2) NEPHRITIC SYNDROME

- Due to damage of glomerulus, large protein leaks through.

↓

Microscopic Haematuria (Cola.-Colored urine).

↓

Mild Proteinuria

↓

Oliguria → (low urine production)

↓

Mild Odema (Peripheral, Periorbital).

↓

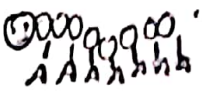
Azoetomia), Hypertension.

Causes:

i) Post streptococcal glomerulonephritis
ii) IgA nephropathy
iii) Other postinfective disease.
iv) Infective Endocarditis.
v) Visceral abscess
vi) SLE
vii) Shunt nephritis

1) POSTSTREPTOCOCCAL GLOMERULONEPHRITIS

- Caused by Group A β-Hemolytic streptococci.



Specific antigen on surface Group A

↓

Streptolysin

↓

Complete destruction of RBC : (β)

(α) is partly damaged.

Causes:

- Usually affects children
- 6 weeks after skin infection - Impetigo.
- 1-2 weeks after Throat infection - pharyngitis.
- Usually resolves in 1 month.

Pathogenesis:

Streptococci → Type III hypersensitivity (Immune complexes)

↓

Deposits in GBM & Subepithelial space

↓

Inflammation & destruction due to

(C_3 complement, cytokines, oxidants, proteases)

↓

Nephritic Syndrome (Haematuria, proteinuria, odema, oliguria, hypertension)

Diagnosis:

① Light Microscopy

↓

Enlarged Hypercellular

② Electron microscopy:

- Subepithelial deposits.
- "HUMPS"

③ Immunofluorescence

↓

"Starry Sky" Granular.

④ Antibody against strep → Anti DNase B, ↑ ASO titre.

Treatment → Supportive

Complication: Rapidly progressive GN → Renal failure

ii) Ig A Nephropathy:

Also called Berger's disease.

- Abnormal Ig A formed and deposited in the kidney glomerular mesangium.

Ig A, two — Ig A_1 → Milk, Tears, Salaive
Ig A_2 → Mucus (GIT, RS, UT).

- There is abnormalities in the Glycosylation of Ig A result in ↓ Ig A_1 clearance.

↓
Body Produce Ig G antibody (Anti-Glycan antibody)
↓
Immune Complex (Deposit Ig A_1 & Ig G)
↓
Travel & Deposit in Mesangium (Type III Hypersensitivity)
(Space between Blood vessels)
↓
Complement system & Inflamatory cytokines.
↓
Damage to glomerular and mesangium
↓
Hematuria (Nephritic syndrome).

C/F :- 1) Presents in childhood.
2) Microscopic or Gross Haematuria
3) Associated during & Infection of Mucosal lining (Synpharyngitic Haematuria) D/D PSGN.
4) Patient present with Haematuria with or without proteinuria

Investigation: 1) Light Microscopy → Mesangial cell Proliferation
2) Electron Microscopy → Immune Deposits
3) Immunofluorescence → Ig A, Immune Complexes

Henoch-Schonlein Purpura (HSP)

- The same findings of IgA Nephropathy are seen, which affects the kidney and
  1) Skin - Rashes in buttocks
  2) Connective Tissue
  3) GIT
  4) Joints, 5) Scrotum
- It is a systemic Vasculitis that arises after infection.

iii) Rapidly Progressive Glomerulonephritis (or) Crescentic Glomerulonephritis.

- Rapid loss of Renal Function causing acute nephritic syndrome with Crescent shaped proliferation.

3 types

1) Type I Anti GBM Disease / Goodpasture syndrome

- It is a autoimmune disease that affects the lungs and the kidneys causing haemoptysis and haematuria.

→ It affects the Basement membrane (made up of protein (collagen) → Triple Helix structure. Each chain is 1 of 6 types (α_1 to α_6). Type IV collagen is usually affected. of α_3 chain ($\alpha_3, \alpha_4, \alpha_5$)

Pathogenesis

Autoantibodies deposit in α_3 chain (Type II hypersensitive reaction)
↳ IgG1
↓
Damage to BM, Endothelium & Capsule. ← Activate Complement system

Risk Factors

Genetic → HLA DR15

Environmental → (Injection, Smoking, Oxidative stress, Hydrocarbons Based solvents).

C/F: → Lungs are affected first.

Lungs → Hemoptysis, Cough, (Restrictive Lung Disease). (Alveoli)

Kidney → Haematuria, Proteinuria, (Nephritic syndrome)

2) Type II

- Immune Complexes ~~are~~ mediated with
  1) Post Streptococcal GN
  2) IgA Nephropathy
  3) Henoch-Schonlein purpura
  4) Lupus Nephritis (Person with SLE @, 40% Suffer with this) So always tested for this.

↓ Types → Mesangial Lupus
Focal Lupus
Segmental Diffuse Lupus
Membranous Lupus.

3) Type III

- Pauci-Immune → No Anti-GBM
  No Immune complexes.

- ANCA in Blood

Anti Neutrophilic cytoplasmic Antibody. → cANCA, pANCA

C ANCA → Associated with Wegner's Granulomatosis
Cytoplasm.

P ANCA → Associated with Microscopic polyangitis.
Perinuclear

Churg-Strauss syndrome
↓
[Granulomatous inflammation.
Asthma, Eosinophillia].

Pathogenesis of RPGN (Crescentic GN)

Complexes → Damage to BM → Leakin Bowman Capsule [plasma protein, Monocytes & Macrophage, Fibrin, Epithelial cell] ↓ Crescentic shaped.

Crescentic shaped. → Haematuria → Nephritic syndrome.

Investigation

Light Microscope → Crescent shaped.

Electron Microscope → Type I → Linear Pattern of Immune Complexes. (Anti GBM)
Type II → "Granular" Patter of Immune Complexes.
Immuno Fluorescens → Type III → No Ant Complexes. ANCA are seen.