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9-12 Nisan 2025
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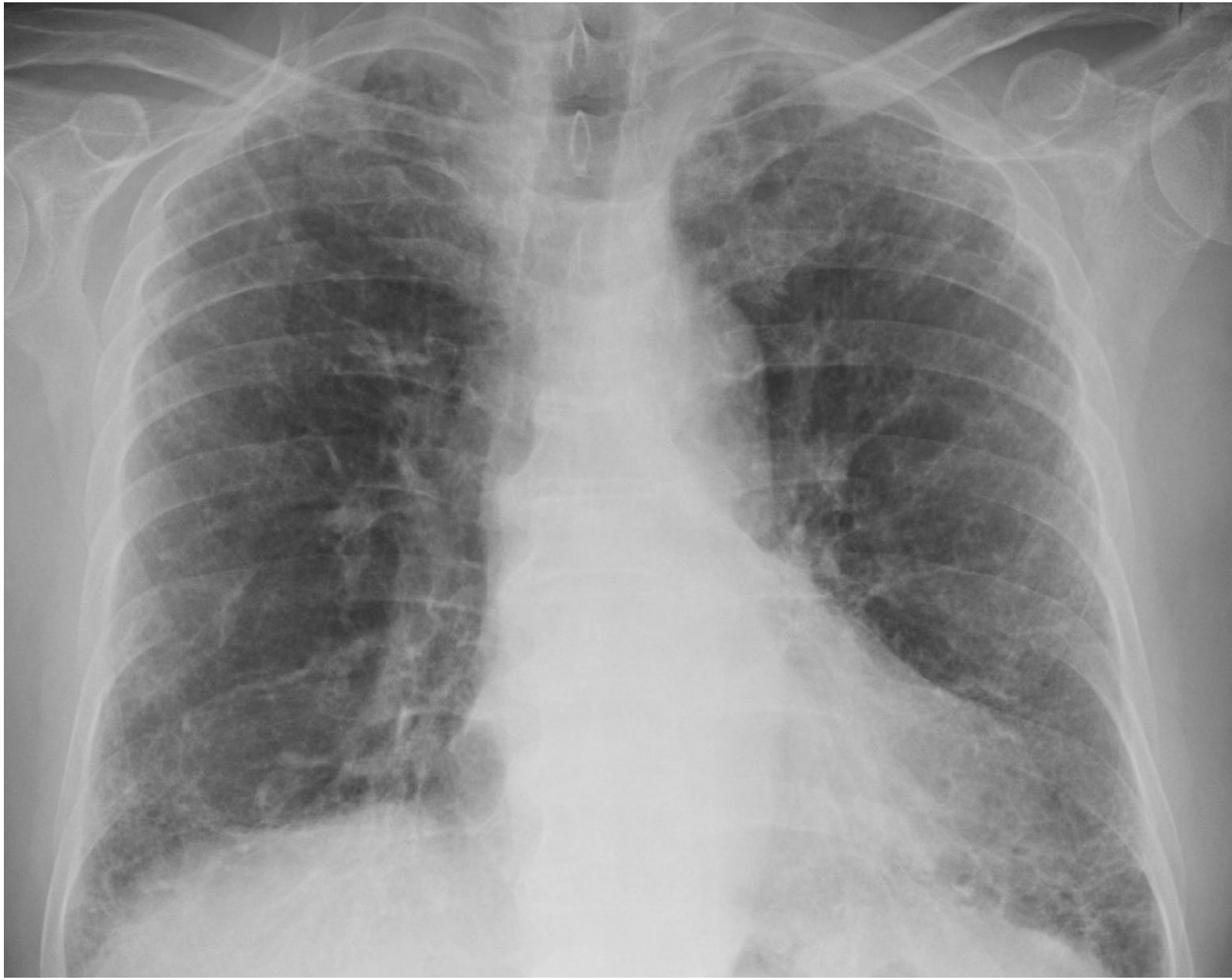
OLGULARLA FİBROTİK VE KİSTİK AKCİĞER HASTALIKLARI

Doç. Dr. Fatma Demirci Üçsular

SBÜ. İzmir Tıp Fakültesi Dr. Suat Seren Göğüs Hastalıkları ve Cerrahisi Eğitim ve Araştırma Hastanesi, İZMİR

Olgu-1

- 70 yaş, erkek hasta.
- 1 yıldır kuru öksürük, efor dispnesi,
- Şoför, sigara yok, maruziyeti yok.
- Bibaziler velcro ralleri , clubbing yok.
- SpO2: %95.
- Rutin hemogram ve biyokimya olağan.
- Romatolojik biyobelirteçler negatif.
- FVC: %90, DLCO: %61



