Cryptogenic organising pneumonia

CASE STUDY

What I have learned from this case?

- Cryptogenic Organising Pneumonia is a rare, but significant respiratory disease
- There are a number of classical radiological findings on CXR and CT, but there are also a number of variations of CT patterns
- It is very treatable
  - And therefore it is very important to be aware of the CT patterns of organising pneumonia

CASE: 60-70 y/o Male

- PC - Cough + Fever
- HPC
  - Fever - 38.7℃
  - Cough – >1/52
  - Sees a doctor
    - Dx = infectious pneumonia
    - Rx = antibiotics
  - Cough + fever improving but lung infiltration didn't get better so he consulted another doctor
- PMH - Gastric Ca.

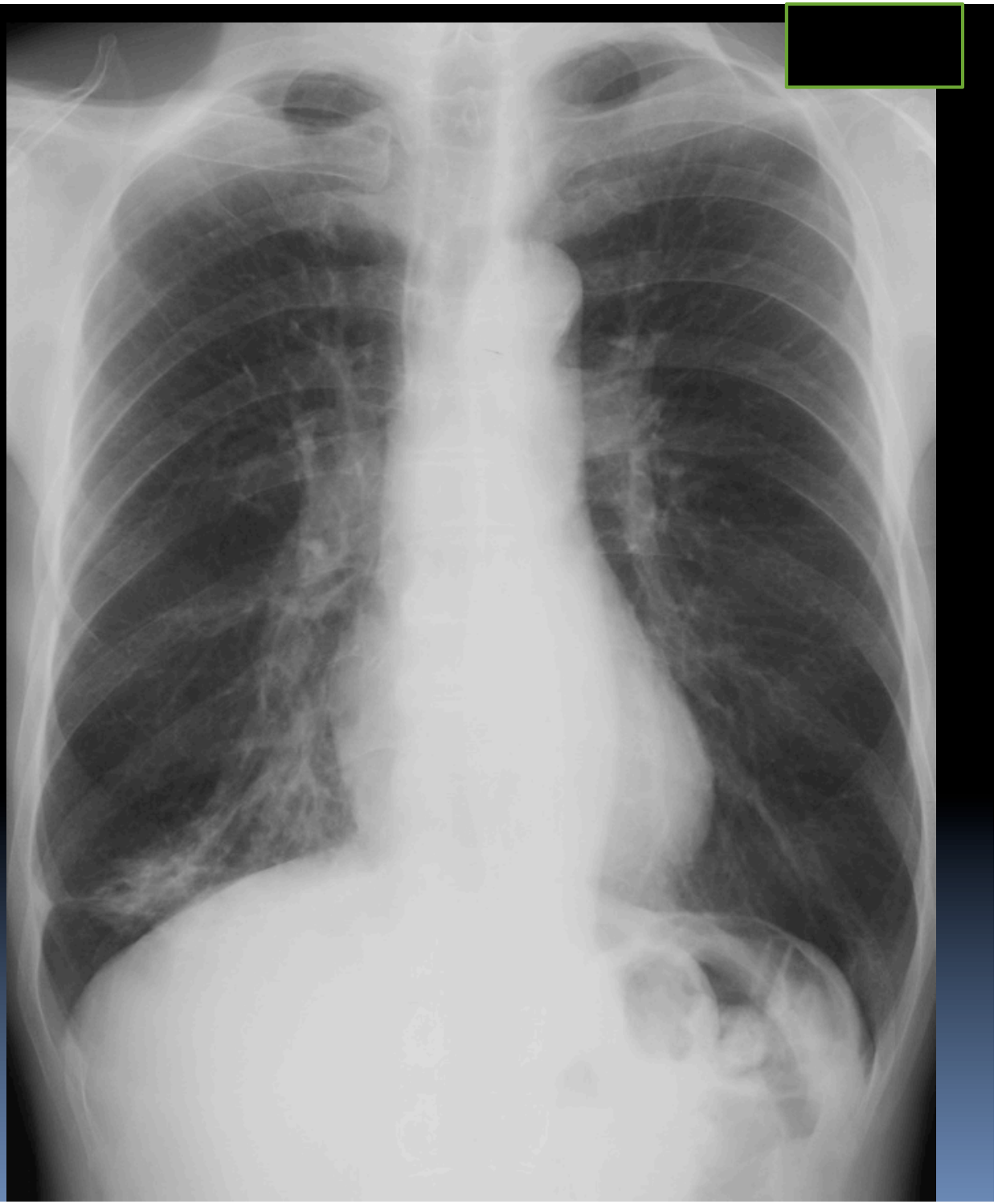
CASE: 60-70 y/o Male

- Smoker – 40(2 packs)/day more than 40 years
  - = ~80 pack/yrs
- Job – sells bicycles
- Medications (-)
- Allergies (-)

