Kidney in systemic diseases

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- The kidneys may be directly involved in a number of multisystem diseases or secondarily affected by diseases of other organs.
- Involvement may be at a
  - pre-renal
  - renal (glomerular or interstitial) or
  - post-renal level.

DIABETES MELLITUS

- Called as “diabetic nephropathy”
- After 20 years of disease, the chance of nephropathy is 30-40% in type I, and 15-20% in type II.
- Progression into nephrotic syndrome – screening: microalbuminuria
- Hypertension develops
- ESRD, renal replacement therapy (hemodialysis) = poor prognosis

Pathology:
- Diffuse glomerulosclerosis is most common, but nodular glomerulosclerosis is pathognomonic (Kimmelstiel-Wilson disease)
- Kidneys are enlarged – due to cellular hypertrophy and proliferation

Patients with diabetes mellitus are prone to other renal diseases:
- Papillary necrosis
- Chronic interstitial nephritis
- Type IV renal tubular acidosis
- Acute renal failure after the use of contrast material

Slower the progression:
- Strict glycemic control
- Treatment of hypertension
- ACEI and ARB
### HEPATIC-RENAL DISEASE

- IgA nephropathy is more common in patients with liver disease.
- Severe hepatic dysfunction may cause a haemodynamically mediated type of renal failure, hepatorenal syndrome.
- It also predisposes the kidney to develop acute renal failure (acute tubular necrosis) in response to relatively minor insults including bleeding and infection.
- Patients with such severe hepatic failure are often difficult to treat by dialysis and have a poor prognosis.

### PULMONARY-RENAL DISEASE

- The pulmonary-renal syndrome is a dramatic presentation with renal and respiratory failure.
  - Goodpasture’s disease
  - Small-vessel vasculitis

### MALIGNANT DISEASES

**Paraneoplastic effects on the kidneys:**

- Immune reaction
  - Glomerulonephritis, especially membranous nephropathy
  - Systemic vasculitis (rarely), usually ANCA-negative
- Metabolic consequences
  - Hypercalcaemia
  - Uric acid crystal formation in tubules: usually in tumour lysis syndrome
- Remote effects of tumour products
  - Light chains in myeloma, amyloidosis
  - Antibodies in cryoglobulinaemia

### TUBERCULOSIS OF THE KIDNEY AND URINARY TRACT

- Tuberculosis of the kidney is secondary to tuberculosis elsewhere and is the result of blood-borne infection.
- Initially, lesions develop in the renal cortex; these may ulcerate into the renal pelvis and involve the ureters, bladder, epididymis, seminal vesicles and prostate.
- Calcification in the kidney and stricture formation in the ureter are typical.

**Clinical features:**

- Haematuria (sometimes macroscopic)
- Malaise, fever, night sweats, lassitude, weight loss
- loin pain
- associated genital disease
- chronic renal failure as a result of urinary tract obstruction or destruction of kidney tissue.
SICKLE CELL DISEASE

- Renal dysfunction is due to sickling of red blood cells in the renal medulla because of low oxygen tension and hypertonicity
- Congestion and stasis lead to hemorrhage, interstitial inflammation and papillary infarcts, hematuria is common
- Diminished concentration ability – isosthenuria (urine osmolality equal to that of serum) - patients can easily become dehydrated, papillary necrosis occurs as well
- Sickle cell glomerulopathy is less common, but will progress to ESRD. Proteinuria is the primary manifestation.
- Optimal treatment is adequate hydration and control of the sickle cell disease

VASCULAR DISEASES AFFECTING THE KIDNEYS

- Hypertension
- Hypotension/ bilateral cortical necrosis
- Thrombotic diseases/DIC
- Vasculitis
- Systemic Lupus Erythematosus

Hypertension:

- Most common cause of renal failure in older adults
- Diastolic BP 95 mm Hg or higher

Two types of hypertension are recognized:

- Benign Hypertension
  - Chronic, long-standing, most older adults
- Malignant hypertension
  - M>F, Younger age group (<40)
  - Diastolic Pressure >115 mm Hg
  - Headache, retinopathy, ARF, CRF

Hypertension can either be primary or secondary:

- Primary hypertension is usually idiopathic (no known cause) in 95% of cases
- Secondary hypertension can be attributed to:
  - 1- Renal causes: Acute GN, Chronic GN, Renal artery Stenosis, vasculitis... etc.
  - 2- Endocrine causes: Adrenocortical hyperfunction... etc.
  - 3- Vascular diseases (atherosclerosis)

SYSTEMIC VASCULITIS

- Inflammation of the blood vessels
- Mostly autoimmune diseases
- Circulating antibodies
Small-vessel vasculitis

- focal inflammatory glomerulonephritis, usually with focal necrosis, and often causes crescentic changes
- associated with a systemic illness with acute phase response, weight loss and arthralgia
- in some patients causes pulmonary haemorrhage, which can be life-threatening
- in other patients it presents as a kidney-limited disorder, with rapidly deteriorating renal function and crescentic nephritis

The most important causes of this syndrome:
- microscopic polyangiitis
- Wegener's granulomatosis (ear, nose and throat involvement and lung disease)
- they are usually associated with antibodies to neutrophil granule enzymes (ANCA), but these antibodies are non-specific and cannot be relied upon to make the diagnosis so biopsy of the affected tissue may be required.
- Henoch-Schönlein purpura is associated with IgA nephropathy and ANCA are usually absent.
- Vasculitis in other organs may give clues to the underlying systemic disorder and its subtype.

SYSTEMIC LUPUS ERYTHEMATOSUS (SLE)

- Subclinical renal involvement, with low-level haematuria and proteinuria but minimally impaired or normal renal function, is common in SLE.
- Usually this is due to glomerular disease, although serologically and sometimes clinically overlapping syndromes (e.g. mixed connective tissue disorder, Sjögren's syndrome) may cause interstitial nephritis.
- SLE can produce almost any histological pattern of glomerular disease and an accordingly wide range of clinical features, ranging from rapidly progressive glomerulonephritis to nephrotic syndrome.

Diffuse proliferative lupus nephritis

- Typically, patients present with subacute disease and inflammatory features (haematuria, hypertension, variable renal impairment), accompanied by heavy proteinuria that often reaches nephrotic levels.
- In severely affected patients the most common histological pattern is an inflammatory, diffusely proliferative glomerulonephritis with distinct features to suggest lupus.
- Controlled trials have shown that the risk of ESRF in this type of disease is significantly reduced by cyclophosphamide treatment, often given as regular intravenous pulses.

Thrombotic Diseases that affect the kidney:

- DIC
- Thrombotic Thrombocytopenic Purpura (TTP)
- Hemolytic Uraemic Syndrome (HUS): E. coli poisoning, metastatic breast cancer, drugs, oral contraceptives