Olgu-1: Toraks BT

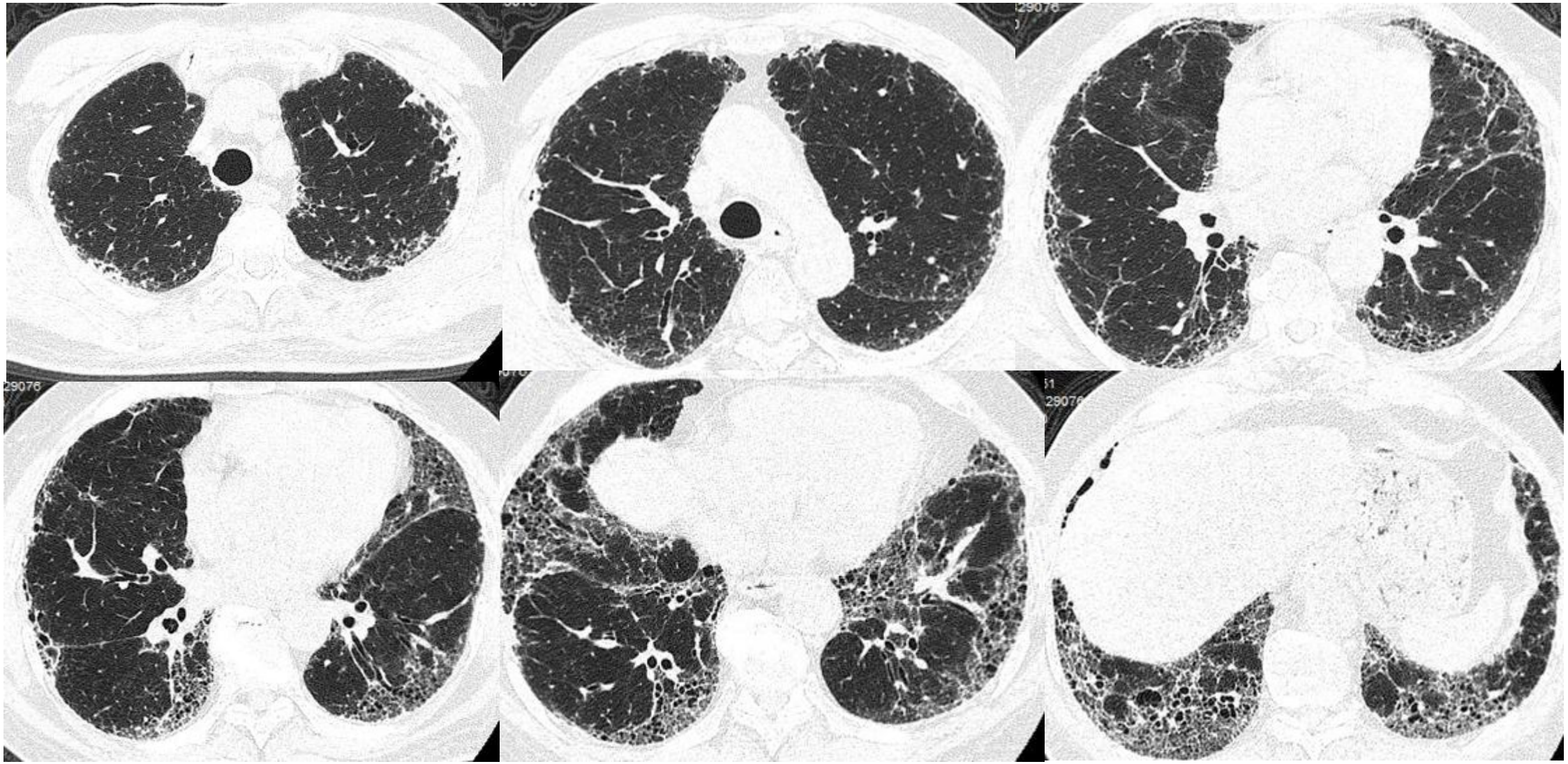


Table 3. High-Resolution Computed Tomography Patterns in Idiopathic Pulmonary Fibrosis

	HRCT Pattern			
	UIP Pattern	**Probable UIP Pattern**	**Indeterminate for UIP**	**CT Findings Suggestive of an Alternative Diagnosis**
Level of confidence for UIP histology	Confident (>90%)	Provisional high confidence (70–89%)	Provisional low confidence (51–69%)	Low to very low confidence (⩽50%)
Distribution	• Subpleural and basal predominant • Often heterogeneous (areas of normal lung interspersed with fibrosis) • Occasionally diffuse • May be asymmetric	• Subpleural and basal predominant • Often heterogeneous (areas of normal lung interspersed with reticulation and traction bronchiectasis/ bronchiolectasis)	• Diffuse distribution without subpleural predominance	• Peribronchovascular predominant with subpleural sparing (consider NSIP) • Perilymphatic distribution (consider sarcoidosis) • Upper or mid lung (consider fibrotic HP, CTD-ILD, and sarcoidosis) • Subpleural sparing (consider NSIP or smoking-related IP)
CT features	• Honeycombing with or without traction bronchiectasis/ bronchiolectasis • Presence of irregular thickening of interlobular septa • Usually superimposed with a reticular pattern, mild GGO • May have pulmonary ossification	• Reticular pattern with traction bronchiectasis/ bronchiolectasis • May have mild GGO • Absence of subpleural sparing	• CT features of lung fibrosis that do not suggest any specific etiology	• Lung findings ○ Cysts (consider LAM, PLCH, LIP, and DIP) ○ Mosaic attenuation or three-density sign (consider HP) ○ Predominant GGO (consider HP, smoking-related disease, drug toxicity, and acute exacerbation of fibrosis) ○ Profuse centrilobular micronodules (consider HP or smoking-related disease) ○ Nodules (consider sarcoidosis) ○ Consolidation (consider organizing pneumonia, etc.) • Mediastinal findings ○ Pleural plaques (consider asbestosis) ○ Dilated esophagus (consider CTD)

Definition of abbreviations: CT = computed tomography; CTD = connective tissue disease; DIP = desquamative interstitial pneumonia; GGO = ground-glass opacity; HP = hypersensitivity pneumonitis; HRCT = high-resolution computed tomography; ILD = interstitial lung disease; IP = interstitial pneumonia; LAM = lymphangioleiomyomatosis; LIP = lymphoid interstitial pneumonia; NSIP = nonspecific interstitial pneumonia; PLCH = pulmonary Langerhans cell histiocytosis; UIP = usual interstitial pneumonia.
The previous term, "early UIP pattern," has been eliminated to avoid confusion with "interstitial lung abnormalities" described in the text. The term "indeterminate for UIP" has been retained for situations in which the HRCT features do not meet UIP or probable UIP criteria and do not explicitly suggest an alternative diagnosis. Adapted from Reference 2.

