Glomerular Diseases Part 2

Nephrotic Syndrome

It characterized by the presence of:

1. Protienuria more than 3.5 gm/day.
2. Hypoalbuminemia
3. Oedema
4. Hyperlipidemia

Nephrotic Syndrome with bland sediment ((pure Nephrotic))

A. Primary Glomerular Disease:

1. Minimal Change Nephrotic Syndrome: ((MCNS)):

- Sudden onset, commonly in children aged 2 – 6 years.
- Less in adulthood
- Upper respiratory tract infection may precedes the onset of the disease
- Some adult with Hodgkin's disease developed MCNS.
- They are usually normotensive.

Investigations:

- Typical Nephrotic Syndrome
- No active sediment in the urine ((No RBC & Cast)).
- Normal renal function
- Normal serum complement
- Renal biopsy : normal light microscopy but the electron microscopy shows fusion of the foot processes.

Treatment:

- Predenselon 1 – 2 mg/kg/day for 4 weeks then 1 mg/kg/day on alternative days for 4 weeks with tapering over the next 4 – 6 months .
- Those with frequent relapses &/or steroid dependent may get benefit from adjective therapy with Cytotoxic alkylating agents.

2. FOCAL SEGMENTAL GLOMERULOSCLEROSIS ((FSGS)):

- More in adult
- Presented with heavy protienuria
- Hypertension and renal impairment are common
- Serum complement levels are normal.
- FSGS may developed in patients with AIDS, Reflux nephropathy & Heroin abusers.

- Minority respond to steroid therapy, majority progress to CRF, the reminder follow a long term courses with relapses and remissions.

3. MEMBRANOUS GLOMERULOPATHY:

- Mostly in adult
- Normal serum complement
- The majority are idiopathic but can be associated with Syphilis, hepatitis B, Ca-stomach, Ca-lung and drugs eg. Captoprill.
- Usually follow slowly progressive course
- Alternated days of steroid regimen may reduce the development of CRF.
- Cytotoxic agents effect is uncertain.

B. SECONDARY GLOMERULAR DISIEASES:

1. DIABETIC NEPHROPATHY:

- 5 years or more of insulin dependence have passed.
- Clinically apparent usually 15 – 20 years after diagnosis of DM
- Initially the protienuria is minimal and transient so it is called microscopic albuminuria but it will progress to constant moderate to severe protienuria within 2 years.
- Once protienuria become constant, a rapid decline in GFR begins with resultant ESRF within 5 years.
- Hypertension accompany 50% of diabetic nephropathy.
- More than 90% of patients with diabetic nephropathy have also retinopathy while only 1/3 of those with retinopathy have nephropathy.
- A non diabetic aetiology suggested in the absence of retinopathy, diabetes duration less than 10 years and presence of microscopical haematuria with or without RBC cast.
- ESRF associated with boats of hypoglycemia.
- Good glycemic control prevent early diabetic microangiopathy
- Antihypertensive therapy appears to slow the rate of renal deterioration.

2. AMYLOIDOSIS:

Primary Amyloidosis:

- Old age, usually 6th decade of life
- Unexplained spleenomegaly
- Enlarged tongue
- Cardiomegaly
- Malabsorption

Secondary Amyloidosis:

- Younger age
- Developed in patients with multiple myeloma, bronchiectasis, chronic suppuration, chronic infectious diseases and FMF.
- Renal involvement is common in all forms of Amyloidosis.
- Protienuria may present for years prior to diagnosis.
- Onset of nephrotic syndrome or fall in GFR signal a rapid progression to CRF within 3 years.

Diagnosis: It is confirmed by Congo – red positive tissue biopsy

Treatment:

- Usually ineffective except the use of colchicine in FMF.
- Renal transplant has been tried.

Nephrotic Syndrome with active sediment ((mixed nephrotic/nephritic))

A. Primary ((Membranoproliferative GN))

- It is a disease of young people
- Almost always there's concurrent haematuria and protienuria
- Low serum complement
- It is a slowly progressive disease progress to RF over 10 years in 50%
- Alternated days prednislone therapy has therapeutic benefit.
- Dipyridamole & aspirin decrease cthe rate of decline in GFR.

B. Secondary Glomerular Diseases:

1. SLE:

- It is an acute and chronic inflammatory multisystemic disease of unknown etiology.
- Common in female
- Associated with ANA
- Low serum complement
- Renal involvement carry a poor prognosis and can lead to ESRD.
- Treatment include high dose steroid ((methyl predenselon)) ± cytotoxic

2. Henoch – Schonlein Purpura ((HSP)):

- Mostly in children
- It characterized by purpuric lesions on the buttocks and legs
- Episodic abdominal pain
- Joint pain

- Normal complement level
- It is self limited disease but 10% progress to ESRD.

3. Mixed essential cryoglobulinemia:

- Usually in female
- Presented with purpura, fever and Reynaud phenomenon
- Usually progress to ESRD
- Plasmopheresis may improve prognosis.

4. Sickle cell anemia glomerulopathy:

- Microscopical haematuria is common.
- Only less than 5% develop nephrotic syndrome
- It carry poor prognosis and progress to RF quickly.

Complication of nephrotic syndrome:

1. Infection
2. Hypercoagulibility
3. Hyperlipidemia and its sequences
4. oedema and its sequeli.
5. Hyponatermia
6. Complications of treatment ((steroid, Cytotoxic,.....))

Thank You,,,