3. Vasculitis associated.
   - wegners granulomatosis
   - mc polyangiitis.

RPGN

definition →

pathogenesis →

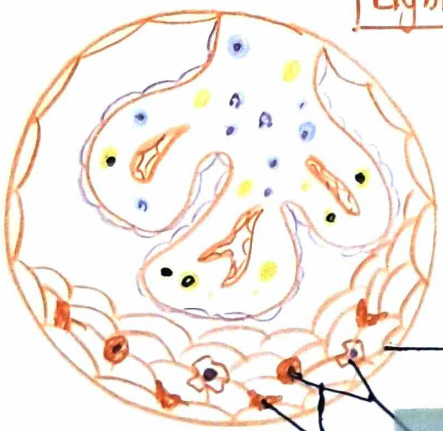
classification

Type I – Anti GBM antibody	Type II : Immune complex	Type III : Pauci Immune
Goodpasture Syndrome.	• PSGN • Lupus nephritis • IgA nephropathy • Henoch-schonlein purpura.	ANCA associated idiopathic Wegner's Granulomatosis
Immunofluorescence m/c ↓ linear GBM fluorescence. ↓ IgG, C_3 deposits.	IF: granular deposits ↓ Starry sky fluorescence IgG, C_3, IgM.	• No anti GBM ab • no Ic deposits in. IF & EM.

GROSS : large, pale kidney [large white K]
flea bitten (may be)

MICROSCOPY – ✓ figure.

Light MC



① Glomeruli

• crescents – parietal epithelial cells
WBC
Fibrin

↓

Indicate severe glomerular injury.

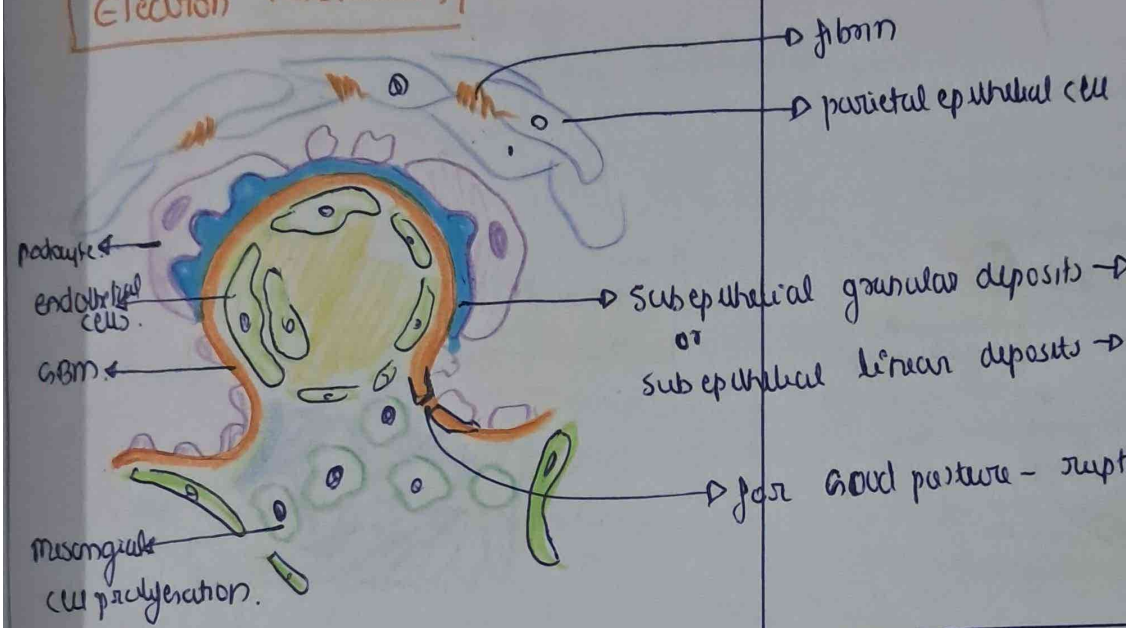
- crescents in most of glomeruli.
- parietal epithelial cells
- monocyte, macrophage infiltrate
- Fibrin.

② Tubules : tubular lumen may show RBC casts & red cells.

③ Interstitium : edema with few inflammatory cells.

don't draw too Many!

Electron Microscopy



Causes of acute Nephritic Sx

I. Primary Glomerulonephritis.

1. acute Glomerulonephritis.
   - streptococcal.
   - non streptococcal.
2. Rapidly progressive GN
3. Membranoso proliferative GN.
4. focus & diffuse proliferative GN
5. IgA nephropathy.

II. systemic diseases.

1. SLE
2. polyarteritis nodosa.
3. wegeners
4. HSP.
5.

Morphology

In other Book

GROSS ✗

- enlarged, pale kidney {large white kidney}
- may show flea bitten appearance.

Microscopy

Glomeruli → Crescents in most of glomeruli.

Composition: Fibrin
Leukocytes
parietal epithelial cells

Tubules - casts

Interstitium - edema + Inflammatory cells

IF

- GIPS → linear GBM fluorescence
- Type II → granular fluorescence.
- type III → little/no deposits.

EM

- Break in GBM.
- Type II - IC deposits.

GIPS

predisposition in HLA DRB1

- Type I RPGN.
- Type II Hypersensitive reaction.
- death due to uremia m/c.

Pathogenesis

Altered α_3 chain of Type IV collagen in GBM → Goodpasture antigen

↓

anti-GBM antibodies

↓

- also cross react with pulmonary alveolar BM.

↓

pulmonary hemorrhage, hemoptysis.

IF

- linear GBM fluorescence.
- crescents.

GROSS ✓

MICROSCOPY ✓ figure (EM) & (LM)

IF → linear deposits

↓

linear GBM fluorescence due to IgG & C_3.

EM

- Rupture in GBM.

CF

- recurrent hemoptysis, life threatening pulmonary hemorrhage.
- uremia is cause of death.