

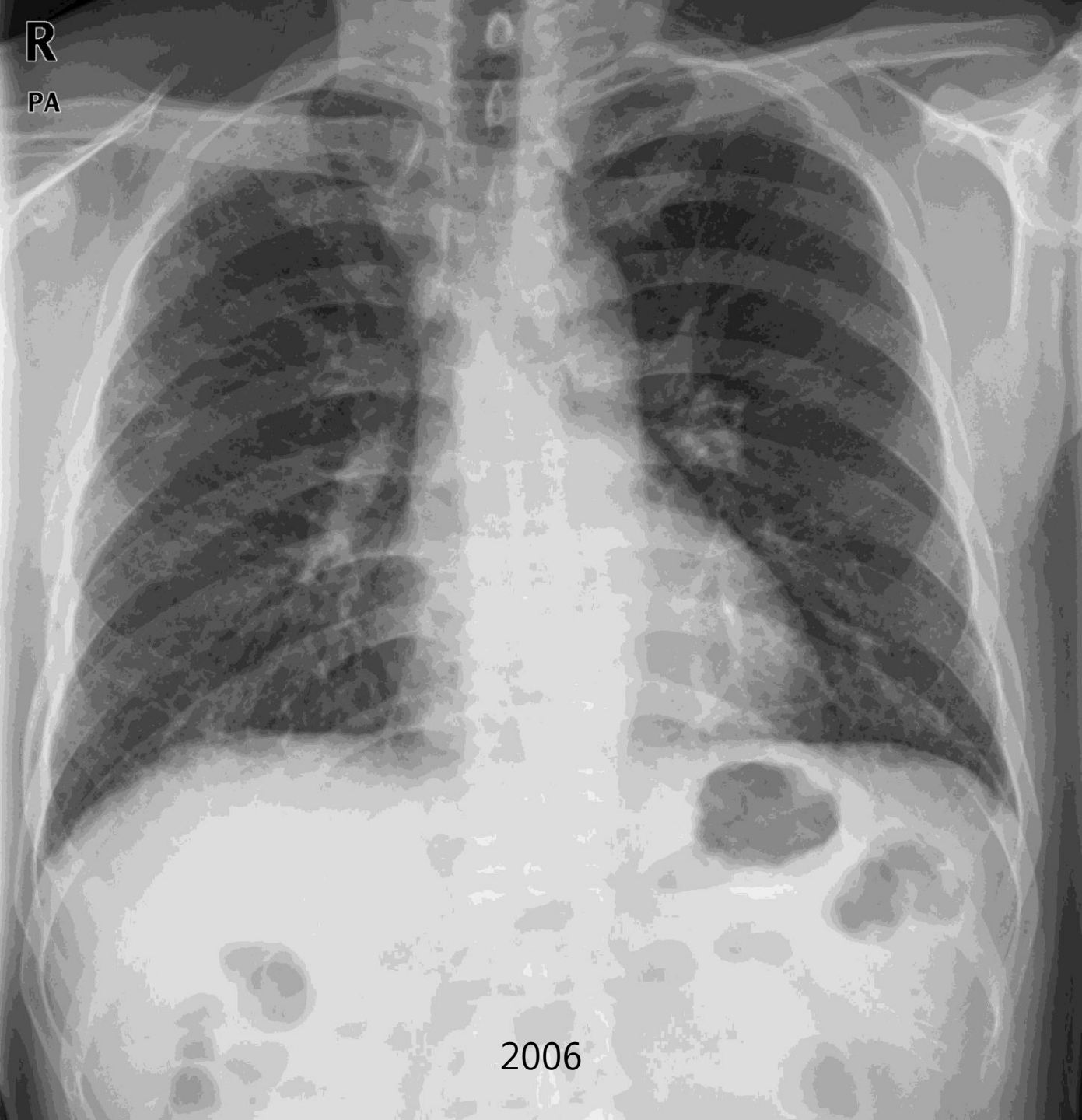
ILD school case

2016.3.19

분당서울대병원 호흡기내과
박종선

Case 1. 유 O 열, M/55

- 2006년 12월, 건강검진 CXR이상 소견으로 내원
- 무증상
- PFT:
 - FVC 3.08L (74%), FEV1 2.62L (88%), FEV1/FVC 85%
 - DLCO 17 mL/mmHg/min (80%)
- PMx> hyperlipidemia
- SHx> 회사원
- Non-smoker
- P/Ex> no definite crackle

