

# CASE ( M / 73 )

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고대 안암 병원  
호흡기 내과  
R3 서민영

# Chief Complaint

Lt. pleuritic chest pain

( onset : 2개월 전 )

# Present illness

- 2013년 1월 Lt. pleuritic chest pain 발생하여 개인병원에서 시행한 CXR 상 특이 소견 보이지 않아 경과 관찰
- 2013년 3월 중순부터 cough, sputum, febrile sense 발생하고 Lt. pleuritic chest pain 악화되어 내원

# History

- Past medical history
  - HTN/DM/TB/Hepatitis(+ / - / + / - )
  - CVA(+)
- Social history
  - Alcohol : 1회/주, 소주 1병/회
  - Ex-smoker : 50PY (10YA cut)

# Review of system

- General weakness / Fatigue ( + / + )
- Weight loss / Poor oral intake ( + / + )
- Cough / Sputum / Rhinorrhea ( + / + yellowish / - )
- Pleuritic pain/ Dyspnea / Palpitation( + / - / - )
- Fever / Chilling ( - / - )
- Night sweating (+)

# Physical examination

- **Vital Signs**

- 160/80 mmHg – 80/min – 30/min – 36.8 °C

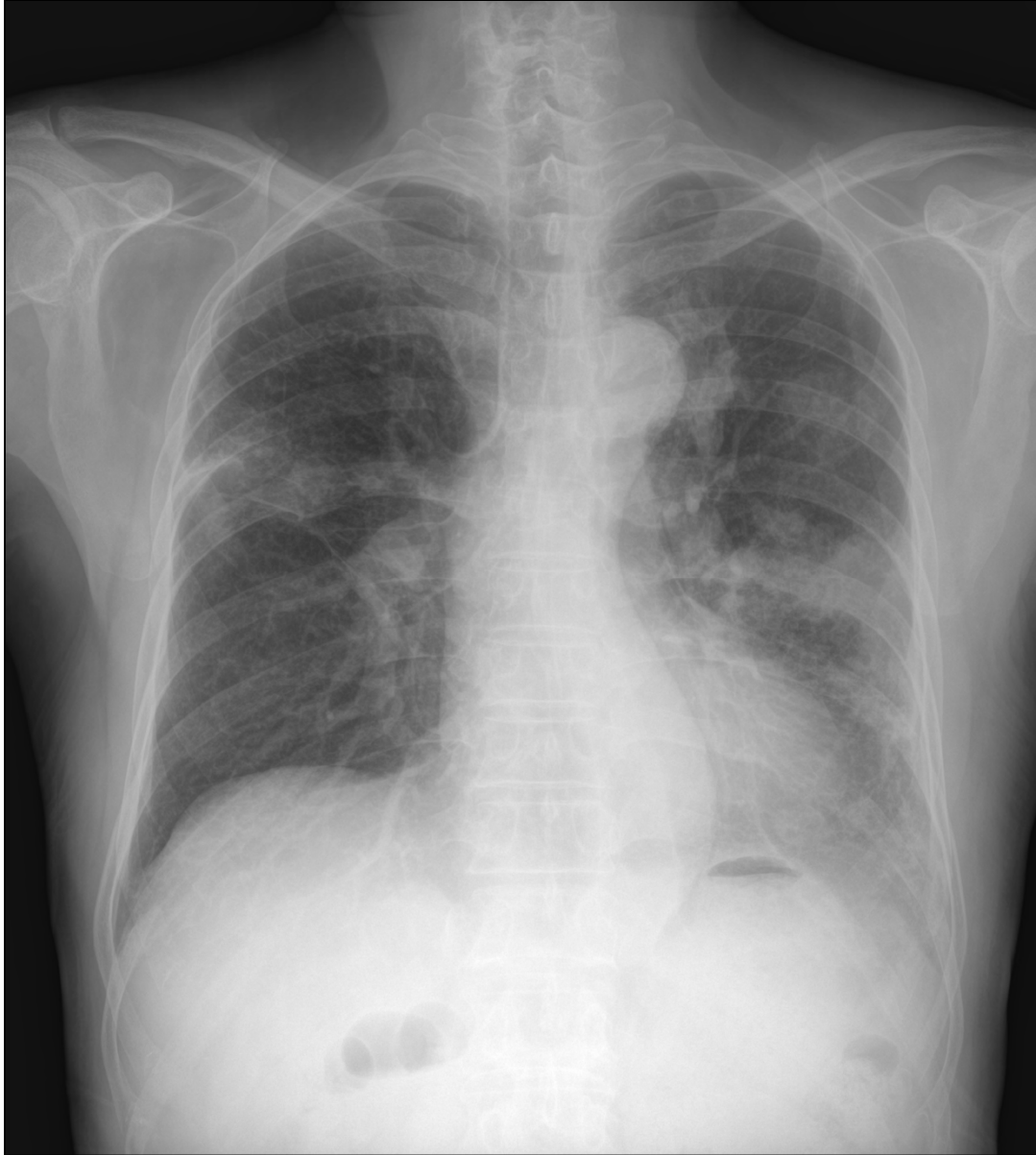
- **Chest**

- **Coarse Breathing Sounds c Crackle & Wheezing at both lung fields**
- Regular heart beat s murmur

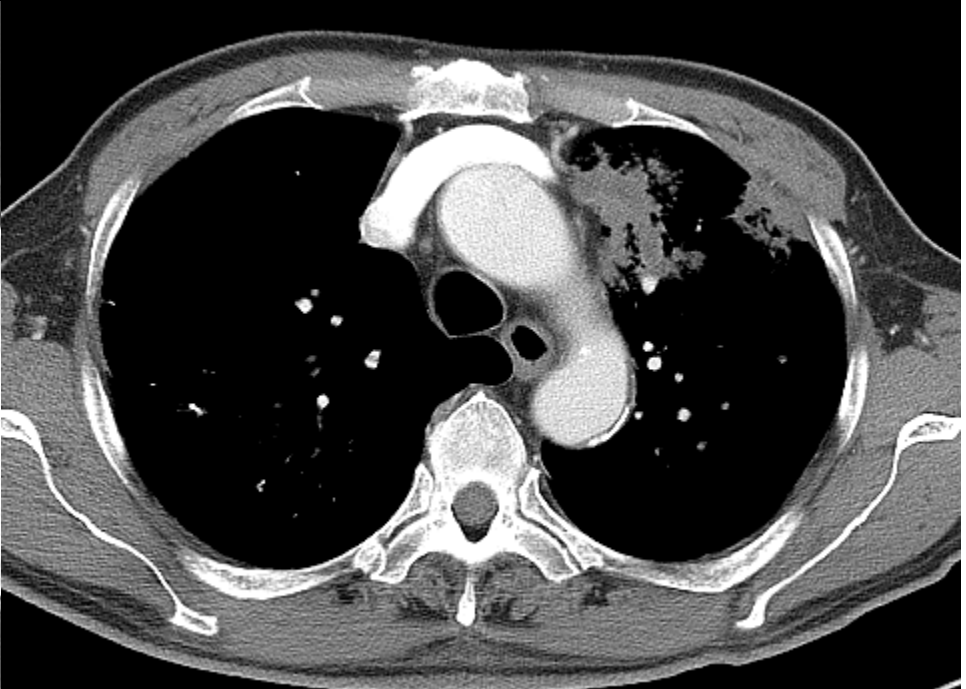
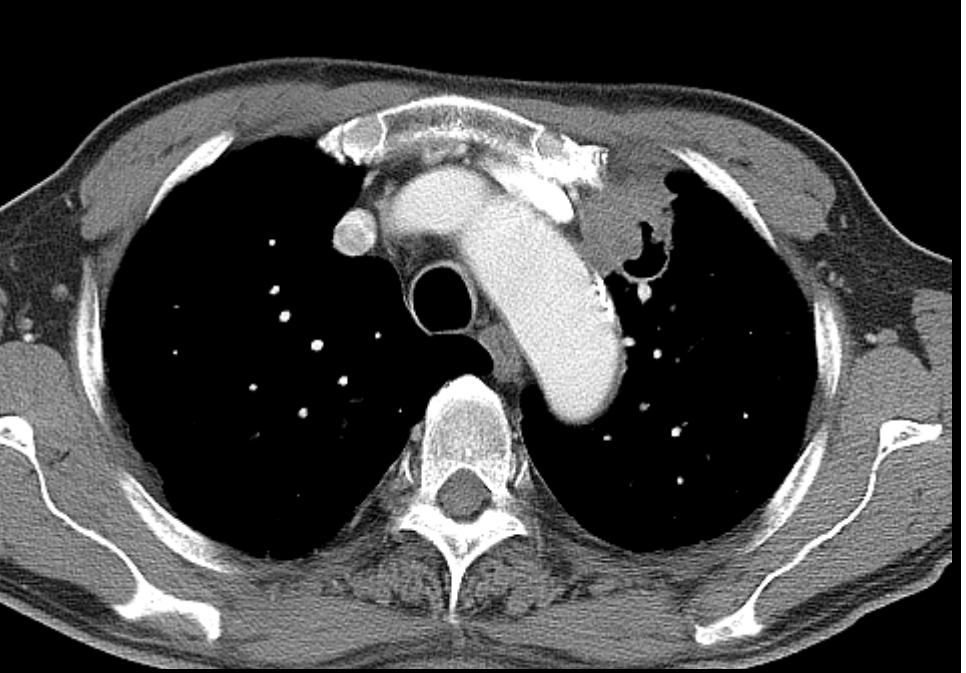
# Lab Findings

