A Multidisciplinary Team Discussion

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Organising an MDT: the role of the MDT co-ordinator

Pre-discussion

Referral Processing

- ✓ Receiving all referral documentation:
  - ILD MDT Proforma
  - PFT Results
  - Referral Letter
- ✓ Pulling over any further information needed
- ✓ Registering the patient onto our system
- ✓ Creating the MDT spreadsheet agenda and emailing to the team

The more detailed the referral package, the faster it can be processed and added for MDT!

Imaging/Pathology

- ✓ Pulling all relevant imaging across prior to discussion
- ✓ Liaising with pathologist on what is needed and their availability
- ✓ Putting in requests for any pathology to be posted for discussion – at least 2 weeks in advance of the MDT

To avoid delay, always list scan locations & future scan details

Post-discussion

The Outcome

- ✓ Documenting the final decision from the discussions
- ✓ Ensuring the outcome is communicated to all relevant teams, including 'next steps' to the patient
- ✓ Ensuring any further investigation or review is followed up

Always keep patients informed of our estimated wait times to help manage expectations

Case 1

- 47 year old man
- Diagnosed with Idiopathic Pulmonary Fibrosis since 2019
- Nintedanib since 2019
- Ex smoker 26 pack year history
- No CTD symptoms
- No exposures
- No family history

- Progressive breathlessness and dry cough over time despite nintedanib
- Long term oxygen prescribed Feb 2023 – 1.5 litres at rest and 8 litres on exertion (previously 4 litres ambulatory)

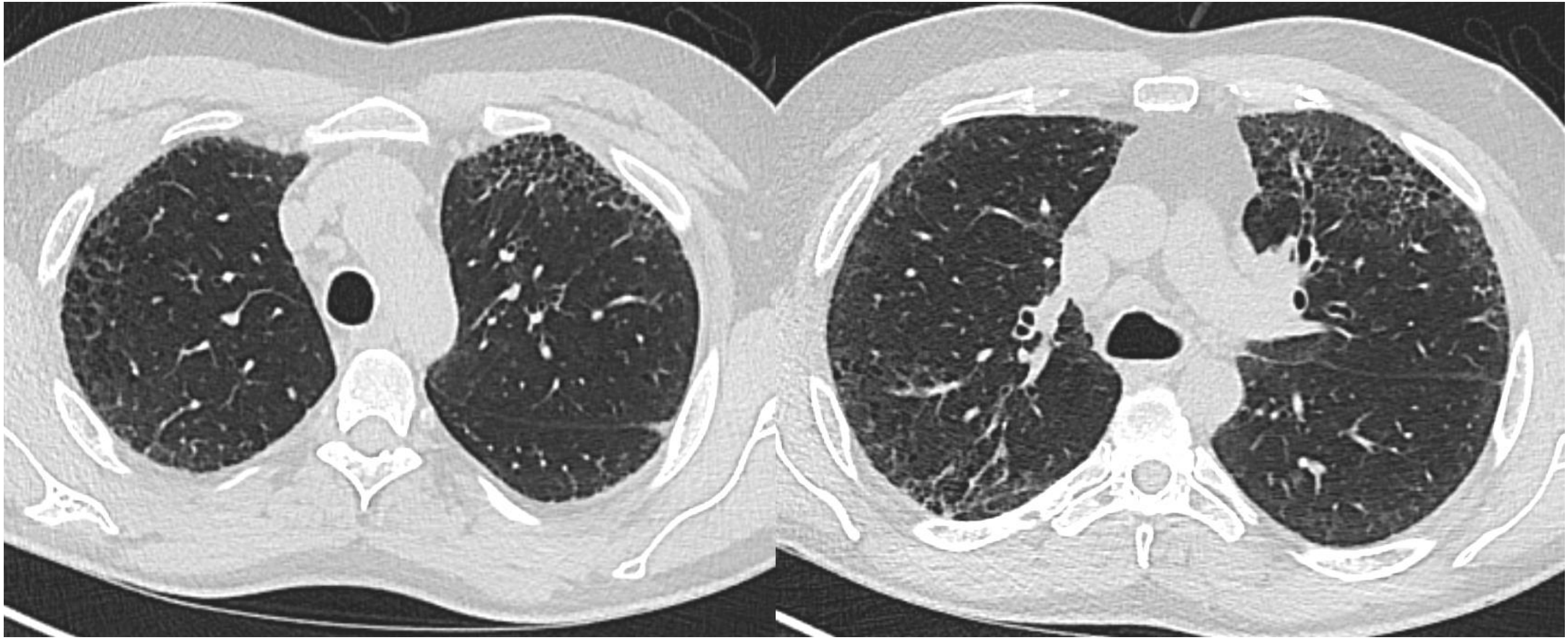
Alarm Bells!

- Diagnosed with IPF at age 43
- No family history of ILD, aplastic anaemia, liver diseases.
- Hair turned grey at age 28.
- Always worked in manufacturing industry – dusty, sawmill, quarries
- No joint pains, no joint swelling. No dry eyes dry mouth. No skin rashes. No symptoms of CTD.
- No exposure to birds , pigeon, moulds.
- Manageable side effects with stomach cramps and diarrhoea once per week
- Noticed breathing getting worse since Dec 22. A year ago ETT was 15-20 minutes on the flat but not up inclines. Now only 5 minutes with oxygen. Productive cough in the morning with white sputum.
- Last chest infection – 1 week ago. Has had further infection requiring 3 courses of antibiotics and steroids – Felt better on steroids from a breathing perspective.

Lung function

Date	BMI	FEV1	FEv1%	FVC	FVC%	DLCO	DLCO%	Change
May 23	28	1.97	57	2.27	54	2.07	21	
Jan 23	31	2.04	59	2.28	54	ND		460ml 10% FVC decline 6m
June 22	30	2.29	66	2.74	64	3.28	33	170ml 4% FVC decline 6m
Nov 21	31	2.44	70	2.91	68	3.07	31	340ml 7% FVC and 4% DLCO Decline 4m
June 21	31	2.66	76	3.25	75	3.36	35	160ml 4% FVC 14% DLCO DECLINE 6 m
Dec 19	32	2.86	81	3.41	79	4.9	49	Stable vs Mar 19
March 19	31	2.86	80	3.46	80	5.1	51	

Baseline CT



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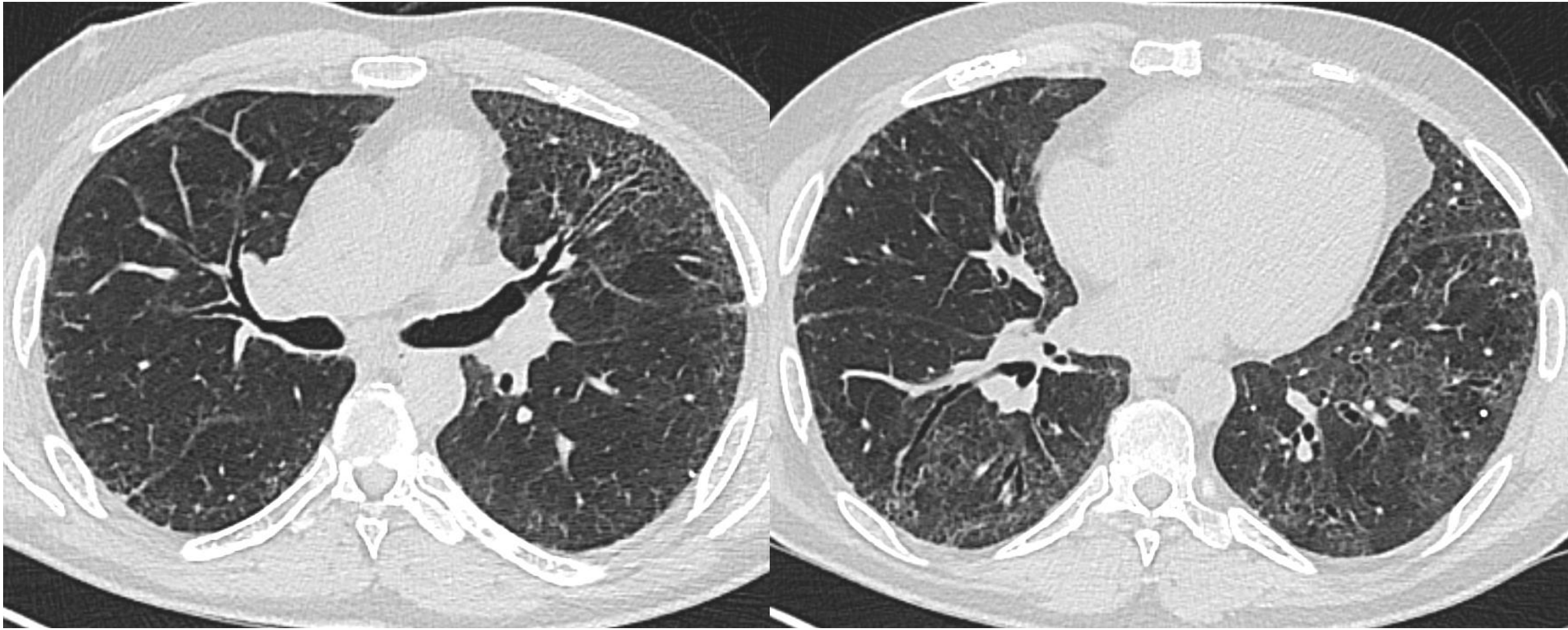