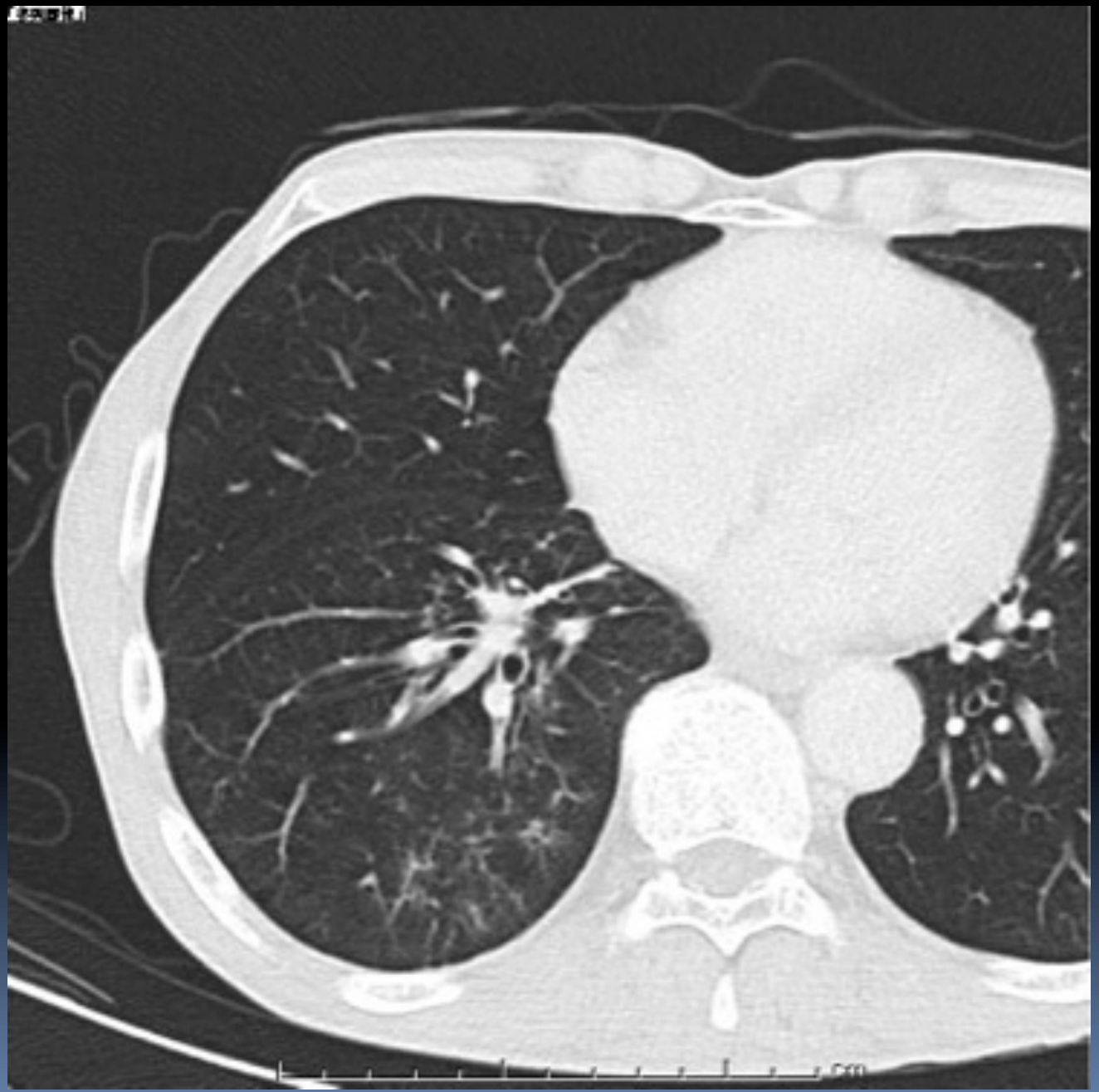
Ix – Lab Studies

- AST – 30
- ALT – 31
- ALP – 383 (H)
- GGT – 80 (H)
- LDH – 156
- Total Protein – 7.2
- Albumin – 4.3
- UN – 13
- Cr – 0.79
- Na – 143
- K – 6.1 (H)
- Cl – 105
- BSL – 121
- CRP – 0.34 (H)
- B-D glucan – 4.0>
- WBC – 4.1 (L)
  - Neutr – 31% (L)
  - Lymph – 59.3% (H)
  - Mono – 7.7%
  - Eosin – 1.5%
  - Baso – 0.5%
- RBC – 3.98 (L)
  - Hb – 12.7 (L)
  - MCV – 101.5 (H)
- Plt – 308

Imaging - CXR



CT



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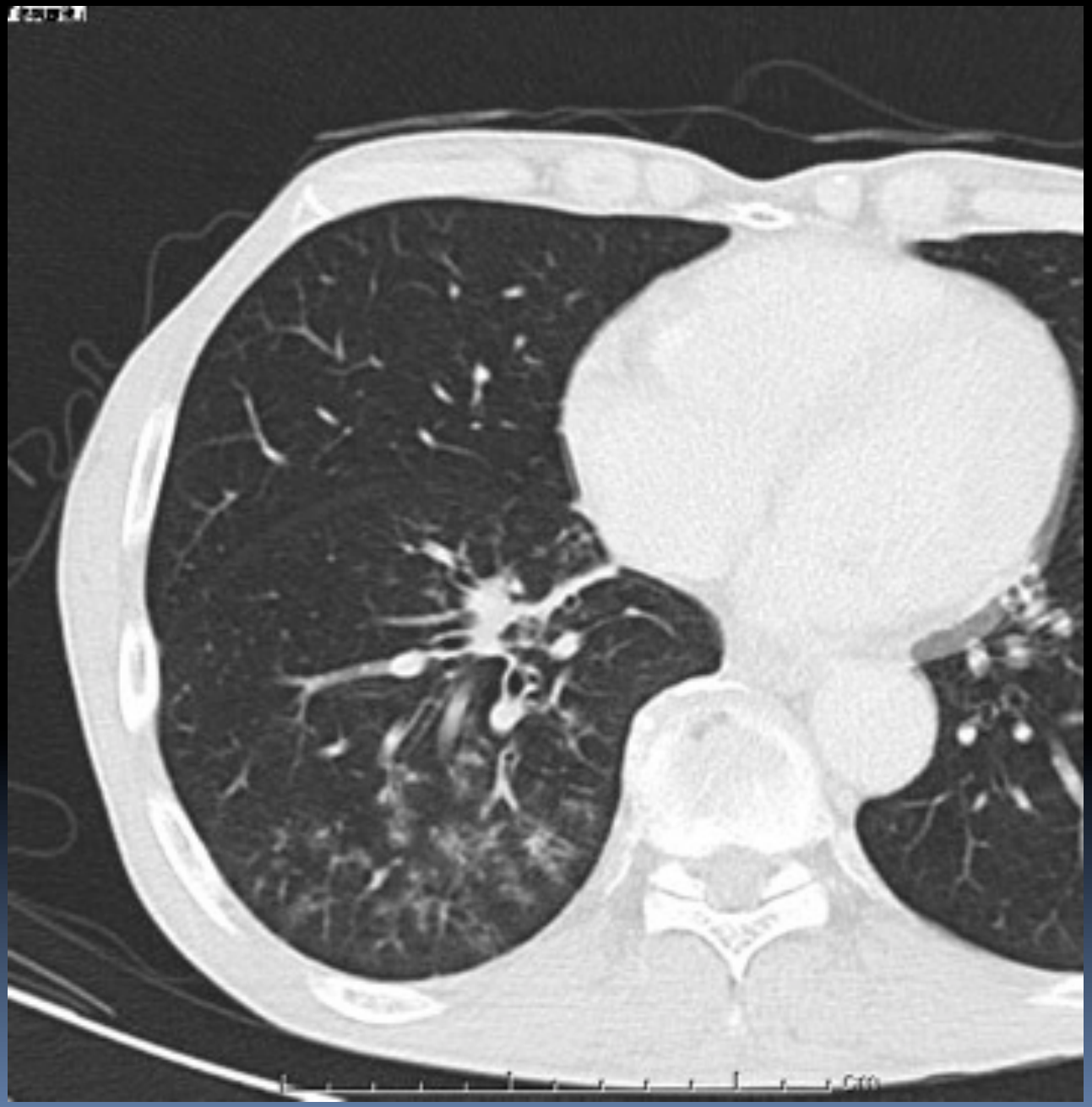

