Definition: Inflammation with necrosis of blood vessels resulting in variable clinical presentations depending upon:

1. **size** of vessel
2. **type** of vessel
3. **histological** infiltrate

Pathophysiology:

Antigen-Antibody Complex Formation → Immune Complex Deposition → Complement Derived Chemotaxis → PMN Lysosomal Enzyme Release → Vessel Wall Injury, Necrosis → VASCULITIS

I. Vasculitis - Types

A. **Hypersensitivity Group** (Leukocytoclastic)

1. Vessel type: small, usually post capillary venules
2. Symptoms: Skin-Palpable Purpura
   a. vesicles, bullae, ulceration
   b. urticaria (collagen vascular disease)
3. Systemic:
a  Joints--arthritis (50%)
b  Renal--hematuria
c  Other--GI tract

4. Etiology:
   a  Infectious (Strep, viral, Rickettsial)
   b  Drug Reaction
c  Collagen vascular disease
d  Cryoglobulinemia
e  Internal Malignancy
f  Henoch-Schonlein Purpura
   i  Palpable purpura (dependent areas-legs and buttocks)
   ii  Abdominal pain
   iii  Arthritis
   iv  Hematuria
   v  Increased IgA
g  Idiopathic

5. Laboratory work-up
   a  CBC, Platelets
   b  Urinalysis, BUN, Creatinine
   c  Chest x-ray
d  ANA
e  Cryoglobulins
f  Serum Protein Electrophoresis
g  ESR (nonspecific)

6. Differential Diagnosis:
   a  Thrombocytopenia
   b  Sepsis

7. Treatment
   a  removal of offending agent
   b  steroids
   c  dapsone

B. Polyarteritis Group (Periarteritis)

1. Polyarteritis Nodosa
   a  Vasculitis of small-medium sized arteries with segmental (skip)
      involvement.
   b  Symptoms:
      i  Constitutional
      ii  Renal--hypertension, glomerulonephritis
      iii  Neuromuscular--arthritis, myalgia, sensorimotor neuropathy
           (mononeuritis multiplex)
      iv  Gastrointestinal--functional
      v  Cutaneous--Livedo reticularis, Nodules (painful shiny red nodules
         in crops usually on lower extremities), Ulcerations
      vi  Other--cardiac, CNS
   c  Laboratory Work-Up: Nonspecific
   d  Treatment:
i Steroids (50% 5 yr. survival)

ii Cyclophosphamide (80% 5 yr. survival)

iii None (10% 50 year survival)

2. **Churg-Strauss**

   a Allergic granulomatous angiitis

   b Asthma, Eosinophilia

   c Pulmonary vessel involvement--100%

   d Splenic involvement--70%

C. **Granulomatous**

1. **Wegener's Granulomatosis**

   a Granulomatous vasculitis of upper and lower respiratory tract, glomerulonephritis, and other small arteries

   b Symptoms:

      i Respiratory Tract--100%

      ii sinusitis, rhinitis, septal destruction, cough, dyspnea

      iii Renal--80% glomerulonephritis

      iv Cutaneous--40-50% nodules, ulcers

      v Other--joints, CNS, cardiac

   c Laboratory work-up

      i ESR markedly elevated

      ii Chest x-ray (cavitary lesions, nodules)

   d Treatment:

      i 90% 2 yr. mortality without treatment
cyclophosphamide and steroids

2. Giant Cell Arteritis
   a. Temporal Arteritis (Polymyalgia Rheumatica)
   b. Giant cell infiltrate of large arteries (temporal, carotid)
   c. Severe, disabling musculoskeletal pain,
   d. Blindness without immediate steroids in high doses
   e. ESR extremely elevated (over 100)

II. SYSTEMIC DISEASE

A. Pruritus
   1. Thyroid (hyper- or hypo) TSH, T4
   2. Renal disease BUN, Creatinine
   3. Hepatic disease LFT's
   4. Malignancy (Hodgkins Disease) Chest x-ray
   5. Polycythemia vera CBC, platelets
   6. Diabetes glucose

B. Endocrine
   1. Adrenal
      a. Addison's Disease
         i. Hypoadrenalism
         ii. generalized bronzing
         iii. dark pigmentation on gums
      b. Cushing's
Hyperadrenalism

moon facies

buffalo hump

hirsutism

acanthosis nigracans

2. Thyroid

a. hyperthyroidism—pretibial myxedema, thyroid acropachy

b. hypothyroidism—myxedema, alopecia, xerosis, macroglossia

3. Diabetes Mellitus

a. Necrobiosis Lipoidica Diabeticorum

b. Diabetic Dermopathy

C. Malignancy

1. Pruritus

2. Dermatomyositis

3. Acanthosis Nigracans

4. Acquired Ichthyosis

5. malignant down - lanugo hairs

D. AIDS

1. Oral hairy leukoplakia—EB virus

2. Kaposi’s Sarcoma

3. Sudden, severe Seborrheic Dermatitis

4. Molluscum contagiosum— refractory

5. Unusual or opportunistic infections
6. Bacillary angiomatosis

7. Chronic cutaneous herpes virus infections

8. Lipodystrophy