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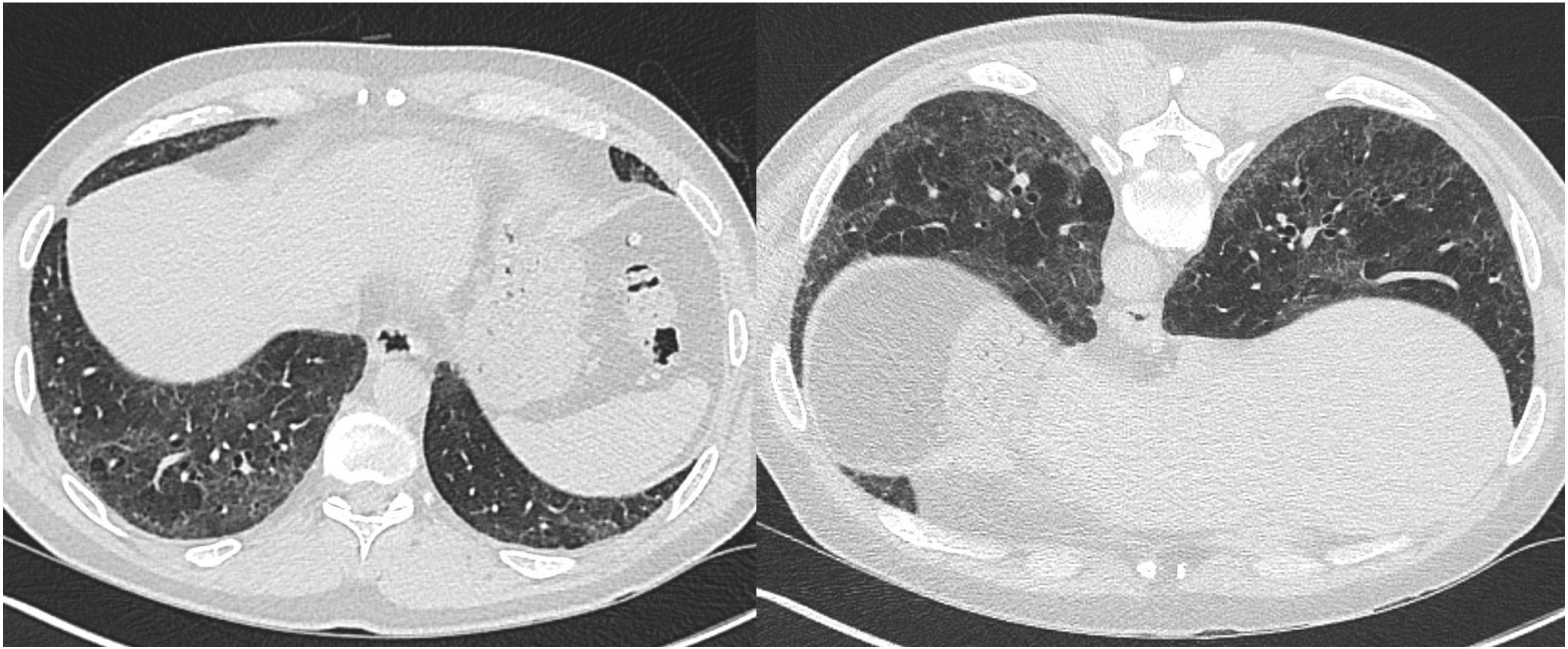
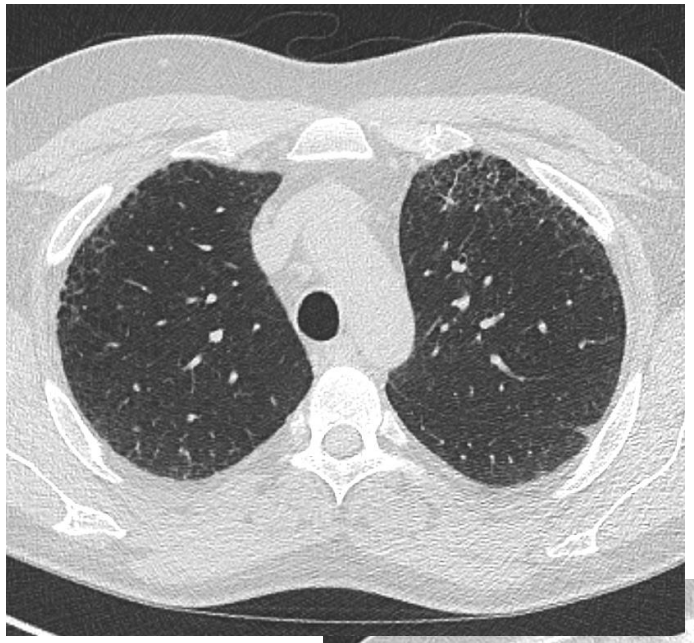
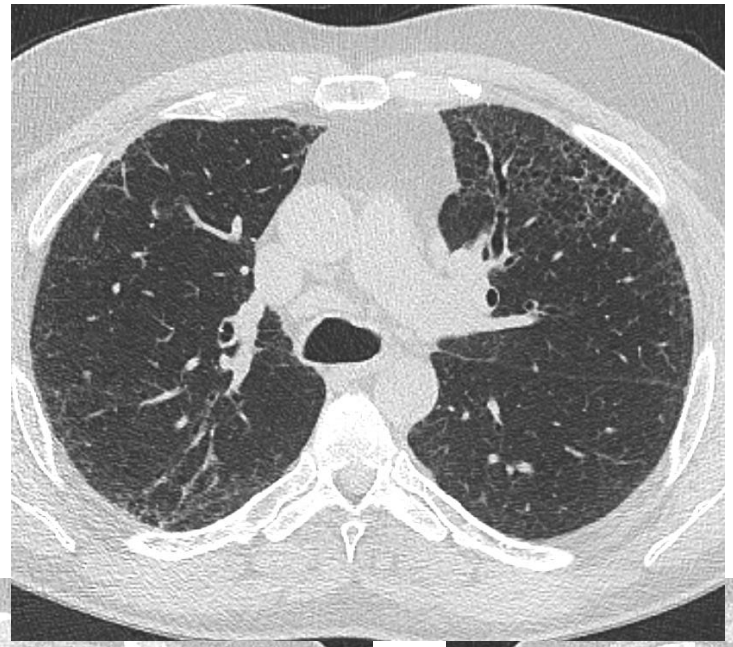
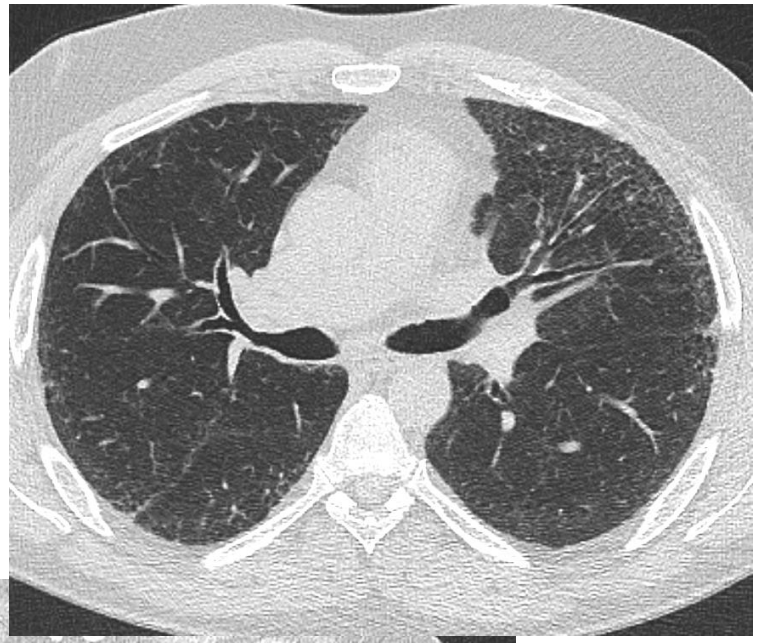
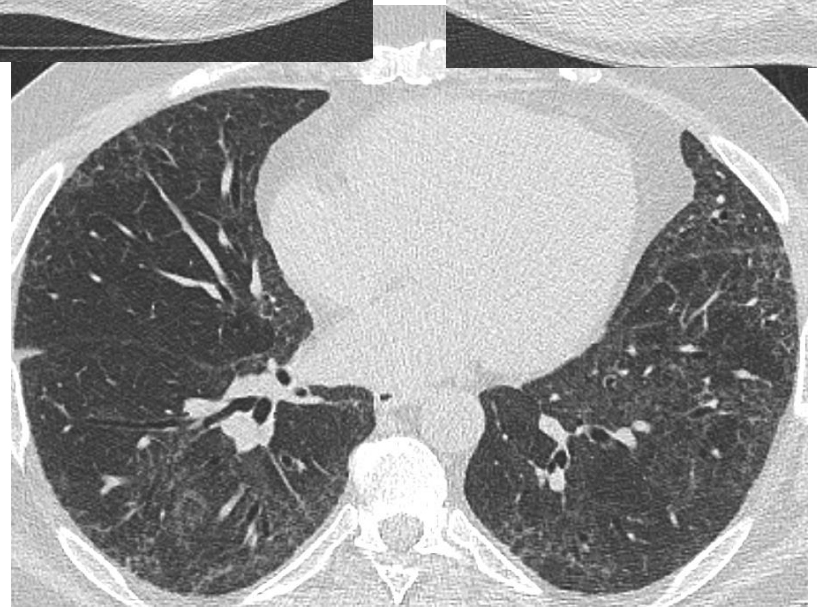
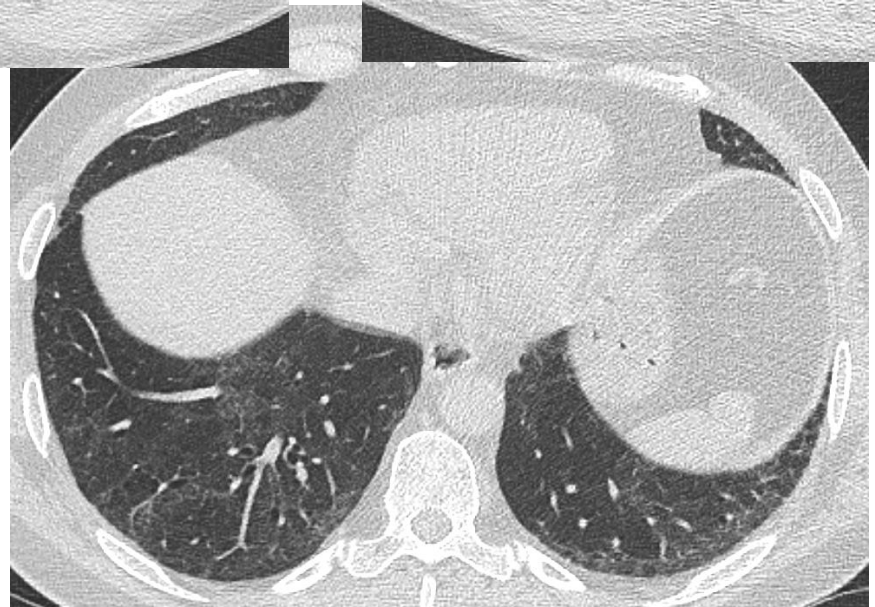
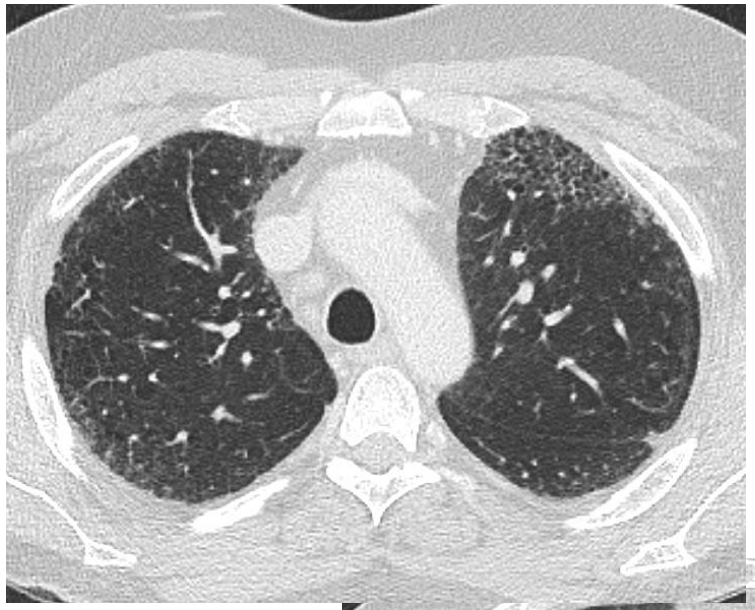
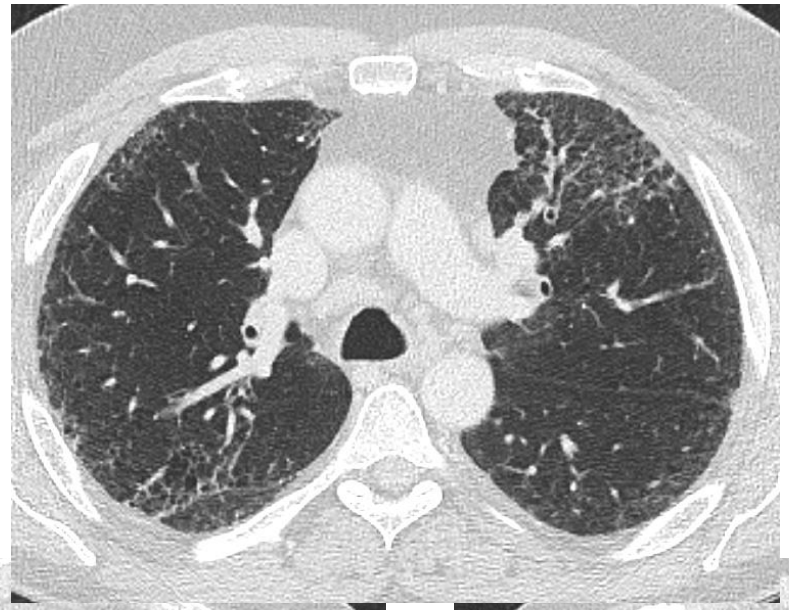
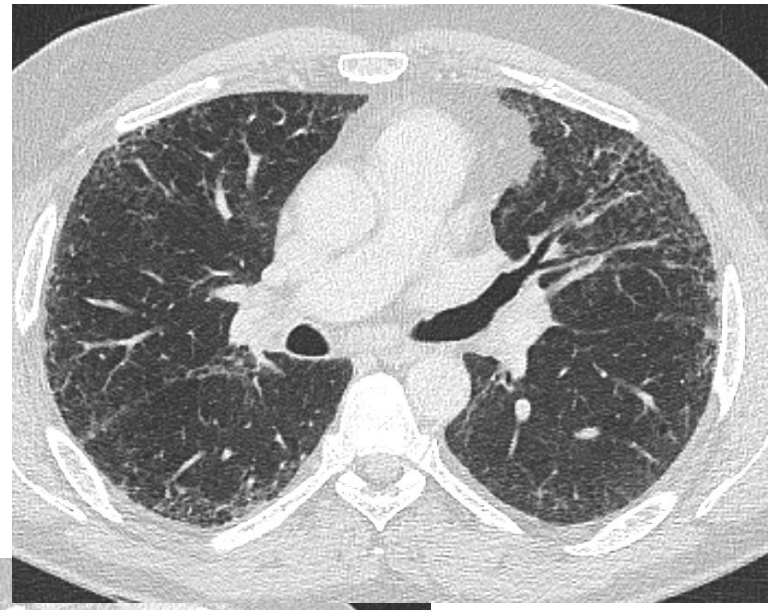
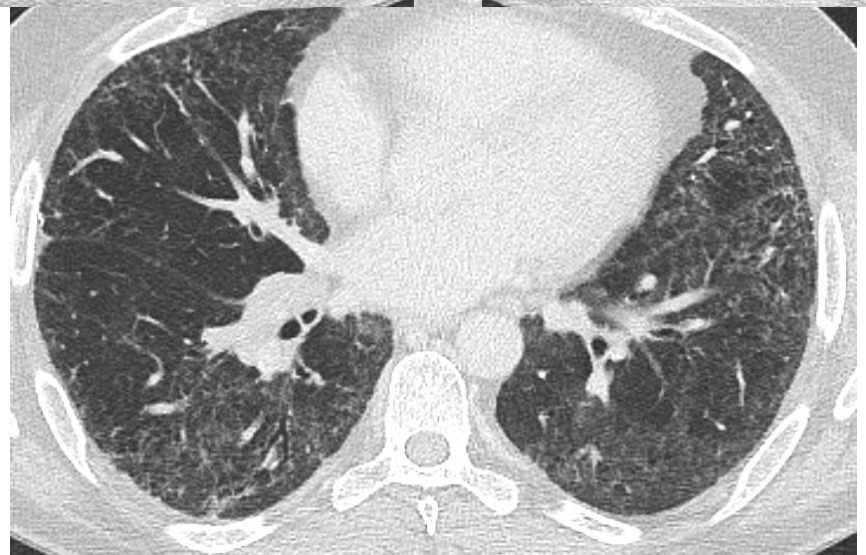
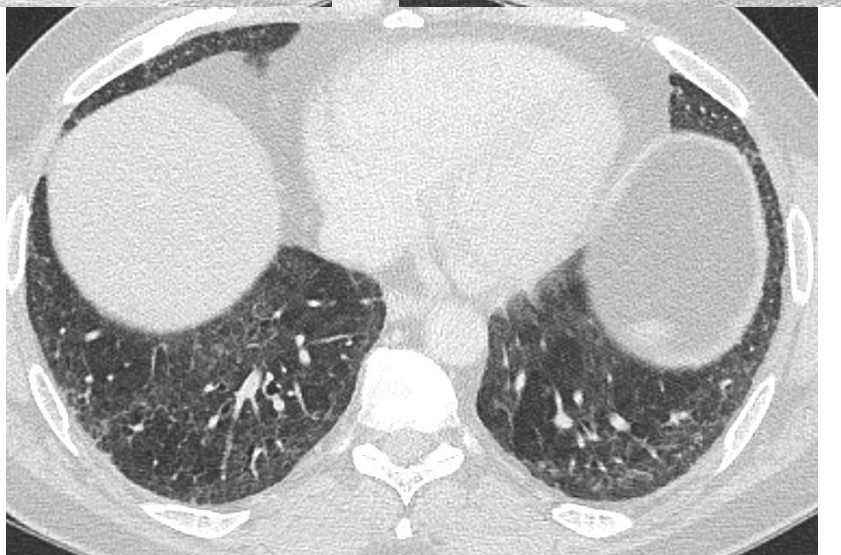