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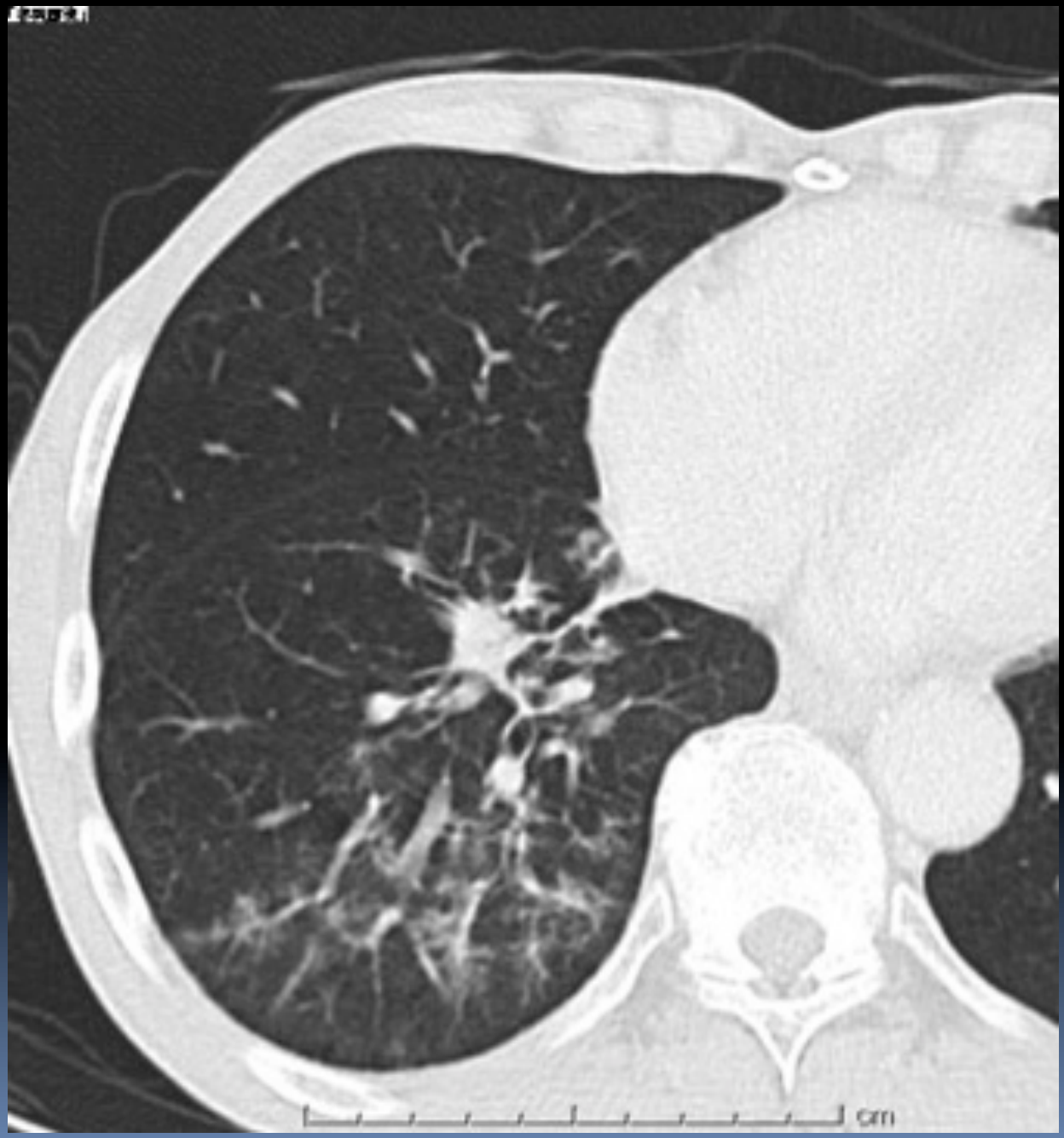

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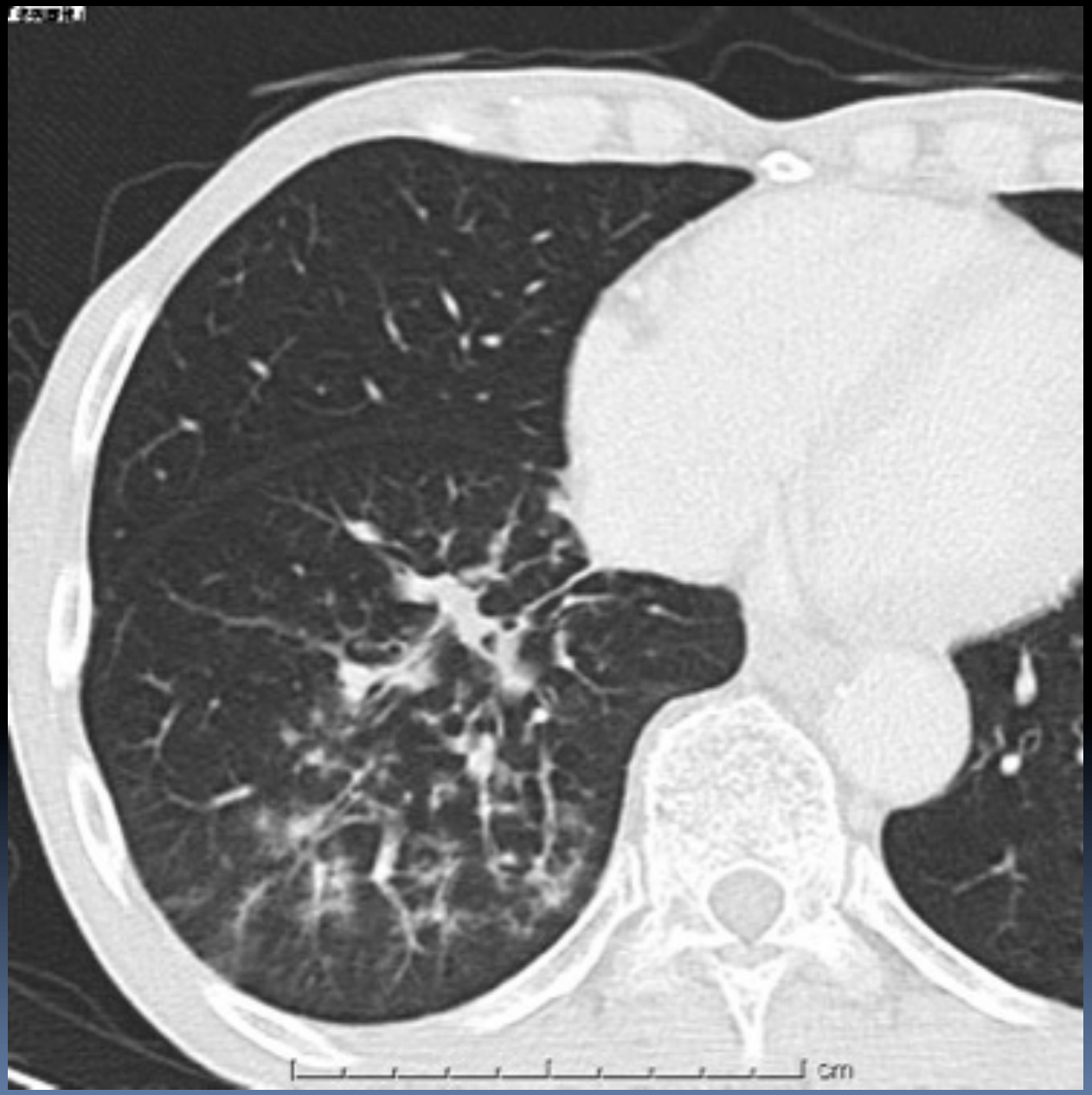