- Kesin UIP
- IPF olarak değerlendirildi
- Antifibrotik tedavi başlandı
- Aşılama
- Pulmoner rehabilitasyon programına alındı

UIP(usual interstitial pneumonia) yapan nedenler:

1. İPF
2. Fibrotik HP
3. RA
4. Sistemik sklerozis
5. Diğer BDH
6. Sarkoidoz
7. İlaçlar (amiodaron vb.)
8. Asbestozis
9. Enfeksiyonlar

Olgu-2

50 yaş, kadın hasta, ev hanımı

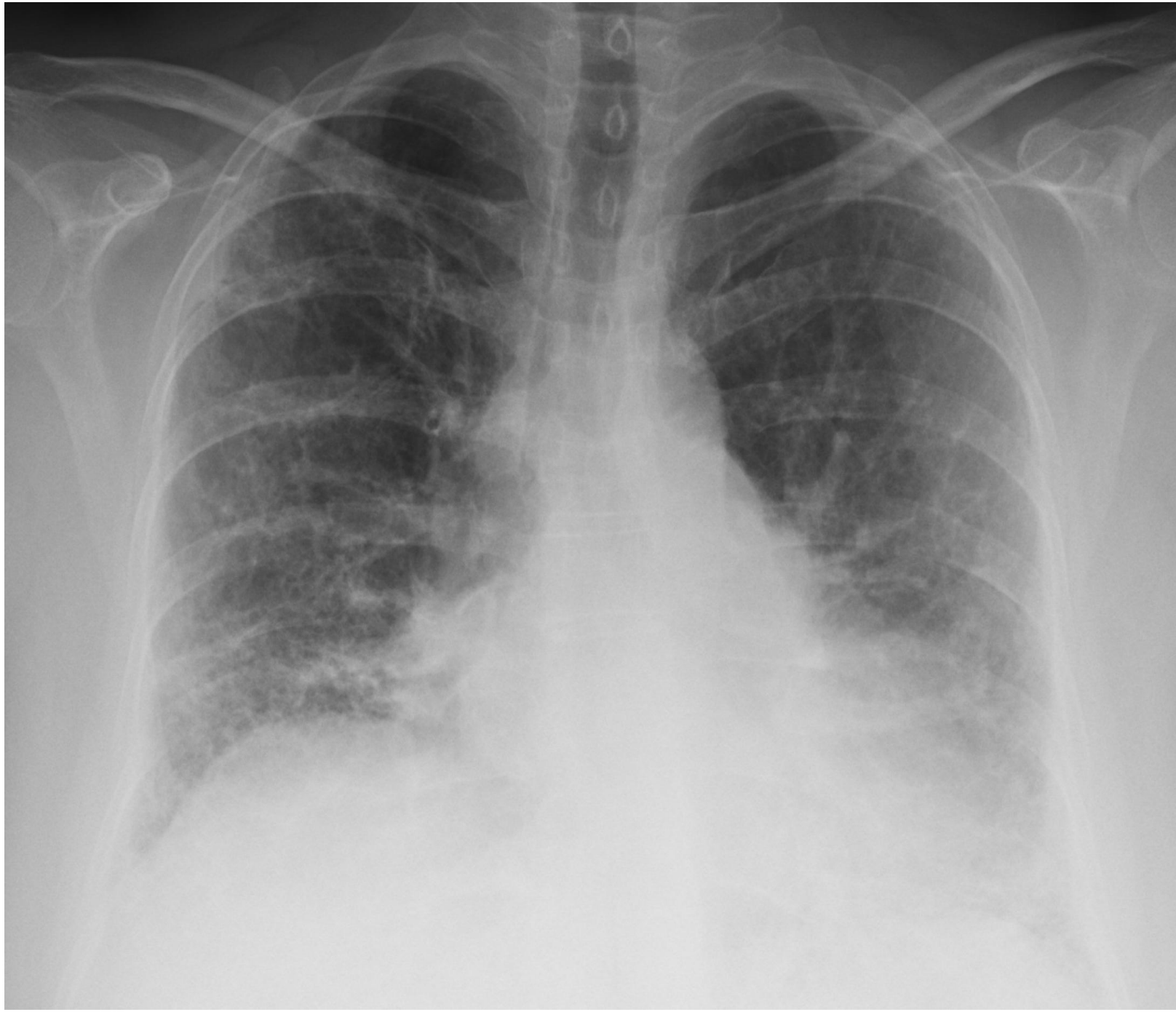
Nefes darlığı, kuru öksürük, yutma güçlüğü, halsizlik.

5 yıldır romatoloji tarafından **sistemik sklerozis-interstisiyel akciğer hastalığı (SSc-İAH)** tanısı ile takipte.

