# Kidney (nephrotic syndrome, IgA nephropathy (Berger disease)

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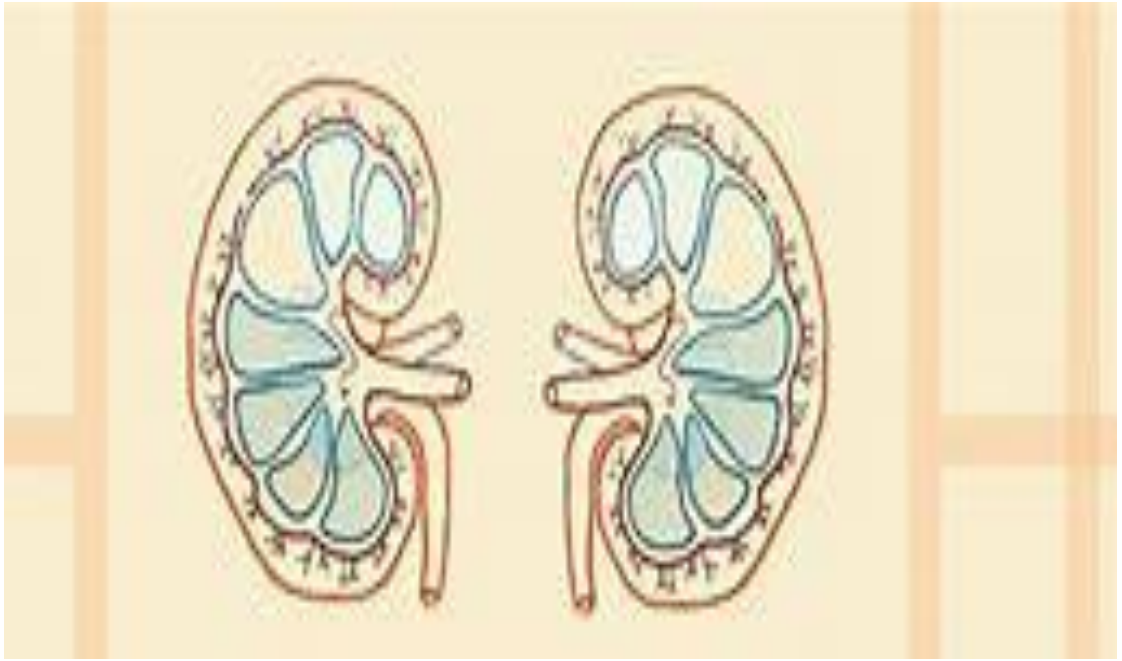

**Histopathology**
المرحلة الرابعة

- **Nephrotic syndrome** is a clinical complex characterized by a number of renal and extrarenal features, most prominent of which are
- Proteinuria (in practice > 3.0 to 3.5gm/24hrs),
- Hypoalbuminemia
- Edema
- Hypertension
- Hyperlipidemia
- Lipiduria

# Etiology

- Idiopathic or Primary
- Secondary.
- Genetic

# Idiopathic

- Mesangial proliferation
- Focal segmental Glomerulosclerosis
- Membranous Nephropathy
- Membrane proliferative glomerulonephritis

- **Mesangial cell proliferation** has been identified as a major factor contributing to glomerulosclerosis, which is a typical symptom of diabetic nephropathy (DN).

**Focal segmental glomerulosclerosis (FSGS)** is a disease in which scar tissue develops on the parts of the kidneys that filter waste out of the blood (glomeruli).

# Membranous nephropathy

- Membranous nephropathy occurs when the small blood vessels in the kidney (glomeruli), which filter wastes from the blood, become inflamed and thickened.

# Membrano proliferative glomerulonephritis (MPGN)

- Membrano proliferative glomerulonephritis (MPGN) is a type of glomerulonephritis caused by deposits in the kidney glomerular mesangium and basement membrane (GBM) thickening and damaging the glomeruli.

# Secondary causes

- Infectious – Hepatitis (B,C), Toxoplasmosis
- Inflammatory –nephritis
- Immunological –Food allergens
- Neoplastic – Lymphoma, Leukemia
- Traumatic ( Drug induced ) – Penicillamine, NSAIDS, Mercury, Lithium

# Genetic causes

- Congenital Nephrotic Syndrome
- Diffuse Mesangial Sclerosis
- Congenital – Oligomeganephronia

# Congenital nephrotic syndrome

- Congenital nephrotic syndrome is a kidney condition that begins in infancy and typically leads to irreversible kidney failure (end-stage renal disease) by early childhood. Children with congenital nephrotic syndrome begin to have symptoms of the condition between birth and 3 months.

# Diffuse Mesangial Sclerosis

Diffuse mesangial sclerosis is a kidney condition that begins early in childhood and rapidly progresses to kidney failure. It is usually associated with other conditions such as Denys-Drash Syndrome

# **Oligomeganephronia** (OMN)

- **Oligomeganephronia** (OMN) is one of rare congenital kidney disease. The number of nephrons reduces and the volume of glomerulus increases.

# PATHOPHYSIOLOGY

- Alteration in glomerular basement membrane
- Altered glomerular protein permeability
- Increased loss of protein in urine
- Decreased colloidal osmotic pressure
- Decreased vascular volume
- Decreased renal blood flow
- Increased secretion of aldosterone
- Na and H2O reabsorption
- Edema

# Nephrotic syndrome clinical features

- Severe swelling (edema), particularly around your eyes and feet
- Foamy urine, a result of excess protein in your urine
- Weight gain due to fluid retention
- Fatigue
- Loss of appetite

# IgA Nephropathy (Berger Disease)

## IgA Nephropathy (Berger Disease)

• Characterized by the presence of prominent IgA deposits in the mesangial regions, detected by immunofluorescence microscopy.

## IgA Nephropathy

• The disease can be suspected by light microscopic examination, but the diagnosis is made only by immunocytochemical techniques

**IgA nephropathy** is typically an isolated renal disease, similar IgA deposits are present in a systemic disorder of children. In addition, secondary IgA nephropathy occurs in patients with liver and intestinal diseases.

# Immunoglobulin A nephropathy occurs as :

1- primary (idiopathic) disease

2- secondary to liver disease (especially alcoholic cirrhosis), and associated with a variety of inflammatory diseases including enteritis inflammatory bowel disease, dermatitis, and HIV infection

**Immunoglobulin A nephropathy probably can result from multiple different etiologies and pathogenic processes, such as :**

(1) Abnormal structure and function of IgA molecules.

(2) Reduced clearance of circulating IgA complexes.

(3) Increased affinity for/or reduced clearance of IgA deposits from the glomerular mesangial.

(4) Excessive IgA antibody production in response to mucosal antigen exposure.

(5) Increased permeability of mucosa to antigen.

# Pathogenesis- Secondary IgA nephropathy

IgA nephropathy occurs with increased frequency in individuals with gluten enteropathy , liver disease, in which there is defective hepatobiliary clearance of IgA complexes (secondary IgA nephropathy).

# Morphology On histologic examination the lesions vary considerably:

1.The glomeruli may be normal

2. mesangial widening

3. focal proliferative glomerulonephritis

4.overt moon glomerulonephritis.

5.The presence of leukocytes within glomerular capillaries is a variable feature.

- Mesangial Proliferation

# Morphology- Immunofluorescence

- The characteristic immunofluorescent picture is of mesangial deposition of IgA, often with C3 and properdin and lesser amounts of IgG or IgM.


THANKS FOR LISTENING