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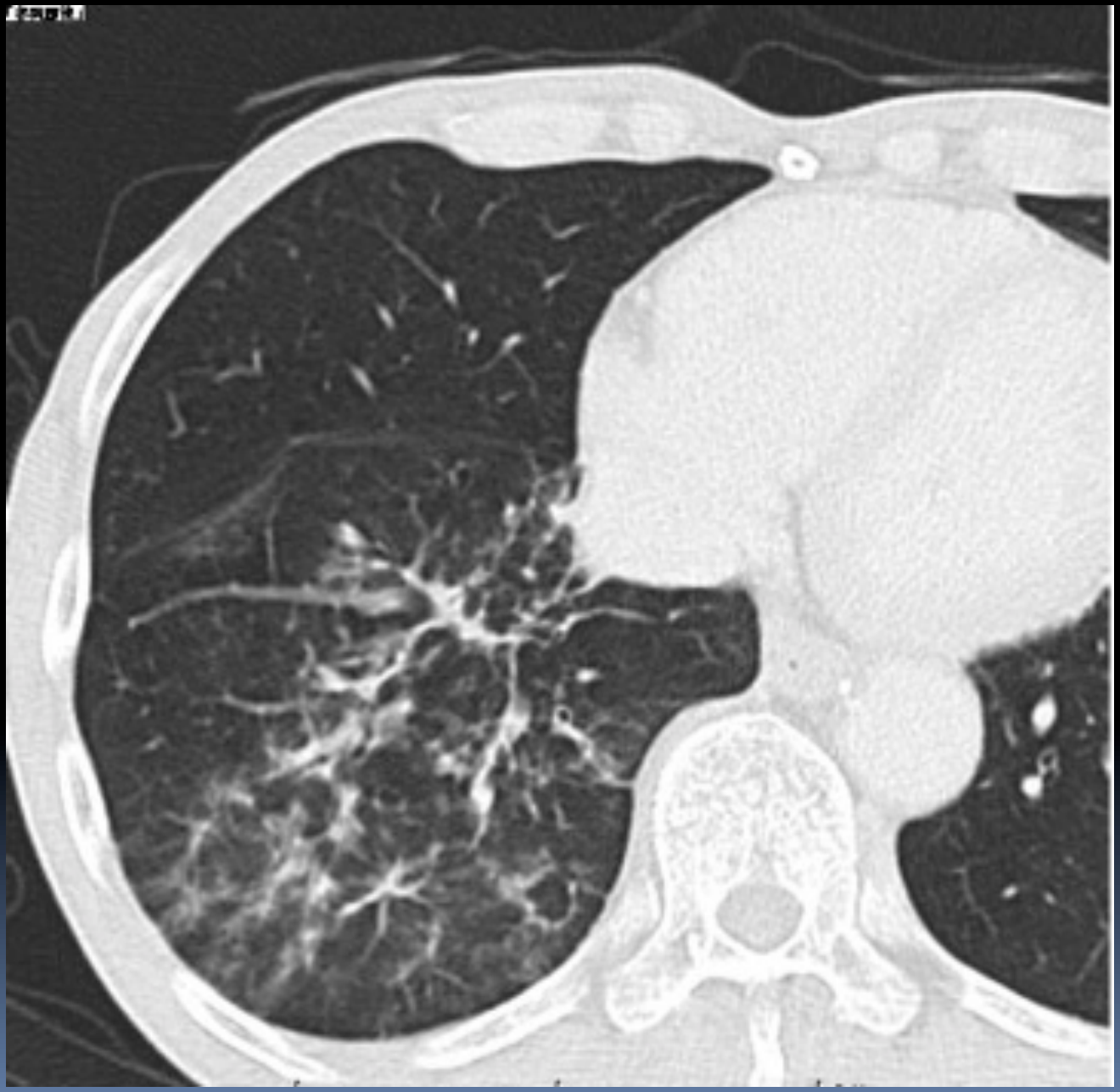