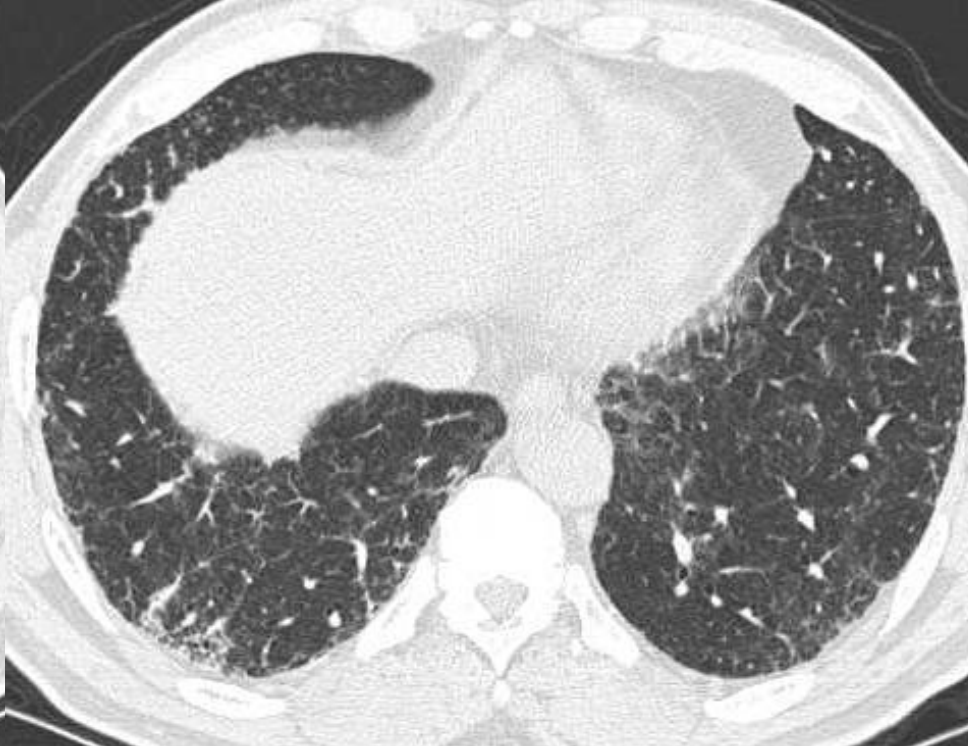


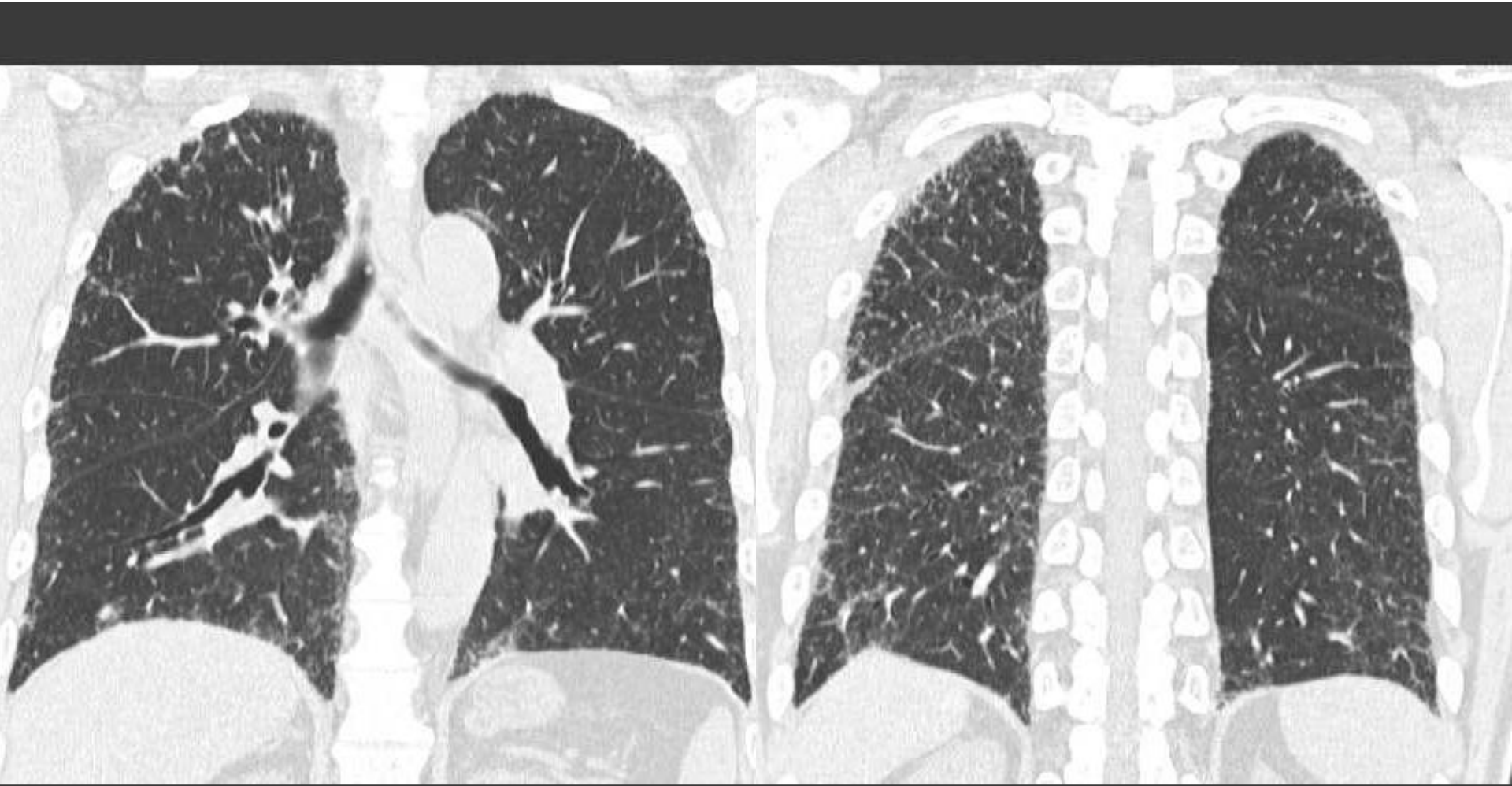
R

PA

2006



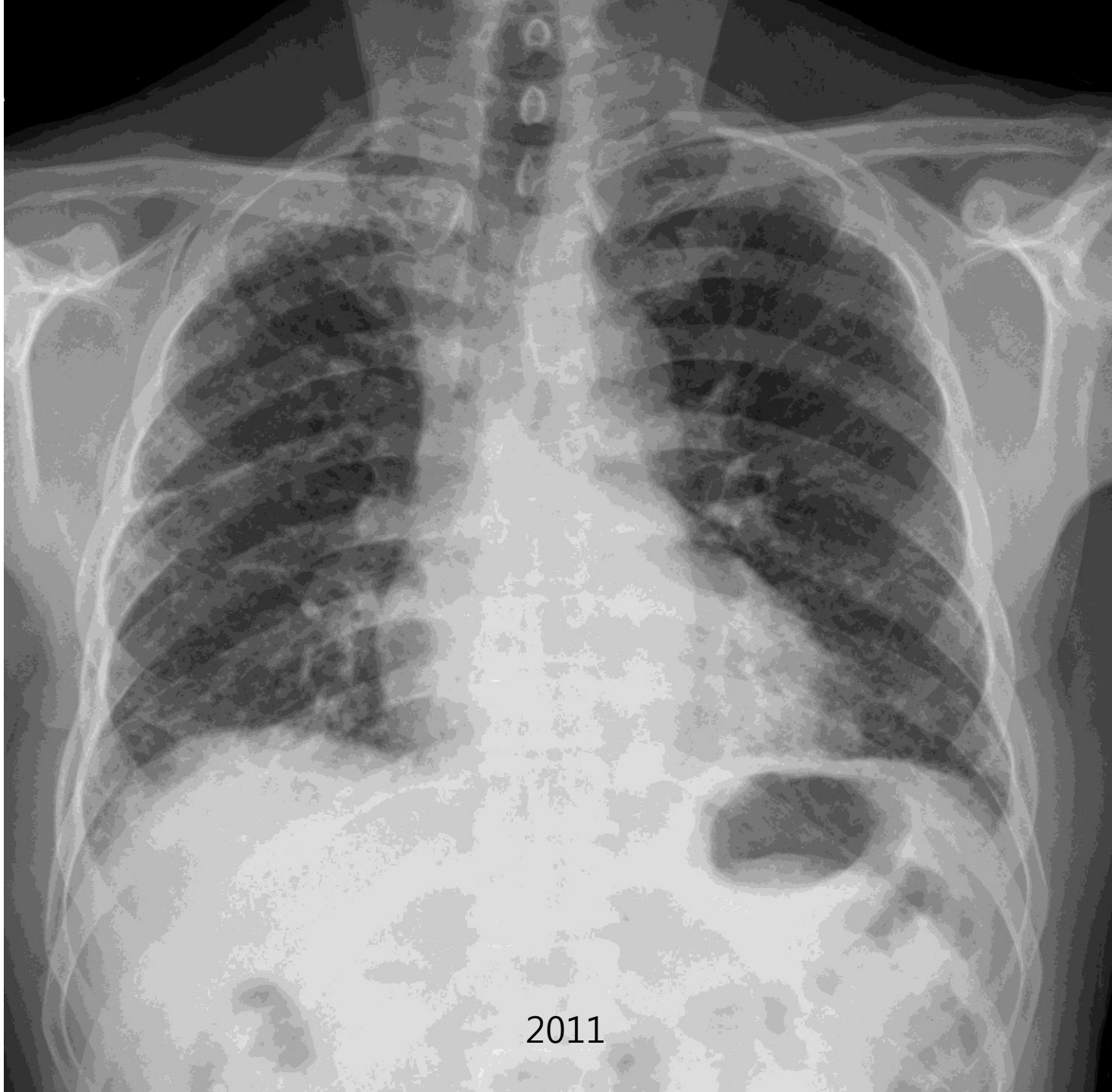




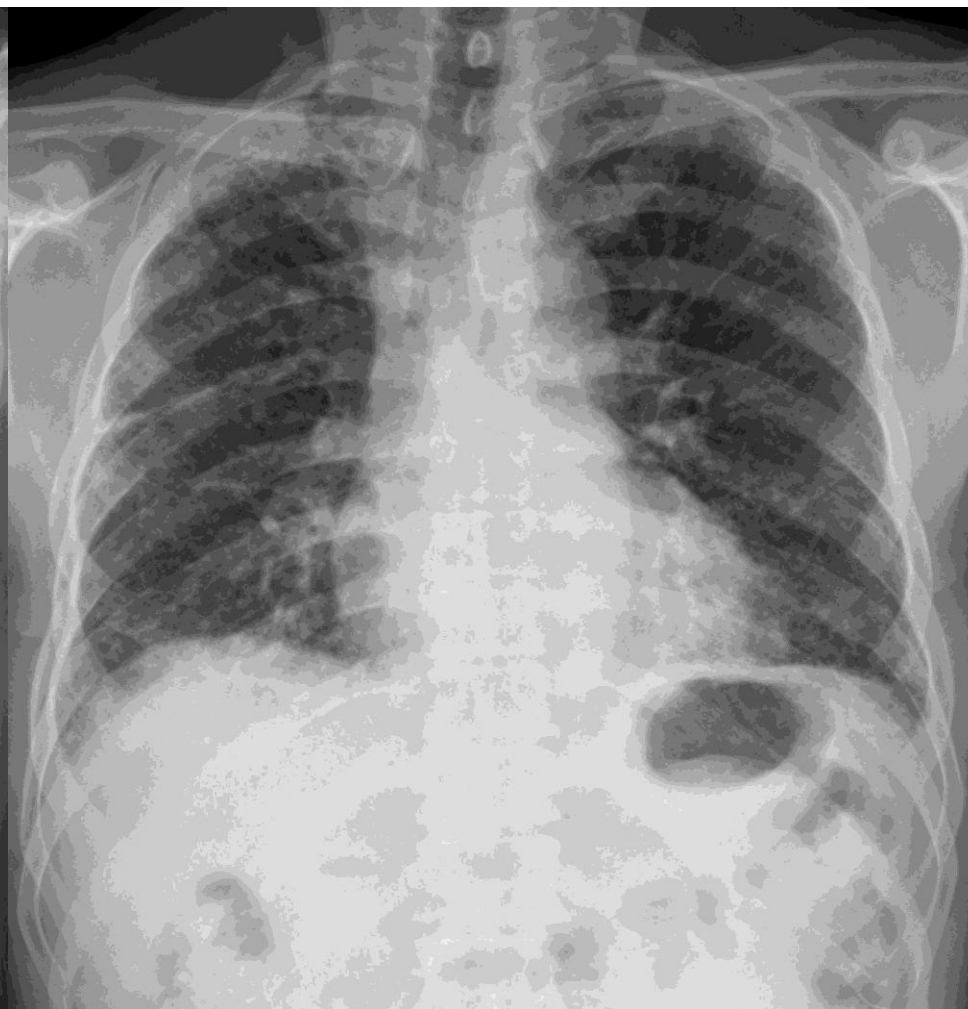
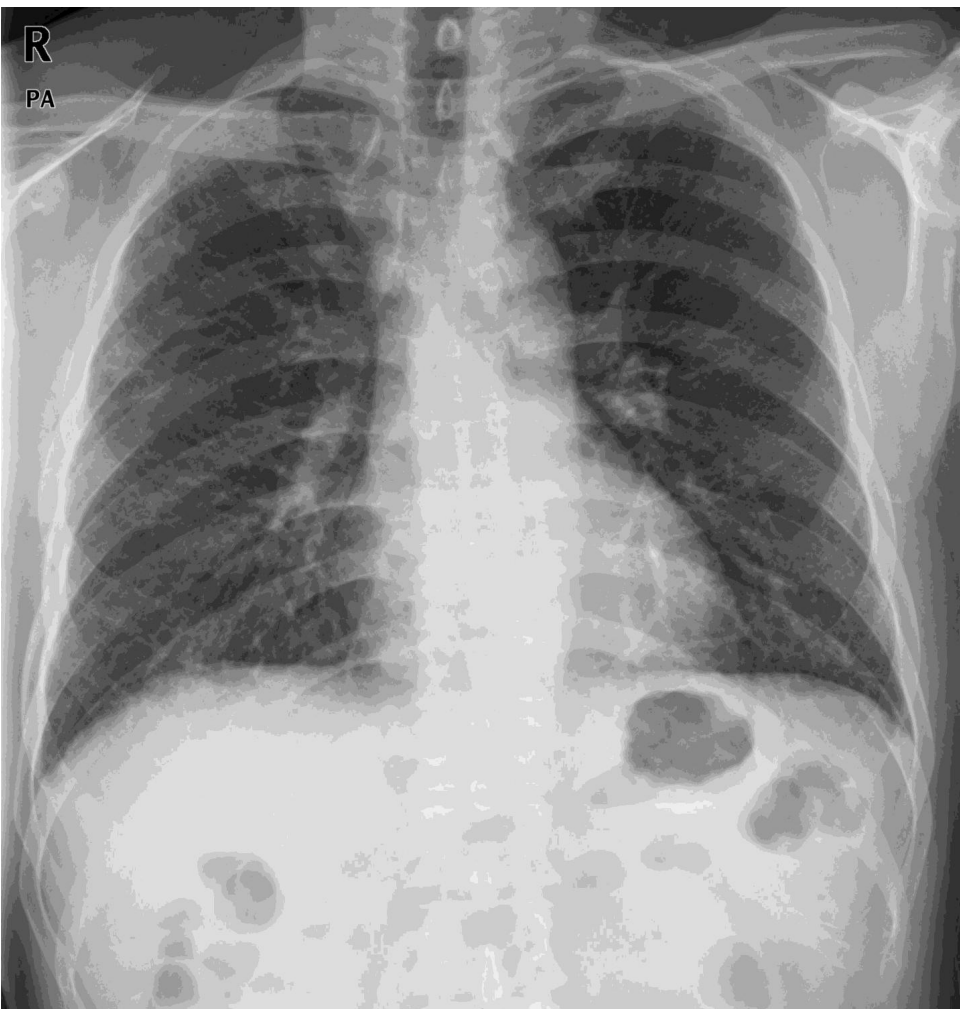


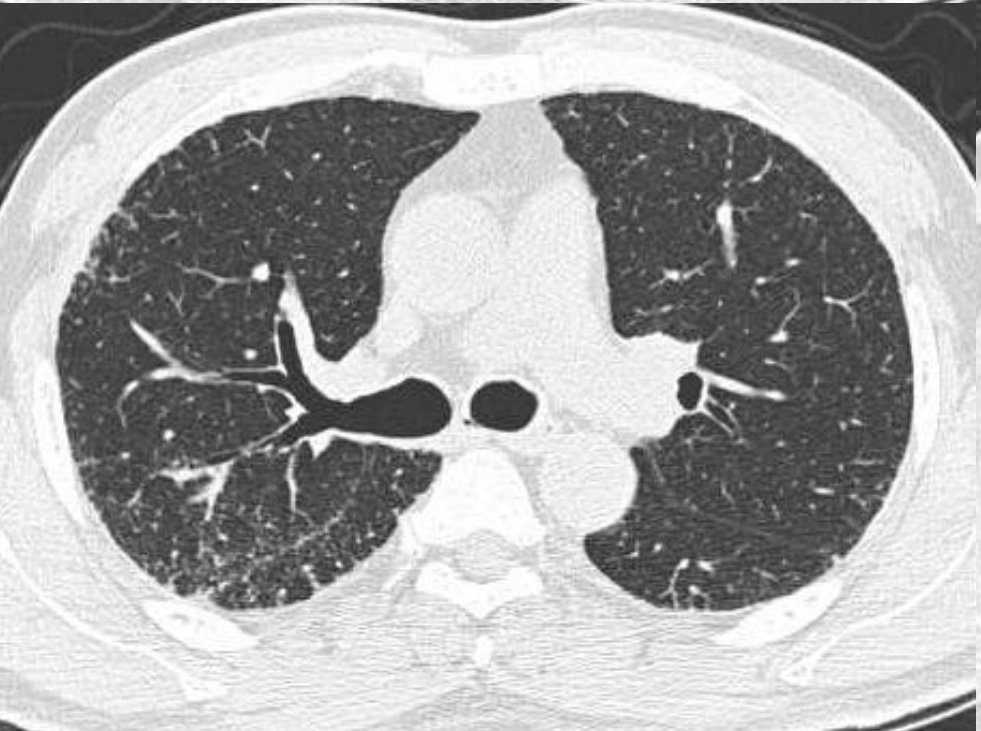
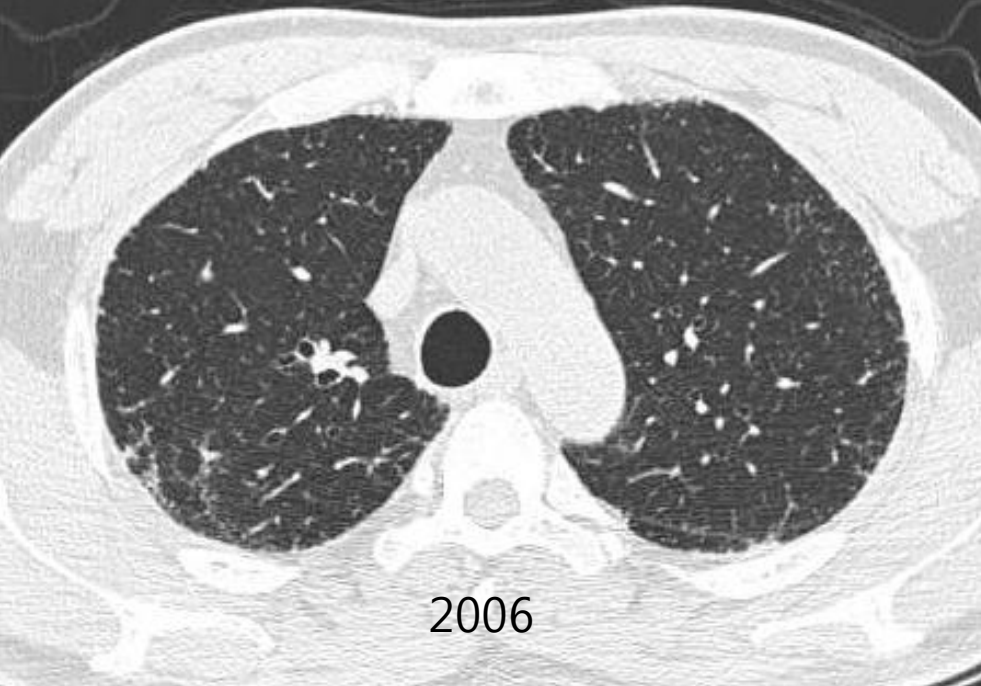
- 2007년 12월, 1yr fu시 no symptom
- fu CT: no definite progression
- P/Ex > BLLF late inspiratory fine crackle
- PFT:
 - FVC 2.92L (71%), FEV1 2.55L (84%), FEV1/FVC 87%
 - DLCO 13.8 mL/mmHg/min (65%)

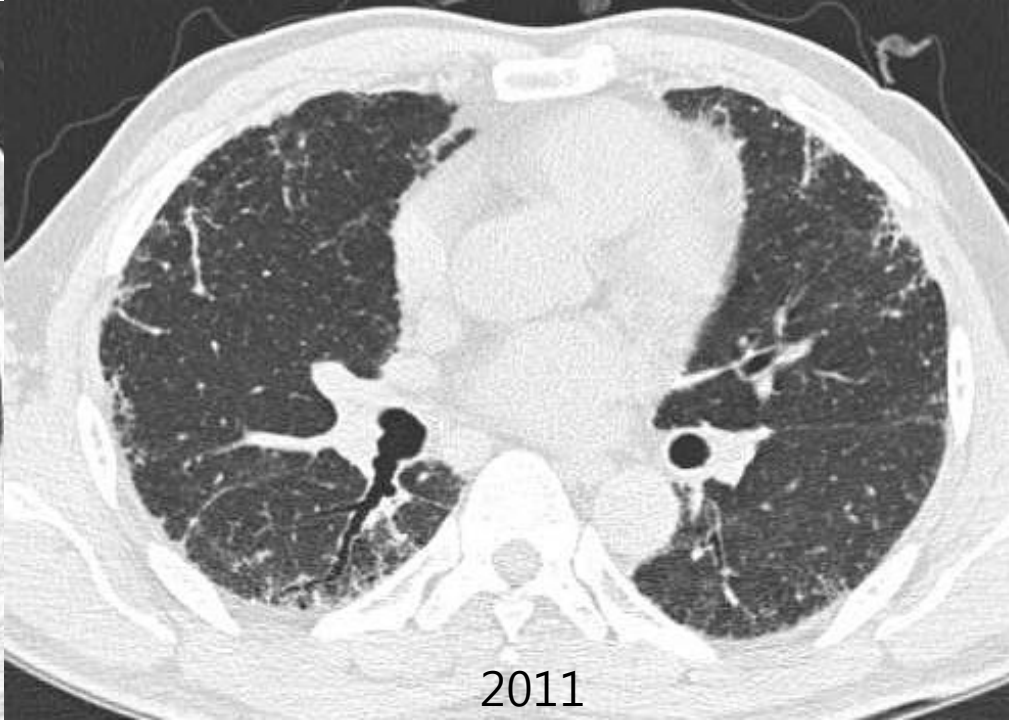
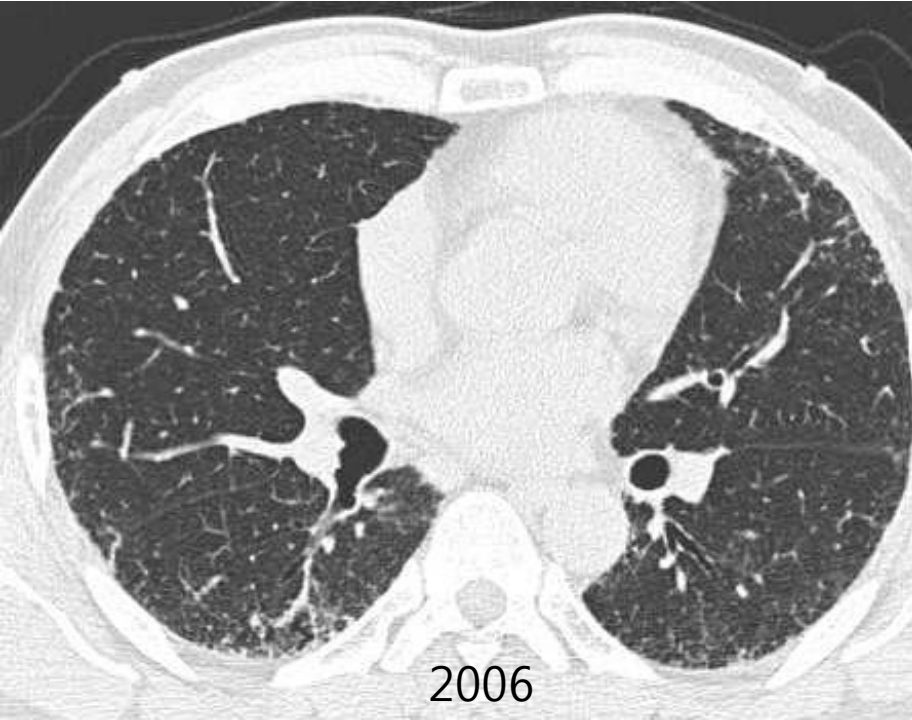
- 2011년 4월, 2개월간 cough, DOE 있어 외래 방문
 - FVC 2.09L (54%), FEV1 2.05L (65%), FEV1/FVC 88%
 - DLCO 13.0 mL/mmHg/min (66%)
- BAL > RBC 120/ $\mu\ell$, WBC 90/ $\mu\ell$, macrophage 84%, neutrophil 11%, lymphocyte 5%
- Lab > RF(-) anti-CCP(-)

