Hematuria & ACUTE GLOMERULONEPHRITIS

Dr Salma Ajarmeh
Associate Professor of Pediatrics
Consultant Pediatric nephrologist
Mutah University

Hematuria

is defined as the presence of >5 RBCs/HPF in 3 of 3 consecutive centrifuged specimens obtained at least 1 week apart.

Transient vs. persistent

Microscopic vs gross

Asymptomatic vs symptoms

Causes of Hematuria in Children

Glomerular Diseases

IgA nephropathy

Systemic lupus Erythematosus

Membranoproliferative glomerulonephritis

Idiopathic (benign familial) hematuria

Alport syndrome

Rapidly progressive glomerulonephritis

Goodpasture disease

Hemolytic-uremic syndrome

Causes of Hematuria in Children

Infection

Bacterial

Tuberculosis

Viral

Hematologic

Coagulopathies

Thrombocytopenia

Sickle cell disease

Renal vein thrombosis

Stones and Hypercalciuria

Causes of Hematuria in Children

Anatomic Abnormalities

- Congenital anomalies
- Trauma
- Polycystic Kidney

Tumors

Drugs

History taking for AGN

Very detailed hx to ddx the renal from systemic GN and from other non GN

- age , sex
- ***hematuria gross / clots /pain***
- ***color***
- ***timing***
- UOP
- edema
- headache and blurred vision
- Duration of symptoms
- SOB and chest pain
- first episode or recurrent

History

- uti symptoms
-Hx of URTI (when ??)
 -Cough and Sino pulmonary infections
-Hemoptysis / epistaxis
-Joint pain and arthritis
- Ulcers
-skin Rashes
-purpura and abdominal pain
-Trauma / mass
Family hx

Physical Exam

- **Vitals : BP and Pulse**
- **Weight**
- **Chest exam R/O P. Edema and Heart Failure**
- **Edema**
- **Abdomen ..mass / bladder**
- **Skin rash and arthritis**
- **Neuro exam**
- **Fundoscopy**
- **Look for a source of infection**

GLOMERULONEPHRITIS

GLOMERULONEPHRITIS

A group of renal diseases caused by an immunological trigger that mediates inflammation and proliferation of the glomerulus leading to damage of the mesangium, capillary bed and the GBM.

-*acute GN* is the most important and constitutes about 30% of all GN

- *post streptococcal (post infectious) PIGN /PSGN* is the well known type

Pathophysiology

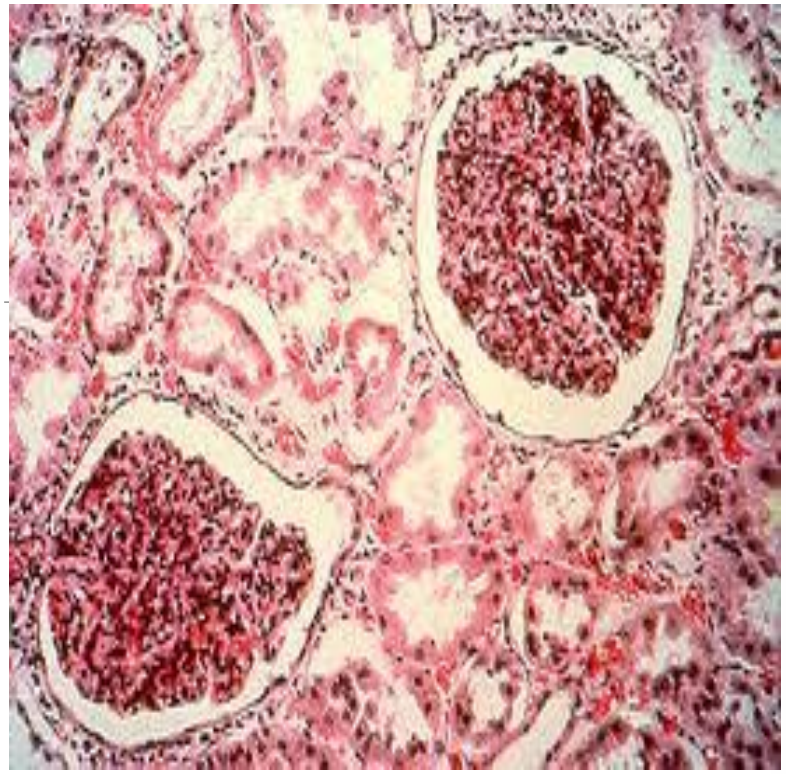
-Results from glomerular deposition of already formed Ig complexes present in serum, **or** formed in sito.