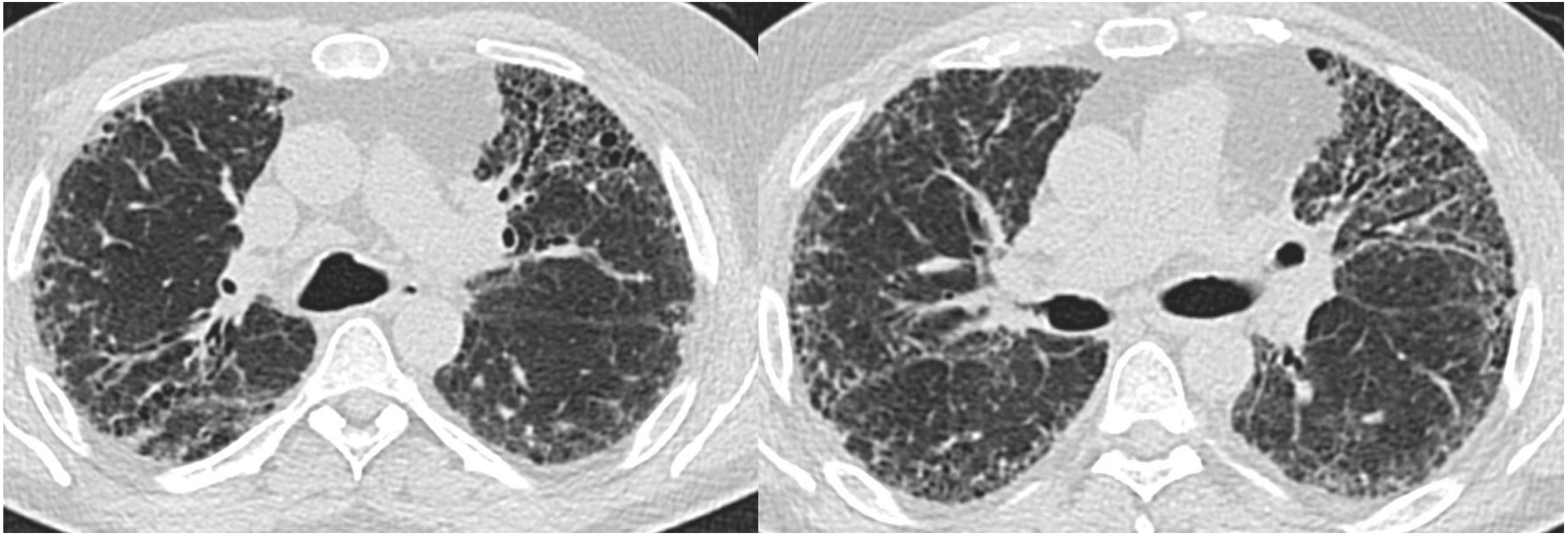
CT 6 months

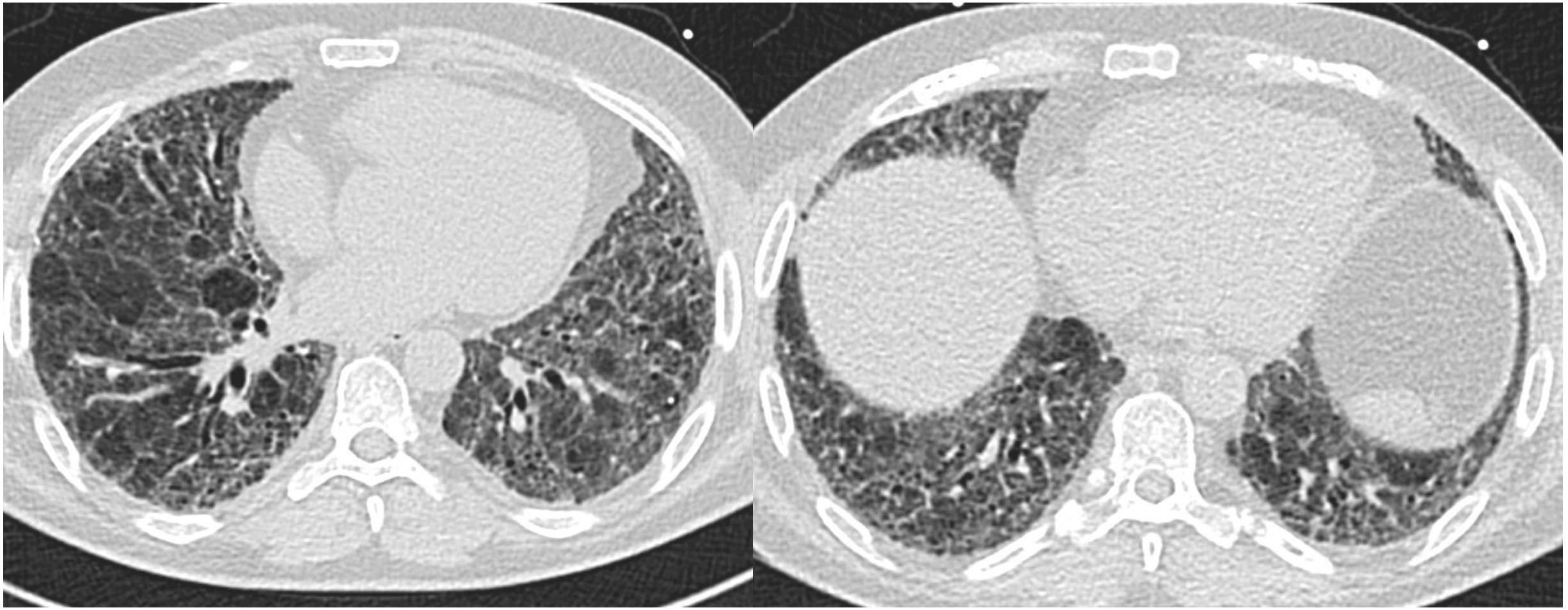
CT 3 years

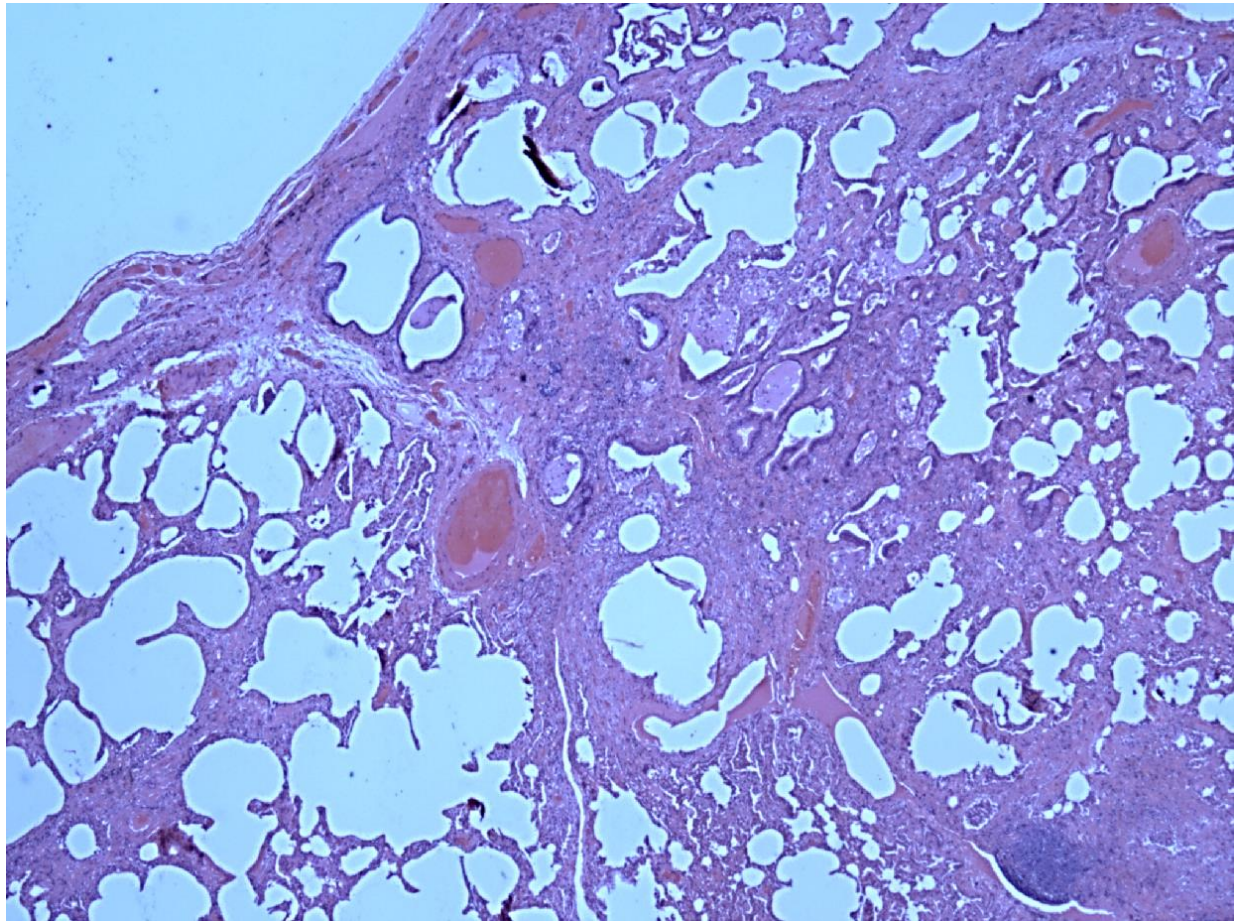
CT 5 years

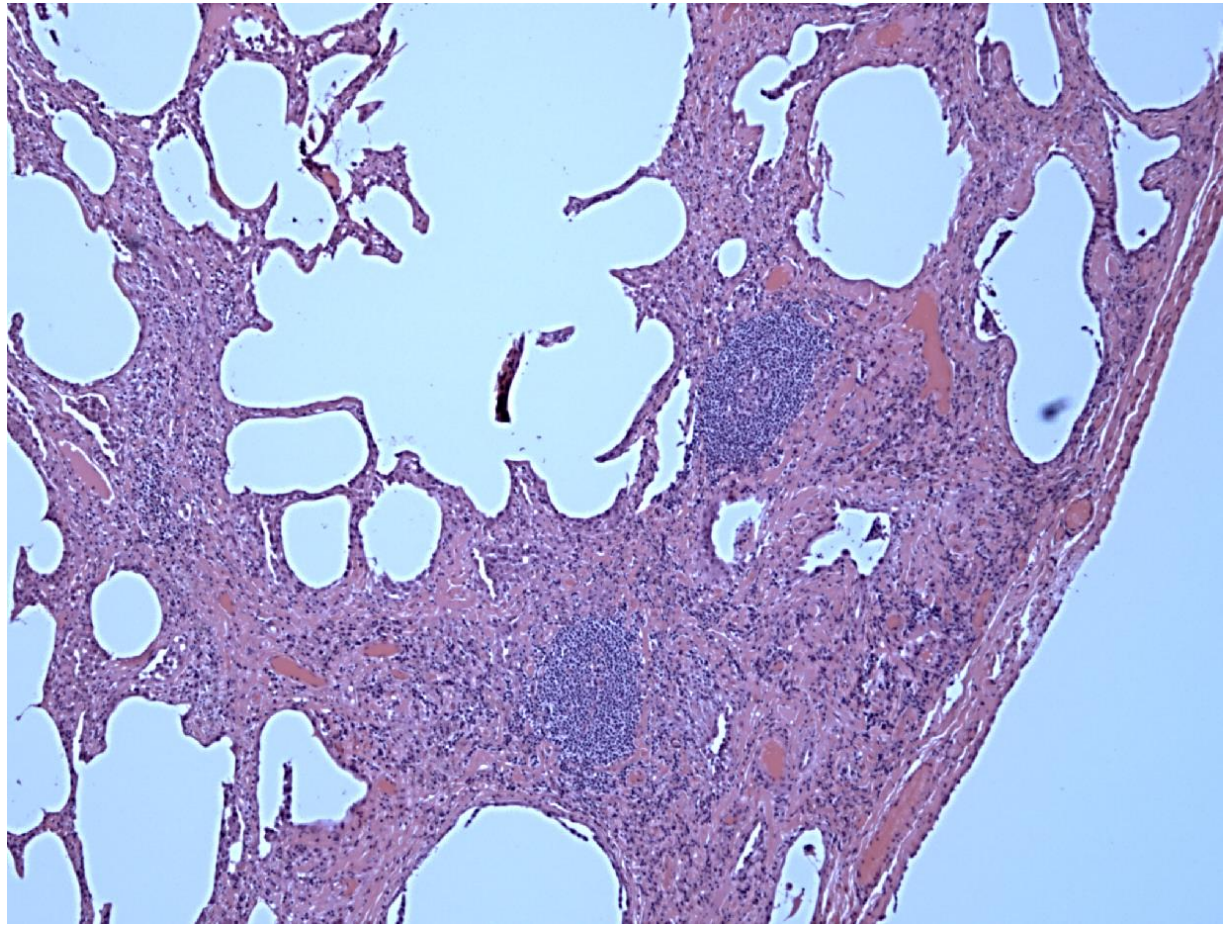


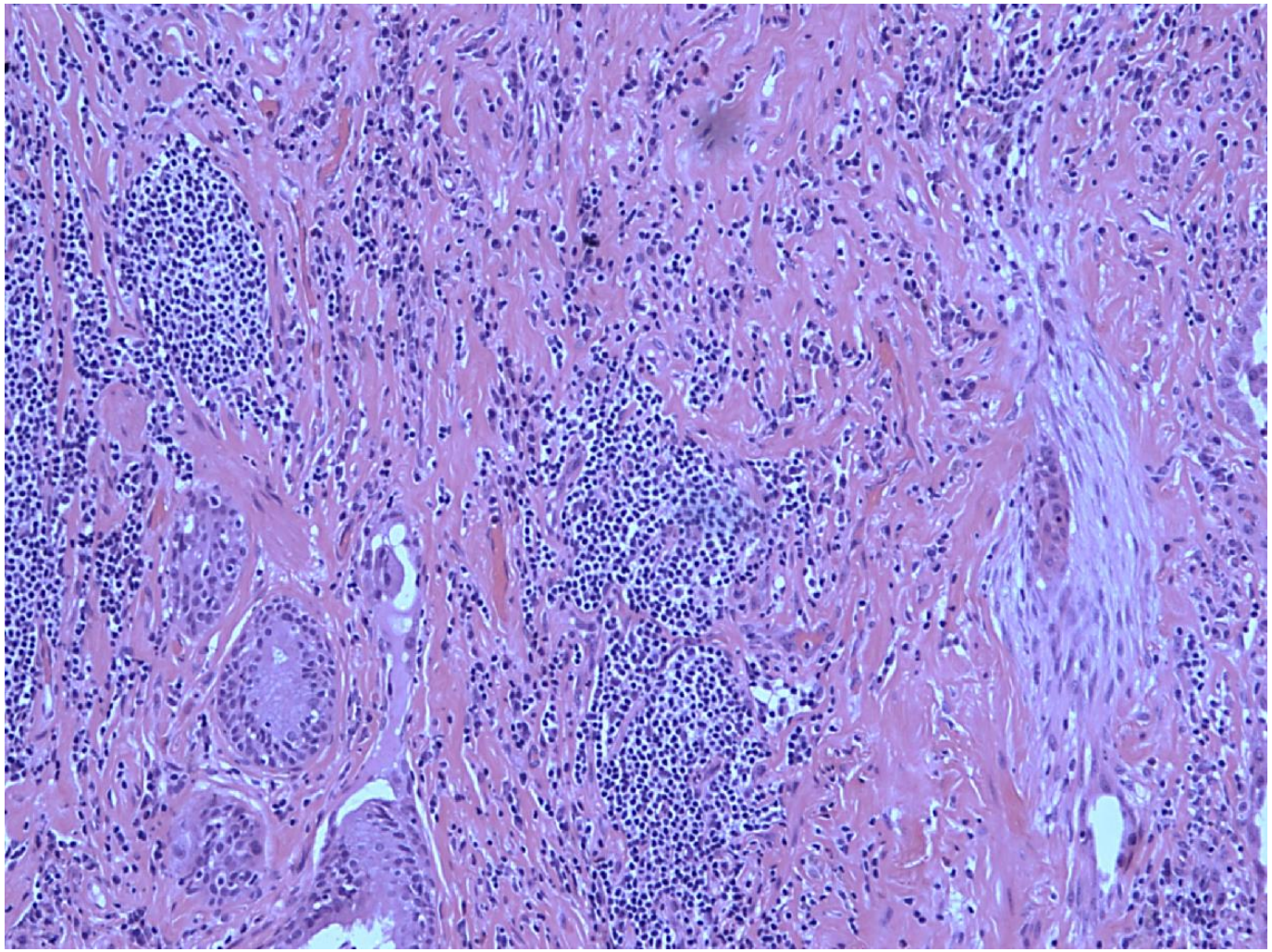
CT 5 years



He had a lung biopsy in 2019







What we did

- Ensure no stone unturned
- Definitely no features of CTD – clinically or AI screen and myositis screen
- Sent to genetics – awaited (anyone <50 year old)
- Switched Nintedanib to pirfenidone
- Transplant assessment – now on the list
- Biopsy sent for 2nd opinion Manchester
- Re MDT review of Biopsy and imaging
- Added a low dose prednisolone – for what its worth! (felt better on prednisolone)

Role of specialist nurse at MDT

Patient advocate with knowledge of the patient

- Give information about the pre-morbid state of the patient.
- Provide information on the social circumstances' patient
- Contribute and communicate patient’s wishes

Patient advocate with no prior knowledge of patient

- Focused on age, weight and lung function
- Focus on patients' stage of disease, comorbidities, suitability for transplantation, nutrition obese or cachectic

General role

- Focus the discussion from patterns to diagnosis, eg. UIP is this IPF, or chronic HP
- Post MDT, ensure patients follow a pathway of care, anti-fibrotic clinic, immunology OPD
- Keep non-drug treatments in discussion, rehabilitation, oxygen assessments, palliative care
- Some may communicate the MDT discussion to patients

Saw our specialist nurse

- Complex poorly patient who is declining rapidly (300mls FVC pa) needs 1hr nursing consultation
- Focus of care
  - Address his psychological needs, fears, family, finances DS1500 PIP, Will
  - Unpack what he understands what he is struggling with,
  - Likely to die soon so needs palliative conversation referral & advanced care plan
  - Assess his symptoms; cough, reduced ETT, breathlessness
  - Address non pharma treatments, oxygen cough physio, rehab, dietitian (his weight)
  - Discuss change of antifibrotic
- Other issues to think about
  - Close contact with transplant centre copy all correspondence
  - Family, children no follow up but remove triggers smoking / risk factors jobs in silica wood

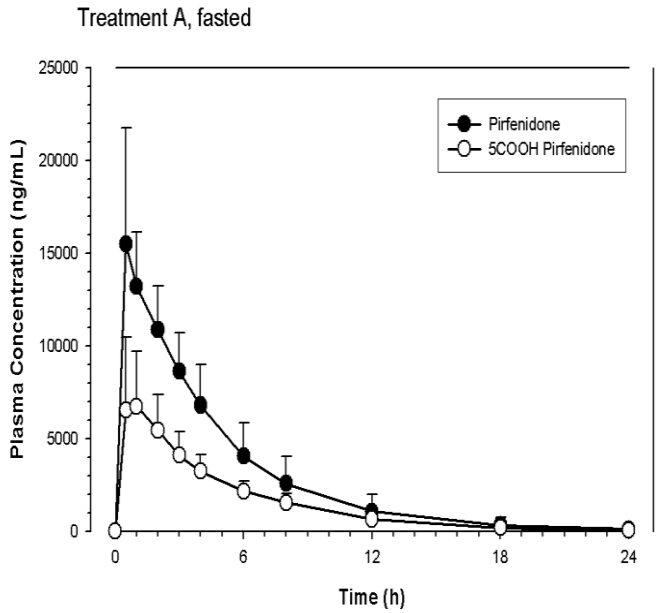
Pirfenidone

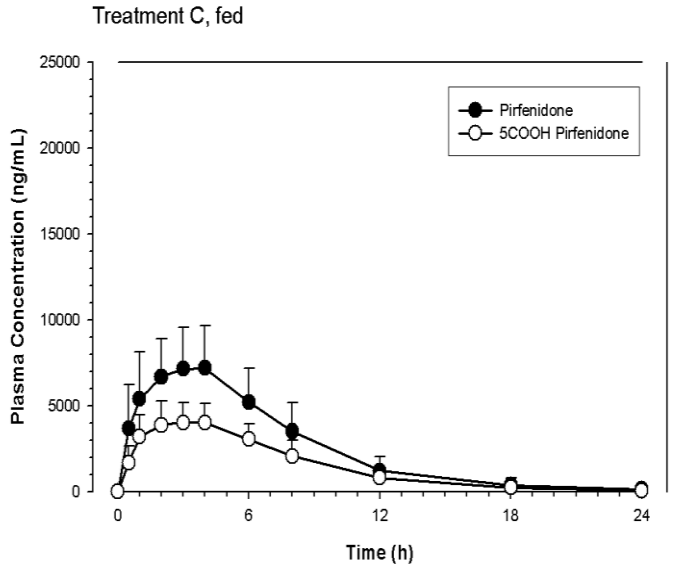