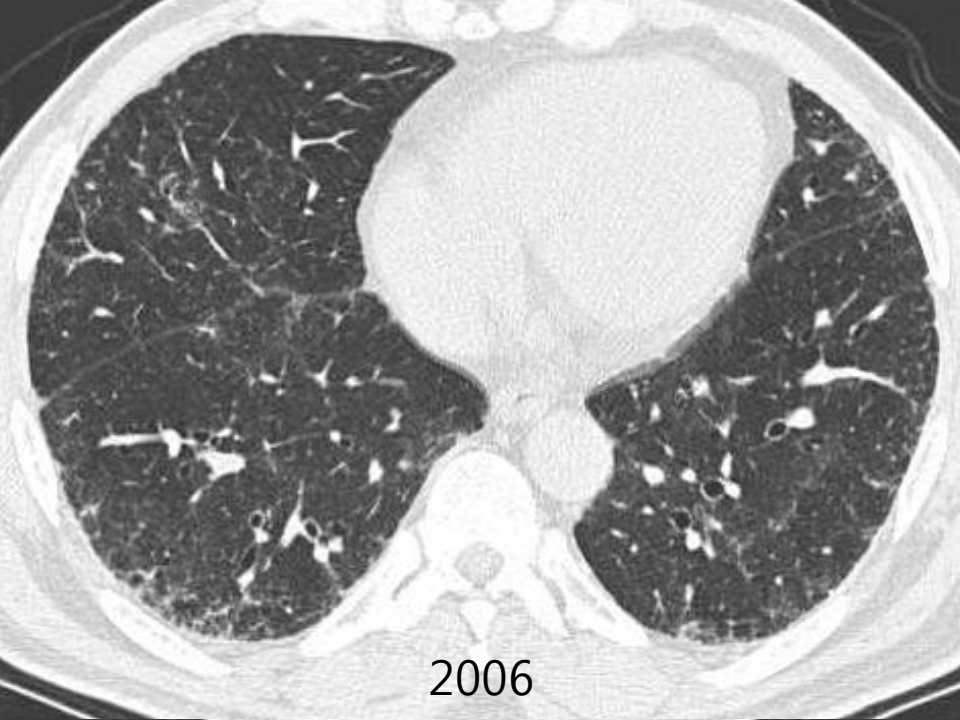


2011







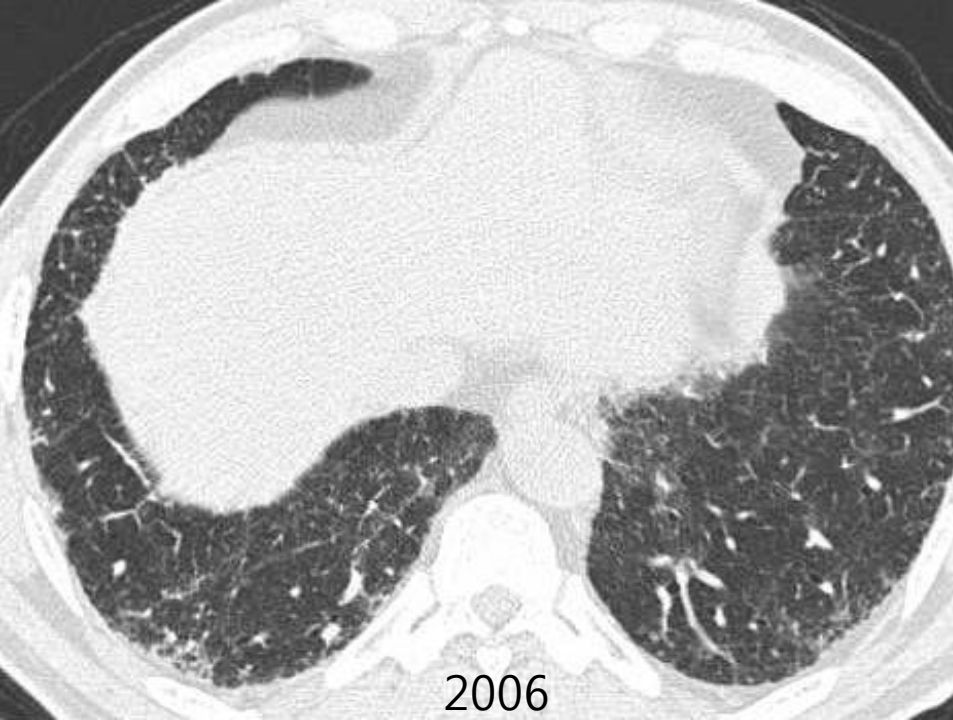


2006

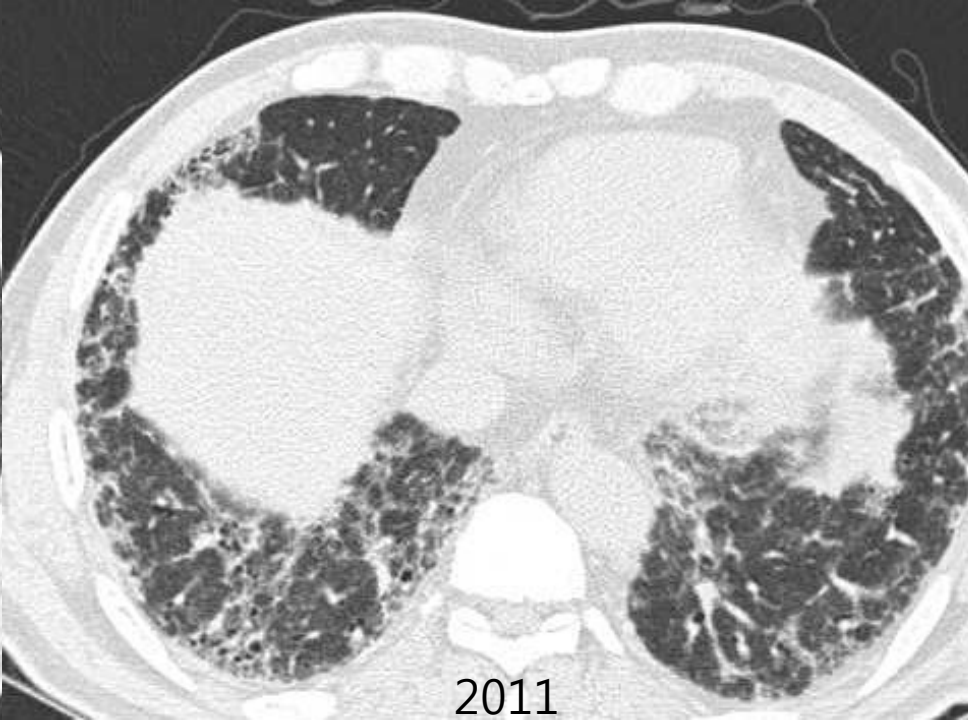


2011





2006



2011





2006

2011



2006



2011

- **2011.11월 VATS lung biopsy**

Lung, RUL & RLL wedge resection;

Diffuse interstitial fibrosis with

- 1) honeycomb change
- 2) fibroblastic foci
- 3) temporal variegation
- 4) alveolar bronchiolization
- 5) type II pneumocyte hyperplasia
- 6) intraluminal necroinflammatory exudate
- 7) smooth muscle proliferation

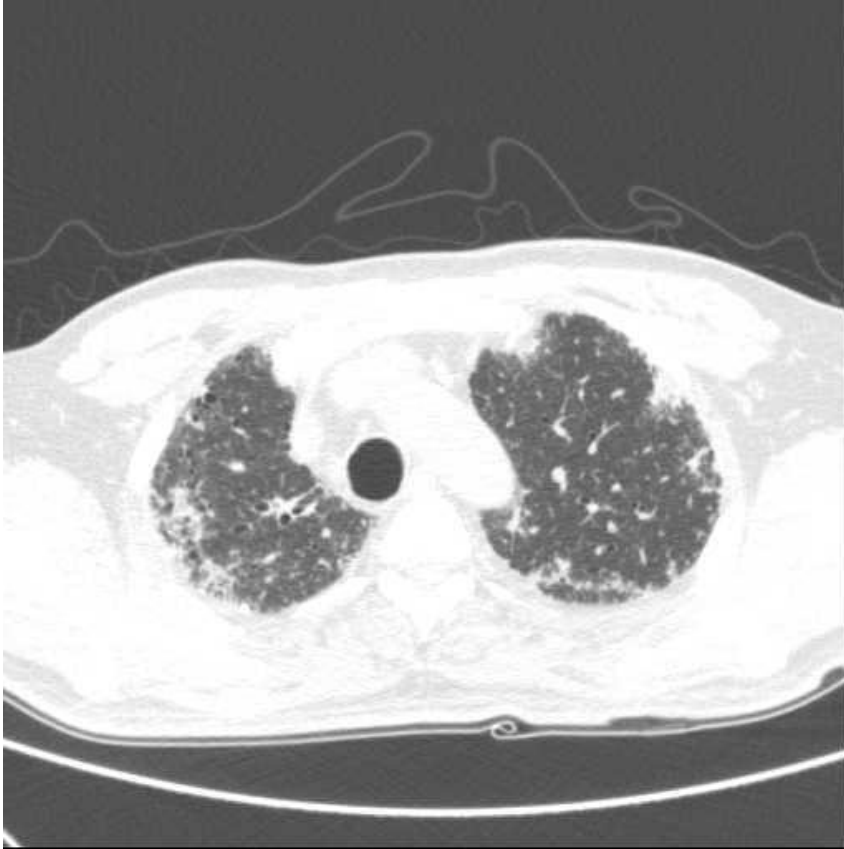
Consistent with usual interstitial pneumonia

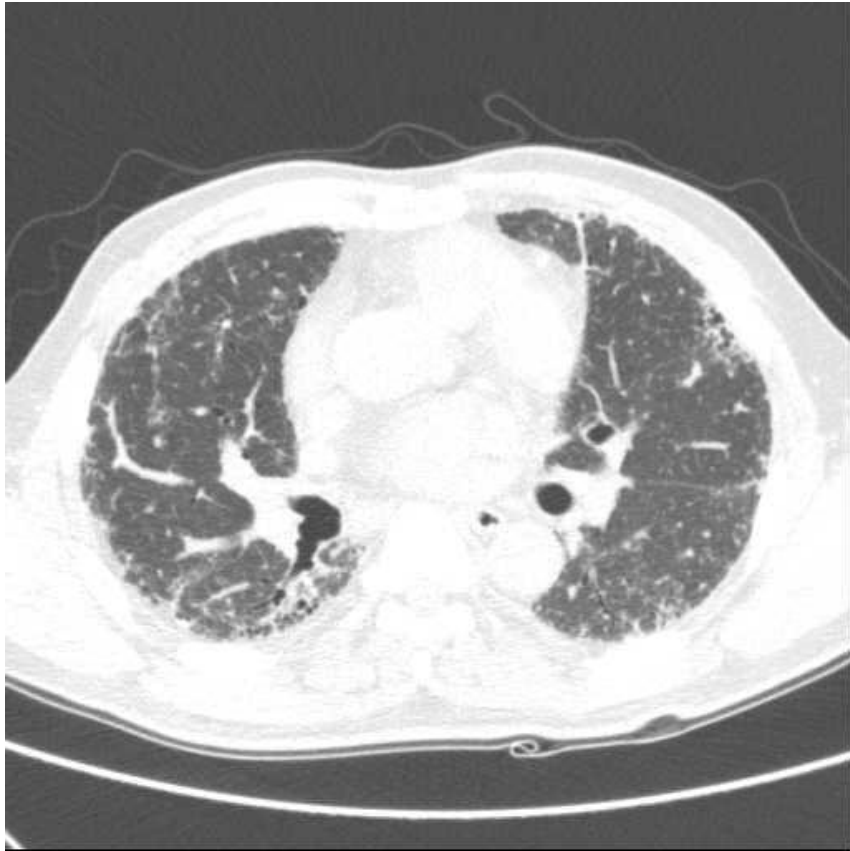
- 2011년 11월
- Diffuse interstitial fibrosis with
 - 1) honeycomb change
 - 2) fibroblastic foci
 - 3) temporal variegation
 - 4) alveolar bronchiolization
 - 5) type II pneumocyte hyperplasia
 - 6) intraluminal necroinflammatory exudate
 - 7) smooth muscle proliferation

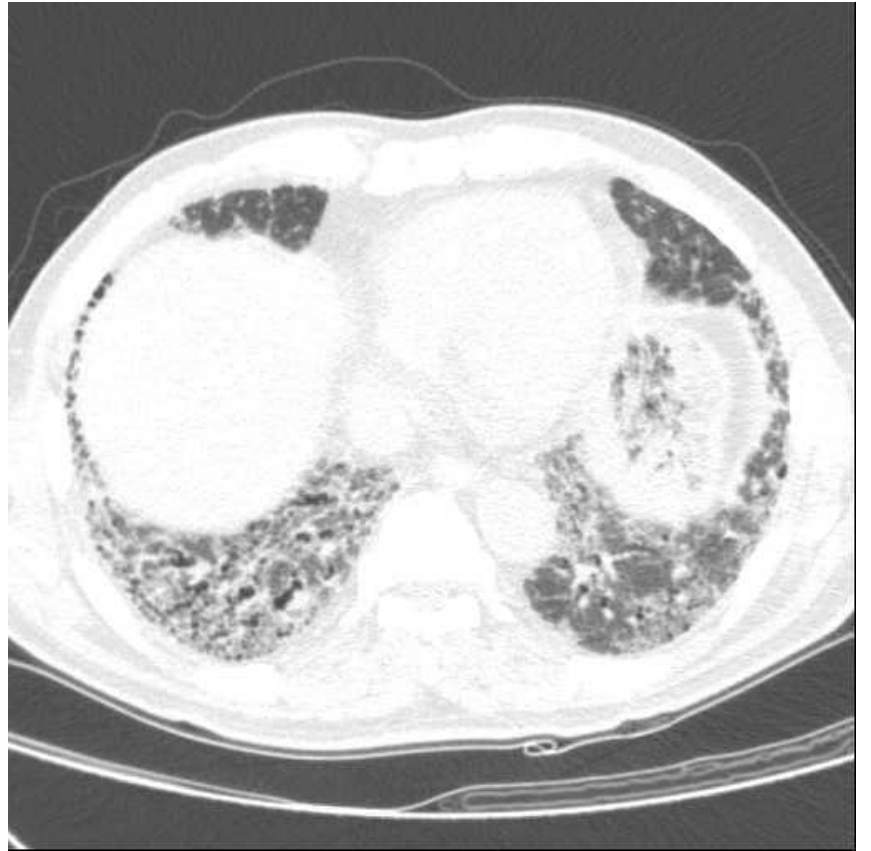
Consistent with usual interstitial pneumonia

- 2011년 11월 BAL
 - RBC 120/ $\mu\ell$, WBC 90/ $\mu\ell$,
 - macrophage 84%, neutrophil 11%, lymphocyte 5%

