Organizing Pneumonia

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<table>
<thead>
<tr>
<th>Histologic Pattern</th>
<th>Idiopathic Clinical Syndrome</th>
<th>Associated Diseases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Usual interstitial pneumonia</td>
<td>Idiopathic pulmonary fibrosis</td>
<td>Connective tissue disease (CTD), drugs, asbestosis</td>
</tr>
<tr>
<td>Nonspecific interstitial pneumonia (NSIP)</td>
<td>Idiopathic NSIP</td>
<td>CTD, drugs, hypersensitivity pneumonitis (HP)</td>
</tr>
<tr>
<td>Desquamative interstitial pneumonia (DIP)</td>
<td>Idiopathic DIP</td>
<td>Smoking, CTD, drugs, toxic inhalation</td>
</tr>
<tr>
<td>Organizing pneumonia (OP)</td>
<td>Cryptogenic OP</td>
<td>CTD, drugs, infections, chronic eosinophilic pneumonia, HP</td>
</tr>
<tr>
<td>Constrictive bronchiolitis (CB)</td>
<td>Idiopathic CB</td>
<td>Post-viral, CTD, drugs, graft vs. host disease, lung transplant rejection</td>
</tr>
<tr>
<td>Diffuse alveolar damage</td>
<td>Acute interstitial pneumonia</td>
<td>Infection, aspiration, trauma, sepsis, pancreatitis, etc.</td>
</tr>
</tbody>
</table>

Pathology of organizing pneumonia

- Characteristic pattern of injury
- The entity formerly known as BOOP
- Insult to the alveolar wall epithelium
- Healing response originates from respiratory bronchiole
- Granulation plugs within alveolar spaces and respiratory bronchioles
- Inflammation -> fibrosis
- Type III collagen, resorption into interstitium
Organizing pneumonia

- 1. Granulation tissue in alveolar spaces/ducts
- 2. Granulation tissue in respiratory bronchioles
- 3. Mild interstitial infiltrate of lymphocytes and plasma cells
- 4. Isolated or associated with other findings:
  - Fibrosis
  - Hyaline membranes
  - Prominent interstitial inflammation
  - Granulomas

Categories

- Cryptogenic organizing pneumonia
- Secondary organizing pneumonia
- Organizing pneumonia as component of another diffuse lung disease

Organizing pneumonia: causes

<table>
<thead>
<tr>
<th>Idiopathic OP</th>
<th>Secondary OP</th>
<th>OP associated with other diffuse lung disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cryptogenic</td>
<td>Drugs</td>
<td>Infection/aspiration</td>
</tr>
<tr>
<td>Connective tissue disease</td>
<td>Connective tissue disease</td>
<td></td>
</tr>
<tr>
<td>Toxic inhalations</td>
<td>Chronic eosinophilic pneumonia</td>
<td></td>
</tr>
<tr>
<td>Graft vs. host disease</td>
<td>Hypersensitivity pneumonitis</td>
<td></td>
</tr>
<tr>
<td>Lung transplant</td>
<td>Vascularitis</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Diffuse alveolar damage</td>
</tr>
</tbody>
</table>
### Organizing pneumonia: clinical

#### Clinical

<table>
<thead>
<tr>
<th>Organizing pneumonia</th>
<th>Cryptogenic organizing pneumonia</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age</strong></td>
<td>Any (pediatric to geriatric)</td>
</tr>
<tr>
<td><strong>Symptom duration</strong></td>
<td>Acute to chronic</td>
</tr>
<tr>
<td><strong>Extrapulmonary symptoms</strong></td>
<td>Yes or no</td>
</tr>
<tr>
<td><strong>Other</strong></td>
<td>Search for other clinical clues to the diagnosis</td>
</tr>
</tbody>
</table>

#### Diagnosis of COP

- Clinical + radiographic
- Exclusion of other causes of similar HRCT findings
- Exclusion of secondary causes of OP
- Response to steroids
- Biopsy in non-classic cases

#### Organizing pneumonia (treatment)

**King et al. Chest 1992; 102: 85**

**Lazor et al. Am J Respir Crit Care Med 2000; 162: 571**

**Lohr et al. Arch Intern Med 1997; 157: 1323**

- Complete clinical/physiologic resolution in 63%
- Response in days to weeks
- 30% relapse within 1-3 months
- Relapse not associated with increased mortality
- HRCT findings of fibrosis: bad prognostic sign
- Lower survival in 2° OP (44%) vs. 1° OP (73%)
- 6-12 months of prednisone followed by slow taper