Drug Name (Brand Name)	Dose Range	Monitoring	Common Adverse Drug Reactions	Common Drug Interaction	Prescribing Considerations
Pirfenidone (Esbriet)	Increasing dose to 2403 mg/day Week 1 – 267mg 1 tablet 3 times a day Week 2 – 267mg 2 tablets 3 times a day Week 3 – 267mg 3 tablets 3 times a day	U&Es, LFTs, weight	GI, photosensitivity, rash, liver toxicity, dizziness, fatigue, weight loss	Avoid grapefruit juice, fluvoxamine and inhibitors of CYP1A2 (e.g ciprofloxacin, amiodarone), cigarette smoking	Administer with food Apply factor 50 suncream (UVA and B) every day

Please see www.medicines.org.uk for further information

Pirfenidone: common AE

- **Gastrointestinal AEs (nausea/indigestion)**
- Slow titration needed
  - Take during/end of meal
  - Stagger during meal
  - Appropriate PPI
  - Anti emetics: ondansetron
  - Consider reduction/interruption
- **Appetite suppression**
  - Ensure nausea/indigestion contro
  - Small regular meals/ snacks
  - Monitor weight
  - Dietician input, supplements
  - Dose reduction/ interruption





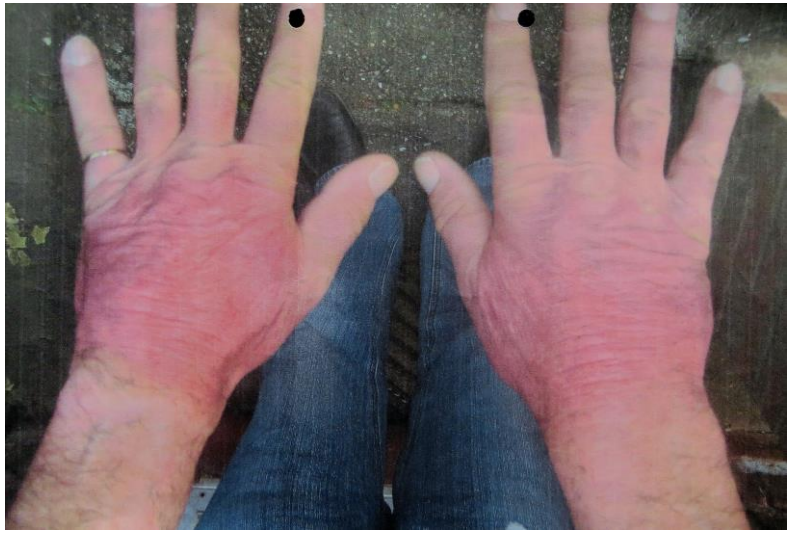
Taking pirfenidone with food reduces the maximum concentration, which may be responsible for adverse events [a] Data on file, InterMune, Inc. 2006.

Pirfenidone: common AE

- Abnormal Liver Function Tests
  - Monitor ALT, AST and bilirubin
    - Up to 3x ULN or pts normal repeat test
    - 3-5x ULN: reduce or interrupt treatment & repeat
    - 3-5x ULN with Hyper bilirubinaemia: interrupt & repeat
    - > 5x ULN: discontinue and don't rechallenge
- Lethargy
  - Behavioural changes
  - Ensure no other cause
  - Dose reduction/interruption

Pirfenidone: common AE

- **Photosensitivity**
  - Education, advice on High factor sunscreen UVA and UVB
  - Sun avoidance, dress appropriately
  - Allergy versus Photosensitivity? Severity
  - Dose reduction
  - Consider Dermatology referral



Managing patients on antifibrotics

- Education
- Prophylaxis
- Reduction/Interruption
- Retitration
- Change therapy

Learning points

- Young people with ILD Think familial ILD / Telomere dysfunction
- Ask about family history, early greying, aplastic anaemia, liver disease, Raised MVC, low platelets
- Always re review diagnosis if natural history not as expected
- Look for CTD if see lymphoid aggregates on biopsy