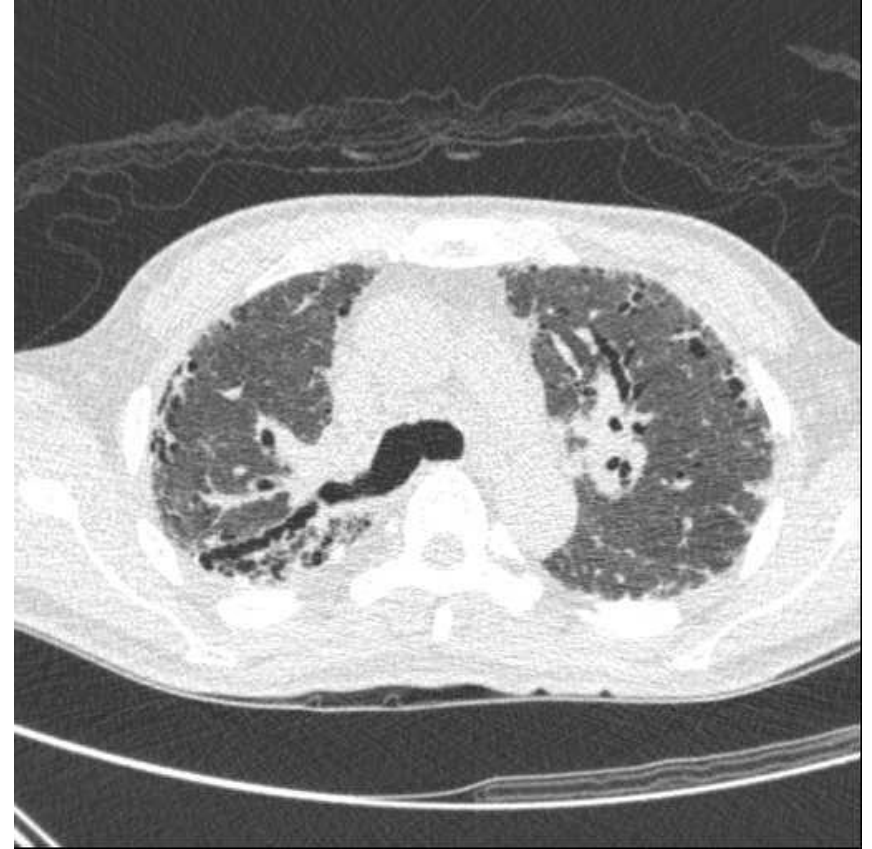
2013

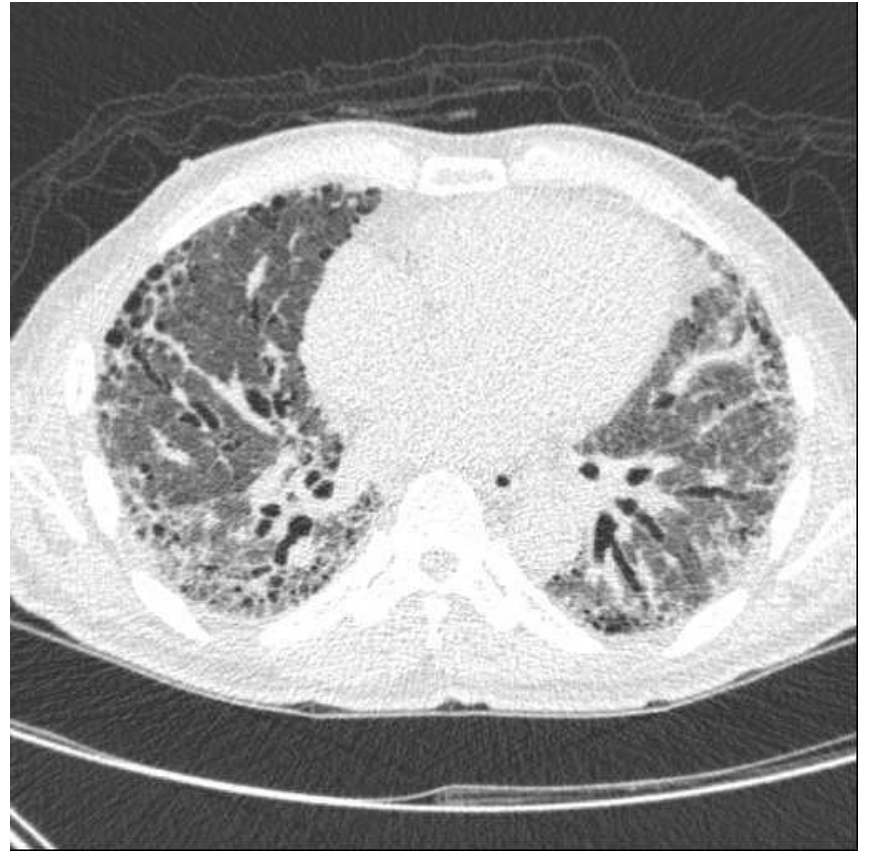


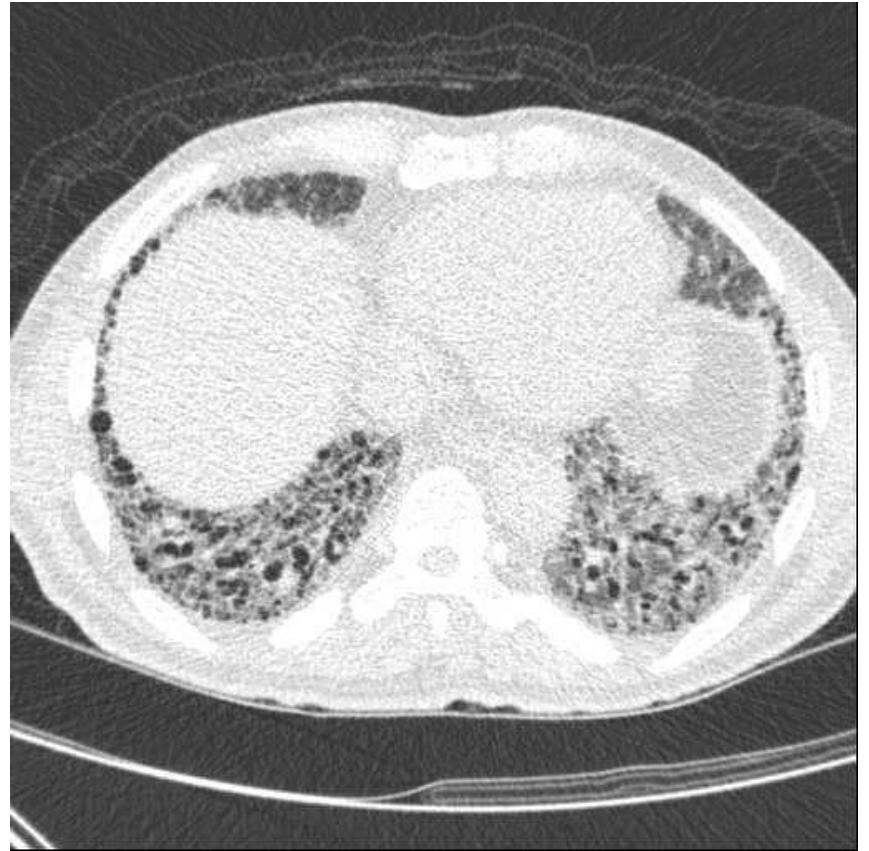


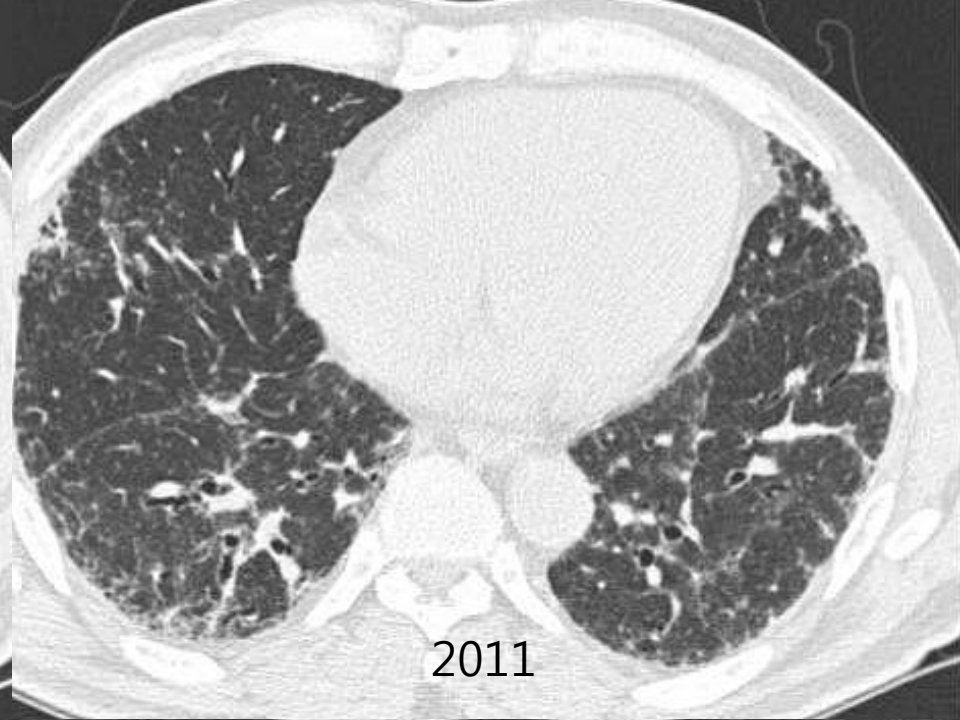
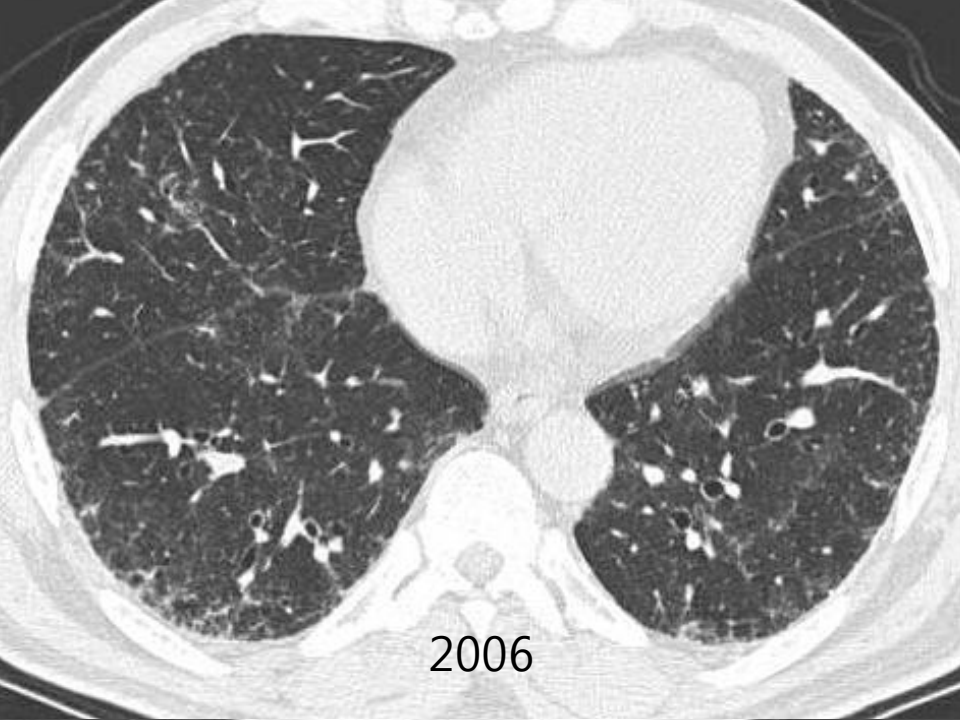


2014

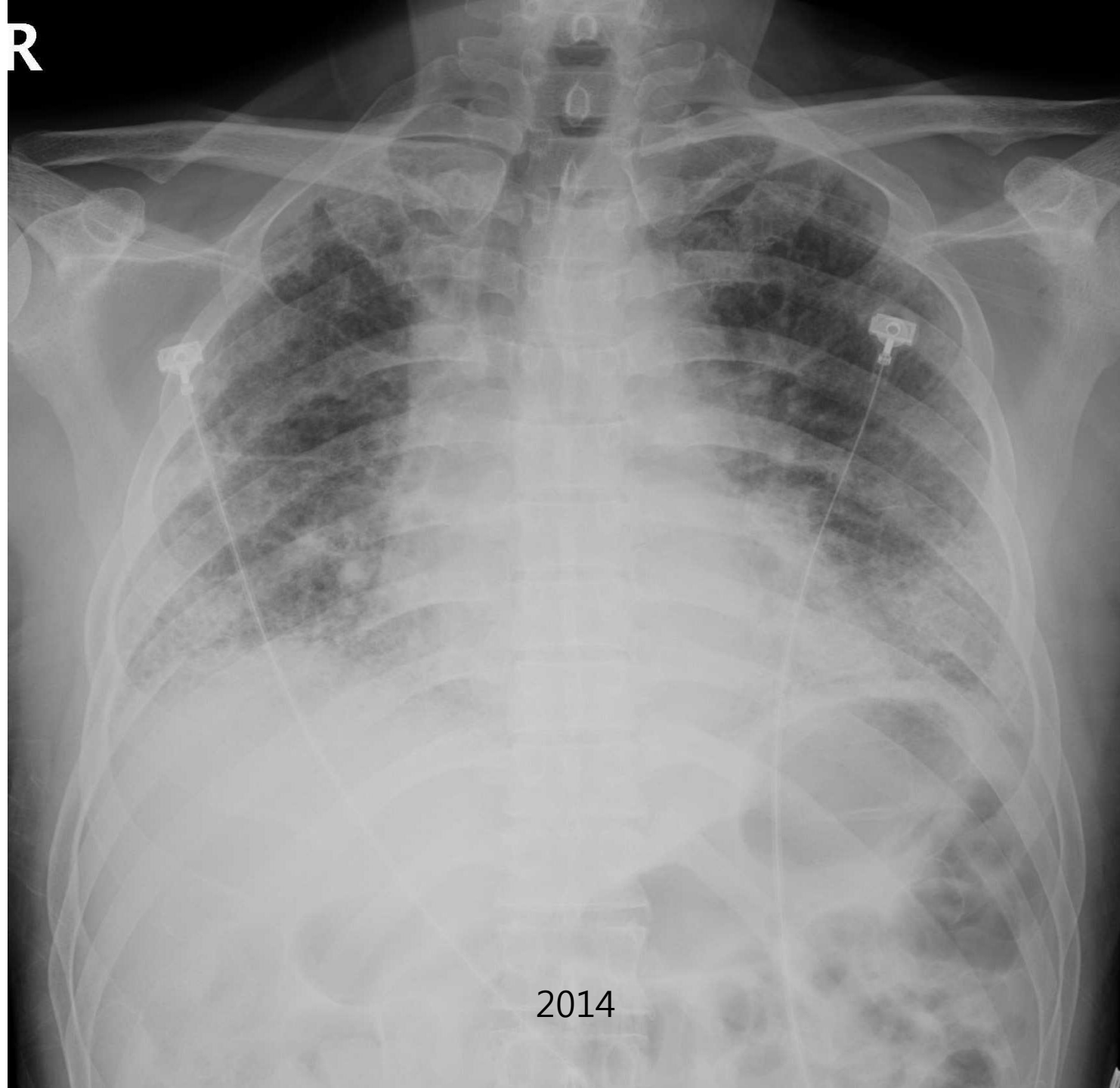








R



2014

Pathology

- 2012.2.24- high dose NAC
- 2013년 1월부터 기침, 호흡곤란 악화, mMRC2, FVC 46%
- 2013.2.25-6.30 pirfenidone
- 2013.11 dyspnea mMRC3, clinical trial enroll
- 2014.4 resting SpO2 90%, FVC 36%, home O2, Lung TPL 등록

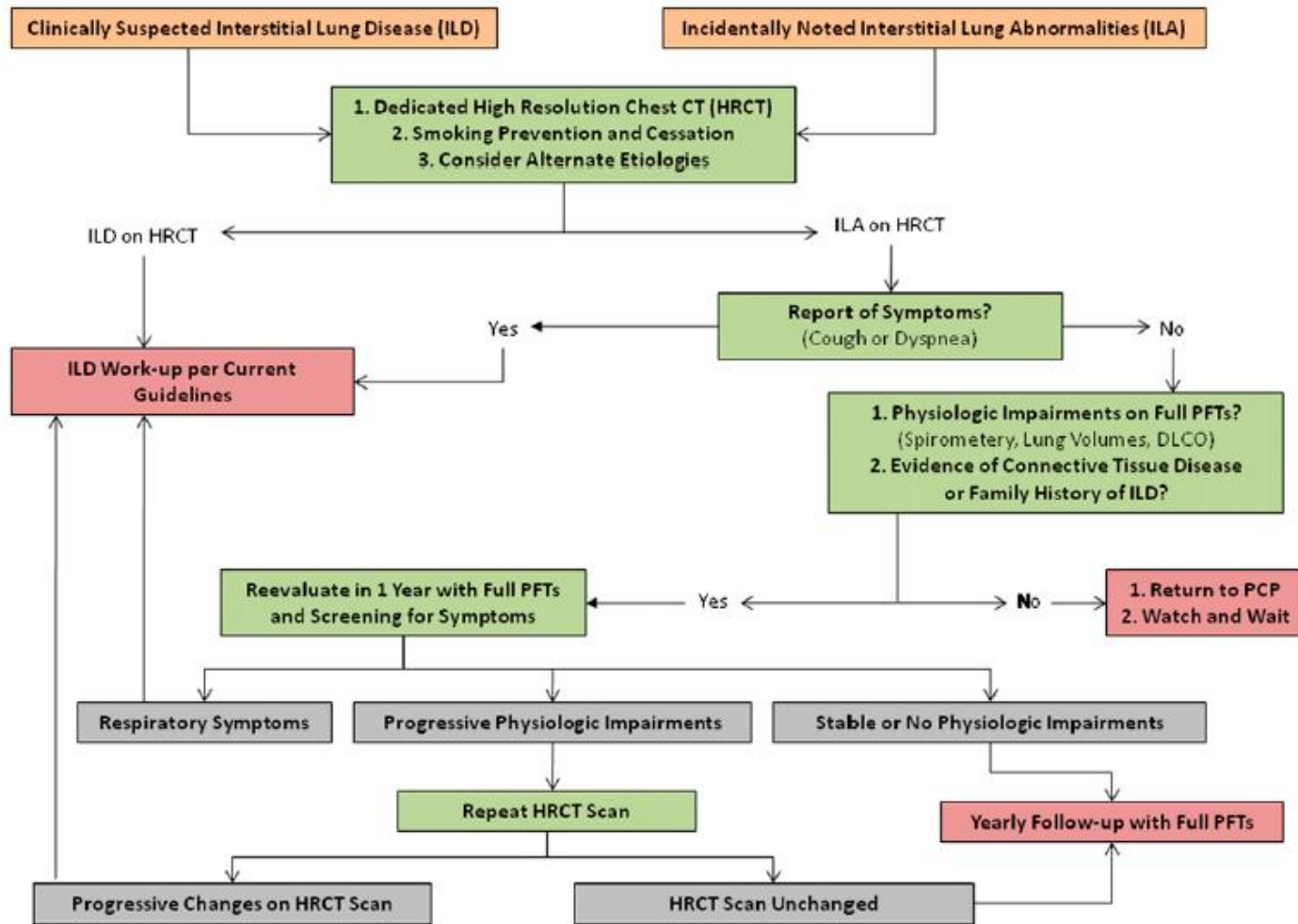
- 2014.7.30 bilateral lung transplantation
- 2014.8.26-28, pd pulse d/t acute rejection
- 면역억제제 복용하며 현재까지 일상생활에 큰 제약 없이 지내고 있음.

Pathology

Discussion

- Diagnosis
 - Asymptomatic period
 - HRCT finding, possible UIP?
 - Timing for lung biopsy?

Diagnostic algorithm and recommended follow-up of individuals with subclinical ILD

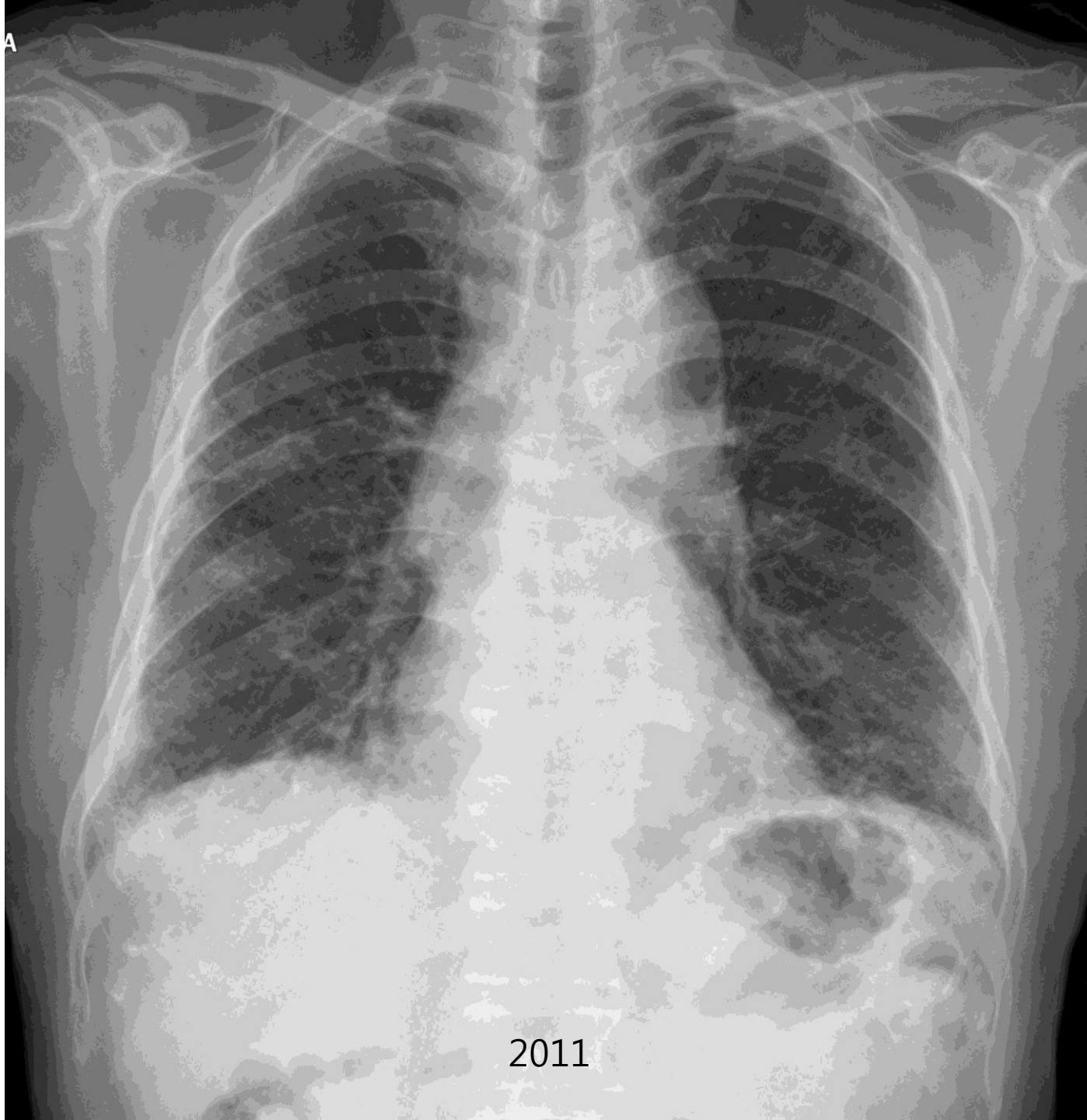


Case 2. 이 O 희, M/70

- 2011년 11월, 6개월간의 cough, sputum, DOE mMRC1
 - FVC 3.04(84%) FEV1 2.40(97%)
 - FEV1/FVC=79%
 - DLCO 13.7(81%)
- PMHx> DM, CKD, Cr 1.9
- SHx> 회사원으로 근무 후 퇴직
- Ex-smoker: 30PYs, 15YA quit