Sigara: 3 pkt yılı ,

ex-smoker ,

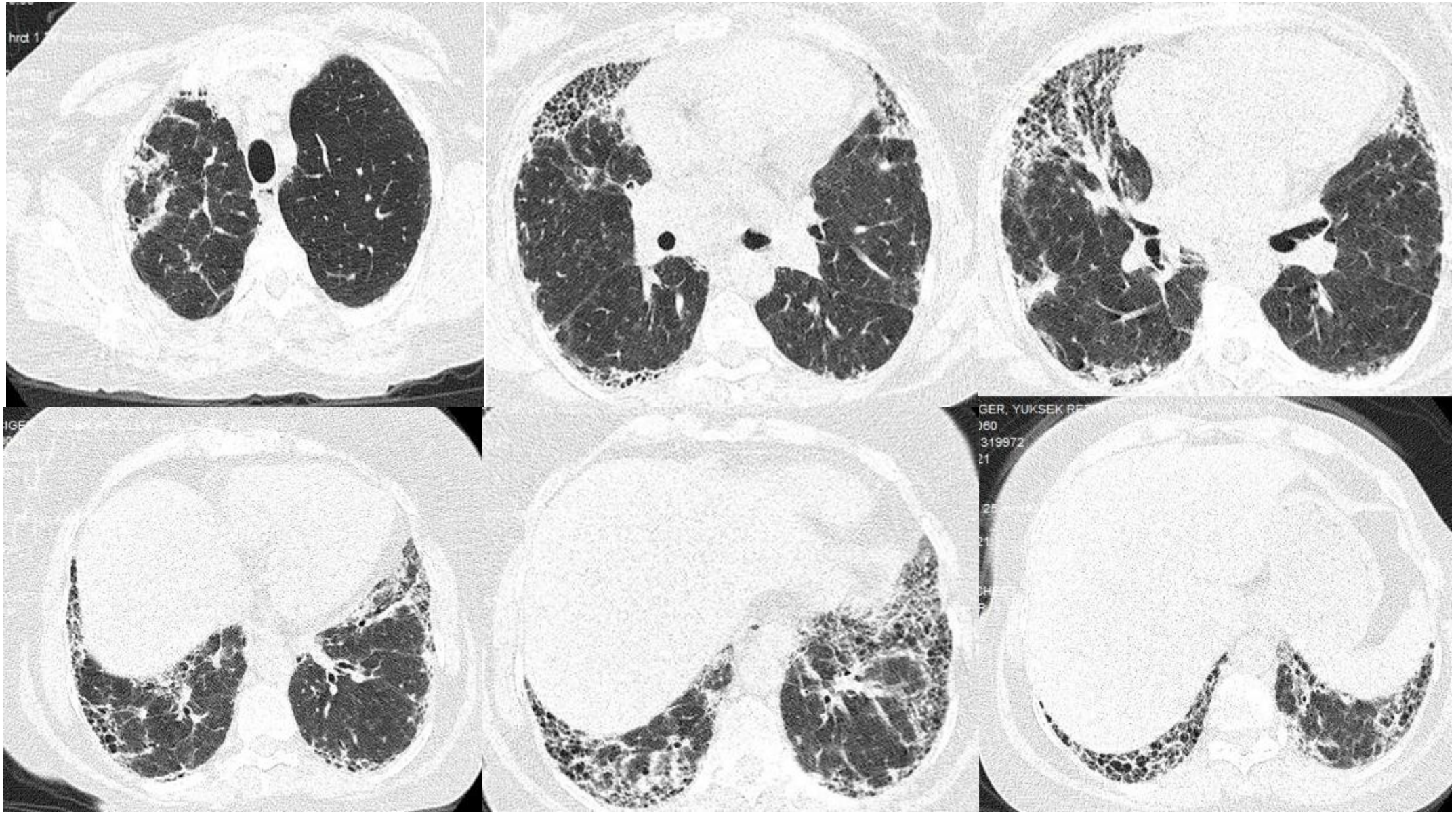
Siklofosfamid, metilprednizolon ve ASA kullanıyor



Olgu-2

- Solunum sesleri: Bibaziler raller
- Sklerodaktili, yutma güçlüğü, raynaud fenomeni
- SPO2:%91
- FVC: % 64, DLCO: % 60
- ANA pozitif,
- Anti-Scl 70+++
- RF:66

Olgu-2: Toraks HRCT



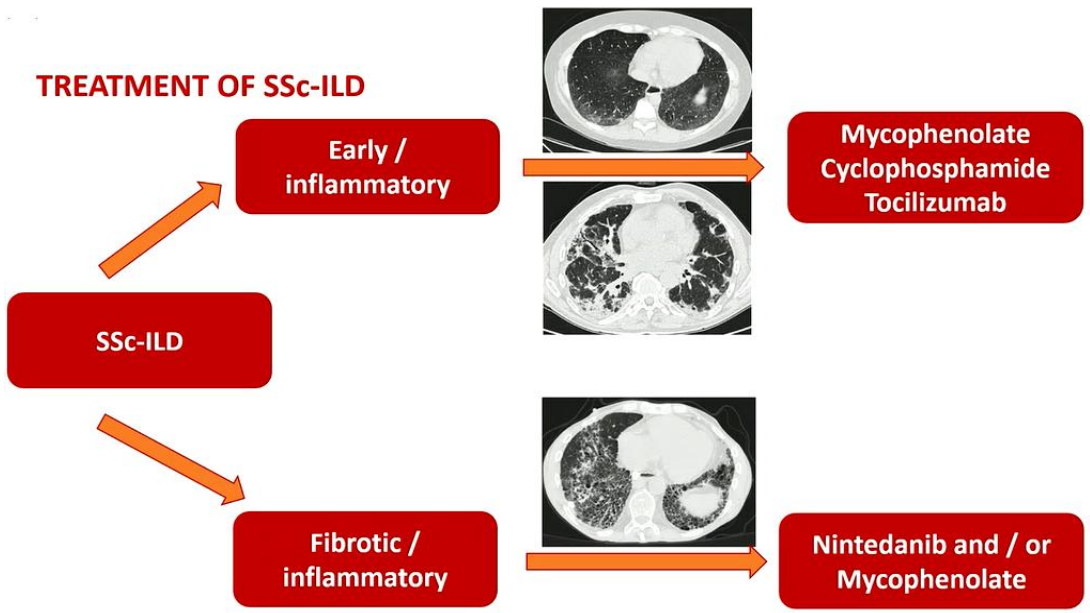
Sistemik sklerozis:

- Etiyolojisi bilinmeyen heterojen hastalık
- Endotel ve fibroblast disfonksiyonu sonucu küçük damar hasarı, artmış kollajen üretimi ve fibrozis ile karakterize

- Sistemik sklerozda akciğer tutulumu hastalığın erken dönemlerinde başlamakta olup mortalite ve morbiditenin en önemli nedenidir
- İAH ve pulmoner arteriyal hipertansiyon en sık akciğer tutulum tipleri ve en sık ölüm sebebidir

Sistemik sklerozis:

- En sık YRBT bulgusu nonspesifik interstisyel pnömoni (NSİP)paterni
- UIP paterni ise hastaların 1/3’ünde görülebilir
- SSc-İAH gelişen hastalara immunsupresif tedaviye ilave antifibrotik-nintedanib tedavisi başlanır

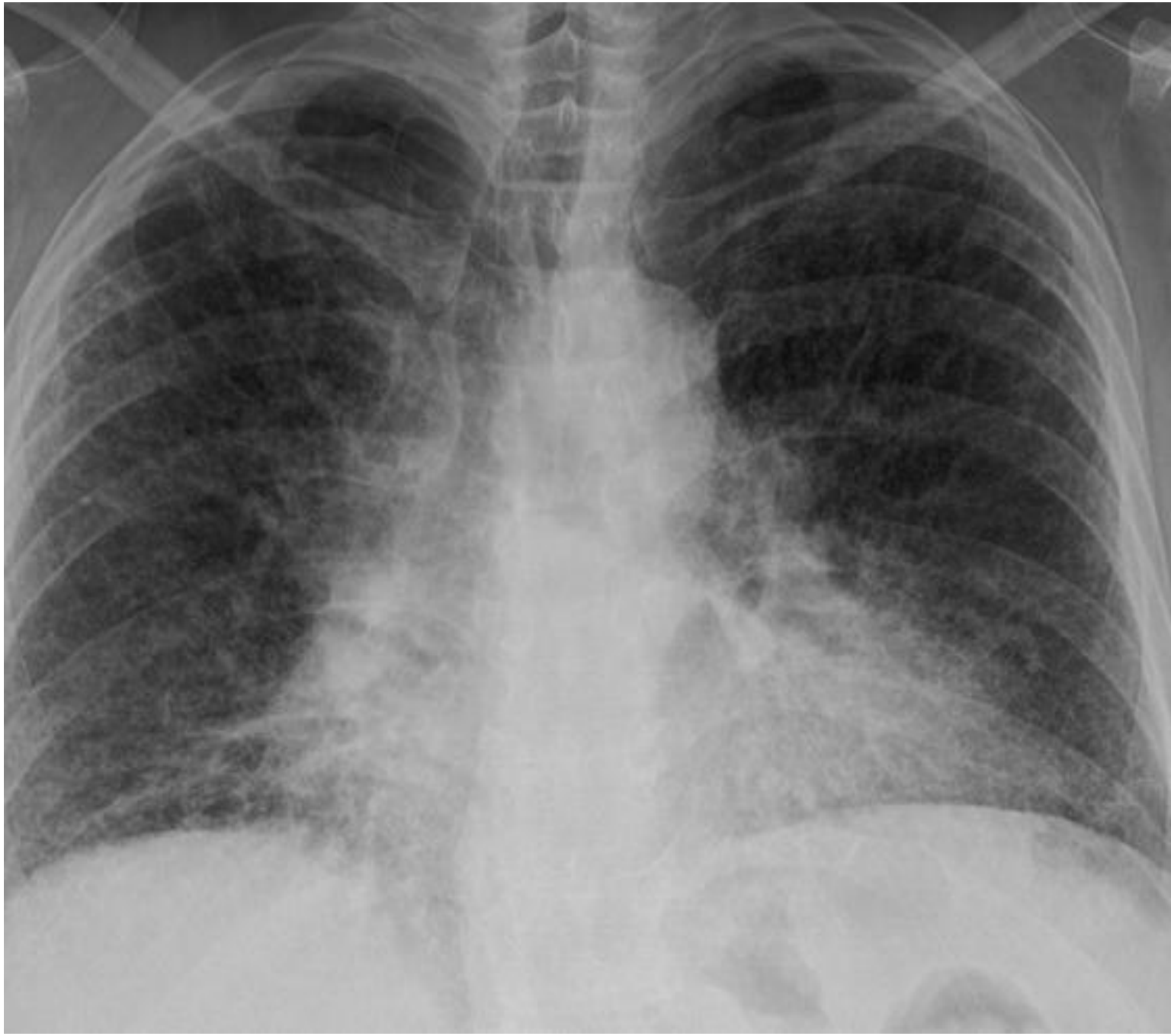


- Sistemik Sklerozis- İnterstisyel Akciğer Hastalığı

Tedaviye mikofenolat mofetil ve nintedanib eklendi.