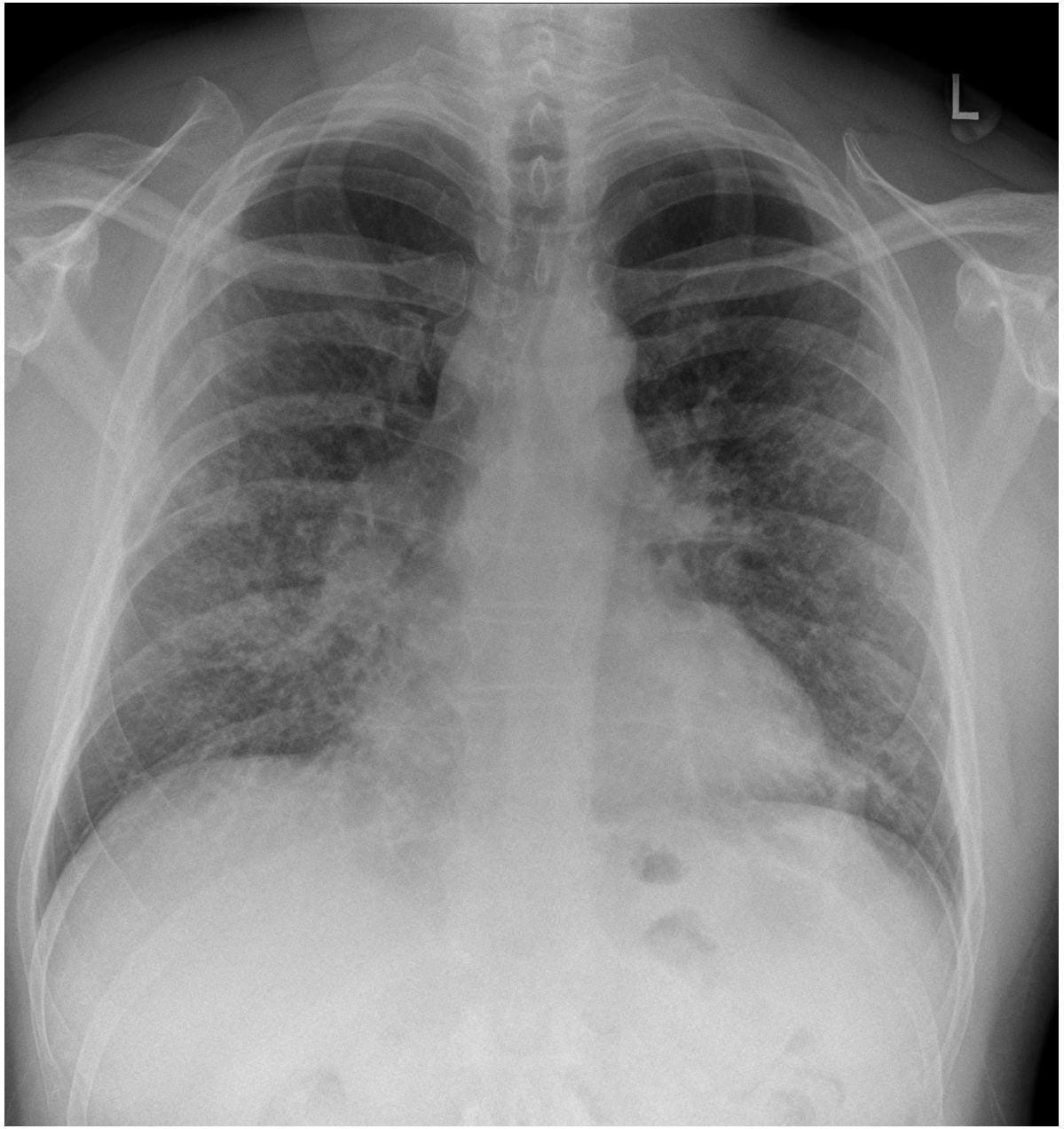
Case 2

- 59 year old man
- Hypercalcaemia and AKI in 2012
- Further episode of Hypercalcaemia and AKI – Summer 2016
- Summer 2017 – AKI
- Summer 2018 - AKI
- 2019 – Basal cell carcinoma of skin x 2
- Bowens disease of skin
- OA hip awaiting replacement
- Gout

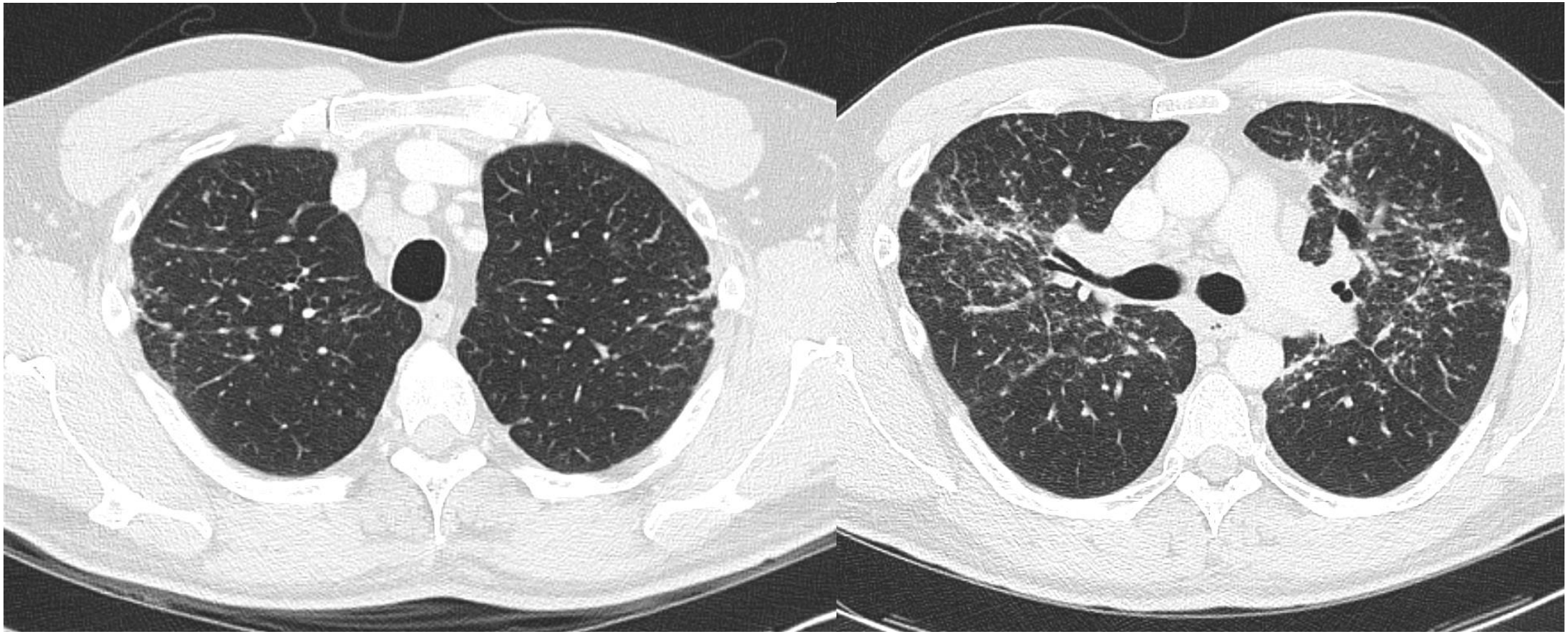
Imaging

- CXR 2012
- CT Chest and abdo with contrast 2012
- CT Chest and abdo with contrast 2014

CXR Baseline



Baseline CT



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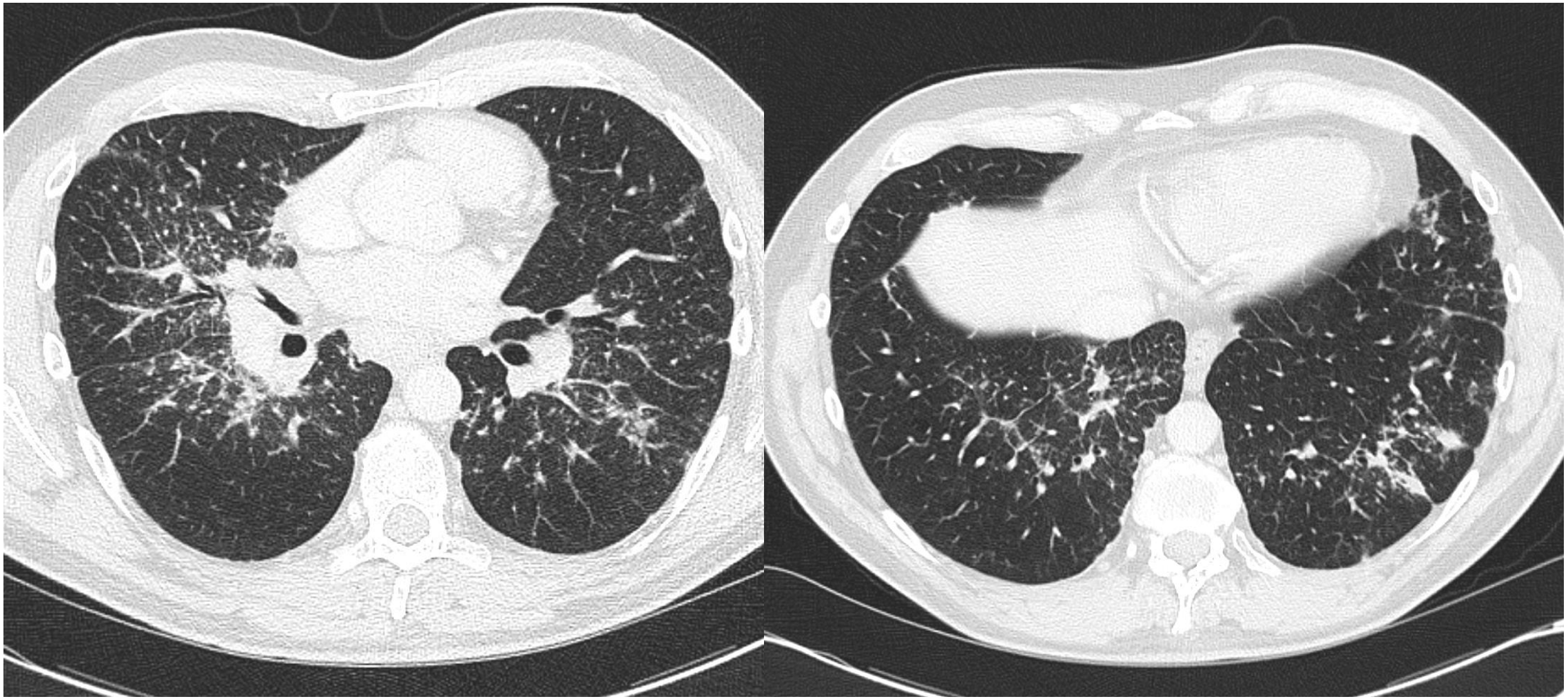
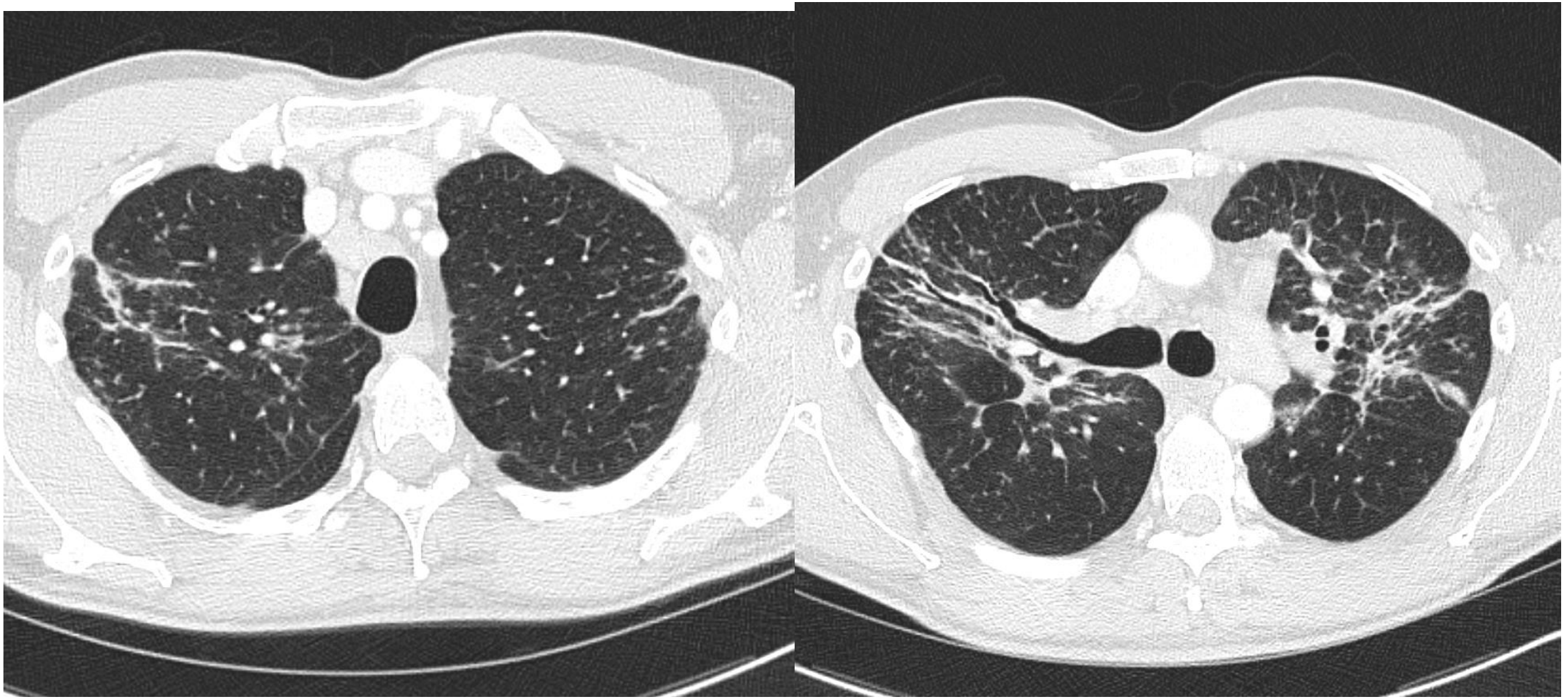