|                            |  |
|----------------------------|--|
| Hb – WBC – PLT             | 10.8 g/dL - 8300 g/dL - 294000 / $\mu$ l |
| BUN / Cr                   | 19.0 / 0.97 mg/dL                        |
| Na - K – Cl                | 139 - 4.0 – 105 mmol/L                   |
| AST / ALT                  | 25 / 34 IU/L                             |
| Protein / Albumin          | 6.0 / 3.3 g/dL                           |
| Bililubin tot              | 0.44 mg/dL                               |
| PT                         | 1.06 INR                                 |
| aPTT                       | 35.0 sec                                 |
| <b>ESR / CRP</b>           | <b>61 mm/hr / 61.48 mg/L</b>             |
| Procalcitonin              | 0.045 ng/mL                              |
| <b>D-dimer</b>             | <b>1.34 mcg/mL</b>                       |
| <b>Lupus anticoagulant</b> | <b>positive</b>                          |

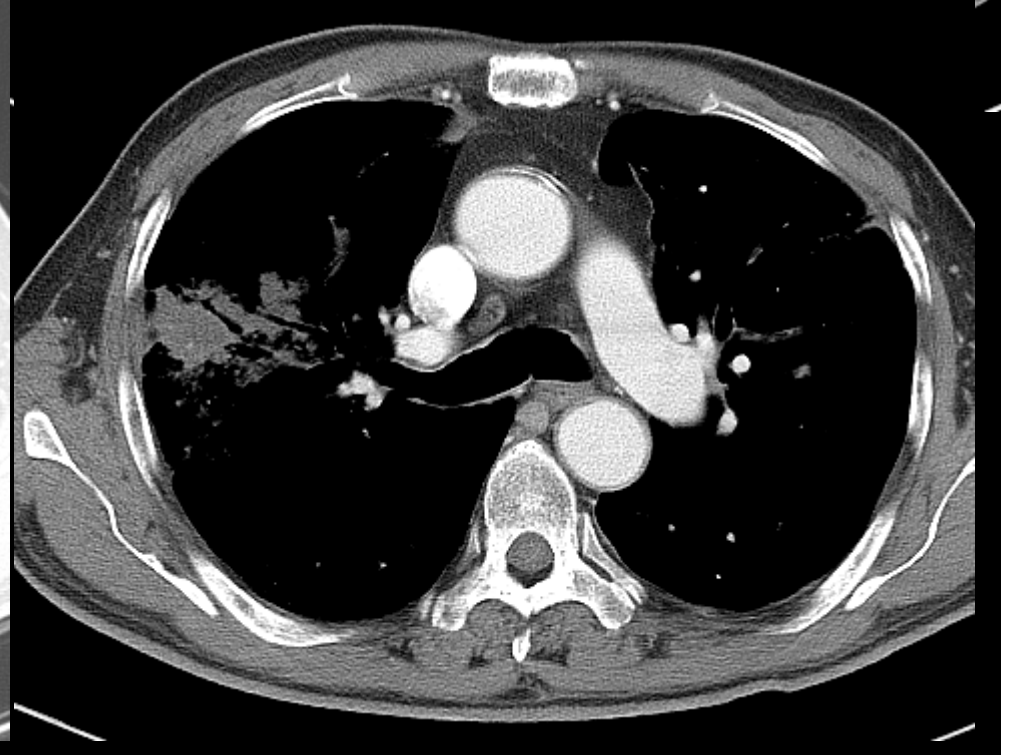
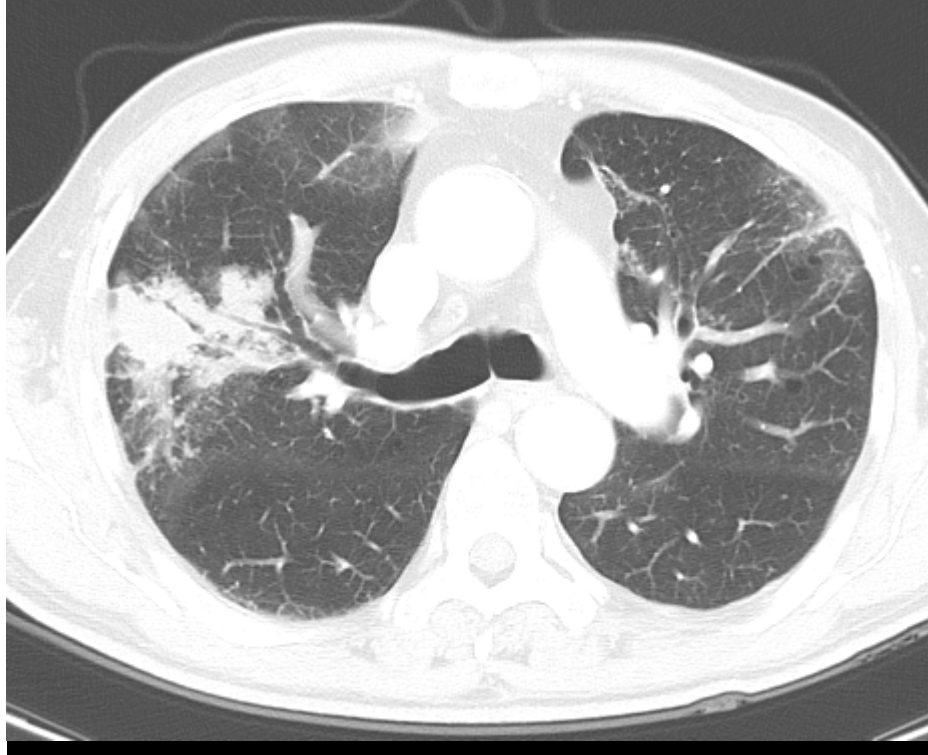
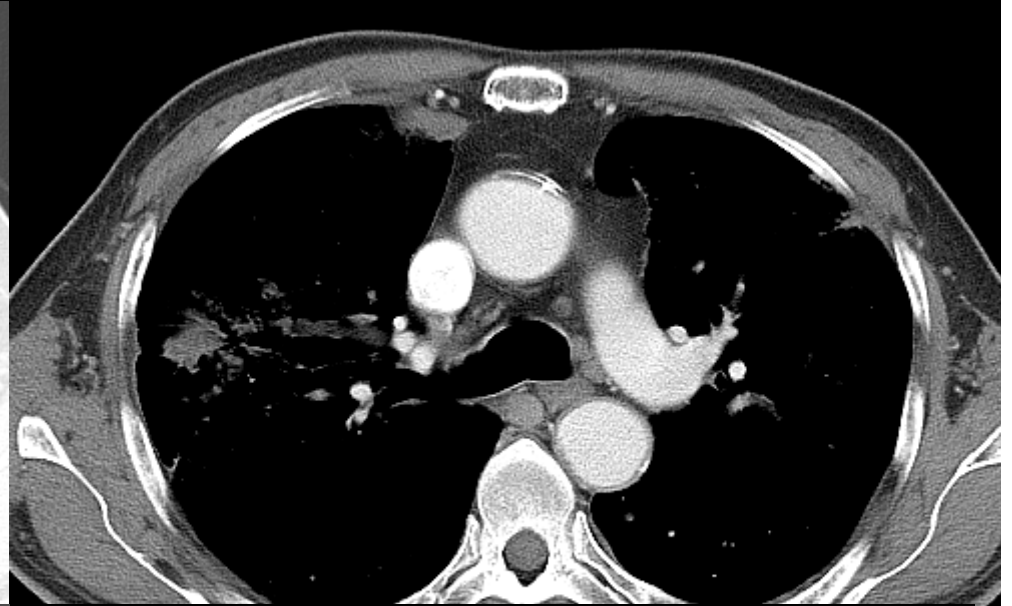
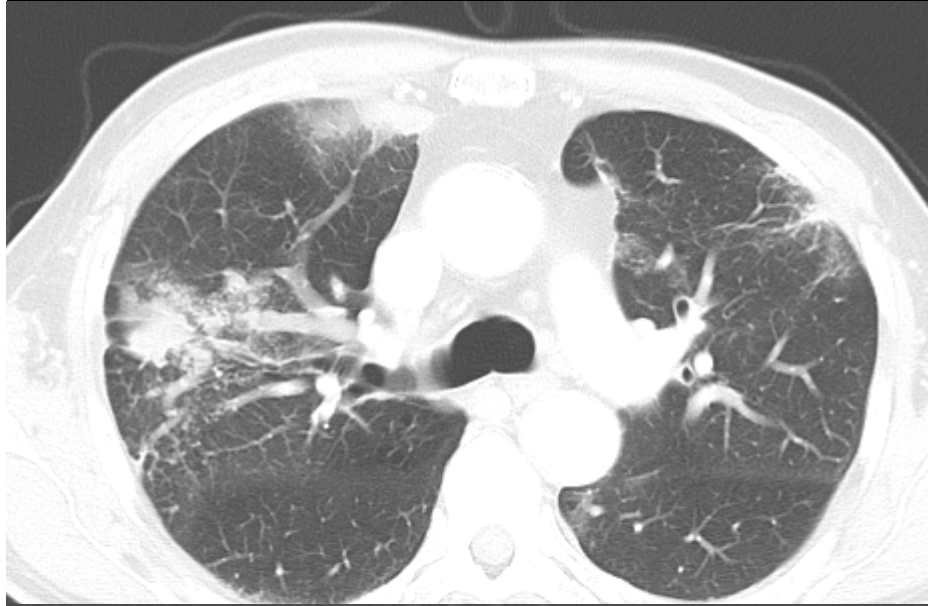
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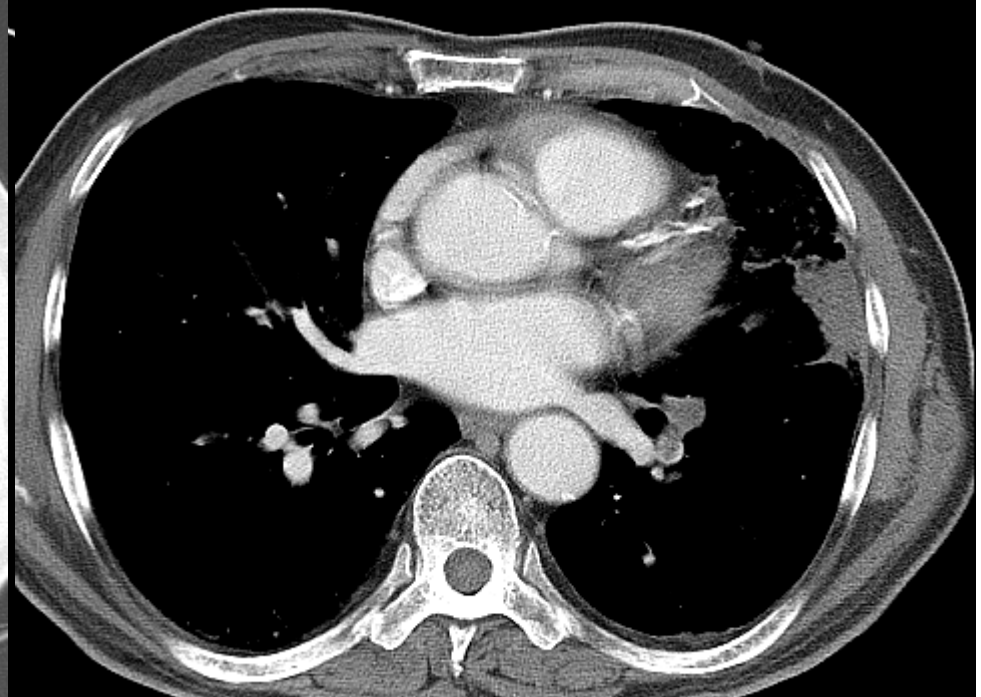
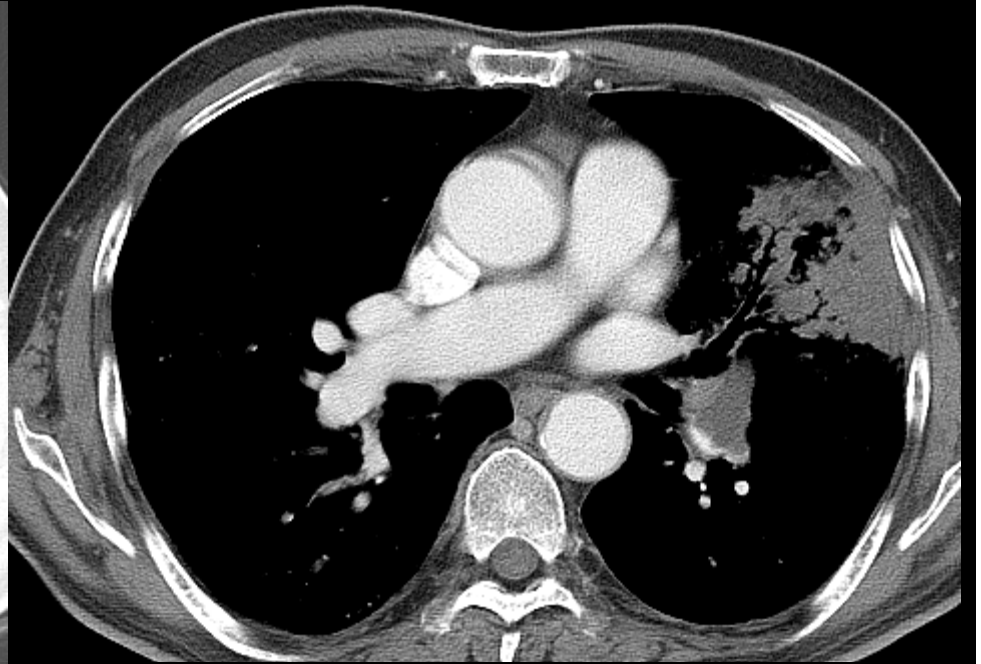
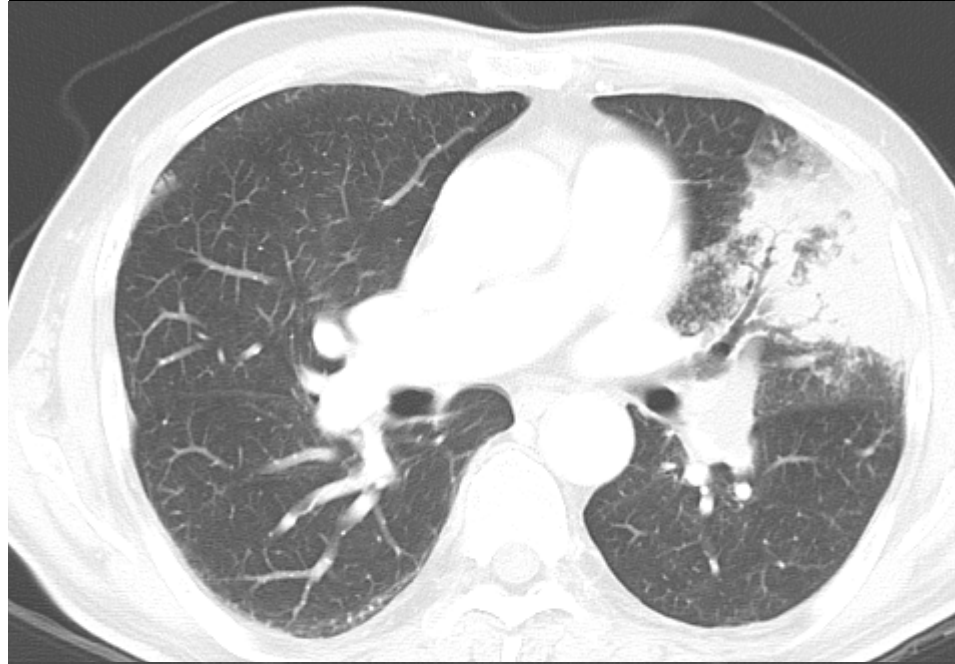
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# Initial Impression