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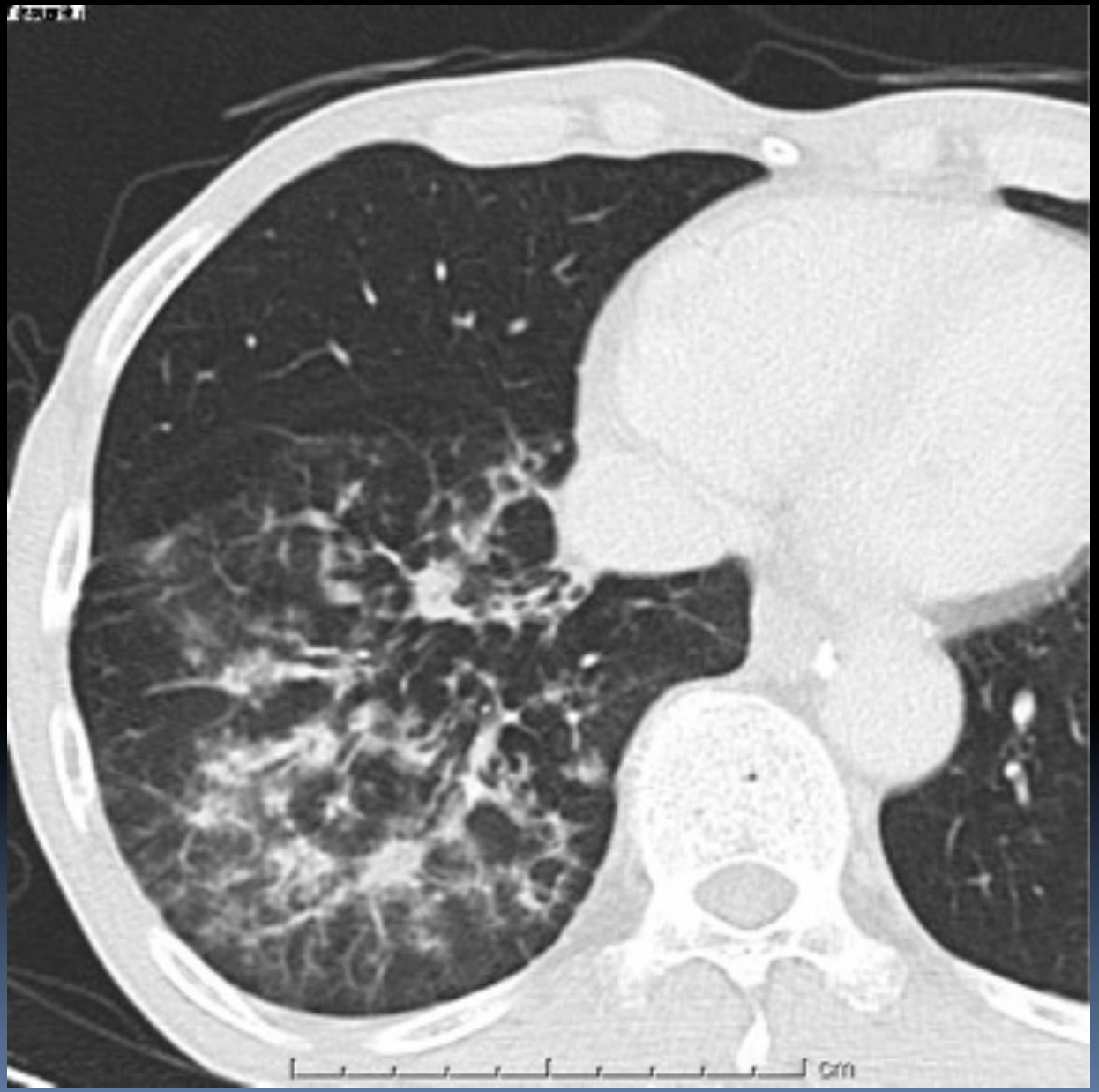

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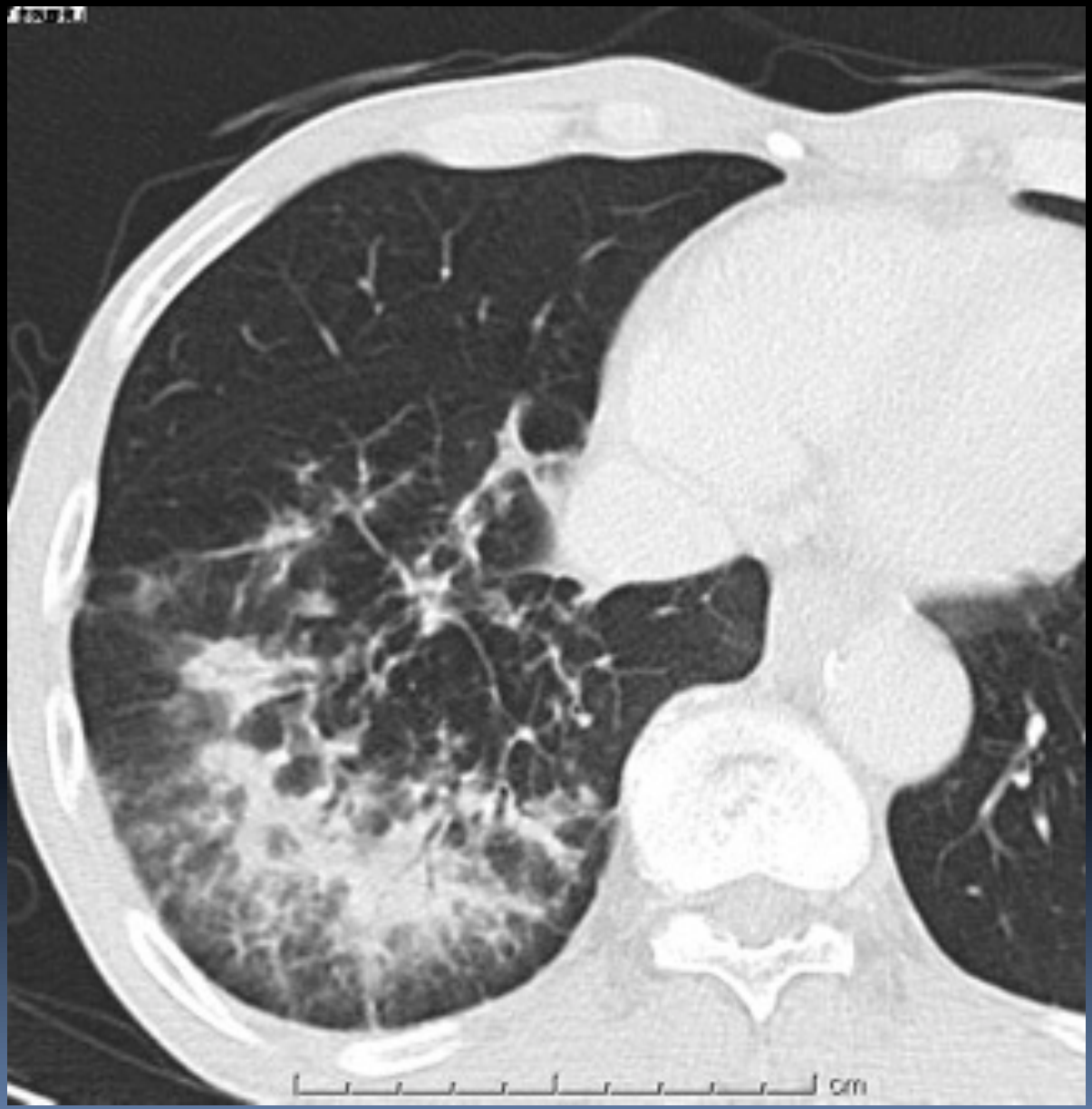