Case 2. 0 | O ± |, M/70

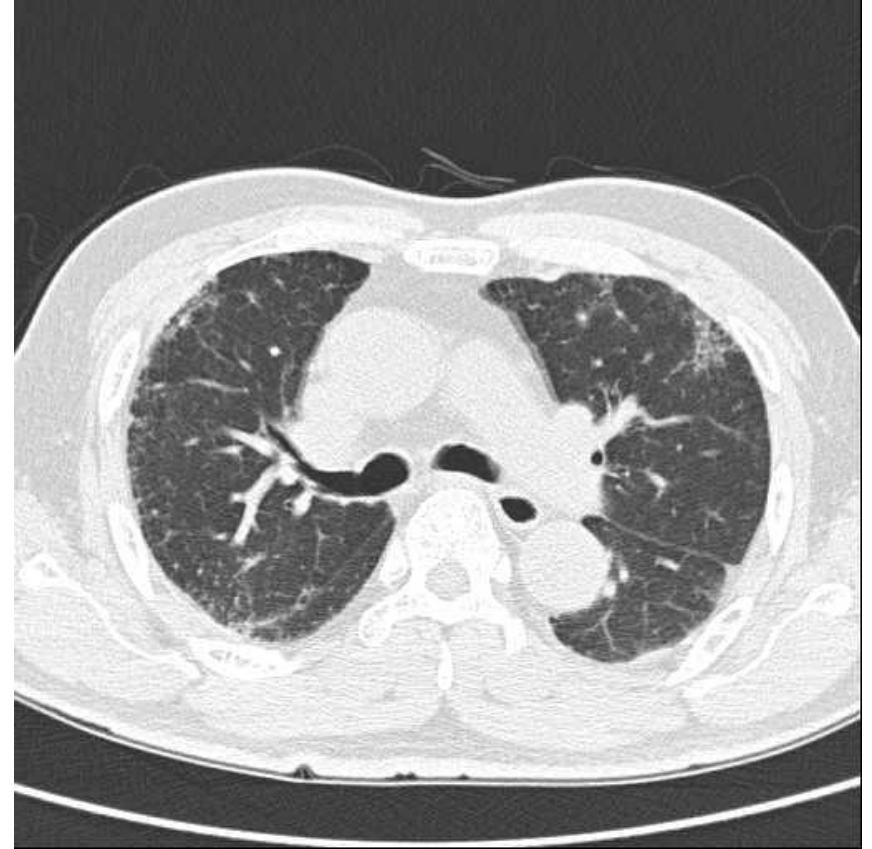
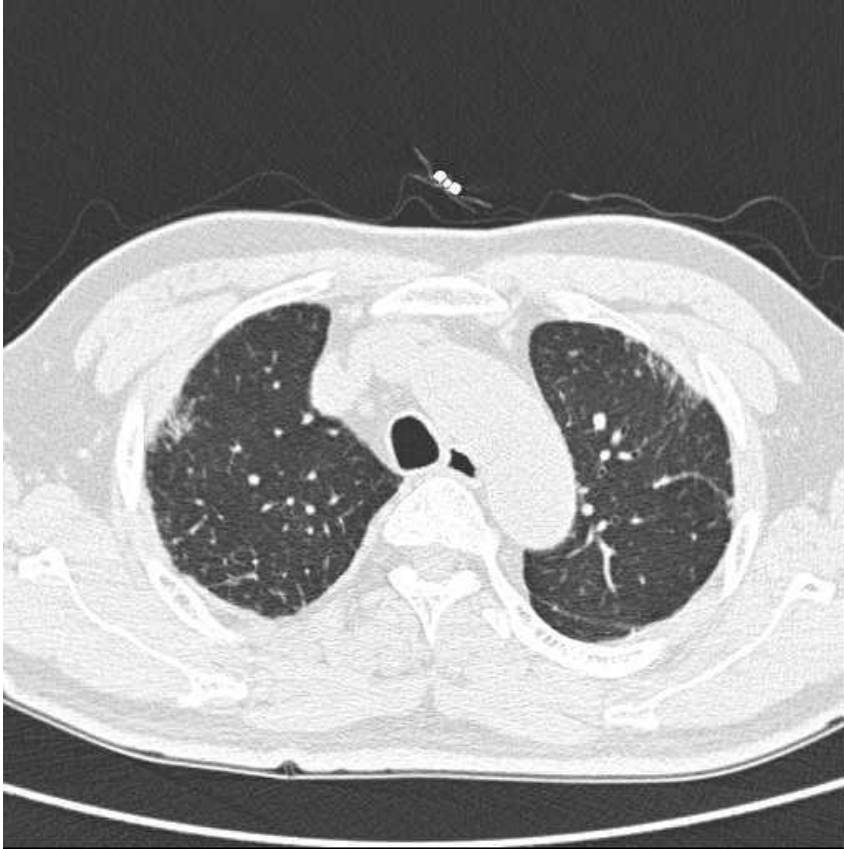
- ROS > C/S(+ / +) arthralgia(-)
no other rheumatologic symptoms
- P/EX > BLLF late inspiratory crackle
- BAL > RBC 270 WBC 250 macrophage 32%,
neutrophil 59%, lymphocyte 9%
- Lab > RF(-) anti-CCP(-)

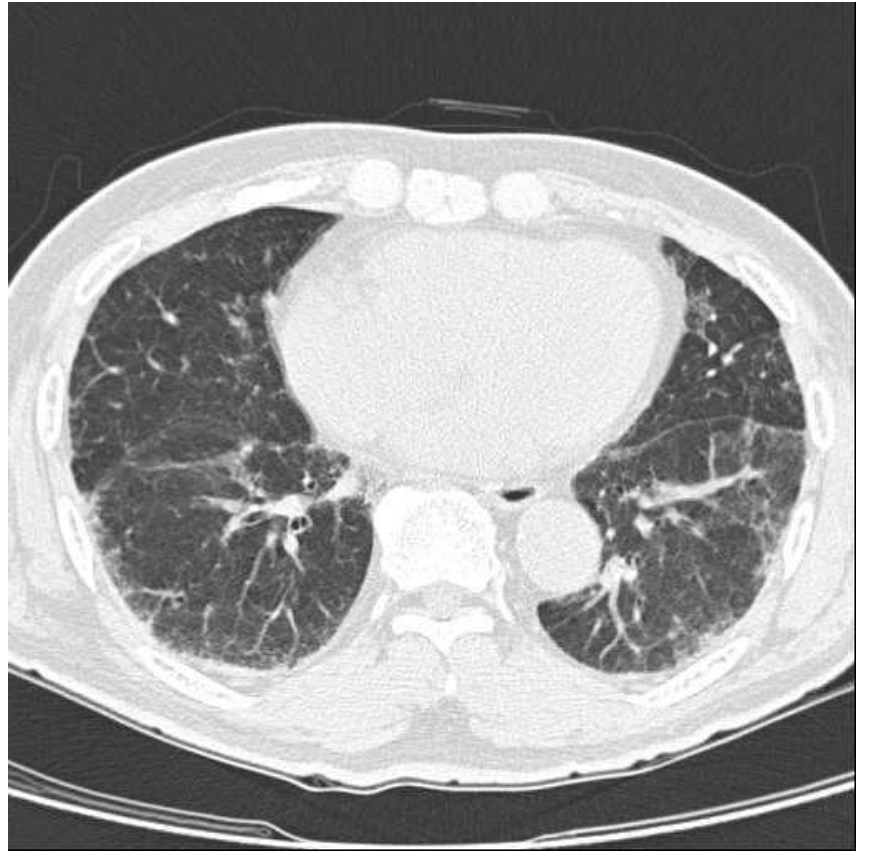


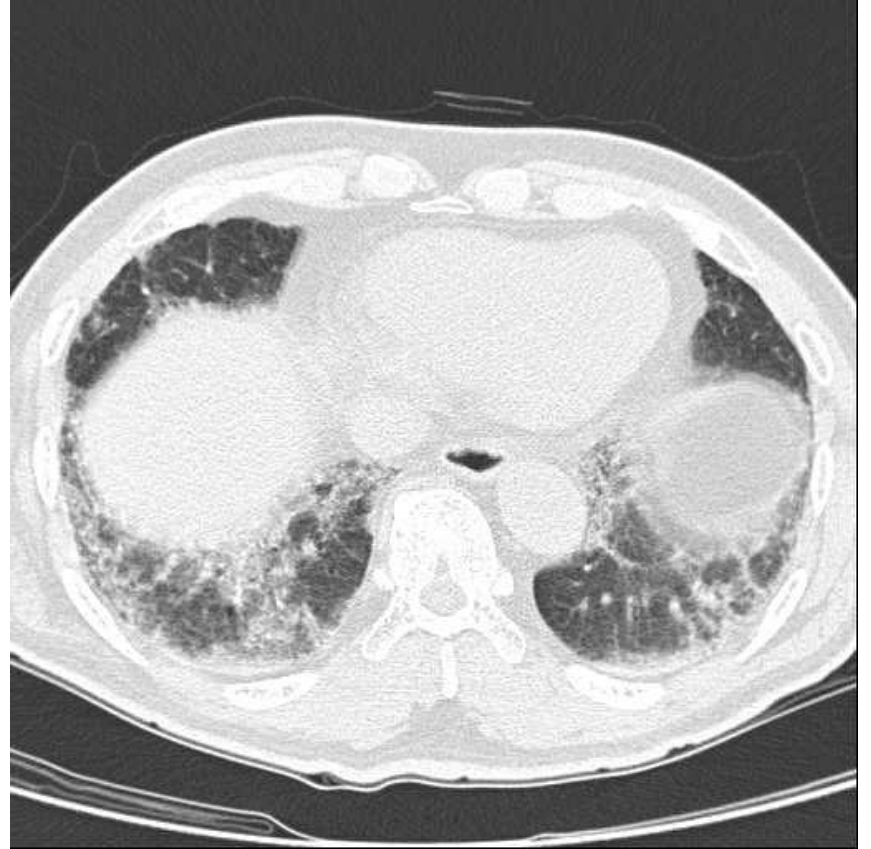
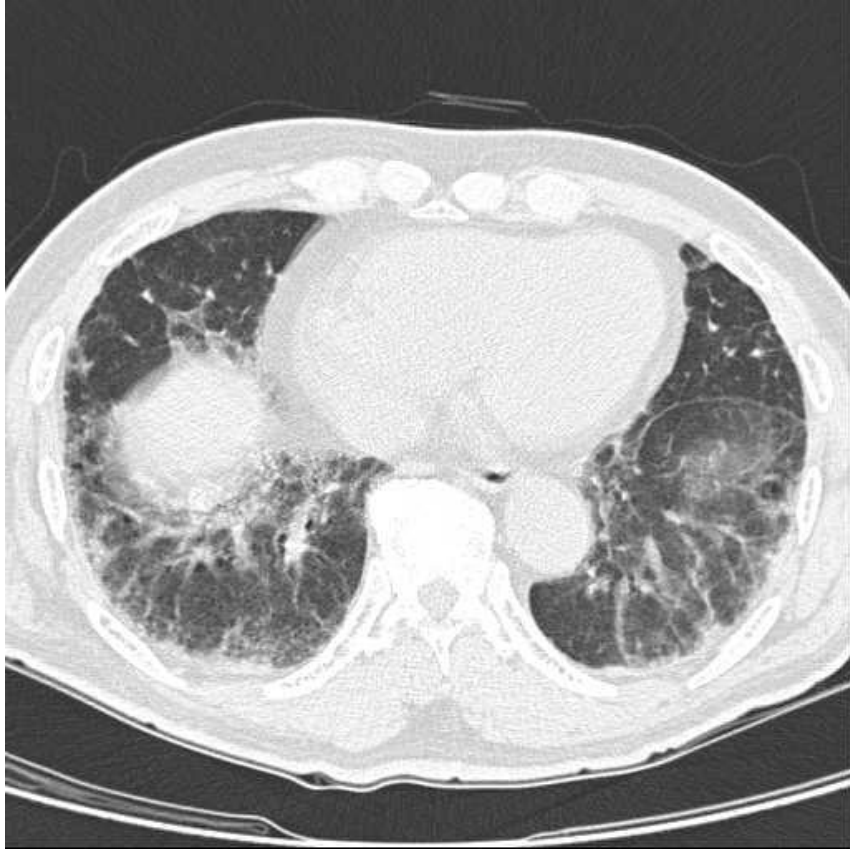
A

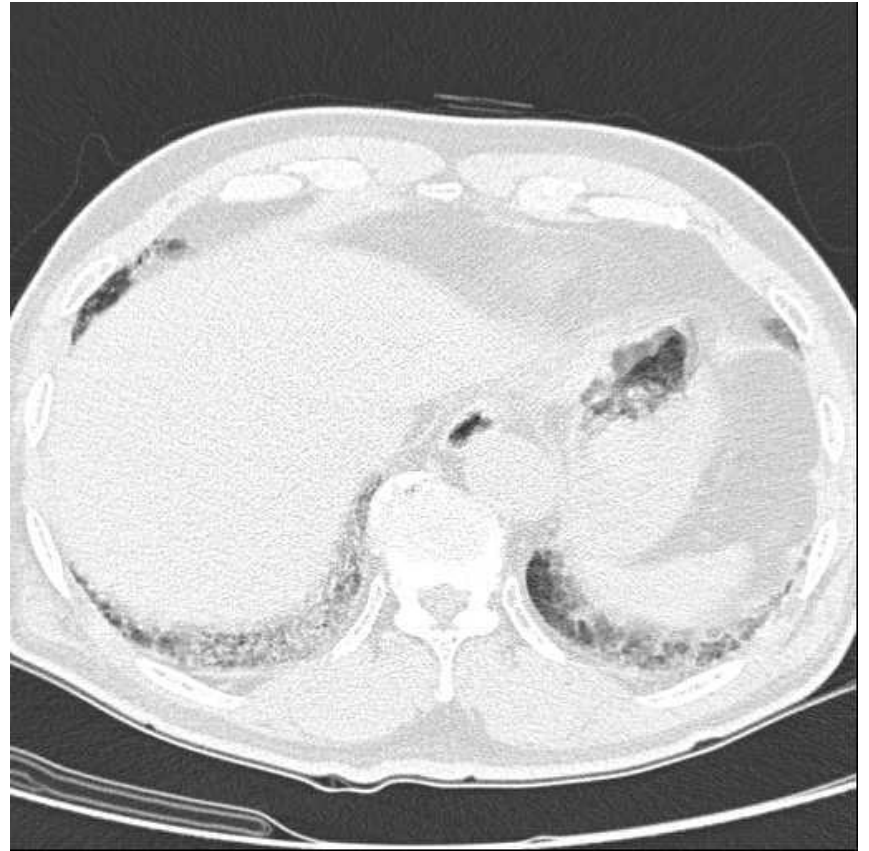
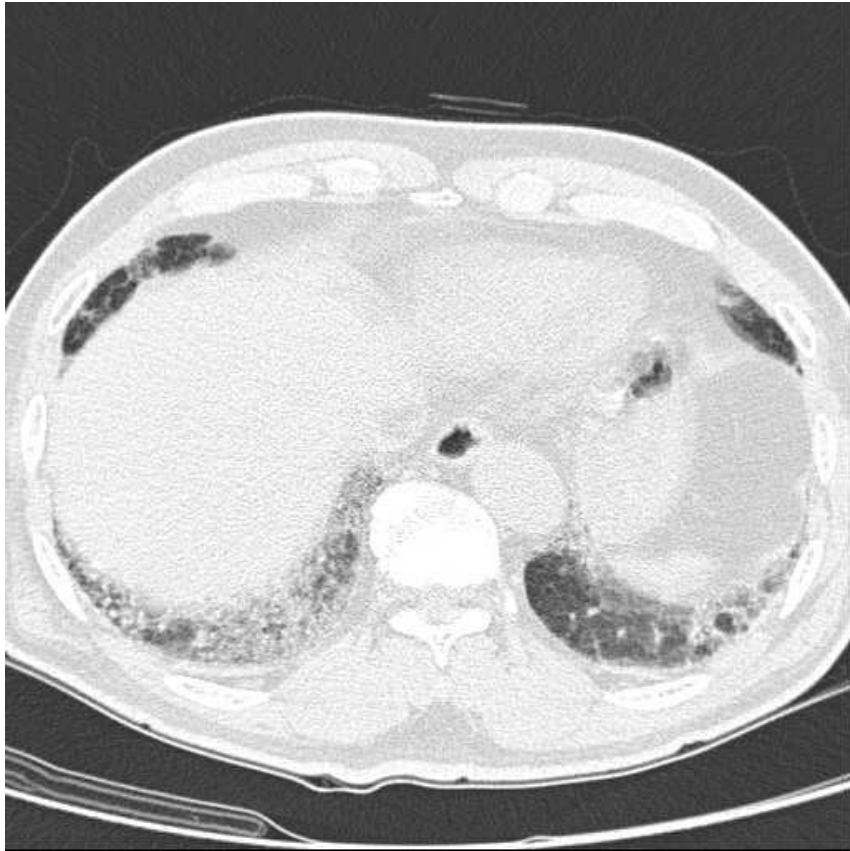
2011