- R/O CAP
- R/O TB
- Pulmonary embolism

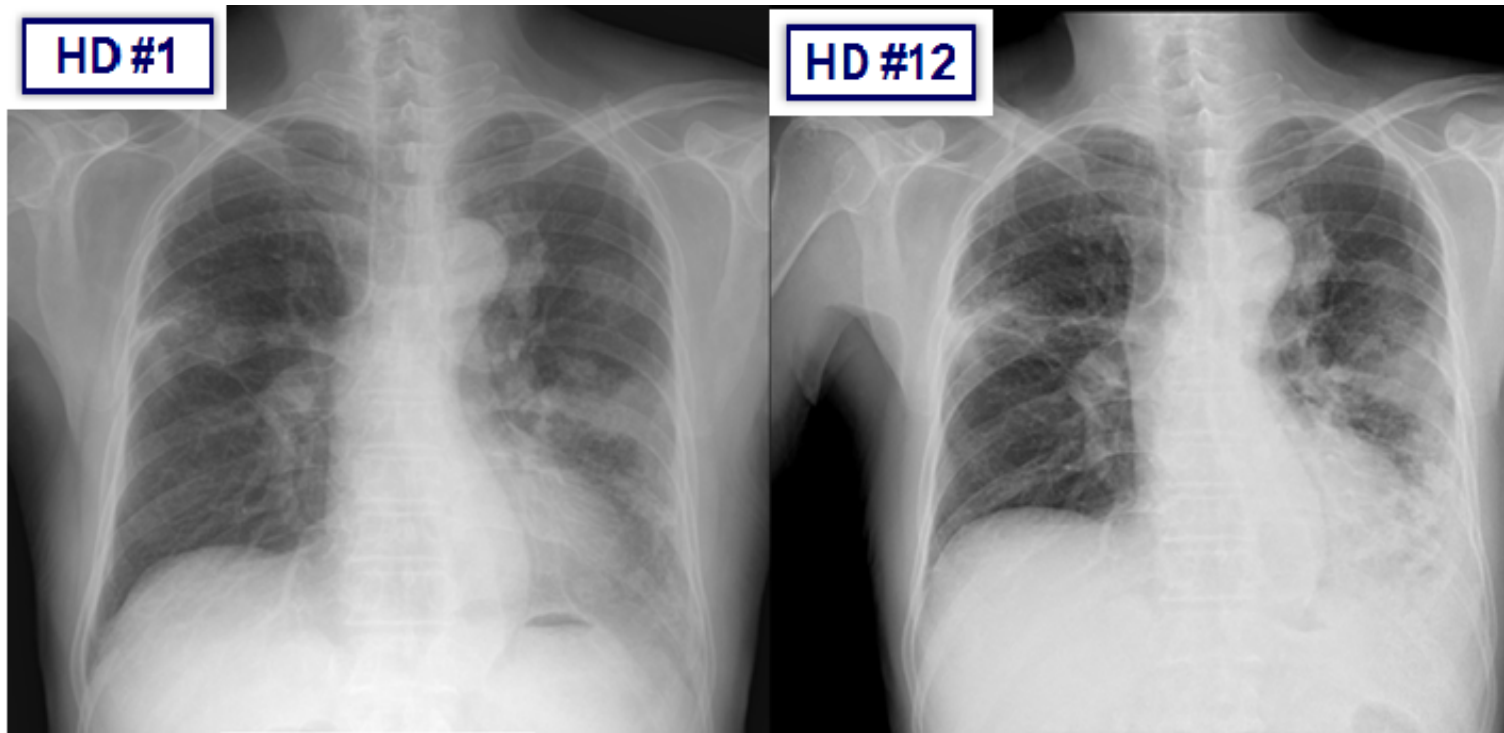
# Progress

- HD #1
  - Antibiotics 투여
  
- HD #6
  - Bronchoscopy & B. washing : No endobronchial lesion
    - ➡ AFB stain (-)
  - TB medication
  - Antibiotics 중단