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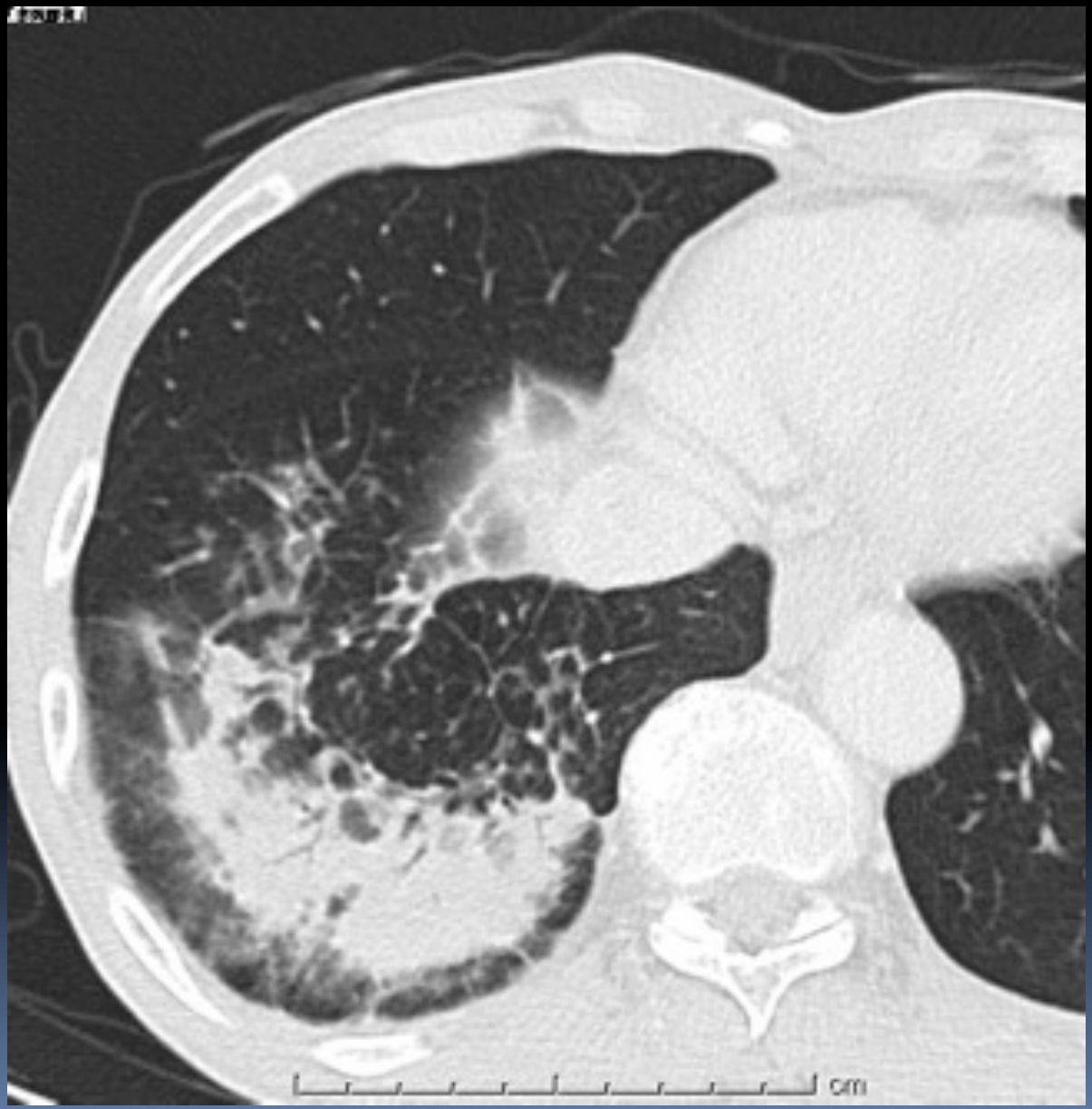

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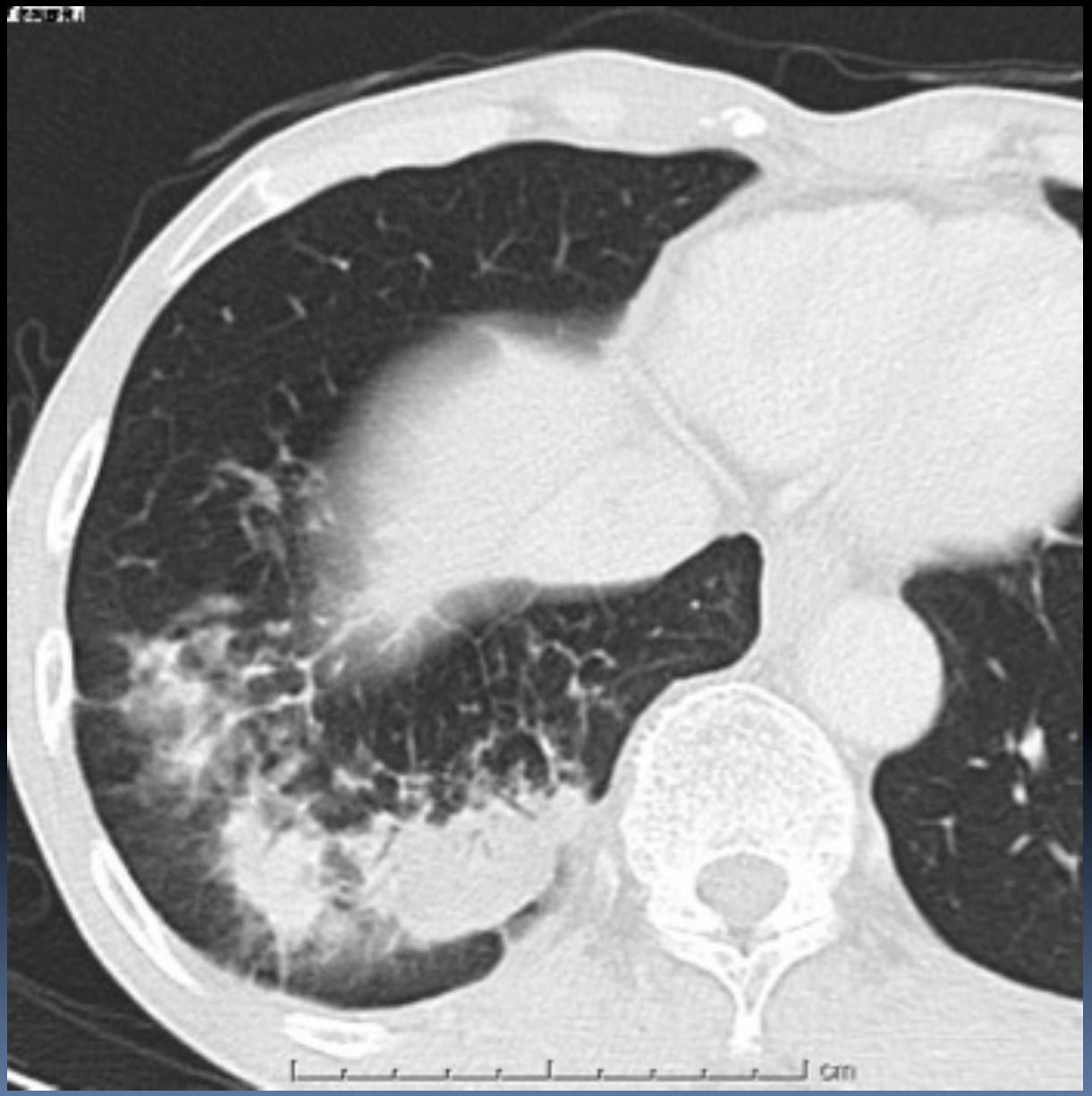