-The kidneys appear enlarged

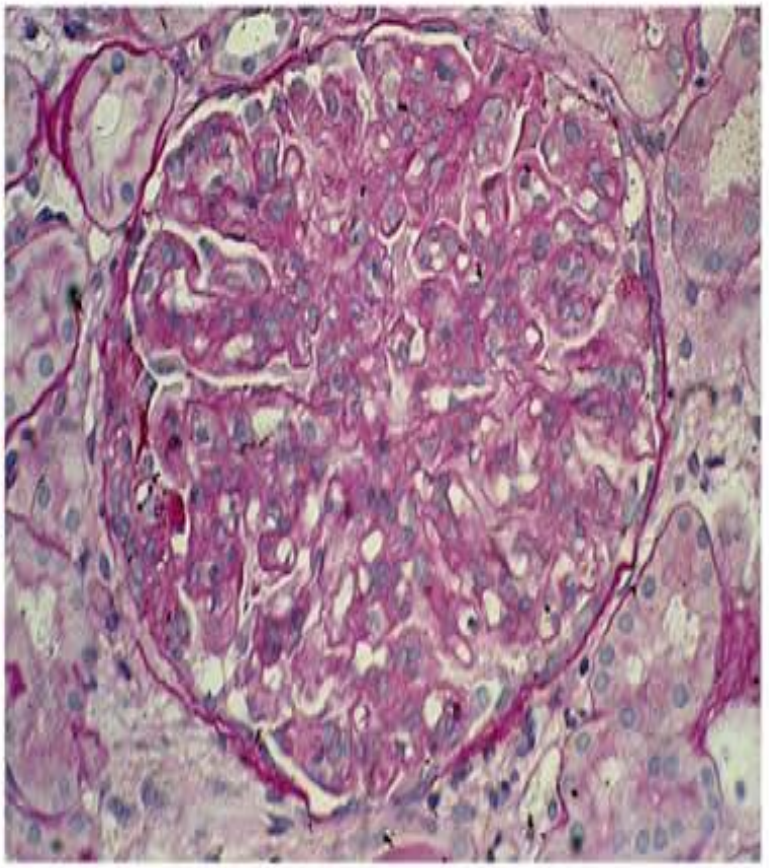
-Swelling of the glomerular tufts and

infiltration with PMN cells

- Deposition of Ig and complements



Acute GN



Clinically ; acute

Nephritic Syndrome

- Hematuria (RBC cast)
- High Blood pressure
- Azotemia (high Cr and BUN) and oliguria
- Edema

Glomerulonephritis is recently classified as

- *Immune-complex glomerulonephritis* (PIGN, IgA nephropathy, lupus nephritis, and cryoglobulinemia)
- *ANCA*–associated (pauci-immune) glomerulonephritis
- Anti-glomerular basement membrane AGBM
- C3 glomerulopathy / MPGN

Glomerulonephritis

classification cont.

- **Systemic vs renal ,,,,,,, idiopathic vs secondary**
- **Nephritic syndrome**
- **Nephrotic syndrome**
- **Nephritic / nephrotic syndrome**

- **Acute vs chronic progressive**

Chronic Nephropathy

Persistent microscopic hematuria

Persistent proteinuria

Chronic renal failure

- **Crescentic /Rapid Progressive (RPGN)**

HSP / MPGN / anti GBM and rarely Post infectious GN

Systemic 2ry GN

Lupus nephritis

Henoch-Schonlein nephritis

Wegener

PAN

Goodpasture (Anti GBM)

Familial Mediterranean fever

Drug nephropathy

(e.g D-penicillamin)

Renal GN

- Ig A nephropathy (Burger disease)
- Membranoprolipherative GN / Mesangiocapillary GN

Post Infectious

-Streptococcal PSGN most common ,

-Bacteria :***Staphylococca***l, or mycobacterial.

Rare :*Salmonella and Brucella*

Viral: CMV, EBV, Hep

Rickettsia , fungal have been reported

PSGN

Acute Poststreptococcal Glomerulonephritis

Is the classic example of the acute nephritic syndrome:

- sudden onset of gross hematuria
- edema
- hypertension
- renal insufficiency.

PSGN used to be the most common cause of gross hematuria in children; now IgA nephropathy is most common ..

ETIOLOGY AND EPIDEMIOLOGY of PSGN

follows infection of the *throat* or *skin* with certain "nephritogenic" strains of group A beta-hemolytic streptococci.

Pharynigitis mostly in winter and skin infection in summer

ETIOLOGY AND EPIDEMIOLOGY

Epidemics of nephritis have been described in association with both

throat (serotype 12).

skin (serotype 49) infections.

the disease is most commonly sporadic.

PATHOLOGY.

Crescents and interstitial inflammation may be seen in severe cases..

Immunofluorescence : deposits of immunoglobulin and complement on the glomerular basement membranes (GBMs) and in the mesangium.

On electron microscopy, electron-dense deposits, or "humps," are observed on the epithelial side of the GBM

PATHOGENESIS.

The precise mechanisms by which nephritogenic streptococci induce the disease remain to be determined.

A depression in the serum complement (C3) and deposition of Ig strongly suggest that poststreptococcal glomerulonephritis is mediated by immune complexes.