# Progress

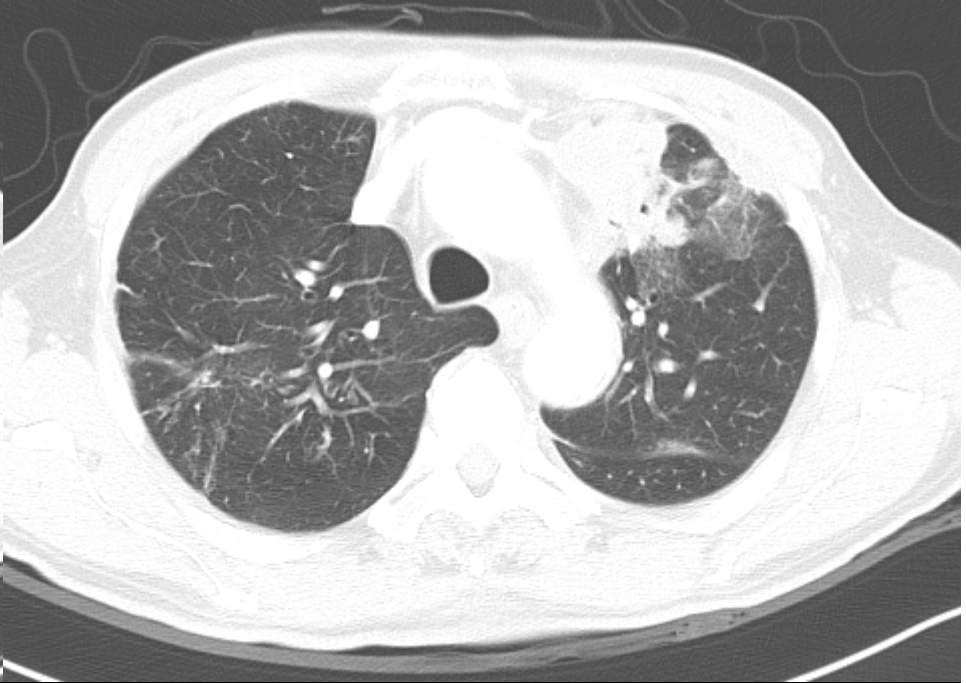
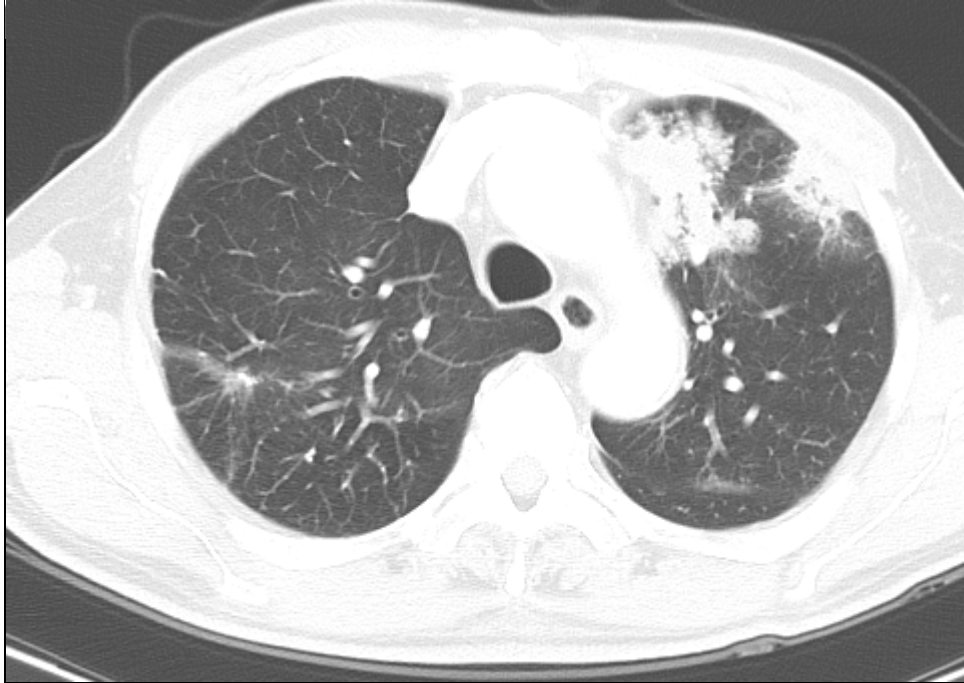
- HD #12