#### Organizing pneumonia: radiology

- HRCT by itself is not diagnostic of OP
- Clinical + HRCT may be sufficient for diagnosis
- Radiologist may be the 1st to suggest the diagnosis
- Distinguish OP vs. OP associated with another diffuse lung disease
- Place pathology in context

#### Role of radiology in OP

- HRCT by itself is not diagnostic of OP
- Clinical + HRCT may be sufficient for diagnosis
- Radiologist may be the 1st to suggest the diagnosis
- Distinguish OP vs. OP associated with another diffuse lung disease
- Place pathology in context
### Organizing pneumonia: radiology

<table>
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<tr>
<th>Pathology</th>
<th>Radiology</th>
</tr>
</thead>
<tbody>
<tr>
<td>Granulation tissue in alveolar spaces</td>
<td>Consolidation &gt;&gt; ground glass opacity</td>
</tr>
<tr>
<td>Granulation tissue in respiratory bronchioles</td>
<td>Small centrilobular nodules</td>
</tr>
<tr>
<td>Interstitial inflammation</td>
<td>Ground glass opacity</td>
</tr>
<tr>
<td>Additional findings when associated with another</td>
<td>Significant ground glass opacity, mosaic perfusion/air</td>
</tr>
<tr>
<td>diffuse lung disease</td>
<td>trapping, fibrosis</td>
</tr>
</tbody>
</table>

### Organizing pneumonia

*Bouchardy et al. JCAT 1993; 17: 352*
*Lee et al. AJR 1994; 162: 543*
*Kim et al. AJR 2003; 180: 1251*
*Ujita et al. Radiology 2004; 232: 757*

<table>
<thead>
<tr>
<th>Finding</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Consolidation</td>
<td>80-95%</td>
</tr>
<tr>
<td>Ground glass opacity</td>
<td>60-85%</td>
</tr>
<tr>
<td>Large nodules or masses</td>
<td>40%</td>
</tr>
<tr>
<td>Small nodules</td>
<td>20-50%</td>
</tr>
<tr>
<td>Peribronchovascular sign</td>
<td>60%</td>
</tr>
<tr>
<td>Reversed halo sign</td>
<td>20%</td>
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### Consolidation in OP (amiodarone)

- Bilateral and patchy
- Peribronchovascular and subpleural
- Often rounded
- Irregular borders
- Associated with architectural distortion and bronchiectasis

### Everolimus, organizing pneumonia

### Crack-lung

### Cryptogenic organizing pneumonia
Cryptogenic organizing pneumonia

Organizing pneumonia

OP (immunocompromised)

Finding | Immune compromised | Immune competent
--- | --- | ---
Consolidation | 91% | 45%
Ground glass opacity | 73% | 56%
Nodules | 55% | 22%

Reversed halo sign

- Originally thought to be specific for OP
- Seen in ~20% of COP patients
- May be seen in OP or OP associated with other diffuse lung disease
- Described in many other diseases:
  - Infections (fungal, tuberculosis)
  - Sarcoidosis
  - Granulomatosis with polyangiitis
  - Lymphomatoid granulomatosis

Reversed halo: other causes

- Invasive fungus
- Reticulation inside halo
- Wall thickness >1 cm
- Pleural effusion
- Granulomatous disease
- Nodules inside halo
- Nodular wall

Septic emboli
Perilobular sign
(Ujita et al. Radiology 2004; 232: 757)

- Present in 57% of patients with COP
- Adjacent to pleura in 83% of cases
- No relationship to HRCT signs of fibrosis
- Not yet described outside of COP

OP: serial changes over time
(Lee et al. AJR 2010; 195: 916)

- 27% complete resolution
- 68% improvement
- 5% no change
- Only variable predictive of non-resolution: initially pulmonary function tests
- Post-treatment CT resembled fibrotic NSIP

Amiodarone

OP from inhalational crack cocaine

Initial | 4 months | 9 months

Myositis: NSIP with OP

Initial | 1 year later
OP secondary to another diffuse lung disease

- Significant ground glass opacity
- Mosaic perfusion/air trapping
- Cysts
- Significant fibrosis

Hypersensitivity pneumonitis with OP

HP with organizing pneumonia

Organizing diffuse alveolar damage

Scleroderma: NSIP + LIP + OP

Spectrum of acute lung injury

Beasley et al. Arch Pathol Lab Med 2002; 126:1064
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