Systemic Lupus Erythematosus

- A multi-system autoimmune disease
- Etiology unknown
- Most common in women, young to middle-aged
- Highly variable clinical presentation in individual patients
SLE, Systems Involved-(1)

- Skin: photosensitive erythematous eruption
- Joints: inflammatory arthritis, usually symmetrical without joint destruction
- Kidneys: Glomerulonephritis
- Cardiopulmonary: Pleurisy, pericarditis, pneumonia and pulmonary hemorrhage, myocarditis and coronary artery disease
SLE, systems involved-(2)

- Central nervous system: Seizures, psychiatric symptoms, cerebrovascular accidents
- Blood: Anemia, often hemolytic. Leukopenia. Thrombocytopenia
- Fever
SLE: Anti-Nuclear Antibodies

**PRINCIPLE OF INDIRECT IMMUNOFLUORESCENCE**

- Serum Antibody
- Nucleus
- Cytoplasm
- Fluorescent Labeled Anti-Immunoglobulin

Figure by MIT OCW.
SLE: Prognosis and Treatment

- Prognosis varies from mild to severe or fatal.

- Treatment is non-specific, and is not curative.
  - NSAIDs
  - Hydroxychloroquine
  - Glucocorticoids
  - Cytotoxic, immunosuppressive drugs
Diffuse Systemic Sclerosis (Scleroderma)

- A chronic, progressive inflammatory auto-immune disease leading to fibrosis in several organ systems and in the vasculature
- Etiology unknown
- No specific treatment; only palliative measures with limited efficacy.
CLASSIFICATION OF SCLERODERMA

**Systemic Sclerosis (SSc)**
- Diffuse Cutaneous Scleroderma
- Limited Cutaneous Scleroderma
- CREST Syndrome

**Localized Scleroderma**
- Morphea
- Linear Scleroderma

**Overlap Syndromes**
- Scleroderma-Like Syndromes

Figure by MIT OCW.
Idiopathic Inflammatory Myopathy

- Inflammatory myopathy of unknown etiology
- Probably has an autoimmune pathogenesis
- Usually a chronic progressive disease
- Causes proximal skeletal muscle weakness
- This entity and other diseases of muscle will be discussed by Drs. Brown and Johns
POLYMYOSITIS: CLASSIFICATION

- Adult Polymyositis
- Adult Dermatomyositis
- Inflammatory Myositis Associated with Cancer
- Childhood Dermatomyositis or Polymyositis
- Myositis Associated with Connective Tissue Disease

Figure by MIT OCW.
Idiopathic Inflammatory Myositis Diagnosis

- Elevation of serum levels of enzymes intrinsic to skeletal muscle; creatine phosphokinase is the most sensitive and specific. Transaminases also are elevated.
- Myopathic changes on EMG
- Abnormal muscle biopsy
- Evidence of inflammation on MRI
Idiopathic Inflammatory Myositis

Treatment

- **Glucocorticoids:** Relatively high doses required. Toxicity is frequent.
- **Other immunosuppressive agents:** methotrexate, azathioprine
- **Physical therapy**
- **Search for underlying malignancy** where appropriate
Sjogren’s Syndrome
(Keratoconjunctivitis sicca)

- An inflammatory auto-immune disease involving the salivary and lacrimal glands, sometimes other exocrine glands.
- Etiology unknown
- Causes dryness of the eyes and mouth.
- May be associated with other rheumatic diseases, such as rheumatoid arthritis and SLE.
- Treatment is palliative
Vasculitis

- A bewildering array of clinical syndromes with the common feature of necrotizing inflammation of blood vessels.

- The etiology is often unknown, but some infections, e.g., hepatitis C, can cause vasculitis through immune complex deposition.
Vasculitis may be classified on the basis of the size of the arteries involved.

<table>
<thead>
<tr>
<th>Vasculitis Syndrome</th>
<th>Vessel Involved</th>
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</thead>
<tbody>
<tr>
<td>Polyarteritis Nodosa</td>
<td>Small, Medium Arteries</td>
</tr>
<tr>
<td>Churg-Strauss (Allergic Granulomatosis and Angiitis)</td>
<td>Small, Medium Arteries</td>
</tr>
<tr>
<td>Hypersensitivity Vasculitis</td>
<td>Arterioles, Venules, Capillaries, Rarely Small Arteries</td>
</tr>
<tr>
<td>Vasculitis Syndrome</td>
<td>Vessel Involved</td>
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<tr>
<td>Henoch-Schönlein Purpura</td>
<td>Venules, Arterioles, Capillaries</td>
</tr>
<tr>
<td>Takayasu's Arteritis</td>
<td>Medium, Large Arteries</td>
</tr>
<tr>
<td>Temporal Arteritis</td>
<td>Medium, Large Arteries</td>
</tr>
<tr>
<td>Wegener's Granulomatosis</td>
<td>Small Arteries, Veins, Medium Arteries</td>
</tr>
</tbody>
</table>

Figure by MIT OCW.
Vasculitis: Diagnosis

- Recognition of clinical syndromes caused by vasculitis
- Elevated levels of acute phase reactants
- Positive test for anti-neutrophil cytoplasmic antibodies (ANCA), are present in some syndromes
- Biopsies and radiographic studies
Vasculitis: Treatment

- Immunosuppressive therapy with glucocorticoids and cytotoxic or antimetabolic drugs
- Prognosis is guarded, but most syndromes are treated effectively, although cures are not always achieved