- Fever 지속
- CRP ↑, LLL consolidation ↑,

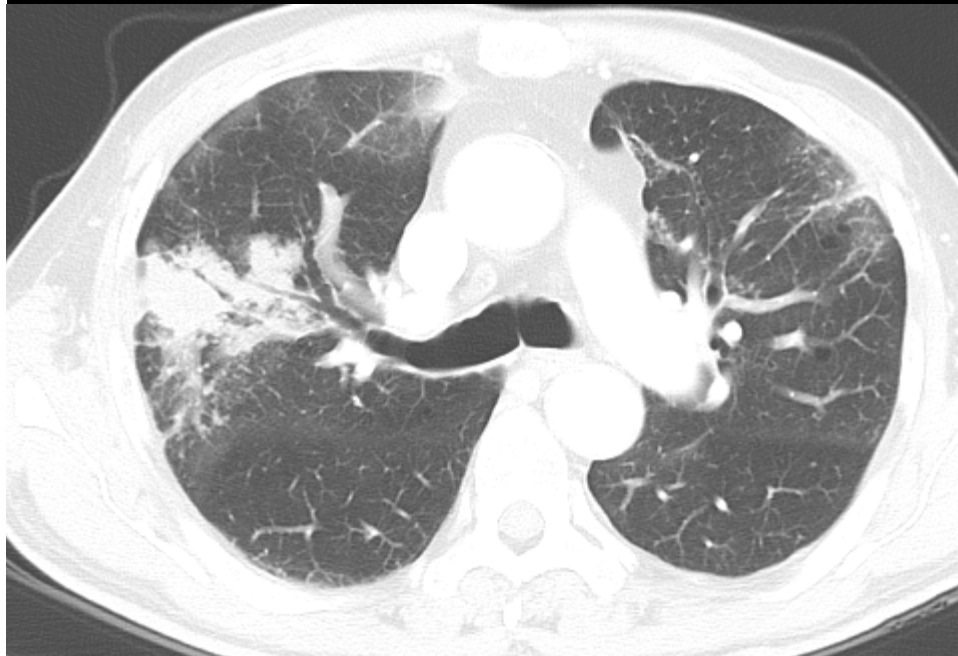


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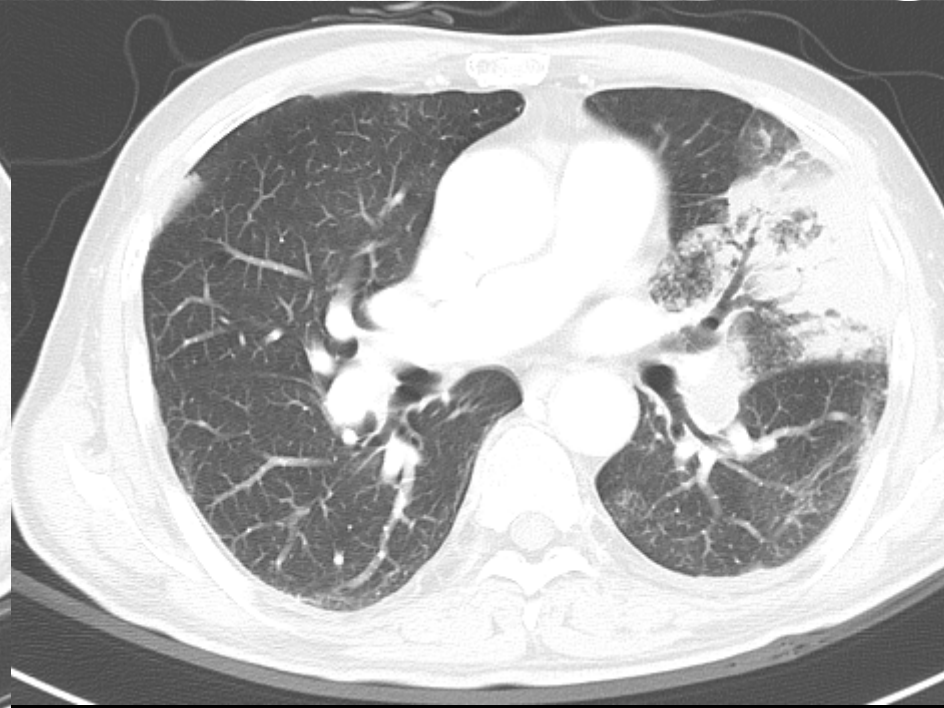
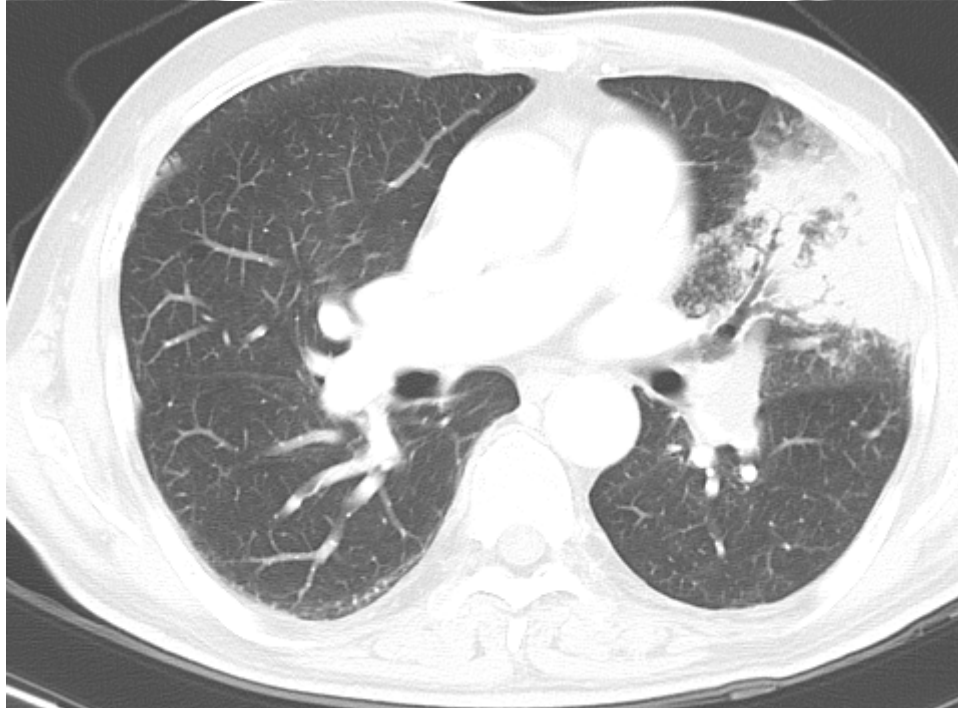
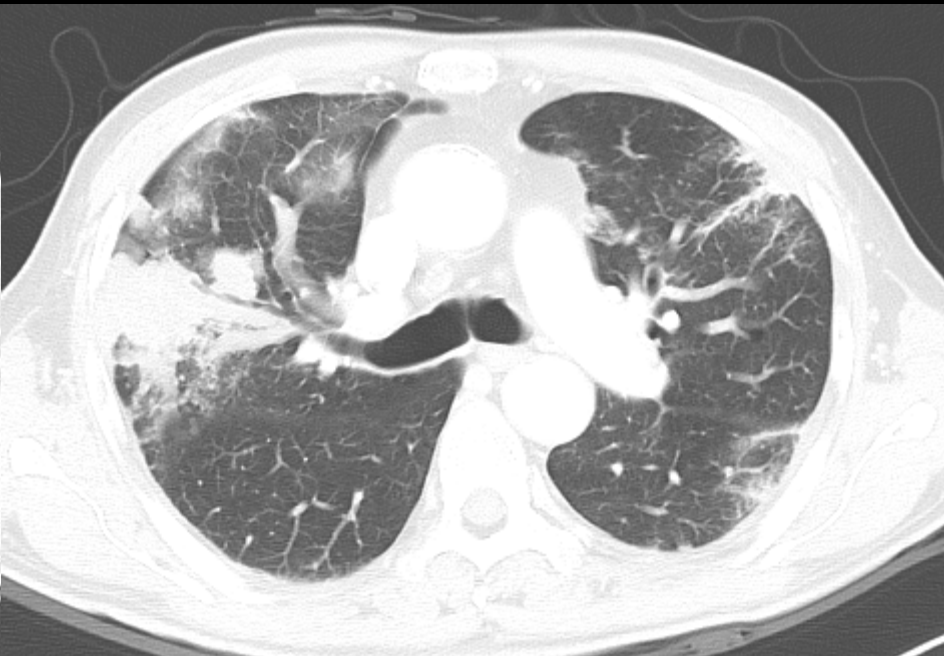
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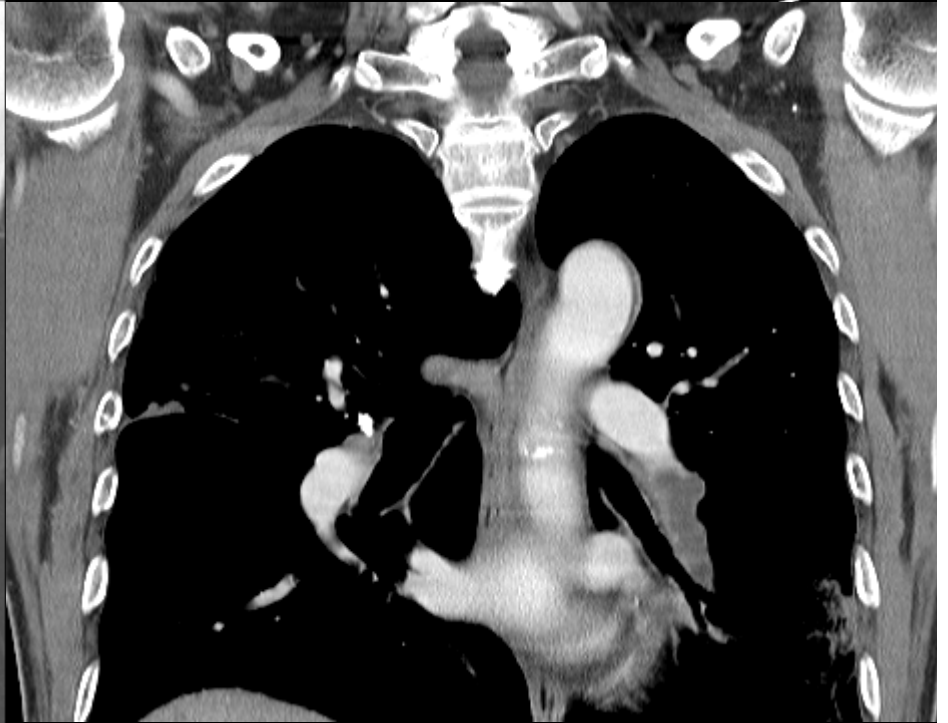
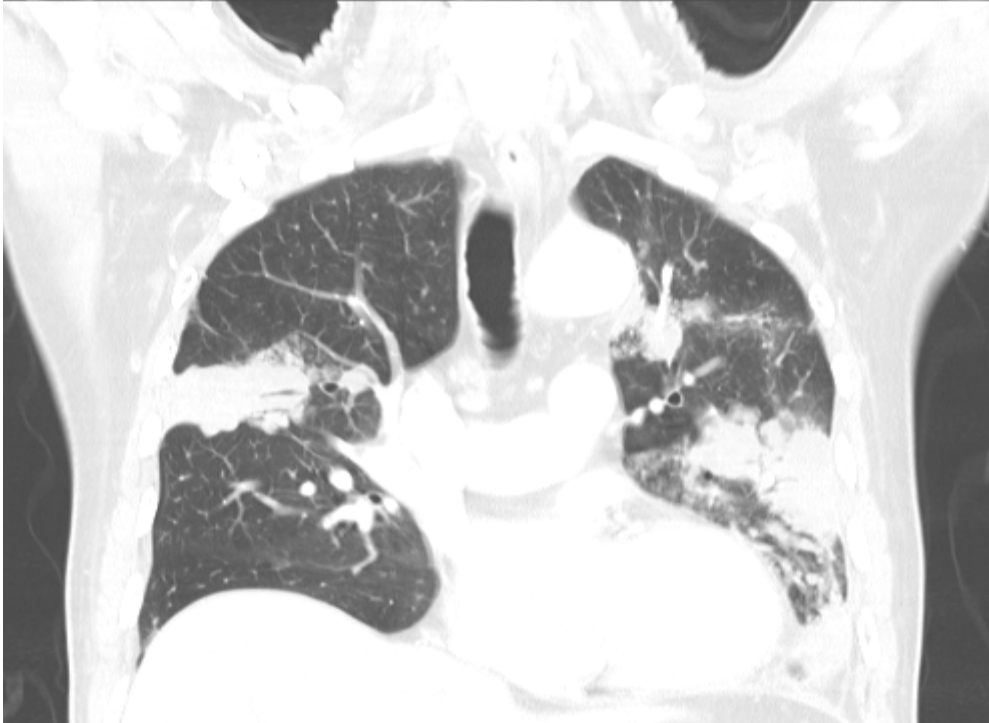
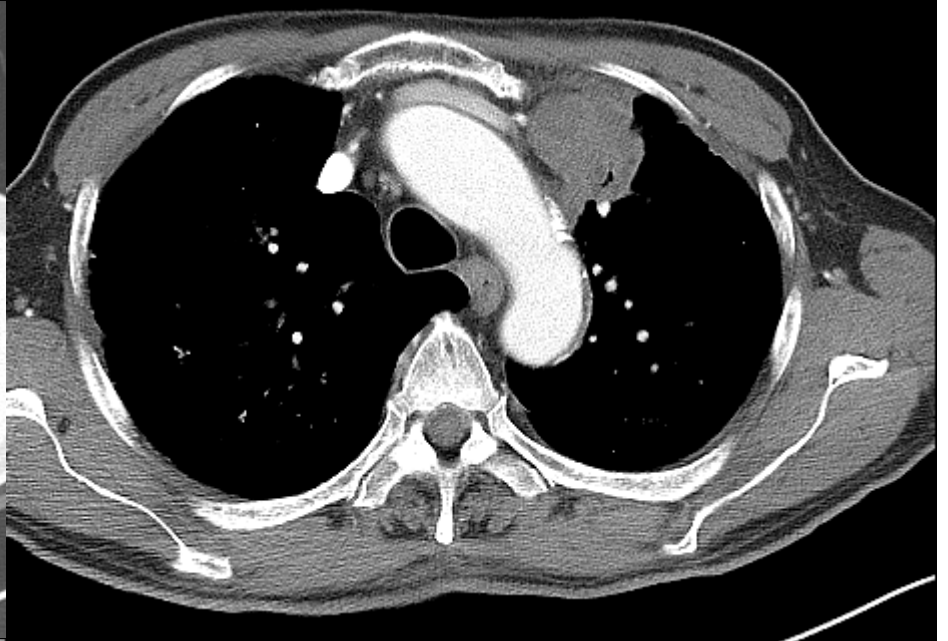
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2013-4-4



