NEPHROTIC SYNDROME.

1° ___/___/___

2° → SLE, AR, Amyloidosis

Definition: It is a clinical disorder caused by a derangement in glomerular capillary walls that leads to ↑ed permeability to plasma proteins.

② ⇒ Massive / heavy proteinuria (>3.5g of protein)

① ⇒ Hypoalbuminemia in serum.

⇒ Generalized edema

⇒ Hyperlipidemia And lipiduria

PATHOGENESIS:

Glomerular damage

↓

Altered golmerular permeability And loss of -ve charge. → ↓ Dg

↓ ↑ Permeability of GBM

MASSIVE PROTEINURIA → Loss of transport protein

↓

Hypo Albumenia

↓ catabolism of lipid

↓ vit D

↓ thyroxine

↓ Plasma volume ← ↓ oncotic Pressure

↓ GFR

↑ Aldo (ADH)

Na+ H2O Retention → Edema

Hyperlipidemia

Hyperlipoproteinemia

Lipiduria

__/__/____

① MASSIVE PROTEINURIA :

→ due to ↑ permeability of Glomerular capillary wall to plasma proteins

Highly selective Proteinuria (low molecular weight) protein
- minimal change disease

Non selective Proteinuria (high / large weight proteins)
- membrane nephropathy

② Hypoalbuminemia : Plasma Albumin level less than 3g/dL.
↓ serum Albumin.
↓ decrease the colloid osmotic pressure

③ Generalized edema : most - periorbital Regions
generalised - is called ANASARCA.
CAUSE : ↓ colloid osmotic pressure of the blood.
Na & H2O Retention.

④ Hyperlipidemia & lipiduria
↳ due to ↑ synthesis of lipoprotein
↳ Abnormal transport of circulating lipid
↳ ↓ catabolism of lipid.

Lipiduria ⇒ The leaked lipoproteins are Reabsorbed by tubular epithelial cell
↓
lipid appear in urine either as free fat or fat oval bodies

Nephrotic < 10
20 → SE
BIA /

COMPLICATIONS:

→ Coronary Atherosclerosis
→ Infection → due to loss of Ig in urine
→ Thrombotic And thromboembolic complications.

PRIMARY NEPHROTIC SYNDROME:

① → minimal-change disease ~75%
② → membranous glomerulopathy ~30% (Adult)
③ → focal segmental glomerulosclerosis (FSGS) elderly

MINIMAL CHANGE DISEASE: (Lipoidonephrosis)

MCQ ⇒ • most frequent NS in children

⇒ Characterized by glomeruli that appear normal appearance by light microscopy. (Inflammation / null deposit)

⇒ ONLY UNDER ELECTRON MICROSCOPY: GI shows diffuse effacement (loss) of foot process of visceral epithelial cell (podocytes).

PATHOGENESIS:

due to injury to the visceral epithelial cells.

→ MCD may follow a Resp infection
⇒ ↑ chance of MCD in certain HLA
→ ↑ MCD in Hodgkin Lymphoma
→ 2ndary to use of NSAID.
→ Respond to corticosteroid And other immune suppressive therapy.

__/__/____

Mechanism: circulation molecules injure podocytes
↓
Proteinuria occurs
2nd to injury to foot process effacement.

MORHOLOGY:

→ kidney midly enlarged
→ cut section show pale to yellow cortex.

LIGHT MICROSCOPY: Immunofluorescence = -ve.

Glomeruli: normal by light microscopy.

Tubules: contain lipid vacuoles and glassy protein droplet
↓
hence lipoid nephrosis.

ELECTRON MICROSCOPY:

Characteristic feature is uniform & diffuse effacement of foot process in visceral epithelial cells.

Urine findings: Proteinuria, oval fat bodies, fatty cast in urine.

C.F → MCD may be preceded by upper Respiratory tract infection or immunization.

→ seen in Adults. __/__/____

FOCAL SEGMENTAL GLOMERULOSCLEROSIS:

→ (FSGS) → sclerosis of some glomeruli (focal) that involve only a part of each affected glomerulus.

↙ 1° FSGS ↘ 2° FSGS.

→ 2° FSGS

↳ Maladaptive Response to nephron loss:

↳ type of chronic kidney diseases cause loss of functional Renal mass & may develop FSGS

↳ Infection: occur with HIV.

↳ Drugs: heroin use, α-Actn, TRPC6.

↳ Genetic:

PATHOGENESIS:

: Injury to podocytes:

→ This injury produces a fusion of the foot process And Associated with Increased glomerular permeability.

: Permeability - Inducing factor produced by lymphocytes:

⇒ deposition of hyaline in the glomeruli is due to entrapment of plasma proteins. & lipid in the foci of injury where sclerosis develops.

MORPHOLOGY:

initially involve only the juxtamedullary glomeruli

- The lesion occur in only some tufts within a glomerulus while other tufts appear normal on light microscopy

__/__/____

EM: sclerotic And Nonsclerotic alls show diffuse effacement of foot process of podocytes of epithelial cell

Immunofluorescence Microscopy: show non-specific trapping of Ig usally IgM complement in the alse of hyalinosis.

MICROSCOPIC FEATURES:

(i) Used mesangial matrix
(ii) obliterated capillary lumina.
(iii) deposition of hyaline. of homogenous And eosinophilic material.
(iv) foamy (lipid-laden) macrophages.

Ad stages: sclerosis of the glomeruli involve other glomeruli — Atrophy of tubules fibrosis of the interstitum

⇒ collapsing glomerulopathy: morphologic variant of FSGS characterized by collapse of the glomerular tuft and epithelial cell hyperplasia.

C.F: hematuria And hypertension alls more common in FSGS poor Respon to corticosteroid therapy.

Urine findings: Proteinuria, oval fat bodies, fatty cast