Olgu-3

- 48 yaş, erkek hasta
- 1 yıldır nefes darlığı , öksürük, göğüs ağrısı, balgam.
- 90 paket yılı, 8 aydır kullanmıyor
- Zeytin fabrikasında çalışıyor (Asetik asit, laktik asit ve kostik maruziyeti var)
- 10 yıldır evde kuş besliyor
- Dispneik, taşipneik, SpO2: %85
- Bibaziler velcro ralleri



Olgu-3

- Rutin hemogram ve biyokimya olağan
- *CRP: 67, Romatolojik biyobelirteçler negatif
- *PO2:52 mmHg
- FVC: %45
- FEV1: %50
- FEV1/FVC: % 86
- TLCO :%16

- BAL: Alveoler makrofaj:%75, lenfosit: %13, nötrofil: %12

Olgu-3: İlk başvuru Toraks BT

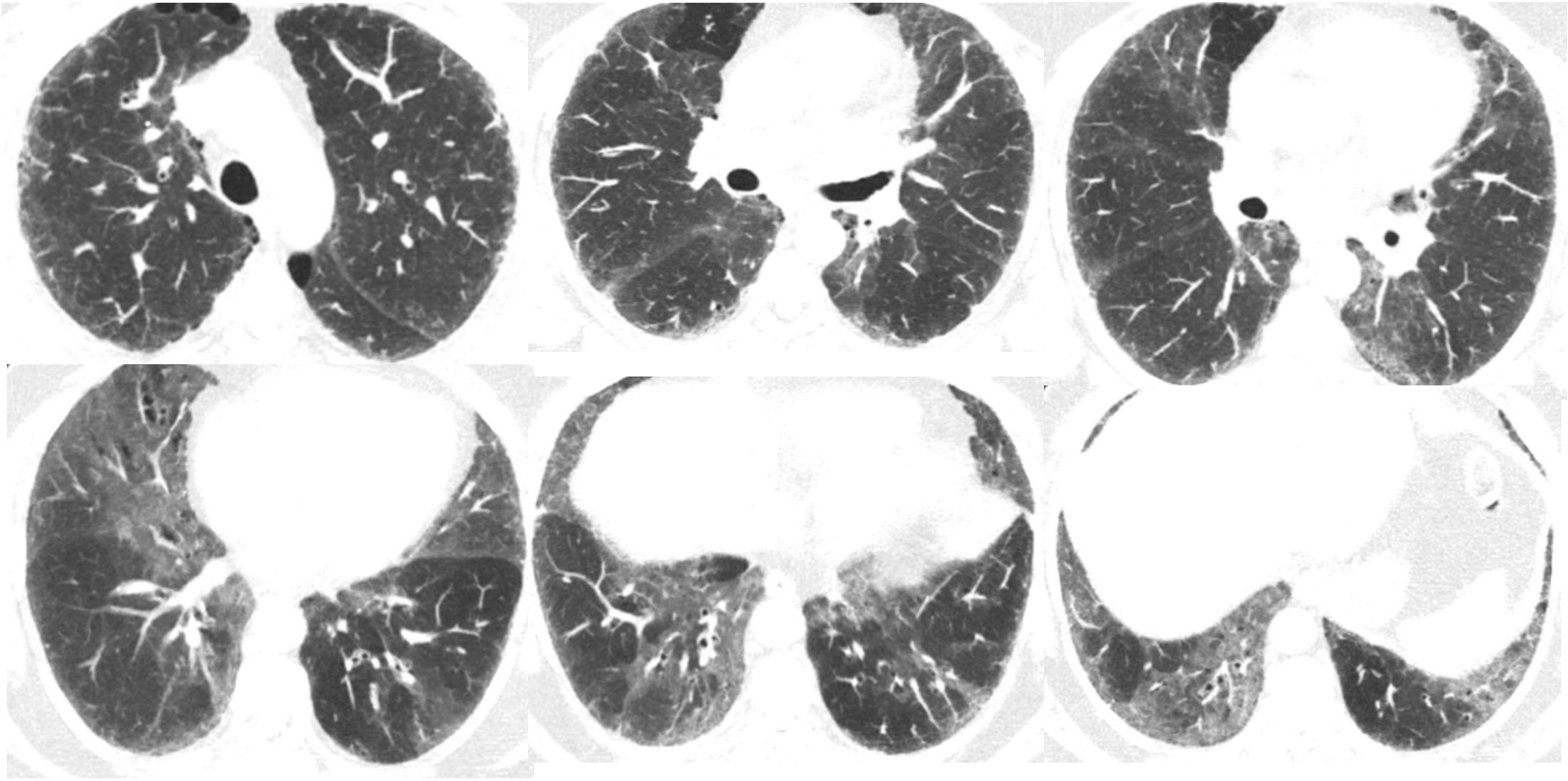


Table 6. Chest HRCT Scan Features of the Fibrotic HP Pattern

HRCT Pattern	Typical HP	Compatible with HP	Indeterminate for HP
Description	The "typical HP" pattern is suggestive of a diagnosis of HP. It requires *a*) an HRCT pattern of lung fibrosis (as listed below) in one of the distributions and *b*) at least one abnormality that is indicative of small airway disease	"Compatible-with-HP" patterns exist when the HRCT pattern and/or distribution of lung fibrosis varies from that of the typical HP pattern; the variant fibrosis should be accompanied by signs of small airway disease	The "indeterminate-for-HP" pattern exists when the HRCT is neither suggestive nor compatible with a typical and probable HP pattern
Relevant radiological findings	HRCT abnormalities indicative of lung fibrosis are most commonly composed of irregular linear opacities/coarse reticulation with lung distortion; traction bronchiectasis and honeycombing may be present but do not predominate The distribution of fibrosis may be: • Random both axially and craniocaudally or • Mid lung zone–predominant or • Relatively spared in the lower lung zones HRCT abnormalities indicative of small airway disease: • Ill-defined, centrilobular nodules and/or GGOs • Mosaic attenuation, three-density pattern,* and/or air trapping (*often in a lobular distribution*)	Variant patterns of lung fibrosis: • UIP pattern: basal and subpleural distribution of honeycombing with/without traction bronchiectasis (*per 2018 diagnosis of IPF guidelines* [20]) • Extensive GGOs with superimposed subtle features of lung fibrosis Variant (predominant) distributions of lung fibrosis: • Axial: peribronchovascular, subpleural areas • Craniocaudal: upper lung zones HRCT abnormalities indicative of small airway disease: • Ill-defined centrilobular nodules, or • Three-density pattern* and/or air trapping	Lone patterns (i.e., not accompanied by other findings suggestive of HP) of: • UIP pattern (*as per 2018 IPF diagnosis guidelines* [20]) • Probable UIP pattern (*as per 2018 IPF diagnosis guidelines* [20]) • Indeterminate pattern for UIP (*as per 2018 IPF diagnosis guidelines* [20]) • Fibrotic NSIP pattern • Organizing pneumonia–like pattern • Truly indeterminate HRCT pattern

Definition of abbreviations: GGO = ground-glass opacity; HP = hypersensitivity pneumonitis; HRCT = high-resolution computed tomography; IPF = idiopathic pulmonary fibrosis; NSIP = nonspecific interstitial pneumonia; UIP = usual interstitial pneumonia.