2013-4-4



# Progress

- HD #19
  - PCNB was performed on the lesion in the left upper lobe for d/dx tuberculosis, fungal infection, malignancy,..



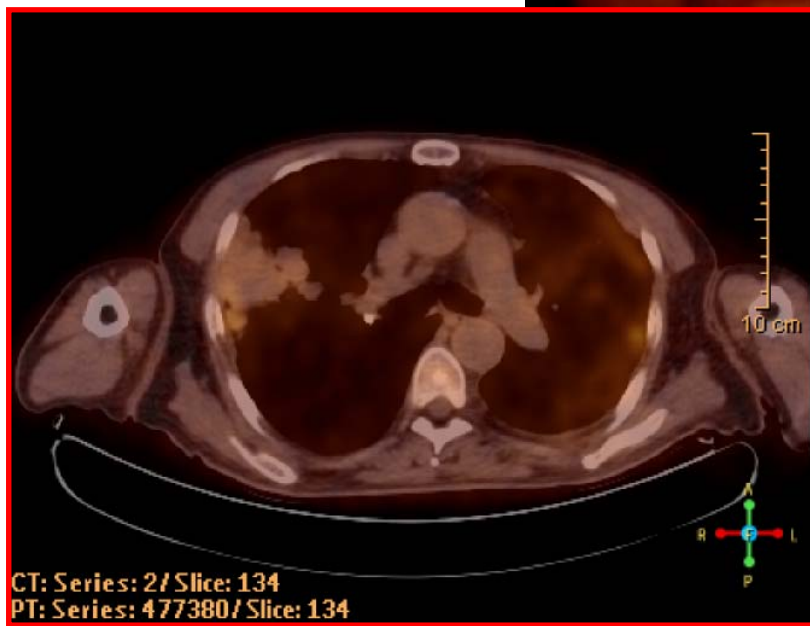
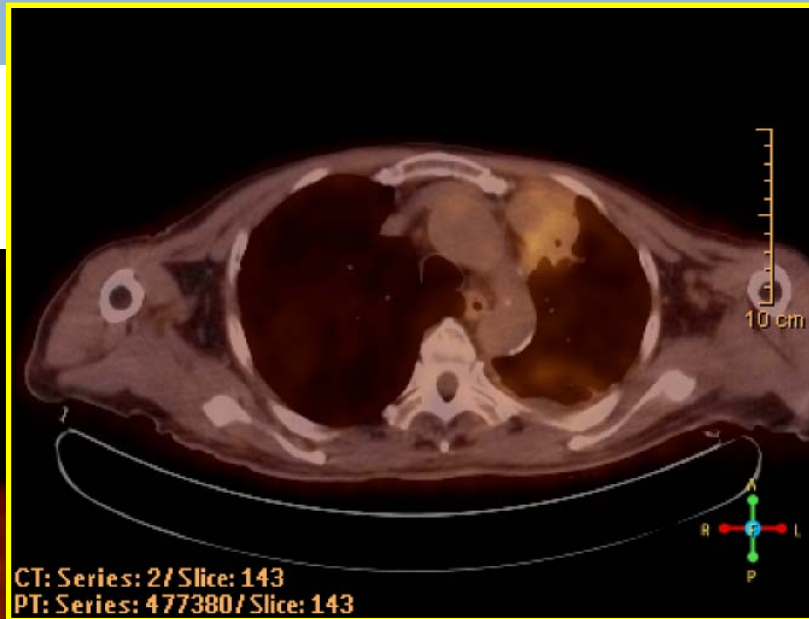
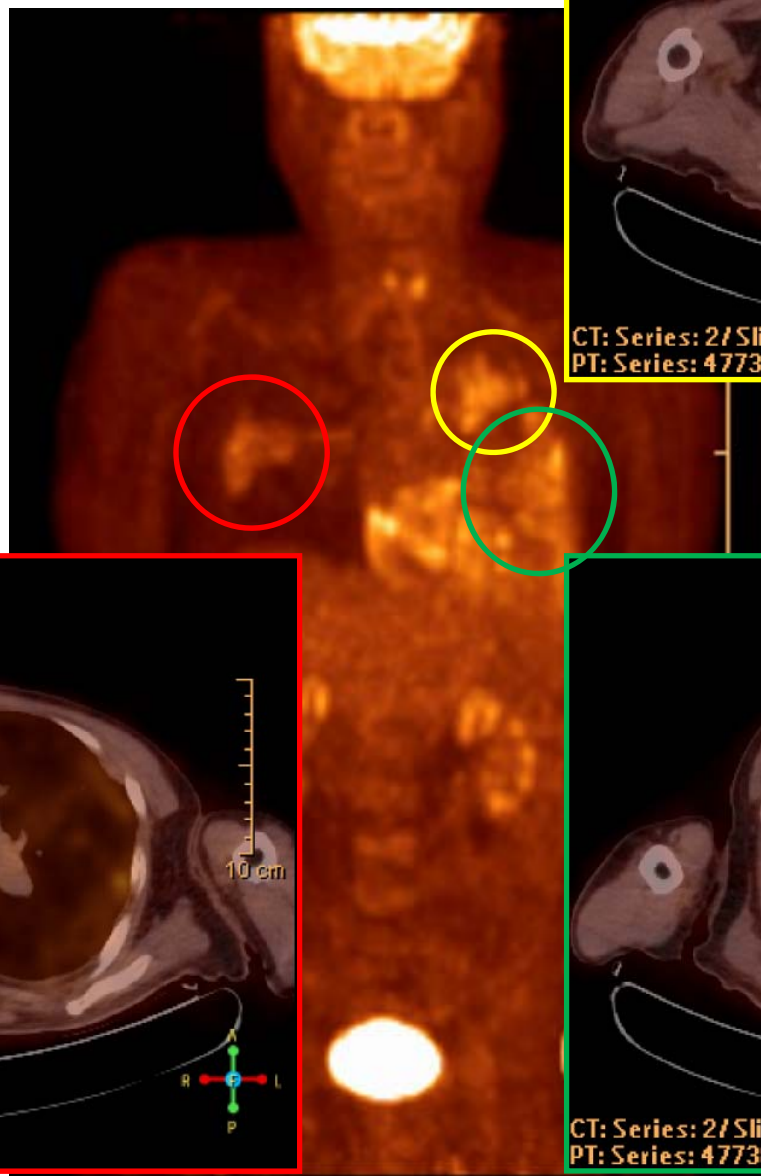