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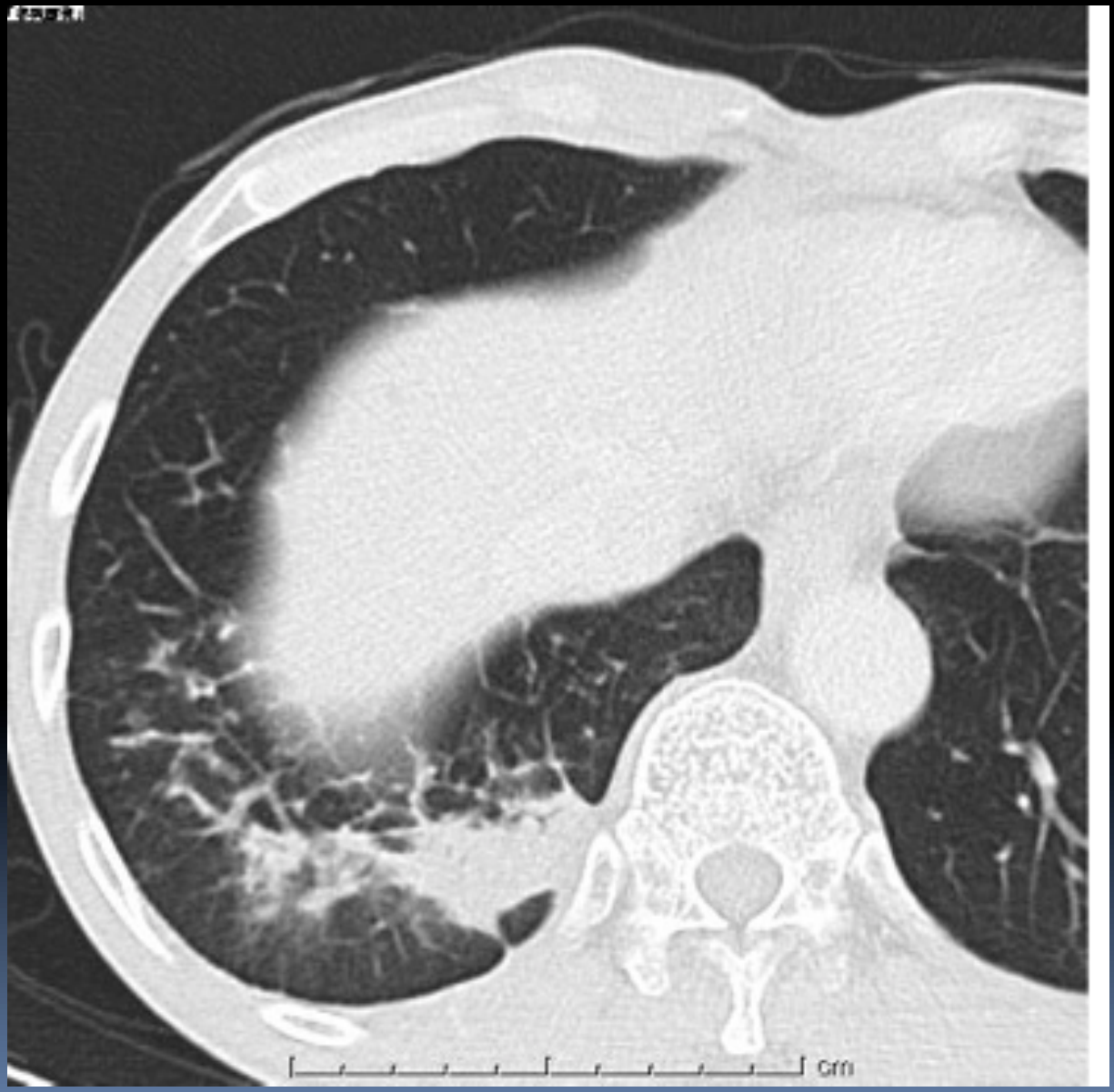

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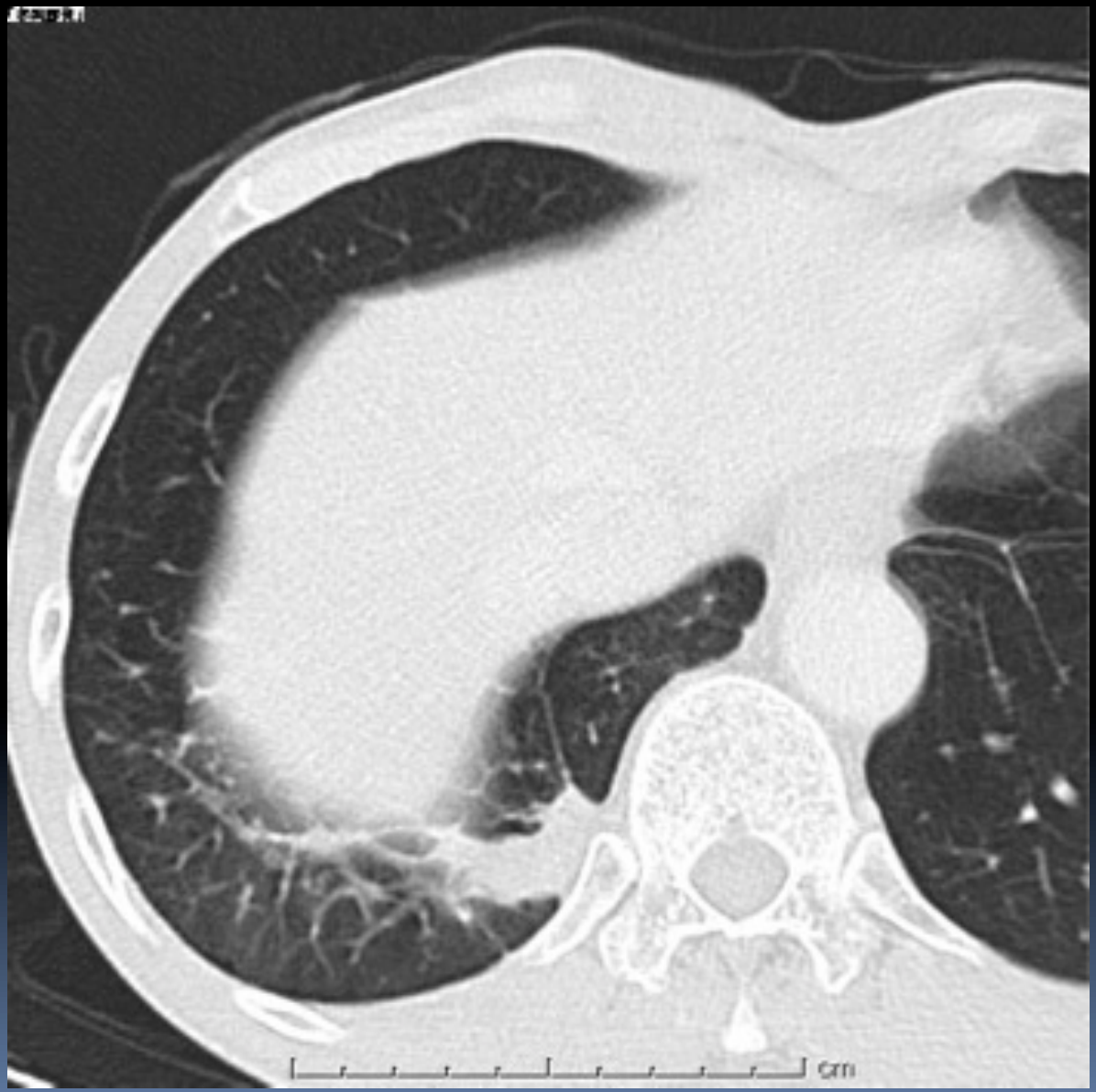