Rarely, fibrotic HP may be seen *1*) as a component of combined pulmonary fibrosis and emphysema or pleuroparenchymal fibroelastosis with emphysema, *2*) as a pure emphysematous form of HP, or *3*) in acute exacerbation.

*The three-density pattern was formerly called the "headcheese sign." It is described in detail in Table 4.

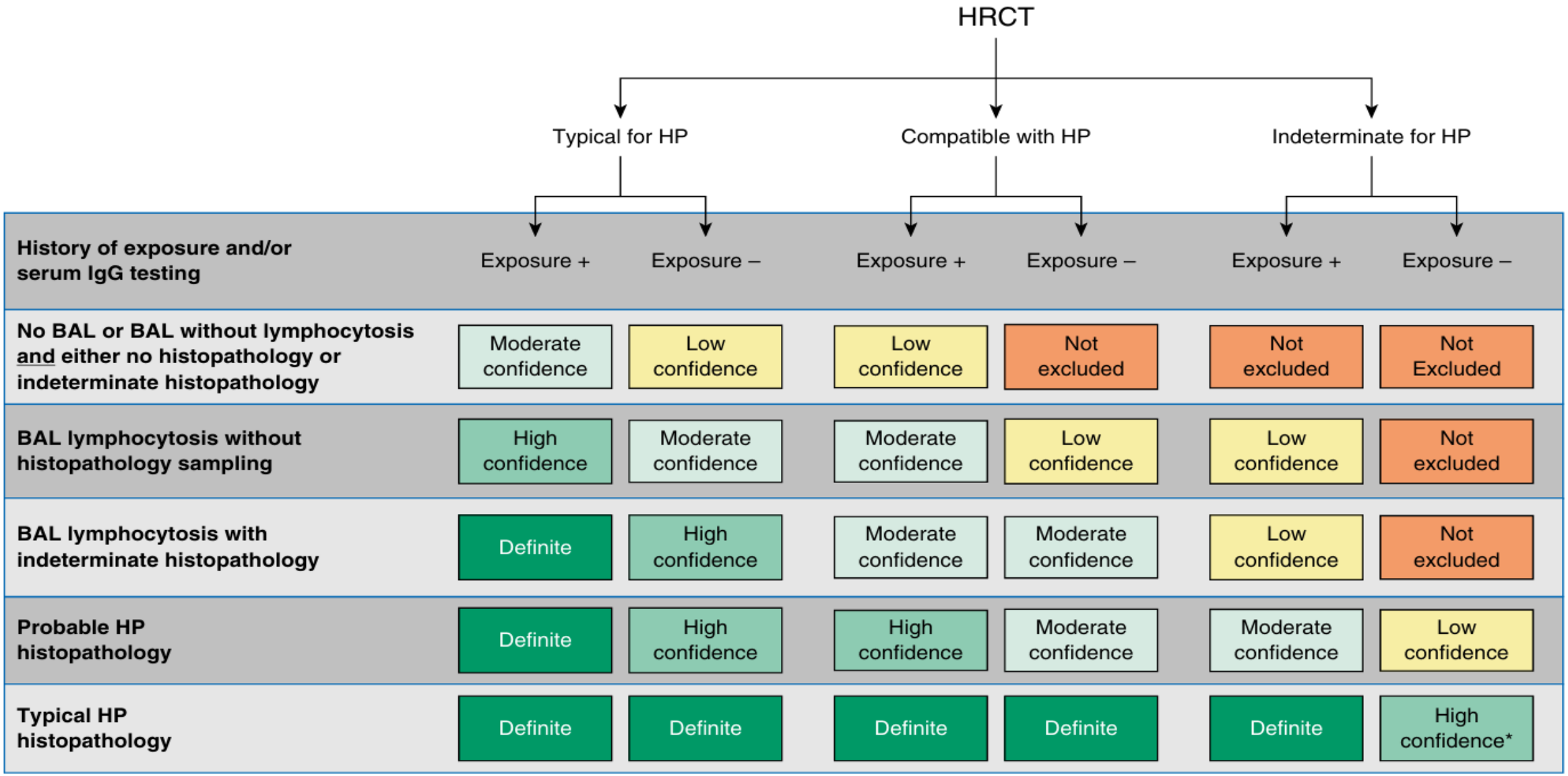


Figure 6. Hypersensitivity pneumonitis diagnosis based on incorporation of imaging, exposure assessment, BAL lymphocytosis, and histopathological findings. All confidence levels are subject to multidisciplinary discussion. *Confidence may increase to "definite" if the pathologist's conclusion persists after reevaluation in the context of additional clinical information or an expert second opinion on histopathology. HP = hypersensitivity pneumonitis; HRCT = high-resolution computed tomography.