# Histopathology

# Histopathology

- **Myxoid mesenchymal tumor**
  - Favor primary pulmonary myxoid sarcoma
- **EWSR1 rearrangement result (FISH)**
  - 60% Rearrangement

# PET-CT [ HD #29 ]





# Final Diagnosis

Primary Pulmonary Myxoid sarcoma

# PRIMARY PULMANARY MYXOID SARCOMA

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# Introduction

- Extremely rare disease
  - only 12 cases have been reported to 2012
- Morphologically Resembling extraskeletal myxoid chondrosarcoma
- Potentially metastasizing tumor entity with a predilection for the pulmonary parenchyma

# Clinical Data

- 10 cases of Primary pulmonary myxoid sarcoma (2011)
  - 7 women and 3 men
  - Age 27 - 67 (mean, 45 yr)
  - 4 current or ex-smokers/ 5 patients with known smoking history
  - Symptoms
    - Cough (3) / Hemoptysis only (1) / Weight loss only (1)
    - Cough, hemoptysis, fever and weight loss (1)
    - Neurological symptoms due to histologically proven cerebral metastasis (1)
  - Presentation
    - Incidental mass on CT for investigation of viral illness
    - Incidental thoracic masses on CXR
    - HIV+ Incidental mass on CXR for infection screen

# Clinical Data

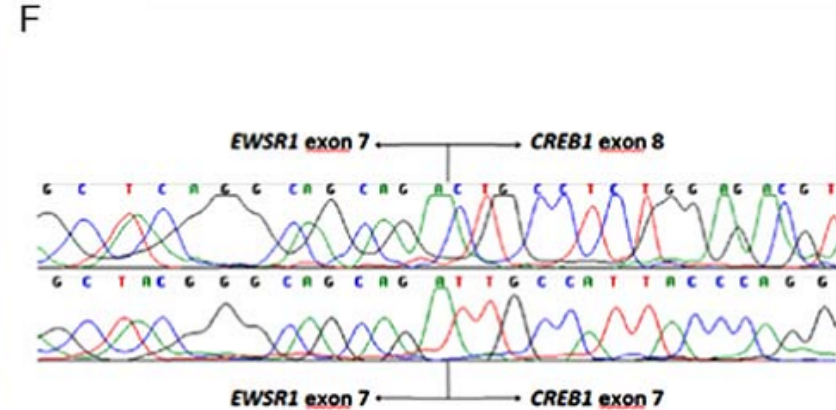
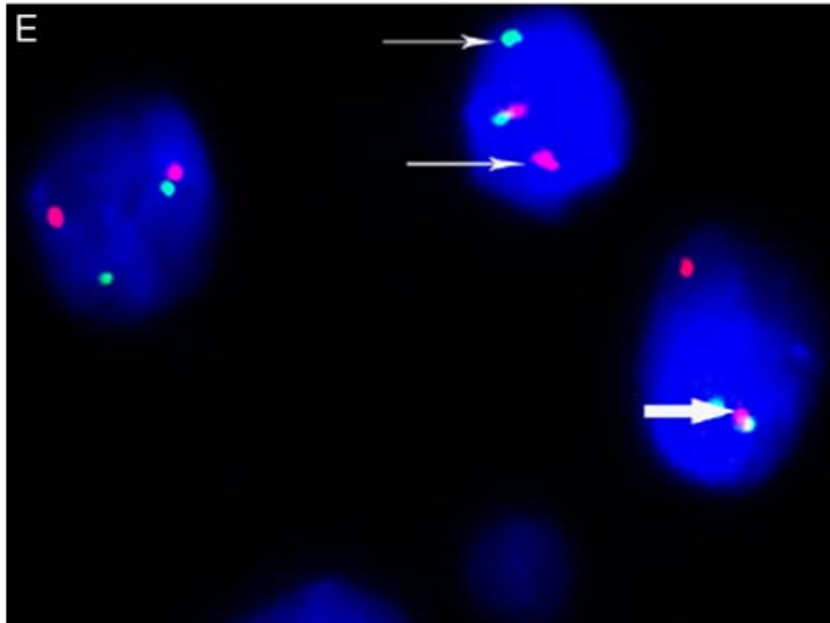
- 10 cases of Primary pulmonary myxoid sarcoma (2011)
  - Site: Rt.(5)/Lt.(5)
  - Predominantly endobronchial(8)
  - Metastasis: kidney(1), brain(1)
  - Tx : Surgical resection
  - Follow up (6)
    - 4 free of disease : 1 - 15(mean, 8 yrs)
    - 1 Left renal metastasis : alive and well after 3 yrs
    - 1 brain metastasis : died a few months after resection

# Molecular Genetics

- 10 cases of Primary pulmonary myxoid sarcoma (2011)
  - FISH: EWSR1 rearrangement(7)
  - RT-PCR: **EWSR1-CREB1** fusion transcript(9)

TABLE 5. Molecular Genetic and FISH Findings

| Case No. | RT-PCR Result      | <i>EWSR1</i> Break Point | <i>CREB1</i> Break Point | FISH Result              |
|----------|--------------------|--------------------------|--------------------------|--------------------------|
| 1        | <i>EWSR1-CREB1</i> | Exon 7                   | Exon 7                   | <i>EWSR1</i> break point |
| 2        | <i>EWSR1-CREB1</i> | Exon 7                   | Exon 7                   | <i>EWSR1</i> break point |
| 3        | None               | None                     | None                     | <i>EWSR1</i> break point |



## Tumor with EWSR1-CREB1 fusion

| Tumo                                      | Site  | Histology   | Immunohistochemistry   |
|---|---|---|--|
| Angiomatoid fibrous histiocytoma          | Extremities (usually superficial)                 | <ul style="list-style-type: none"> <li>• Nodular</li> <li>• bland ovoid or spindle cells</li> </ul>                                 | 50% desmin positive  |
| Clear cell sarcoma                        | Extremities (deep soft tissue)                    | <ul style="list-style-type: none"> <li>• Uniform ovoid to spindle cells with clear to granular cytoplasm</li> </ul>                 | Diffuse S100 protein, HMB45, MelanA and MiTF in most               |
| Clear cell sarcoma-like tumor of GI tract | Wall of small bowel, stomach, or large bowel      | <ul style="list-style-type: none"> <li>• Monomorphic round to ovoid cells with clear to eosinophilic cytoplasm</li> </ul>           | Diffusely S100 protein positive, but negative for HMB45 and MelanA |
| Primary pulmonary myxoid sarcoma          | Lung; most tumors have an endobronchial component | <ul style="list-style-type: none"> <li>• Reticular pattern; cords and trabeculae of cells in prominent myxoid background</li> </ul> | Positive only for vimentin and focally for EMA                     |



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