2011









- **2011.12.2 VATS lung biopsy**

Lung, RUL & RLL, wedge resection :

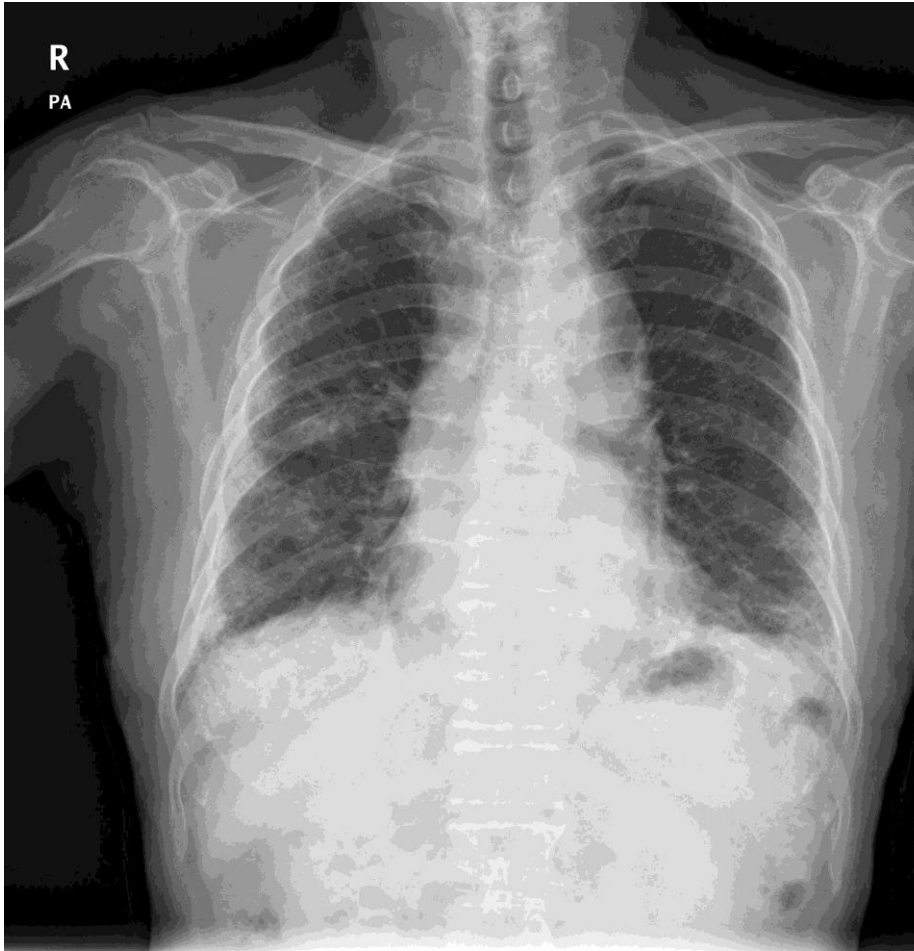
Subpleural interstitial fibrosis with

- 1) honeycomb change
- 2) bronchiolar metaplasia
- 3) focal temporal variegation and fibroblastic foci
- 4) lymphoid follicle and lymphoid aggregate, multifocal
- 5) intraalveolar mucopurulent exudate

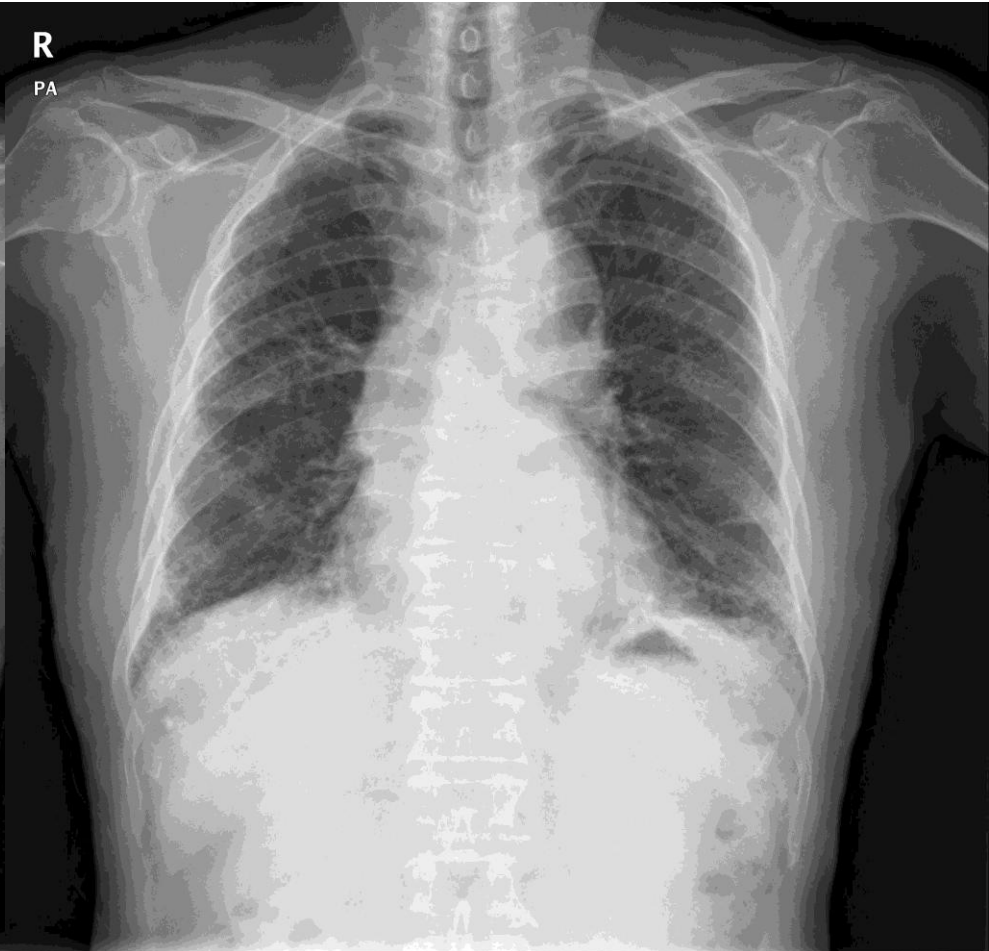
: Consistent with usual interstitial pneumonia

Pathology

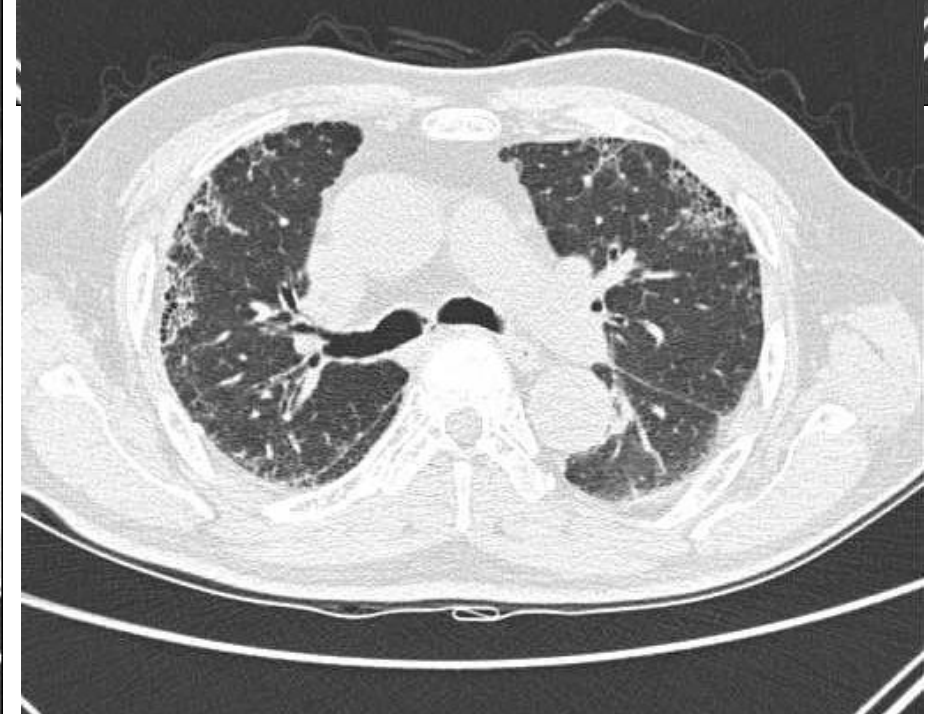
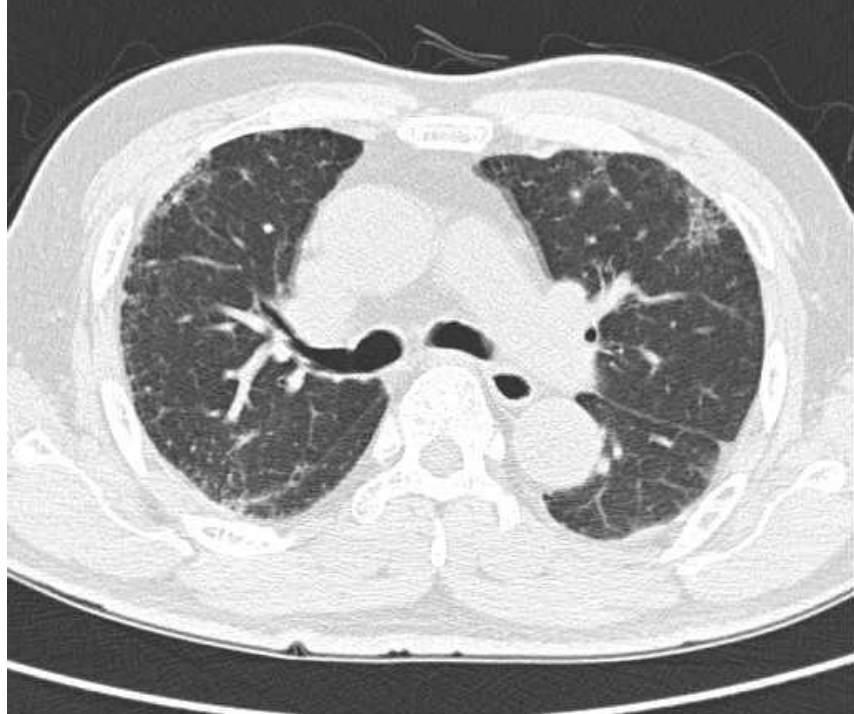
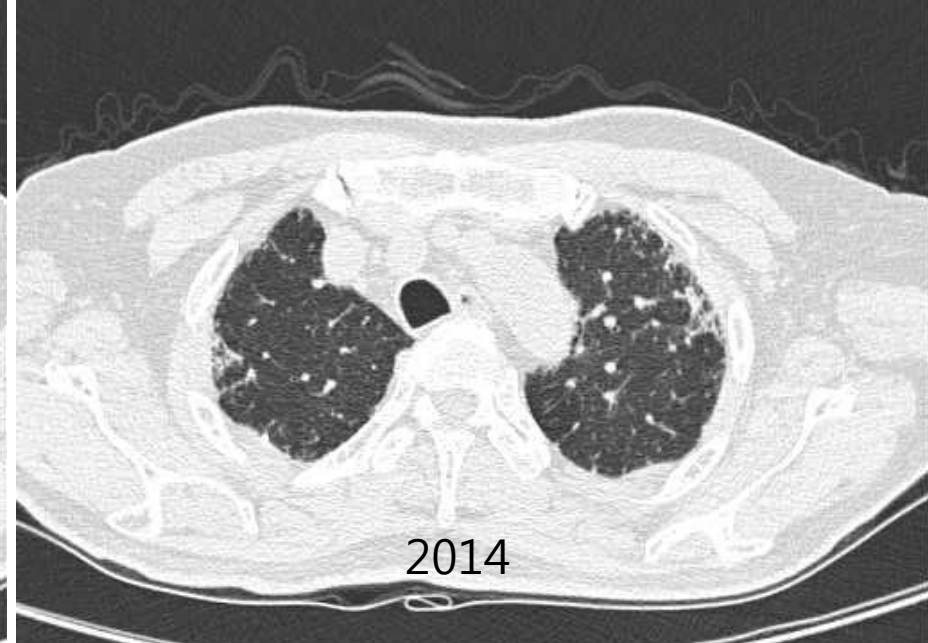
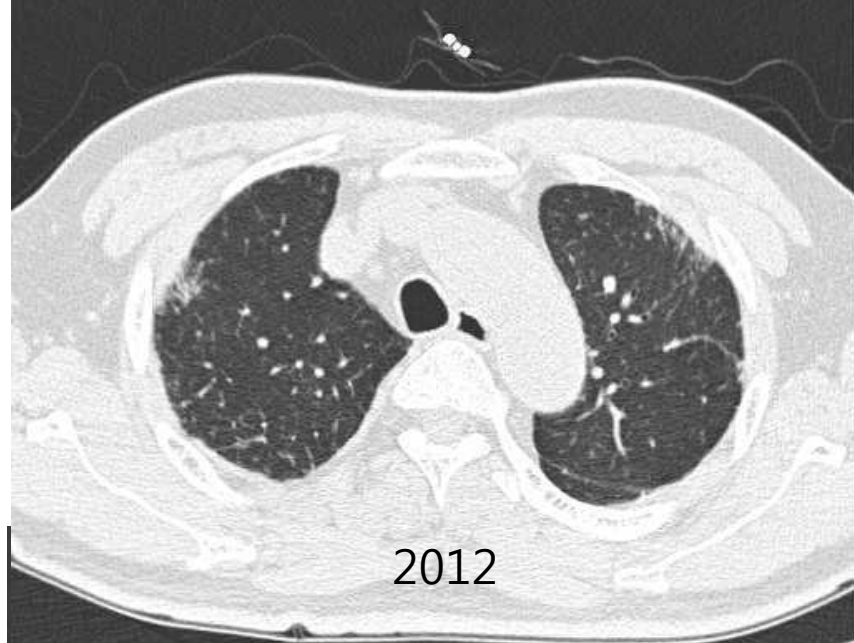
- High dose NAC 복용하면서 stable
- 2012.12월 PIP joint pain & swelling
 - RF+, anti-CCP+ (2011년 VATS시행 당시에는 negative)
 - 류마티스내과에서 RA로 진단받음.
- PFT (2013.4)
 - FVC 2.58 (71%), FEV1 1.93(71%)
 - DLCO 9.3 (61%)