CT 2 years



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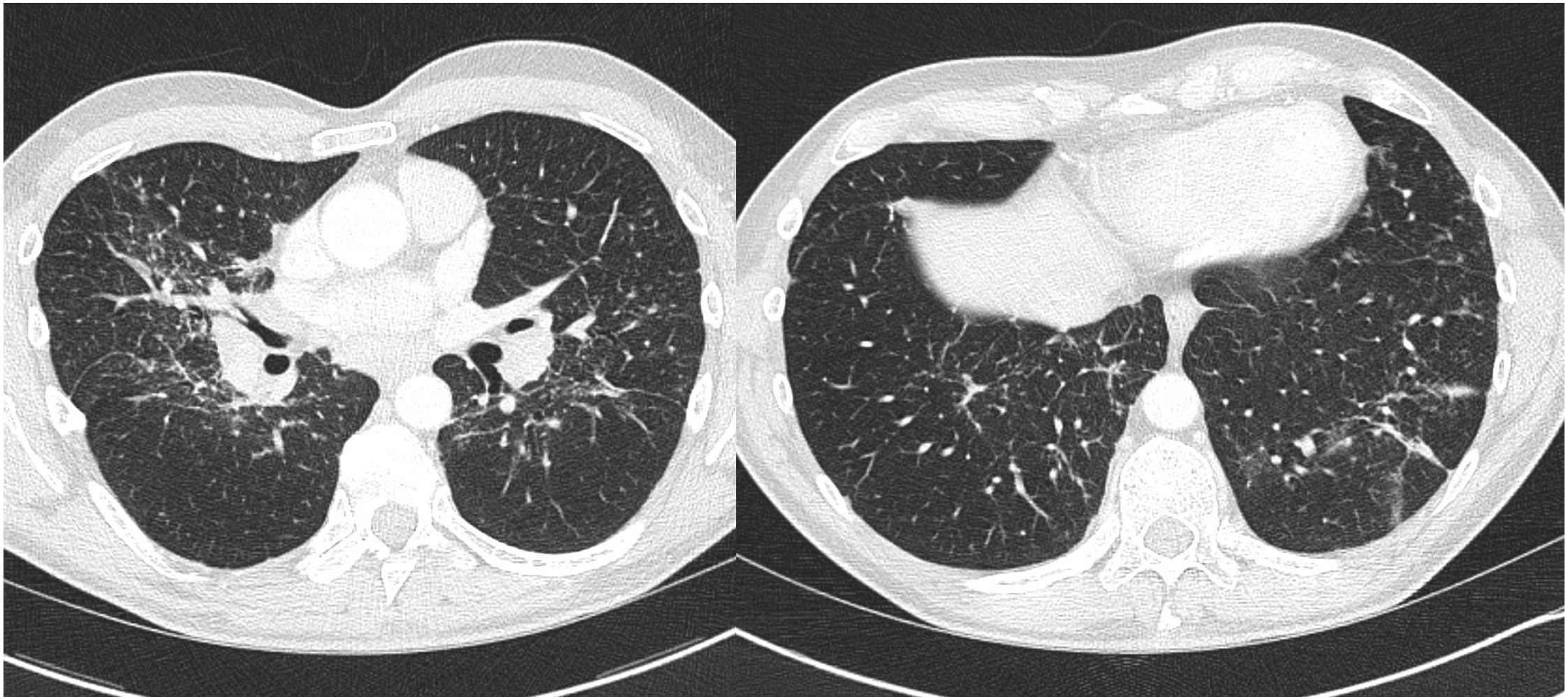
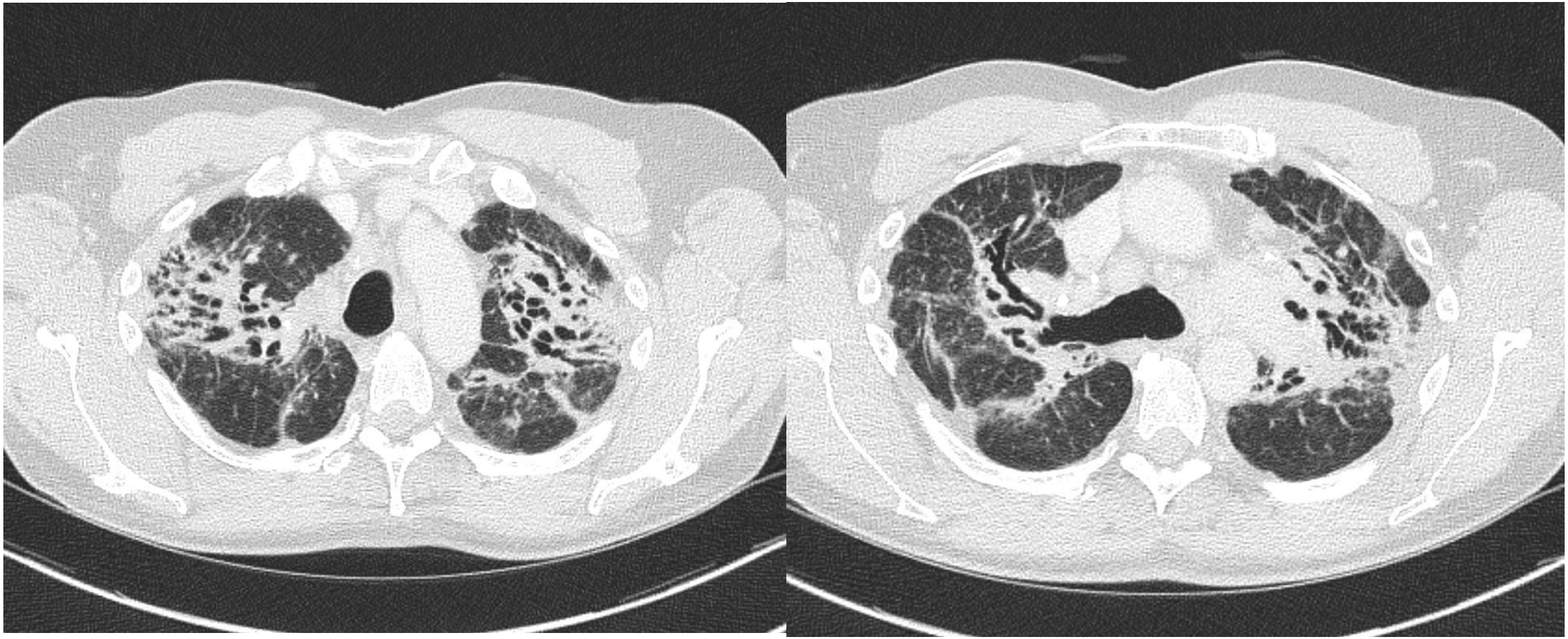
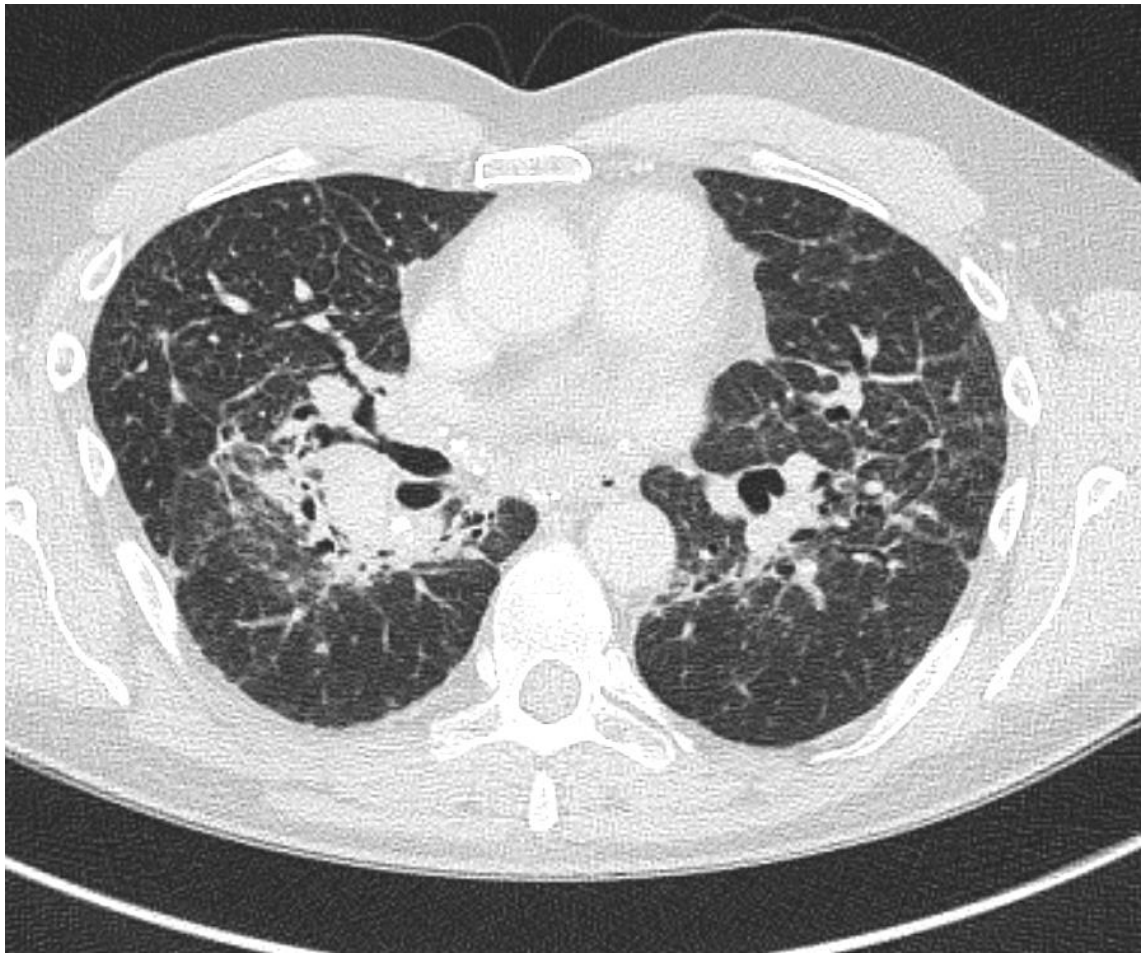
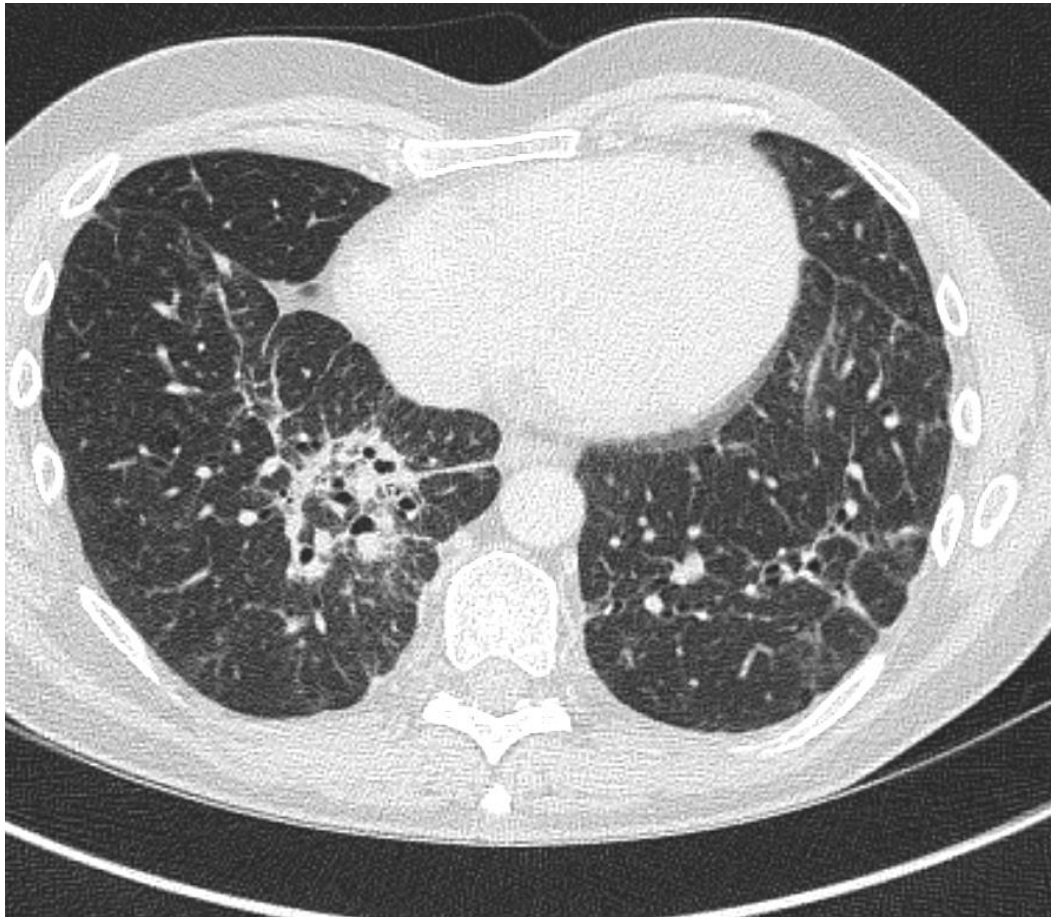