DDx

- Bacterial pneumonia
- Organising pneumonia
- Chronic eosinophilic pneumonia
- Malignancy (including lymphoma)
- Pulmonary drug reaction
- Pulmonary disease associated with a connective tissue disorder

Diagnosis

- TBLB confirmation
- Organizing pneumonia
  - Interstitial infiltrate of plasma cells, lymphocytes
  - Fibromixoid plugs in alveoli
  - No granulomas or necrosis

Cryptogenic Organizing Pneumonia (COP)

- Idiopathic form of organizing pneumonia
- A non-specific response to various forms of lung injury
- One of the main reparative reactions to acute injury by the lung
- Restrictive airways disease pattern
- Diagnosis of exclusion

Triggers

- **Infection**
  - *Bacterial*
    - *Streptococcus pneumonia*
    - *Legionella pneumophila*
    - *Mycoplasma pneumonia*
    - *Coxiella burnetti*
    - *Nocardia asteroides*
    - *Chlamydia pneumonia*
  - *Viral*
    - Adenovirus
    - CMV
    - Influenza + Parainfluenza
    - HIV
- **Drugs**
  - *Antibiotics*
    - Amphotericin B
    - Cephalosporins
    - Minocycline
    - Nitrofurantoin
  - Others
    - Sulfasalazine
    - Bleomycin
    - Amiodarone
    - Acebutolol
    - Busulfan
    - Barbiturates
    - Paraquat
    - Cocaine
    - Gold
    - Phenytoin
- **Connective tissue disorders**
  - Systemic lupus erythematosus
  - Rheumatoid arthritis
  - Sjogren syndrome
  - Polymyositis
  - Dermatomyositis
  - Polymyalgia rheumatica
- **Immunological disorders**
  - Common variable immunodeficiency syndrome
  - Essential mixed cryoglobulinaemia
- **Organ transplantation**
  - Bone marrow
  - Lung
  - Renal
- **Miscellaneous**
  - Inflammatory bowel disease
  - Primary biliary cirrhosis
  - Polyarteritis nodosa
  - Haematological malignancies
  - Myelodysplastic syndrome
  - T-cell leukaemia
  - Lymphoma
  - Seasonal syndrome with cholestasis
  - Radiotherapy
  - Environmental exposure (textile printing dye)
  - Penicillium mould dust

Oikonomou A et al. Eur Radiol 2002

Epidemiology

- 1 -2 cases per 100,000 population
- Ages 40-60
- M = F
- 75% patients symptomatic for <2/12 prior to presentation
  - Subacute onset
- 4%-12% of cases of idiopathic interstitial pneumonias
- Not related to smoking
  - Non-smokers 2x risk

Up to date

Pathogenesis

- Aetiology largely unknown
- Reversible response to injury
- Most important process underlying clinical + radiographic manifestations of COP:
  - excessive proliferation of granulation tissue within small airways (proliferative bronchiolitis) + alveolar ducts
  - associated with chronic inflammation in surrounding alveoli
  - minimal disruption of the lung architecture
  - buds of granulation tissue contain fibroblasts and fibrin exudates

Up to date

Clinical Presentation

- Can mimic community acquired pneumonia
- Symptoms:
  - Fever, malaise + fatigue
  - Persistent non-productive cough
  - Dyspnoea with exertion
  - Anorexia + wt loss >5kg

Up to date

Chest Radiograph

- Classical manifestations quite distinctive:
  - Bilateral, diffuse/patchy areas of air-space consolidation and ground-glass opacities
  - Peripheral + lower zone distribution
  - Normal lung volumes
  - Tendency to progress, recur and/or migrate
- Rarer changes:
  - Unilateral distribution,
  - pleural effusion,
  - pleural thickening,
  - + cavities

Up to date

CT Chest

- Classical CT patterns
  - patchy air-space consolidation
    - Peripheral + lower zone distribution
  - ground-glass opacities
  - small nodular opacities
  - air bronchograms + bronchial wall thickening with dilation in consolidated areas
- CT findings mirror CXR appearance, but often reveal more extensive

Cordier. Eur Resp Jr 2006

Variant CT Patterns

- Increasing awareness that imaging patterns of OP may deviate from the typical picture:
  - Focal lesion
  - Nodular pattern
  - Bronchocentric pattern
  - Linear + band-like pattern
  - Perilobular patten
  - Progressive fibrotic pattern

Oikonomou A et al. Eur Radiol 2002

Treatment

- Treatment of choice = corticosteroids
- Response is prompt in most cases
- Spontaneous improvement is rare

Prognosis

- Complete recovery occurs in 2/3 of patients treated with glucocorticoids in 1-3 months
- Relapses may occur
- 1/3 of patients demonstrate persistent disease

Up to date

Fuji-san

Fuji-san