A number of vasculitic processes can involve the lungs, we are mostly dealing with GPA/Wegner's, microscopic polyangitis (MPA), and eosinophilic granulomatosis with polyangiitis (EGPA/Churg-Strauss).

### Pulmonary Vasculitis

### Granulomatous with Polyangiitis (GPA, Wegner's) – small vessel vasculitis

#### Extra-Pulmonary Organ Involvement

<table>
<thead>
<tr>
<th>Joints</th>
<th>Nervous system</th>
</tr>
</thead>
<tbody>
<tr>
<td>myalgias, arthralgias, arthritis</td>
<td>mononeuritis, cranial neuropathies, hearing loss, tinnitus, ophthalmoplegia</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Eyes</th>
<th>Cardiac</th>
</tr>
</thead>
<tbody>
<tr>
<td>conjunctivitis, episcleritis, uveitis</td>
<td>pericarditis, myocarditis, conduction system</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Skin</th>
<th>Also- GI, GU, parotid, thyroid, liver, breast</th>
</tr>
</thead>
<tbody>
<tr>
<td>vesicular, purpura; subcutaneous nodules</td>
<td></td>
</tr>
</tbody>
</table>

#### Pulmonary Manifestations

- **Chronic rhinosinusitis** – pain; purulent, bloody discharge; hearing loss, nasal perforation
  - May be present without other manifestations for weeks, months before progressing to vasculitic disease
  - Key difference vs EGPA is that EGPA has nasal polyposis and is less likely to have nasal perforation

- **Tracheobronchial disease** – stenosis, inflammatory pseudotumors, malacia, TE fistula
  - Subglottic stenosis in up to 50% - may be the sole manifestation and require tracheostomy

- **Pulmonary nodules** – cavitation in 25-50%

- **Interstitial disease** – fibrosis and honeycombing – commonly a UIP pattern; ILD most common with MPA

- **Alveolar hemorrhage** – presenting ~25%, commonly associated skin, renal, and neurologic involvement

- **Pulmonary artery stenosis** – rarely, seen on CTA, PA pressures may not be elevated

Over 90% of GPA or MPA with lung and renal involvement are cANCA (PR3) positive. Minority will have pANCA (MPO). Persistently elevated ANCA is associated with relapse.

#### Biopsy

- Biopsy is important because need to exclude infection and metastatic disease affecting the lung
  - In general you want a surgical lung biopsy – not recommended in DAH or ILD.
  - Nasal and sinus biopsies are very low yield- usually do not show vasculitis

#### Treatment

**Induction**: usually cyclophosphamide (more experience) or rituximab (less toxicity); for mild disease without renal involvement- may try MTX

**Maintenance**: usually azathioprine or rituximab, again- if no renal disease may try MTX

#### Eosinophilic Granulomatosis with Polyangitis (EGPA, Churg-Strauss)

Three Phases of disease:

- **Prodromal** – atopy, allergic rhinitis, asthma
- **Eosinophilic** – peripheral eosinophilia, eosinophilic infiltration of lung and GI tract in particular; this is the triad of pulmonary opacities, asthma, and eosinophilia that presents prior to systemic vasculitis (true ~40%)
- **Vasculitic** – medium and small vessels; associated with constitutional symptoms

**EGPA Clinical Manifestations**

<table>
<thead>
<tr>
<th>Asthma – in 90% and precedes vasculitis by 8-10 years</th>
<th>Neurologic – usually mononeuritis – up to 75%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Upper Airway – nasal polyps; looks like aspirin</td>
<td>Renal – common, biopsy shows necrotizing</td>
</tr>
<tr>
<td>exacerbated respiratory disease</td>
<td>glomerulonephritis</td>
</tr>
<tr>
<td>Skin – subcutaneous nodules on extensor surfaces,</td>
<td>GI – abdominal pain, bleeding, diarrhea- may</td>
</tr>
<tr>
<td>biopsy shows granulomas</td>
<td>precede vasculitis</td>
</tr>
<tr>
<td>Cardiac – due to high eosinophilic infiltration</td>
<td>Myalgias and arthritis are less common</td>
</tr>
<tr>
<td>Increased thromboembolic risk</td>
<td>Eosinophilic lymphadenopathy in ~40%</td>
</tr>
</tbody>
</table>

**EGPA Diagnostic Criteria**: need 4 or more of the following (sensitivity 85%, specificity 99%)

<table>
<thead>
<tr>
<th>Asthma</th>
<th>Migratory or transient pulmonary opacities</th>
</tr>
</thead>
<tbody>
<tr>
<td>&gt;10% eosinophils</td>
<td>Paranasal sinus abnormality</td>
</tr>
<tr>
<td>Mononeuropathy or polyneuropathy</td>
<td>Biopsy showing extravascular accumulation of eosinophils</td>
</tr>
</tbody>
</table>