CT 10 years

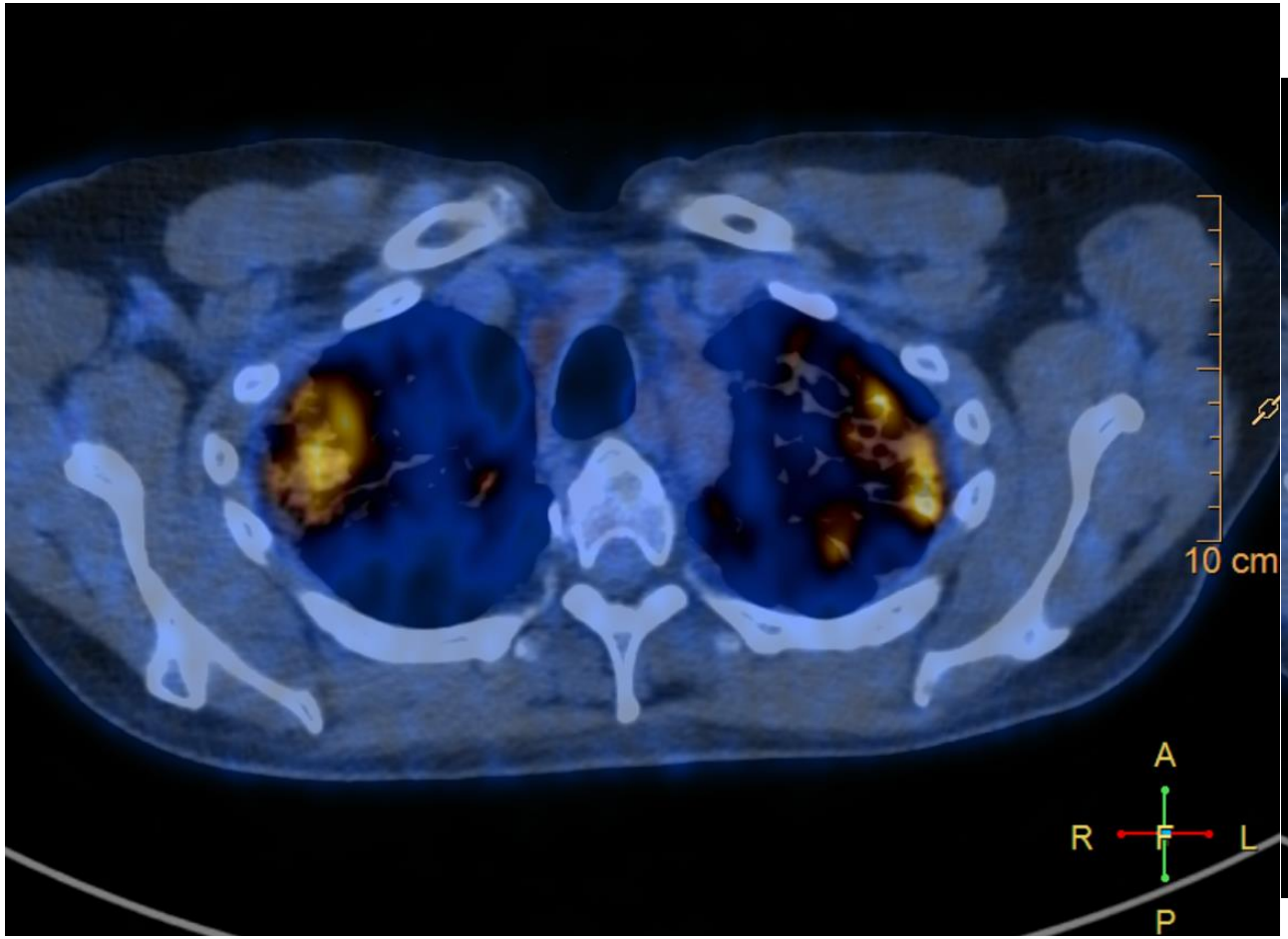


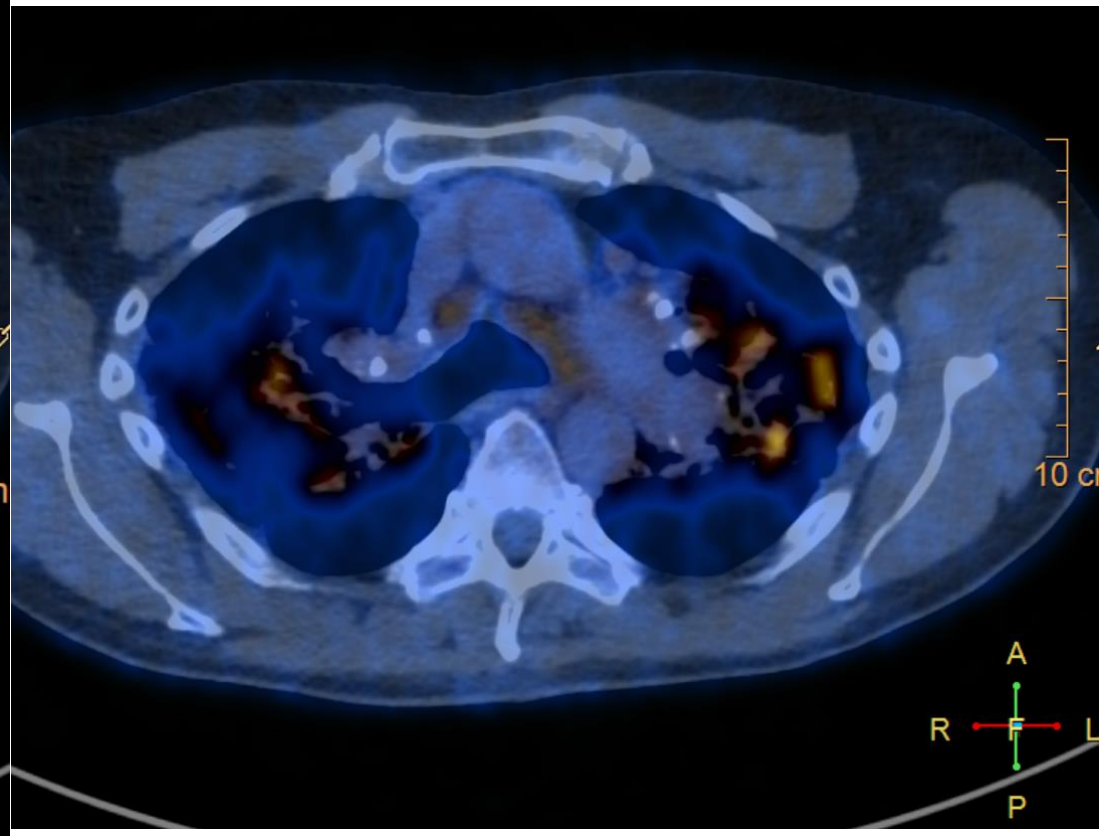
CT 10 years



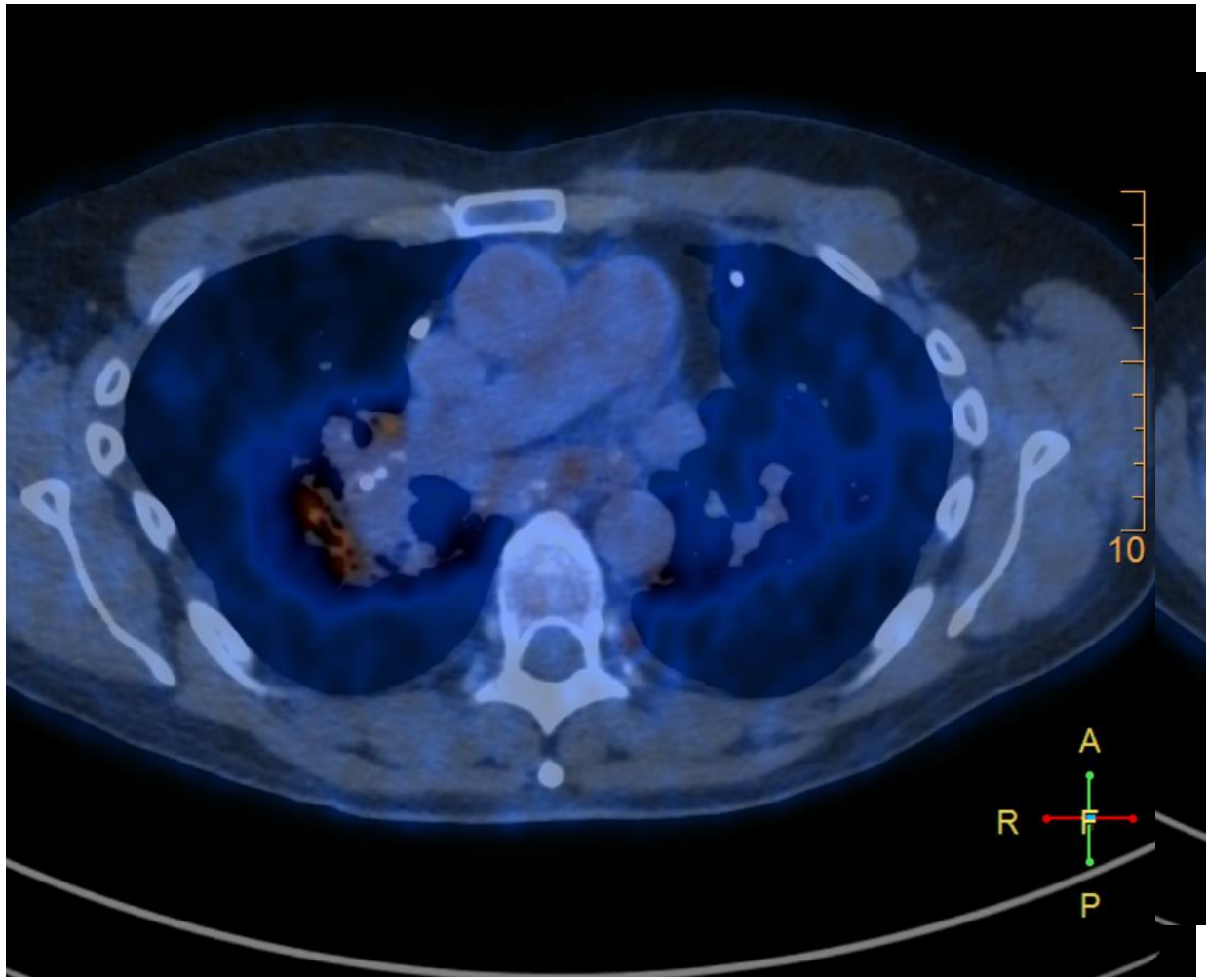


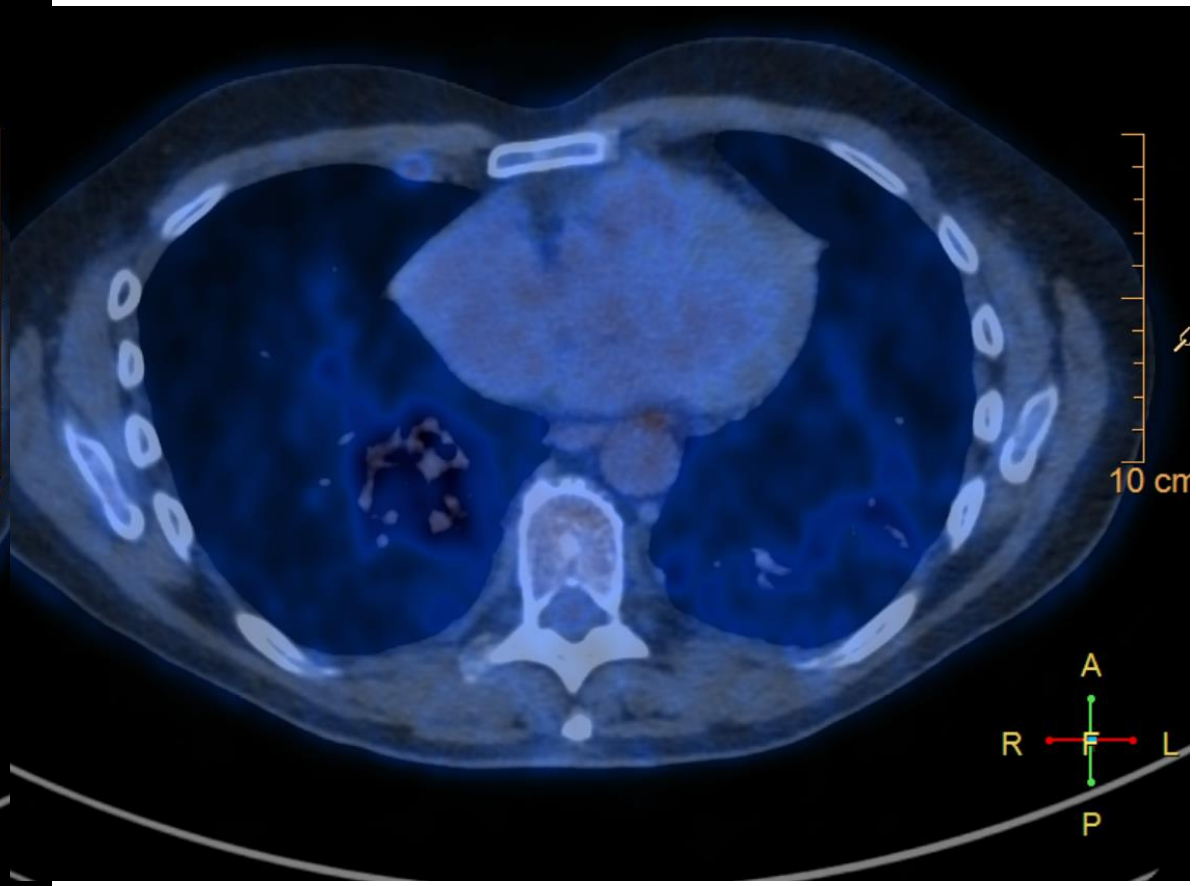
PET CT 10 years





PET CT 10 years





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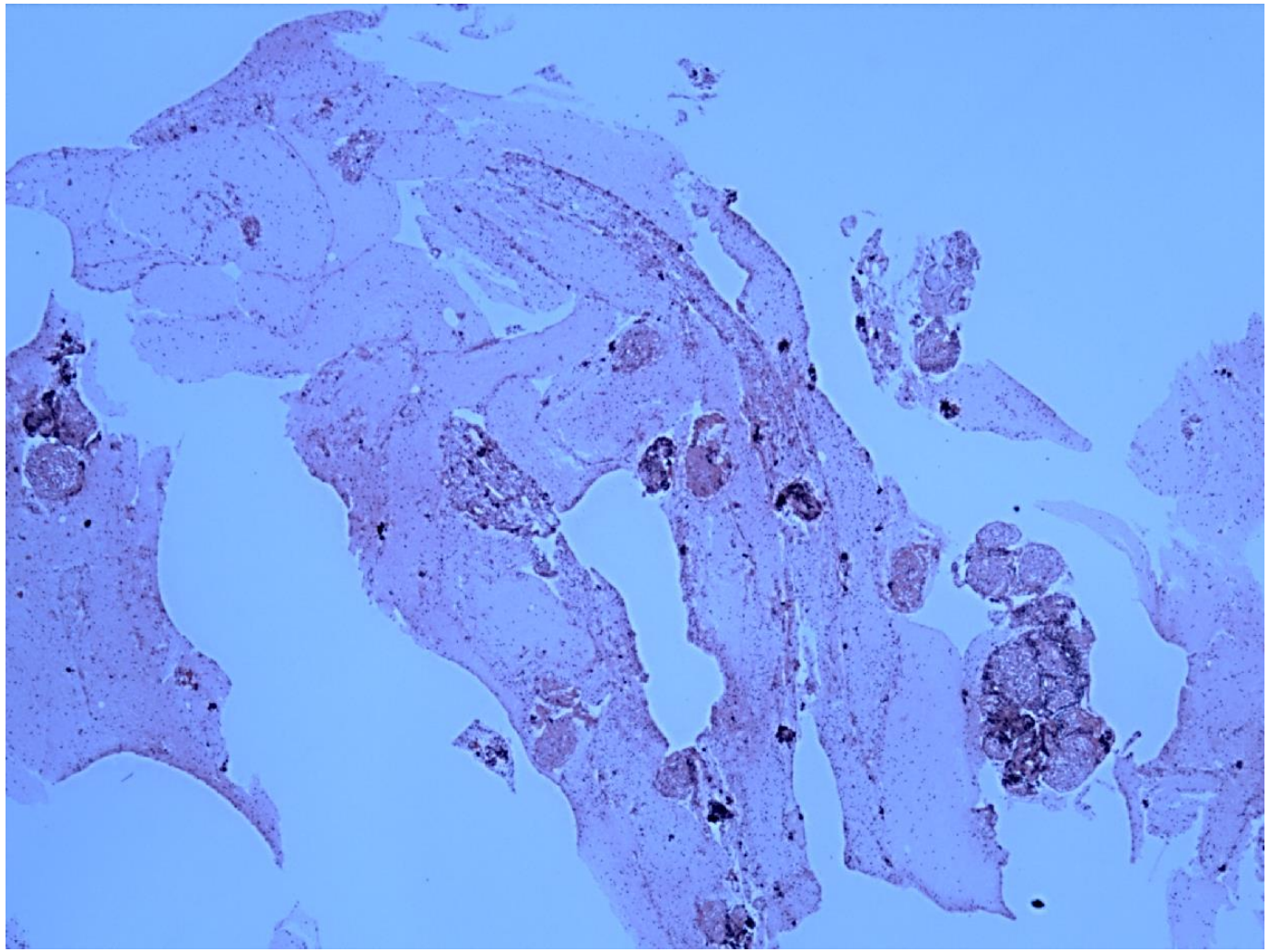

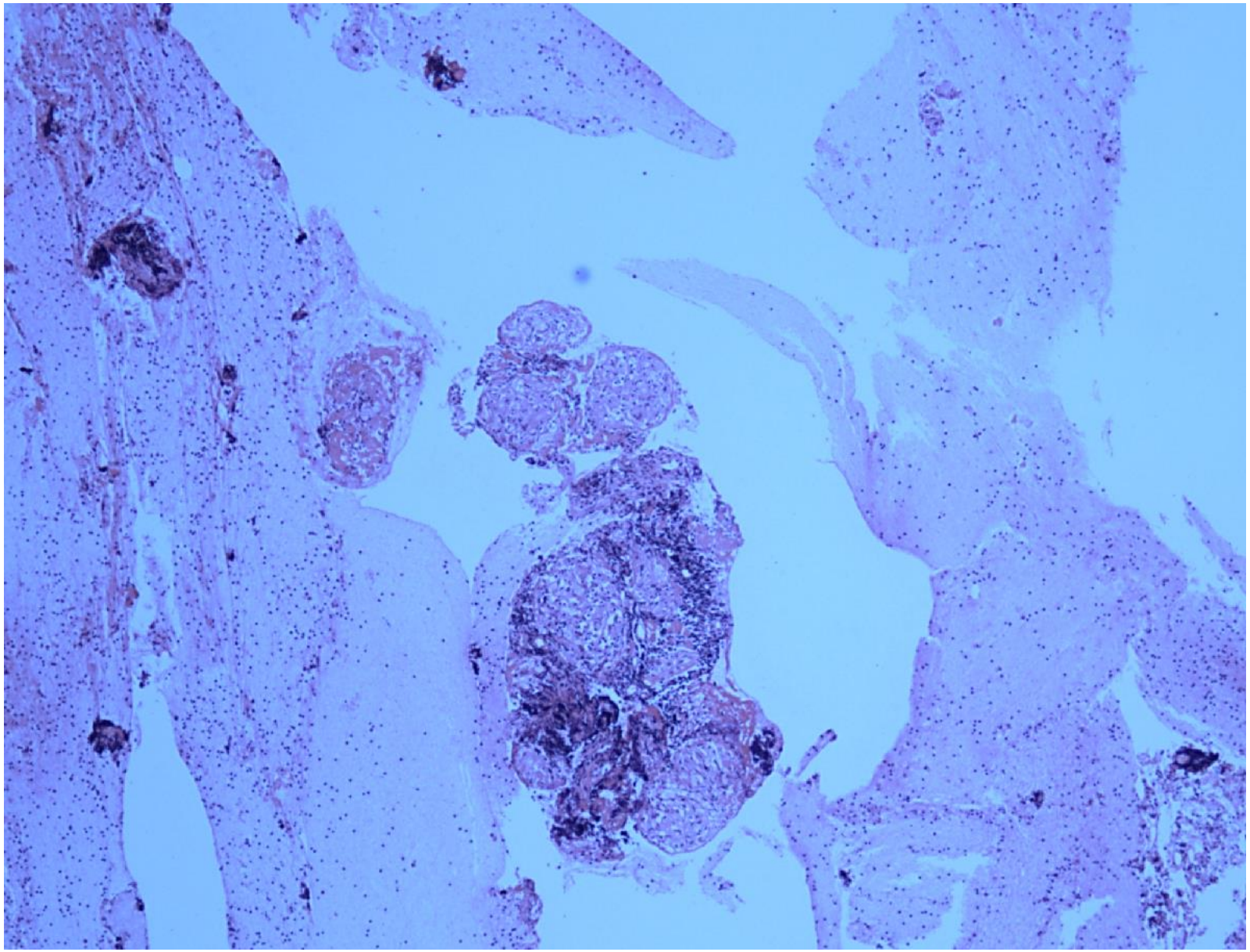

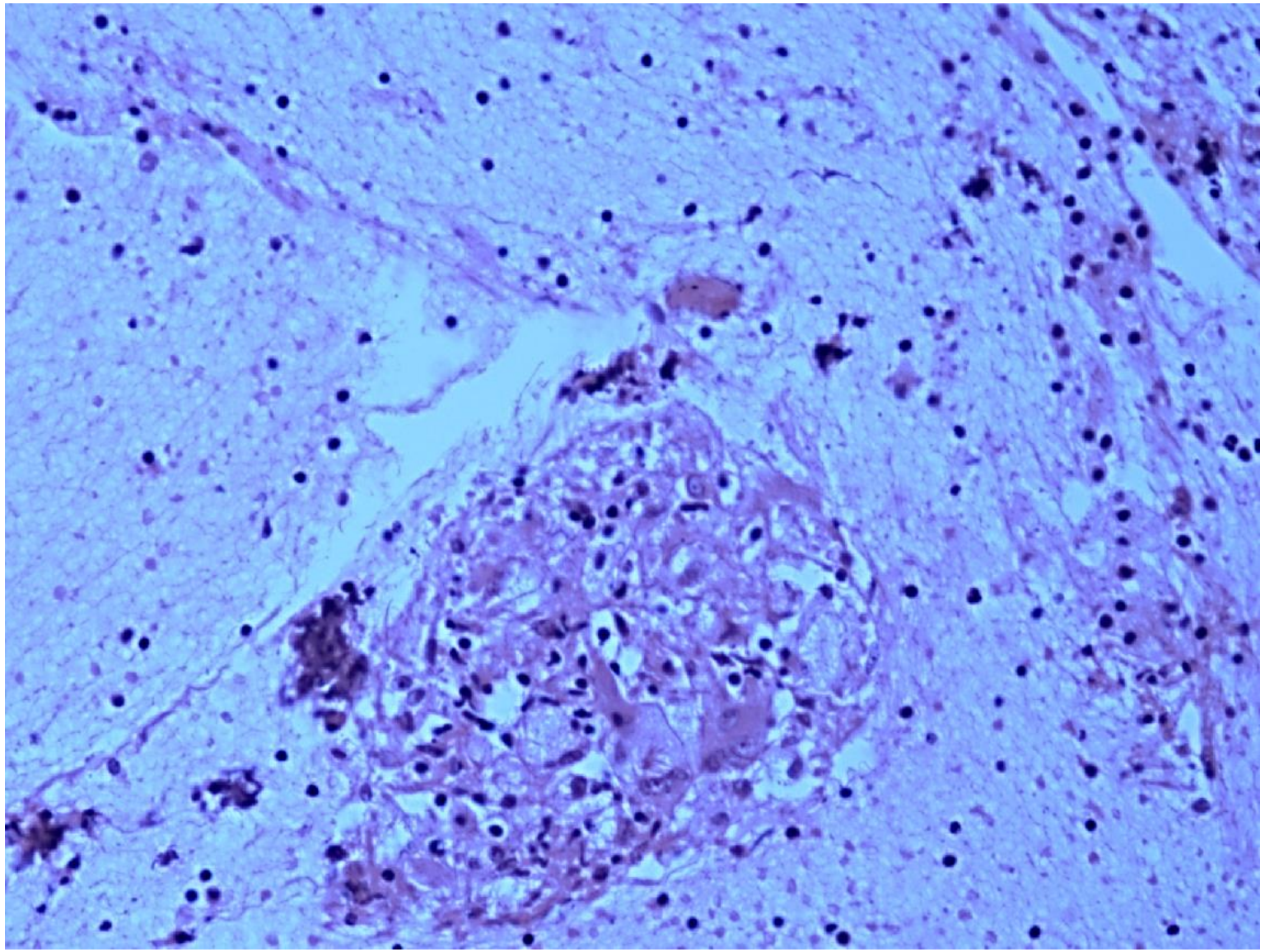

Sarcoidosis

- Brother age 61 had stage 4 sarcoid
- Construction worker
- Unlimited ETT on flat
- Stops after 4 steps up the stairs
- Progressively worsened breathlessness over a year
- Prednisolone on and off for hypercalcaemia

Lung function

Date	BMI	FEV1	FEv1%	FVC	FVC%	DLCO	DLCO%	Change
July 23	24	2.15	67	3.04	74	4.28	51	130ml FVC 9% DLCO improvment
March 23	28	2.24	75	2.91	78		42	350ml 7% FVC decline in 12months
Nov 22								6MWT 515m desat 92%
March 22	27	2.47	81	3.26	85	3.56	40	490ml 12% FVC decline in 12months
Feb 21	26	2.83	92	3.75	97			

Repeat imaging

- CT August 2022
- PET CT August 22
- ECHO no Pulmonary hypertension
- Lymphocytes 0.5 Creatinine 121 Calcium normal

Management

- Started Mycophenolate 1g bd Nov 2022 (not MTX due to skin cancers and due to renal disease)
- Started prednisolone 30mg Nov 22 Current dose 5mg note improved PFT when Pred increased to 20mg for high calcium
- Noticed some improvement in breathing
- Cough returns as steroids weaned
- Insomnia with steroids
- Nintedanib for PF ILD
- Infliximab IFR but opted to change to MTX first

Saw our specialist nurse

- Complex patient with multiple comorbidities (FVC decline 700mls 2.5yrs)
- Main role is support, education, unpacking what he understands and what lies ahead
- Explore concerns
    - What is he struggling with? Work, what do they know? Can he be relocated, Finances family support ,Are hospital visits prohibitive
- Explore physical symptoms cough poor sleep skin cancer renal function
- Interventions, refer to cough physio, rehab, fatigue management baseline 6MWT
