CASE 2
Clinical History

- 34 yo woman
- SLE
- Hemoptysis
Venulitis
Diagnosis

INTRA-ALVEOLAR HEMORRHAGE WITH FOCAL NECROTIZING CAPILLARITIS AND DIFFUSE ALVEOLAR DAMAGE (DAD)
Aortic valve
Diagnosis

NON BACTERIAL THROMBOTIC ENDOCARDITIS (NBTE)
Systemic Lupus Erythematosus

- Connective tissue disease
- 9W:1M
- Child-bearing age
- Auto-immune
- Anti-phospholipid syndrome
### Cardiovascular Manifestations

<table>
<thead>
<tr>
<th><strong>Pericardium:</strong></th>
<th><strong>Myocardium:</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Pericarditis</td>
<td>Myocarditis</td>
</tr>
<tr>
<td>Pericardial effusion</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Valves:</strong></th>
<th><strong>Conduction system and arrhythmia:</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>NBTE</strong></td>
<td>Inflammation</td>
</tr>
<tr>
<td>Diffuse fibrosis</td>
<td>Fibrosis</td>
</tr>
<tr>
<td>Infective endocarditis</td>
<td></td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Coronary arteries:</strong></th>
<th><strong>Aorta and peripheral vessels:</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Atherosclerosis</td>
<td>Aortitis</td>
</tr>
<tr>
<td>Vasculitis</td>
<td>Atherosclerosis</td>
</tr>
<tr>
<td>Aneurysm</td>
<td>Thrombosis/embolism</td>
</tr>
<tr>
<td>Thrombosis/embolism</td>
<td>(antiphospholipid syndrome)</td>
</tr>
</tbody>
</table>

**Drug toxicity to:**
- Cyclophosphamide with myocarditis
- Chloroquine with cardiomyopathy
Pulmonary Manifestations

**Pleura:**
- Fibrinous pleuritis with effusion
- Pleural fibrosis

**Parenchyma:**
- NSIP
- BOOP
- DAD
  - Alveolar hemorrhage syndrome
- Amyloidosis
- Lymphoproliferative disorder

**Parenchymal Infections**

**Respiratory Muscle:**
- Shrinking lung syndrome

**Vessels:**
- Acute reversible hypoxemia syndrome
- Pulmonary hypertension
- Vasculitis
- Thromboembolism (APS)

**Upper airway:**
- Epiglottitis
- Hypopharyngeal ulceration
- Subglottic stenosis

**Drug Toxicity to:**
- Cyclophosphamide with DAD, CIP, UIP, BOOP, hemorrhage
- Methotrexate with CIP, HP, BOOP, DAD
Terminology

Non-Infective vegetations

- NBTE=marantic
- Libman-Sacks

OR

NBTE=Libman-Sacks=Marantic

# Pathogenesis

<table>
<thead>
<tr>
<th></th>
<th>PRE-STEROID ERA</th>
<th>POST-STEROID ERA</th>
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<tbody>
<tr>
<td>MV</td>
<td>38%</td>
<td>32%</td>
</tr>
<tr>
<td>AV</td>
<td>29%</td>
<td>19%</td>
</tr>
<tr>
<td>TV</td>
<td>38%</td>
<td>3%</td>
</tr>
<tr>
<td>PV</td>
<td>25%</td>
<td>1%</td>
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<tr>
<td>Chronic</td>
<td>10%</td>
<td>42%</td>
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</table>
Clinical Features

- 13-74%
- Difficult clinical diagnosis
- Complications:
  - Valvular dysfunction 3-25% → Surgery 1-9%
  - Embolization with systemic infarcts 14-91%
NBTE of MV  NBTE of AV
NBTE of MV
Differential Diagnosis

- Infective endocarditis
- Acute rheumatic disease
- Post-inflammatory valvular heart disease
Acute rheumatic fever
Acute Pulmonary Disease

- Acute lupus pneumonitis
- Diffuse alveolar hemorrhage (DAH)
Acute Lupus Pneumonitis

• 1- 4%
• Abrupt onset
  – Fever
  – Dyspnea and cough
• Hypoxemia
• Patchy alveolar infiltrates
Acute Lupus Pneumonitis

• Steroids or immunosuppressants
• Favorable response
• Fatal up to 50%
Differential Diagnosis

- Bronchiolitis obliterans organizing pneumonia (BOOP)
- Non-specific interstitial pneumonia (NSIP)
- Usual interstitial pneumonia (UIP)
BOOP
Diffuse Pulmonary Hemorrhage

- 1-5.4%
- Hemoptysis (42-100%)
- Rapid onset of anemia
- Diffuse alveolar infiltrates
Diffuse Pulmonary Hemorrhage

- Pulmonary-renal syndrome with glomerulonephritis up 94%
- Respiratory failure with death 23-92%
- Immunosuppressants/plasmapheresis
Differential Diagnosis

• Capillaritis
• Fresh blood
• Respiratory bronchiolitis
• Acute necrotizing pneumonia
Capillaritis

Vasculitis
  Wegener granulomatosis
  Microscopic polyangiitis

Connective tissue disease
  Rheumatoid arthritis
  Mixed connective tissue disease

Drugs

Idiopathic/isolated

Other
  Antiphospholipid syndrome
Acute alveolar hemorrhage
Respiratory bronchiolitis
Acute necrotizing pneumonia
Direct Immunofluorescence

- Often unnecessary
- Acute setting
- Appropriate organ
- Linear IgG specific for Goodpasture
- Granular staining IgG, IgM, C1q, C3:
  - not 100% specific
  - not 100% sensitive
<table>
<thead>
<tr>
<th></th>
<th>#</th>
<th>IgG</th>
<th>IgM</th>
<th>IgA</th>
<th>C3</th>
<th>C5b-9</th>
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Goodpasture - IgG
Systemic lupus erythematosus

C3

IgG
Take Home Points

NBTE:

- Common valvular abnormality in SLE
- Increased surgical specimen
- Distinct from infective endocarditis
Take Home Points

**ACUTE LUPUS PNEUMONITIS**
- DAD
- Hyaline membrane
- Rule out infection

**DIFFUSE PULMONARY HEMORRHAGE**
- Capillaritis
- Clinical hx + serology > IF
- Distinct from necrotizing pneumonia
On to Case 3