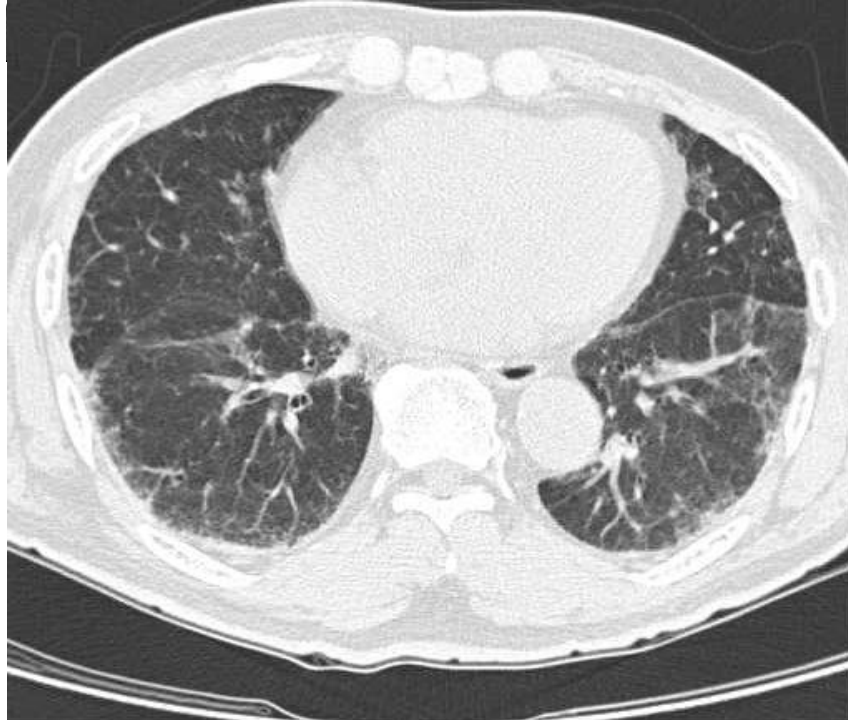
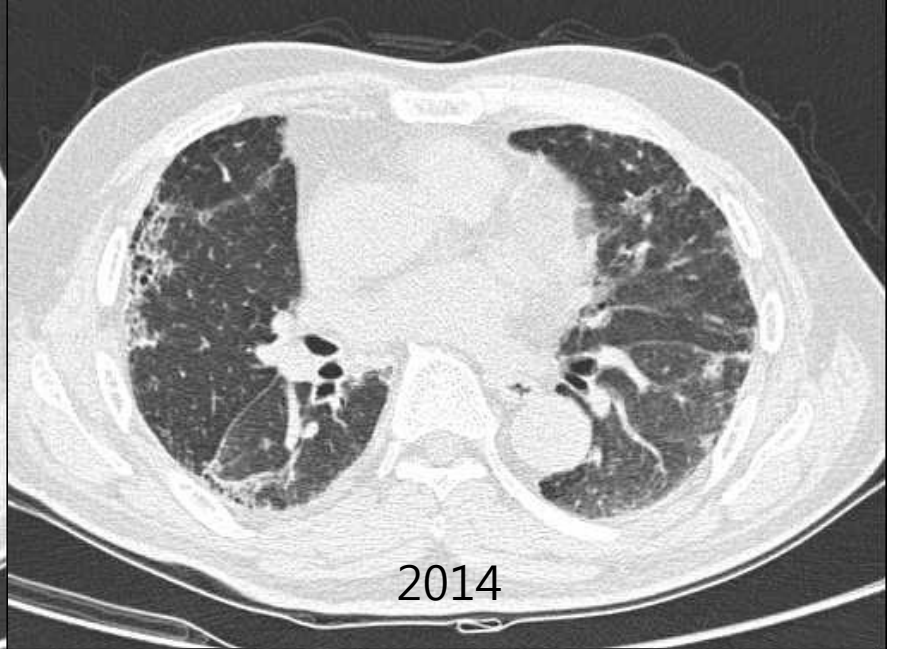
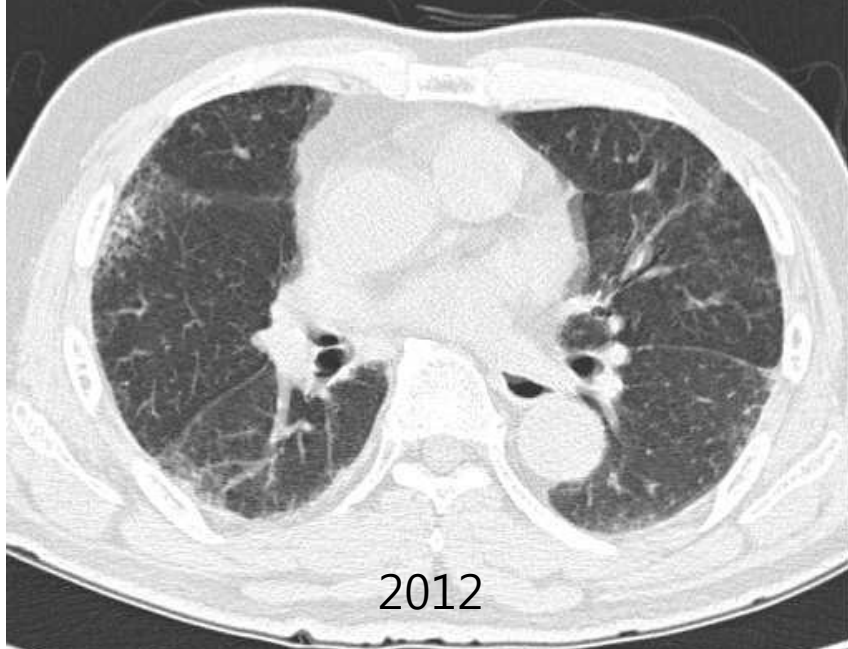


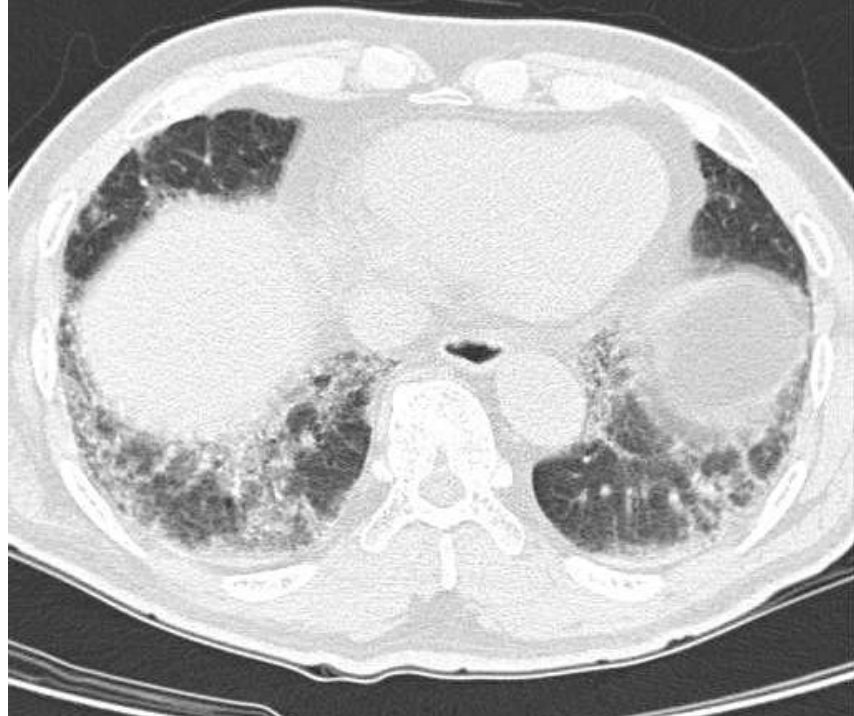
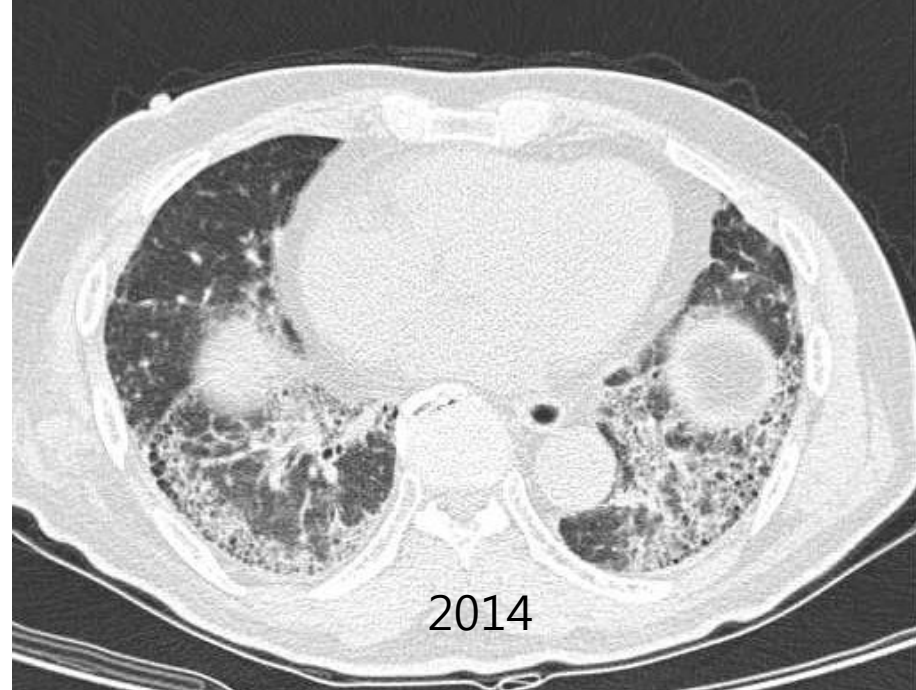
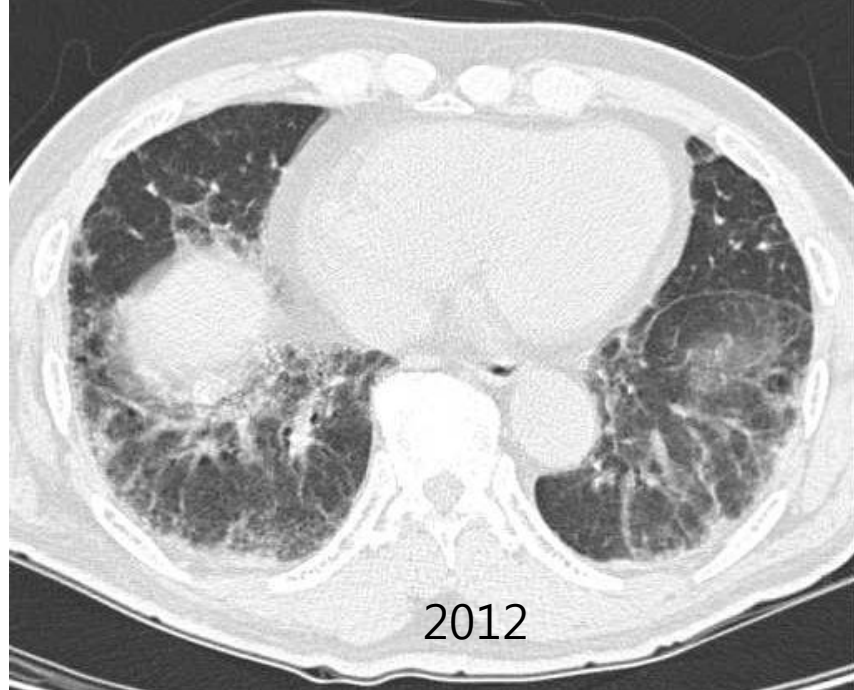
2012