CLINICAL MANIFESTATIONS

most common in children 5-15 yr

rare before the age of 3 yr.

an acute nephritic syndrome 1-2 wk after an URTI or 3-4 wk after skin streptococcal infection.

Renal involvement may vary from asymptomatic microscopic hematuria with normal renal function to full nephritic presentation or rarely progress to acute renal failure requiring dialysis.

Asymptomatic episodes PSGN> symptomatic episodes 3:1

CLINICAL MANIFESTATIONS

- Nonspecific symptoms such as malaise, lethargy, abdominal or flank pain, and fever are common
- Encephalopathy / heart failure if malignant HTN develop.
- The edema is a result of salt and water retention due to decreased GFR
- Nephrotic syndrome may also occur ..
- The acute phase generally resolves within 2 months after onset,
- Urinary abnormalities may persist for 1 yr.

DIAGNOSIS ; AGN

Urinalysis

- demonstrates red blood cells (RBCs), with **RBC casts**
- Proteinuria.

Cultures

- Throat swab

Confirmation of the diagnosis
Blood tests

- CBC
- ASO titer
- Anti DNAase B level
- The serum C3 level is usually reduced

With normal C4!

Confirmation of the diagnosis

Clinical presentation

clear evidence of invasive streptococcal infection by a throat culture or ASO titre.

Positive throat culture may support the diagnosis or may simply represent the carrier state.

ASO titer rarely rises after streptococcal skin infections.

Anti DNAase titer or streptozyme test are more specific

Renal biopsy

- renal biopsy is *not indicated* in a classical PSGN presentation
- It is important if SLE nephritis or MPGN is suspected
- Acute Renal Failure (RPGN)
- heavy proteinuria &Nephrotic syndrome
- the absence of evidence for streptococcal infection
- the absence of hypocomplementemia**, or**

persistent low C3 and C4 for more than 3 mo after onset …..(SLE) ??

COMPLICATIONS.

are those of acute renal failure and include

volume overload,

heart failure,

hypertension,

hyperkalemia,

hyperphosphatemia,

hypocalcemia,

acidosis,

seizures,

and uremia

TREATMENT

no specific therapy for acute poststreptococcal glomerulonephritis.

the management is that of acute renal failure .

Although a 10-day course of systemic antibiotic therapy, generally with penicillin, is recommended to limit the spread of the nephritogenic organisms, no evidence shows that antibiotic therapy affects the natural history of glomerulonephritis.

TREATMENT

Supportive therapy for HTN, electrolyte disturbances

medications (diuretics, Ca Channel Blockers and angiotensin-converting enzyme inhibitors)

PROGNOSIS

Excellent outcome with Complete recovery occuring in more than 95% .

0-5 % mortality was reported in some studies in the acute stage due to acute renal or cardiac failure and hypertension.

Recurrences are **extremely rare**

Urinary abnormalities may pertsist for one year

C3 level is back to normal by 2-3 months .

PREVENTION

Early systemic antibiotic therapy of streptococcal throat and skin infections does not eliminate the risk of glomerulonephritis.

Family members of patients with acute glomerulonephritis should be cultured for group A beta-hemolytic streptococci and treated if culture positive.

IgA Nephropathy

IgA Nephropathy

Most commonly present with recurrent gross hematuria following URTI (1-2 days post infection)

Other modes of presentations:

-Asymptomatic microscopic hematuria

-Proteinuria

-Nephrotic syndrome

-Acute Nephritis

-Mixed Nephritic/Nephrotic

IgA Nephropathy

Mainly present in the 2nd and 3rd decade

Male: Female 2:1

One of the most common causes of GN in the world

Geographical Incidence : more common in Japan ,France and Italy compared to UK and USA

IgA nephropathy results from a galactose-deficient IgA1 that acts as auto-antigen that triggers the production of glycan-specific autoantibodies and the formation of circulating immune complexes that are deposited in renal mesangium.

Depositions of IgA in the mesangium in the absence of any systemic symptoms ….(Vs. HSP nephritis)

Diagnosis IgA Nephropathy

Definite diagnosis is *renal Biopsy*

Increase serum IgA in < 20% of patients

Normal serum C3 and C4

Renal Biopsy:

LM: focal segmental mesangial proliferation

EM and IF : deposition of IgA and sometimes lesser amount IgG, IgM and C3.

IgA Nephropathy

Prognosis :

30 % of patients will progress to ESRD

The recurrence of hematuria and the persistent microscopic hematuria does not correlate with the prognosis

Poor prognosis:

- Hypertension

-Heavy proteinuria > 1g/L

-Diffuse GN with Crescents on Biopsy

Treatment

Fish oil

Steroids : Prednisone & pulse Methyl Prednisone

ACE inhibitors and ARBs for hypertension and control of proteinuria

Tonsillectomy..

THANK YOU