- Drug review and monitoring Methotrexate folic acid check bloods Ca LFTS weight skin hair mouth ulceration, shared care agreements
- Disease monitoring regular lung function tests, annual EGC / Echo
- Check alcohol consumption, no pregnancies, skin protection
- Direct to Sarcoidosis UK website information support
- Discuss Nintedamib check weight diarrhoea renal impairment
- At 59years he may be candidate for transplant discussion

Common drugs used in progressive sarcoidosis

Drug Name (Brand Name)	Dose Range	Monitoring	Common Adverse Drug Reactions	Common Drug Interaction	Prescribing Considerations
Methotrexate	7.5mg-15mg once a week Co prescribe Folic acid Weekly or daily but not on the day of methotrexate	FBC, U&Es, LFTs Raised ALT indicates liver toxicity Raised Alk phos suggests obstruction	Increased risk of infections and skin cancers, leucopenia, thrombocytopenia, anaemia,* mouth ulcers, **liver toxicity,** Lung fibrosis	Extensively protein bound and may displace, or be displaced by, other acidic drugs (check BNF), aspirin/NSAIDS, **folate antagonists**	"off-license" ONCE weekly dose, teratogenic (affects fetal growth)
Nintedanib (Ofev)	150mg BD 100mg BD	U&Es, LFTs, BP, weight	Diarrhoea, nausea and vomiting, Liver toxicity, hypertension, increased risk of thromboembolic events	Rifampicin, carbamazepine, phenytoin, and St. John's Wort	Administer 12 hours apart

Please see www.medicines.org.uk for further information

* Patients should be advised to report immediately any evidence of infection, unexpected bruising or bleeding

Saw the specialist nurse

The biggest challenge in the long-term management of patients with progressive ILD

- Uncertainty around the trajectory of the disease
- The impact of the disease on their psychological and emotional stability
- Dealing with decreasing functional ability
- Adherence to treatment, as this is can be worse than the disease

Learning points

- Progressive sarcoidosis may fit PF-ILD criteria for nintedanib
- PET-CT helpful for IFR for infliximab – but high infection risk
- Regular PFT monitoring is important