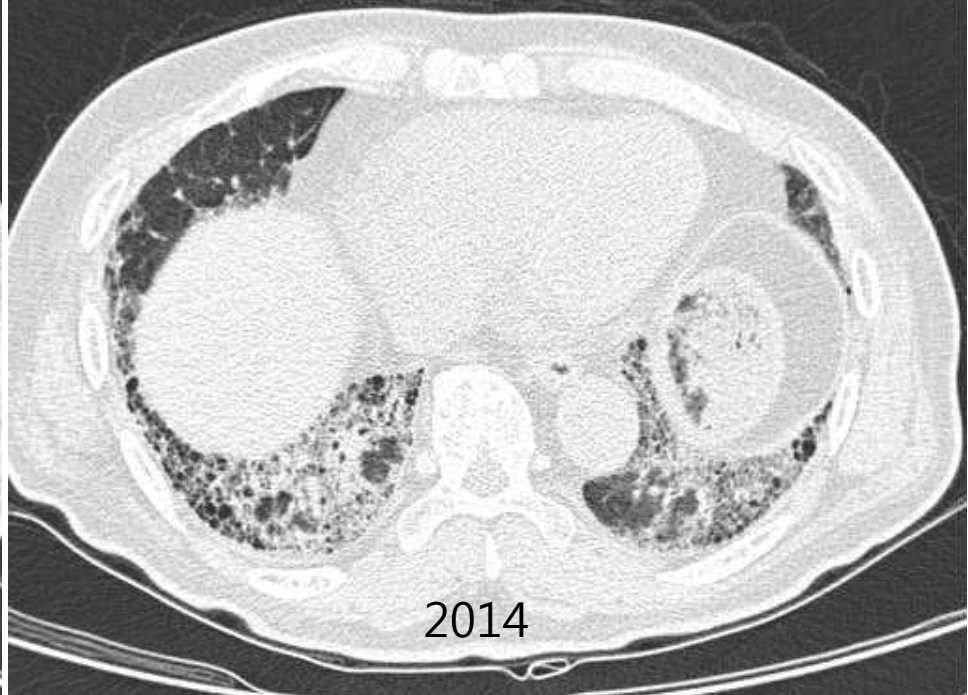


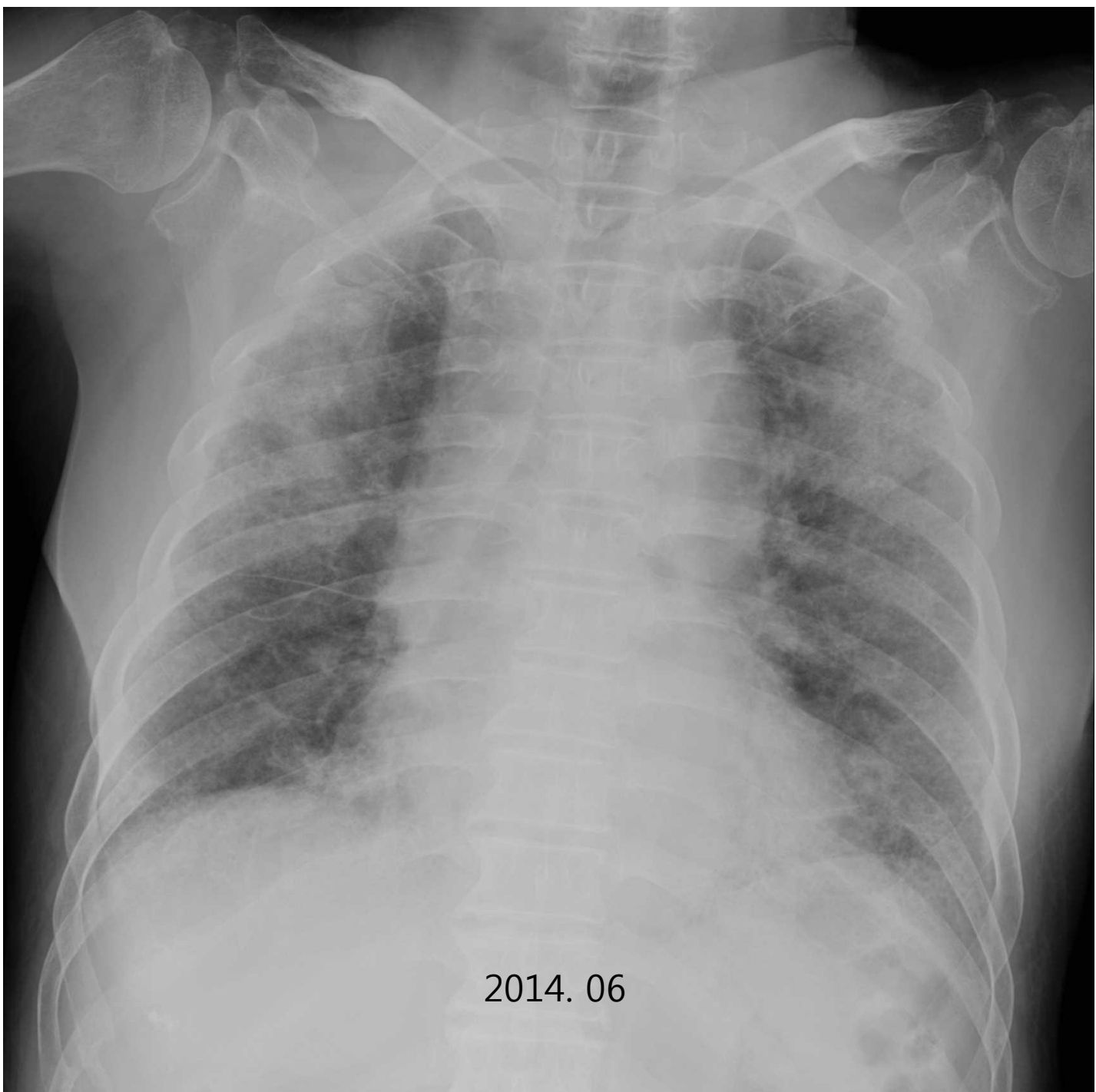
2014



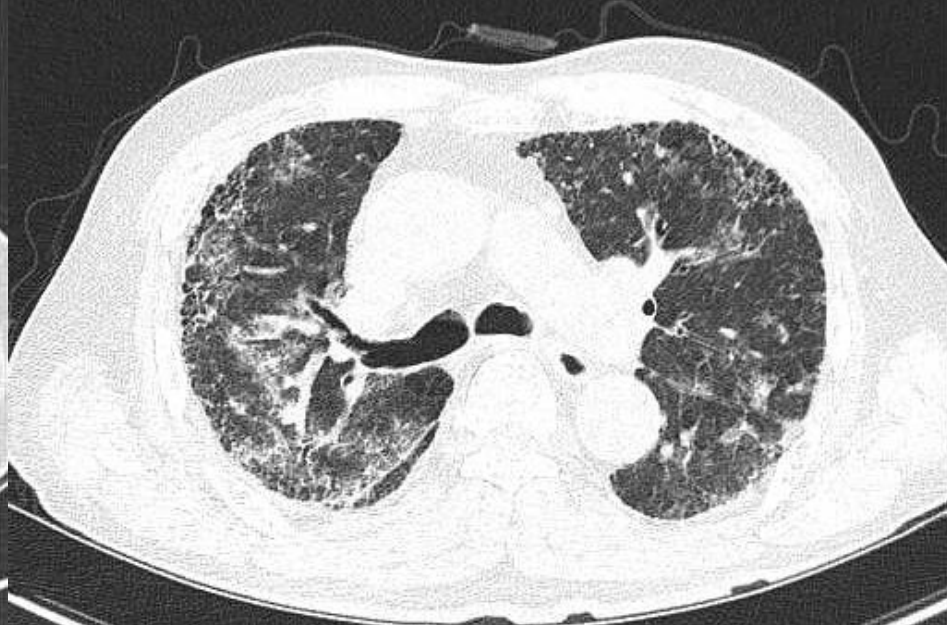
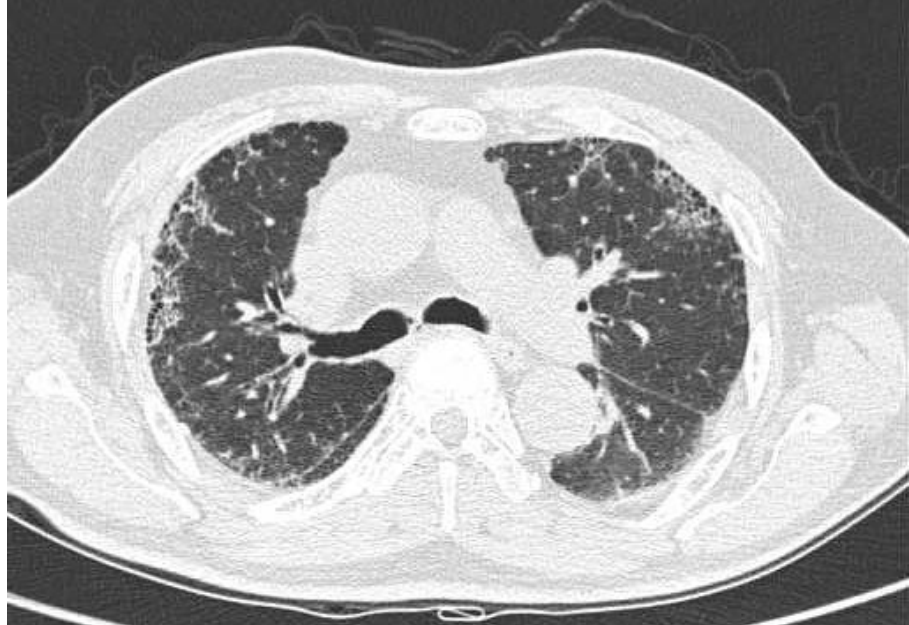
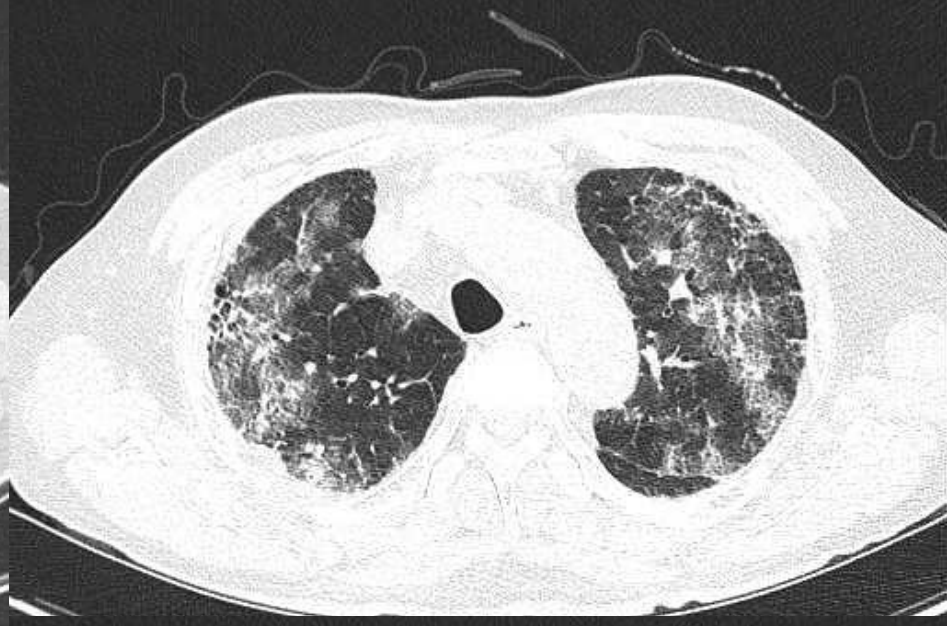
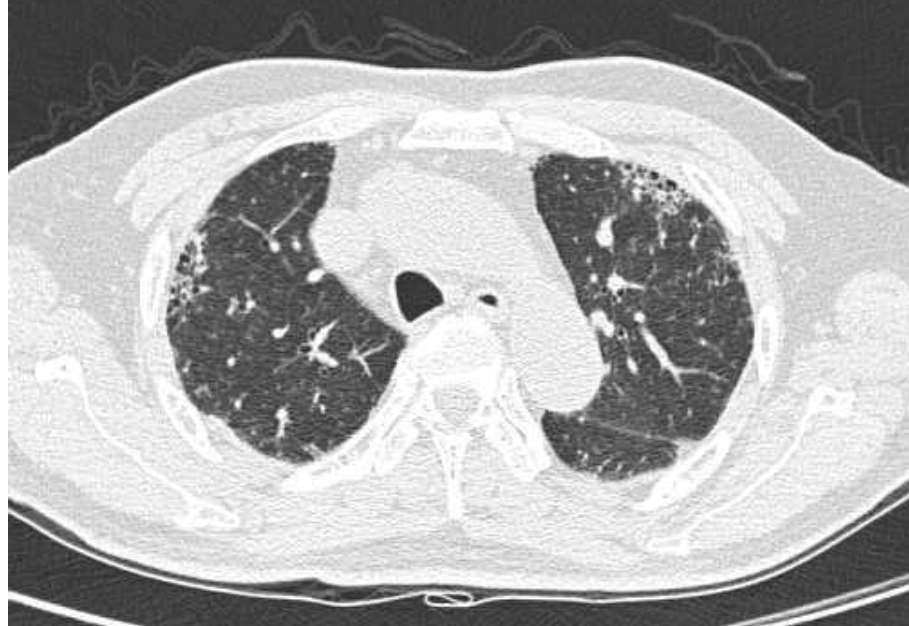


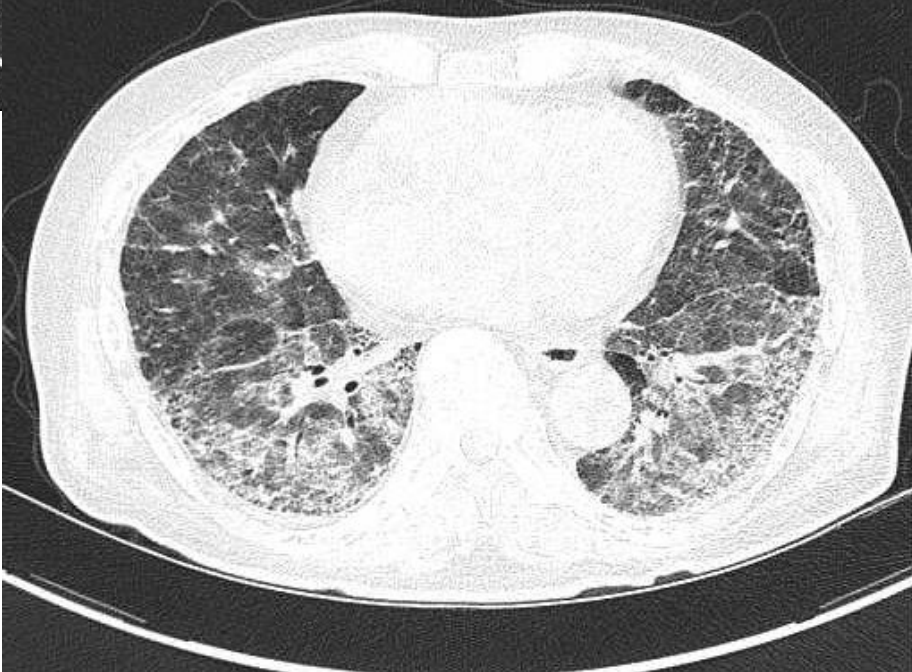
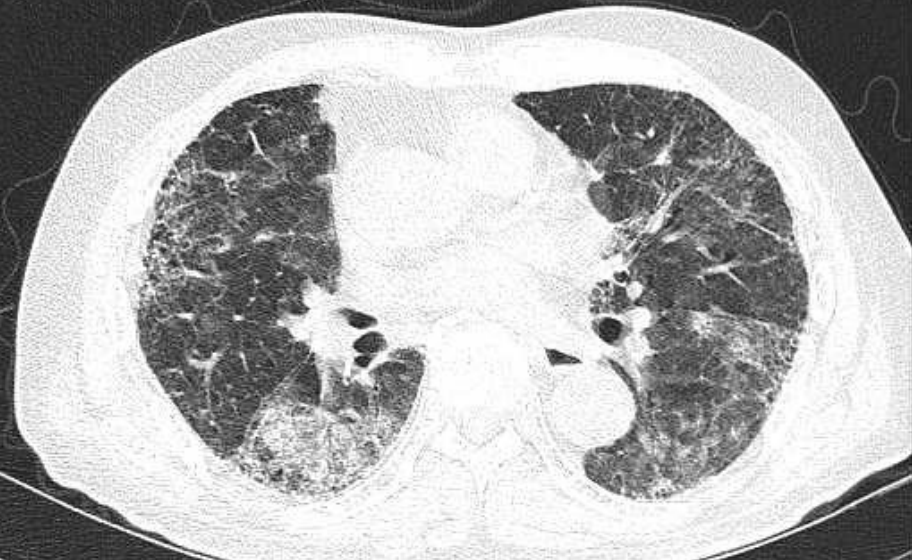






2014. 06





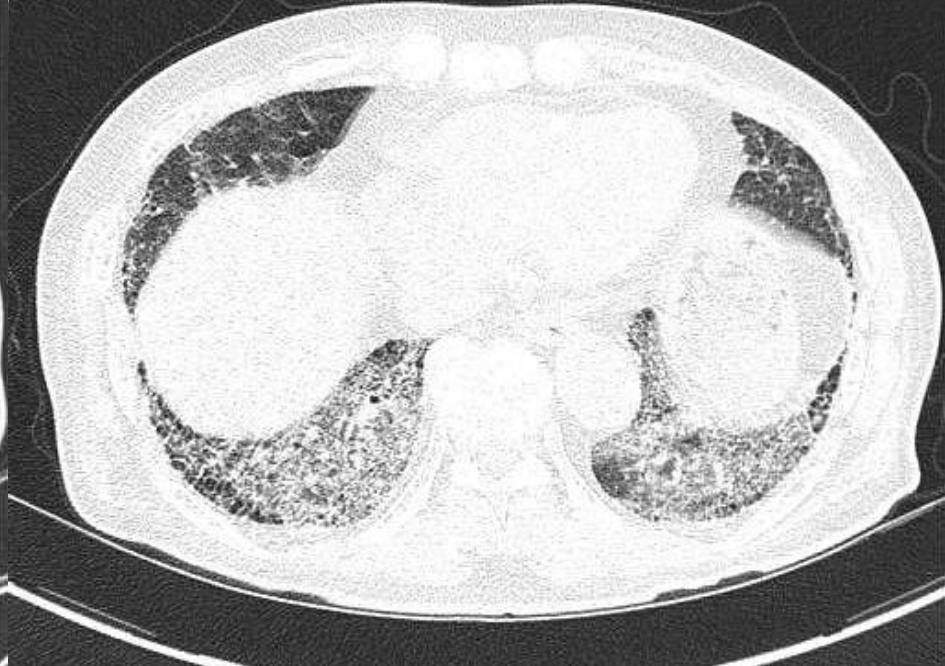
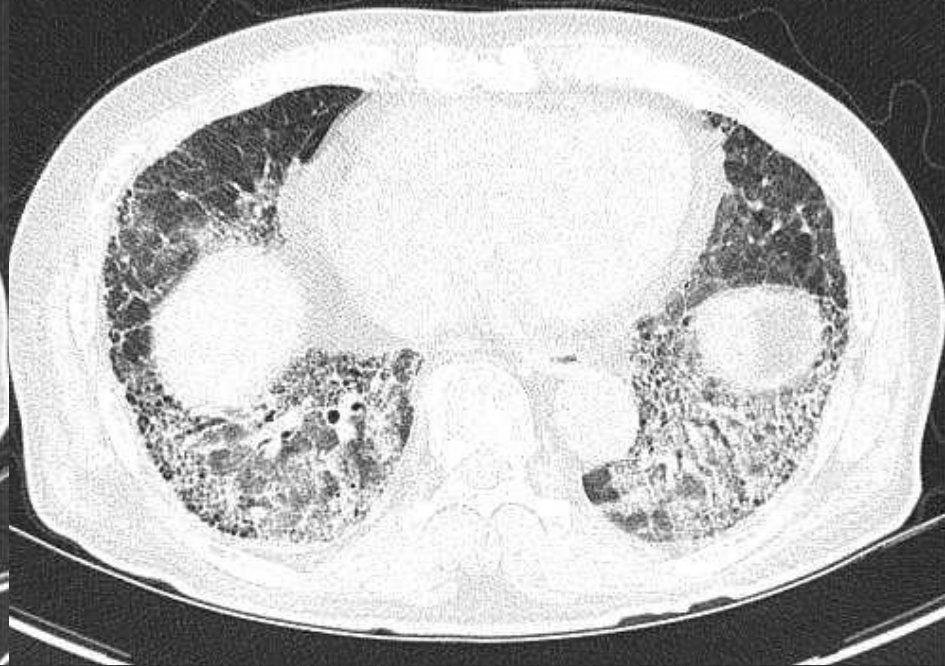
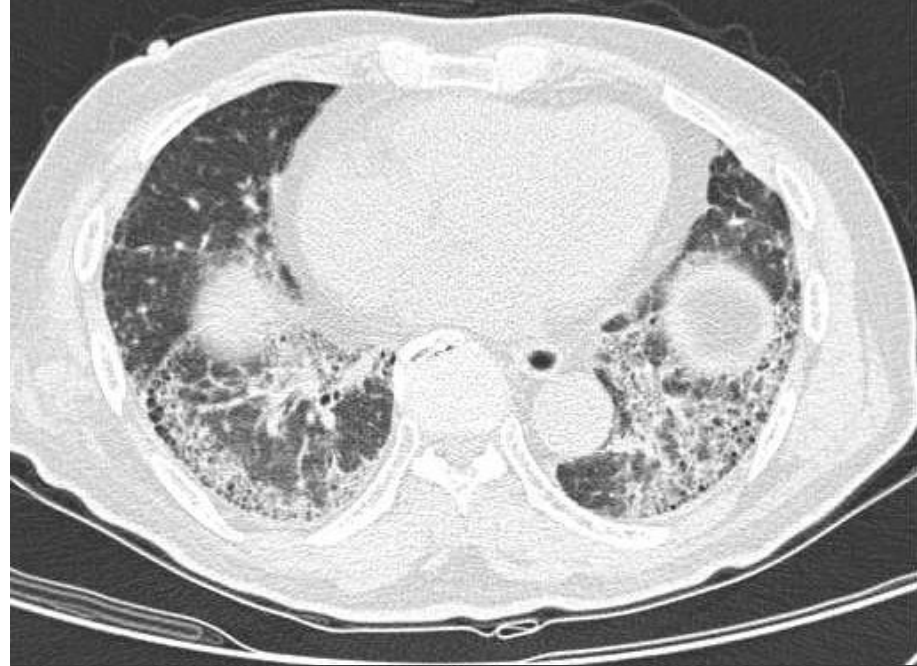


TABLE 6-1
RELATIVE FREQUENCY OF PATTERNS OF ABNORMALITY IN COLLAGEN VASCULAR DISEASES

Pulmonary Disease	Rheumatoid Arthritis	Progressive Systemic Sclerosis	Systemic Lupus Erythematosus	Polymyositis-Dermatomyositis	Mixed Connective Tissue Disease	Sjögren Syndrome	Ankylosing Spondylitis
Nonspecific interstitial pneumonia	++	+++	+	+++	++	+++	
Usual interstitial pneumonia	+++	+	+	+	+	+	
Organizing pneumonia	+		+	+++	+	+	
Lymphoid interstitial pneumonia			Rare		+	++	
Diffuse alveolar damage	+	+	++	+			
Hemorrhage			+++				
Mosaic perfusion and air trapping	+++					++	+
Pleural effusion or thickening	++		+++				

+ Uncommon; ++ common; +++ most common pattern.

TABLE 6-2

HRCT FINDINGS IN RHEUMATOID ARTHRITIS

Bronchiectasis without fibrosis^a

Findings of fibrosis (i.e., traction bronchiectasis and bronchiolectasis, intralobular interstitial thickening, irregular interlobular septal thickening, irregular interfaces)^a

Honeycombing

Ground-glass opacity^a

Peripheral and subpleural predominance of fibrosis or ground-glass opacity^{a,b}

Lower lung zone and posterior predominance^{a,b}

Pleural thickening or effusion^{a,b}

Small centrilobular nodules (follicular bronchiolitis)

Large (rheumatoid) nodules

Findings of bronchiolitis obliterans (i.e., air trapping, mosaic perfusion)

^aMost common finding(s).

^bFinding(s) most helpful in differential diagnosis.

- 2014.6월 acute exacerbation 으로 ICU care & mechanical ventilation
- MethylPd pulse 시행하였으나 Acute MI로 사망

Discussion

- Diagnosis
 - Clue for CTD-ILD?, radiology, pathology
- Treatment
 - Optimal timing?