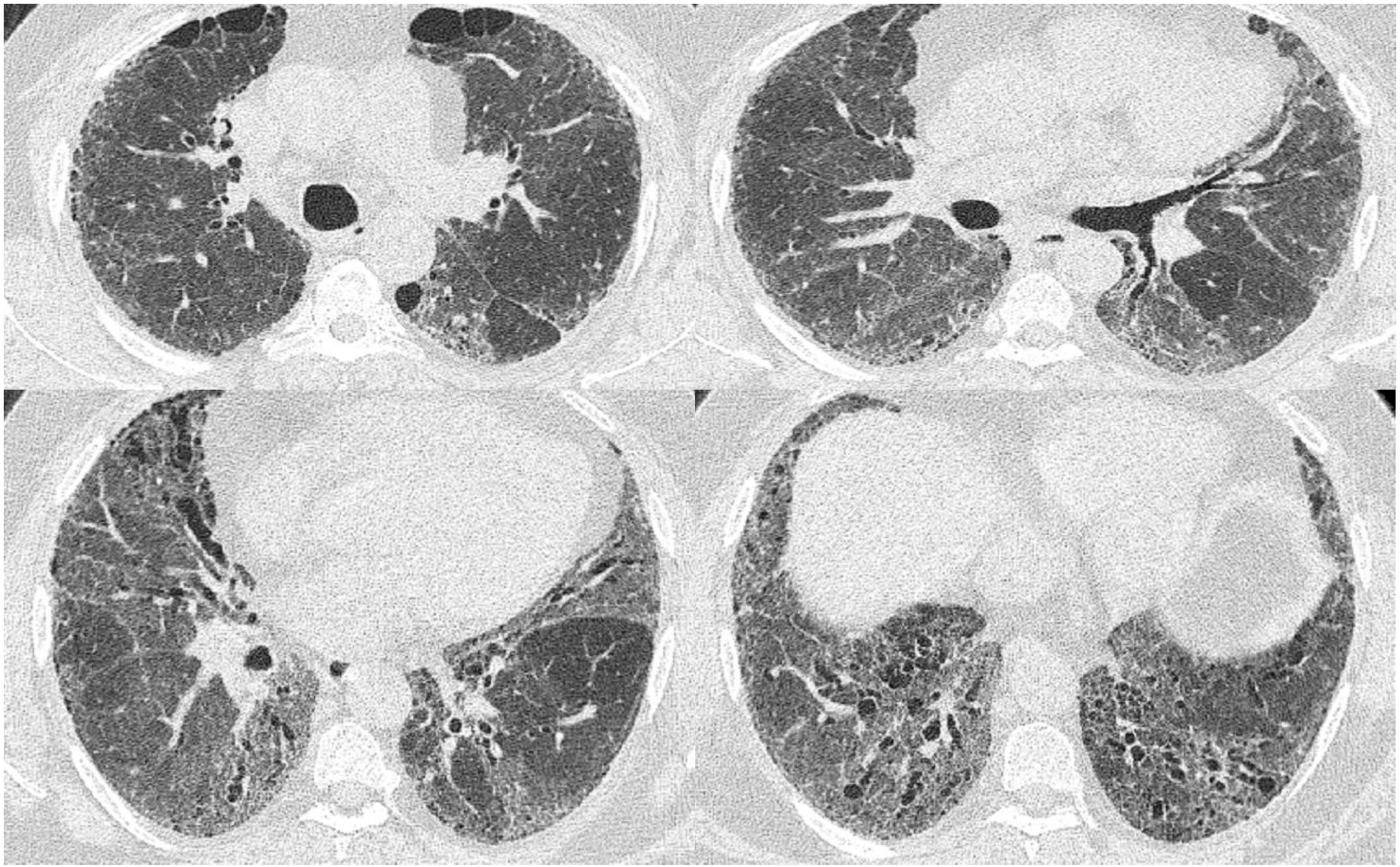
- Mesleki maruziyetin, kuş besleyiciliğinin ve radyolojik bulguların uyumlu olması nedeniyle olgu ***fibrotik hipersensivite pnömonisi*** olarak değerlendirildi
- Tekrar 0,5 mg/kg metil prednizolon başlandı.

Olgu-3

- Hipersensivite pnömonisi tanısı ile 1 yıl kortikosteroid tedavisi aldı.
- Yanıtsızlık nedeniyle tedaviye mikofenolat mofetil eklendi.

18 ay sonra Toraks BT

Olgu-3



- Fibrotik hipersensivite pnömonisi, progresif pulmoner fibrozis olarak değerlendirildi.

- Tedaviye nintedanib (2x1, PO) eklendi.

2022 ATS/ERS Kılavuzuna göre Progresif Pulmoner Fibrozis Tanımı

Radyolojik olarak pulmoner fibrozis kanıtı bulunan, İPF dışında etiyolojisi bilinen veya bilinmeyen interstisyel akciğer hastalığı mevcut olan bir hastada, PPF (progresif pulmoner fibrozis), **alternatif bir açıklama olmaksızın son 1 yıl içinde aşağıdaki üç kriterden en az ikisinin gerçekleşmesi olarak tanımlanmaktadır*:**

1. Solunum semptomlarında kötüleşme

2. Hastalık progresyonunun fizyolojik kanıtı (aşağıdakilerden herhangi biri):

a. 1 yıllık takip süresince FVC'de ≥%5'lik mutlak düşüş

b. 1 yıllık takip süresince DLCO'da (Hemoglobin açısından düzeltilmiş) ≥%10'luk mutlak düşüş

3. Hastalık progresyonunun radyolojik kanıtı (aşağıdakilerden bir veya daha fazlası):

a. Traksiyon bronşektazisi ve bronşiyolektazinin yaygınlığı veya şiddetinde artış

b. Traksiyon bronşektazisi ile birlikte yeni buzlu cam opasitesi

c. Yeni ince retikülasyon

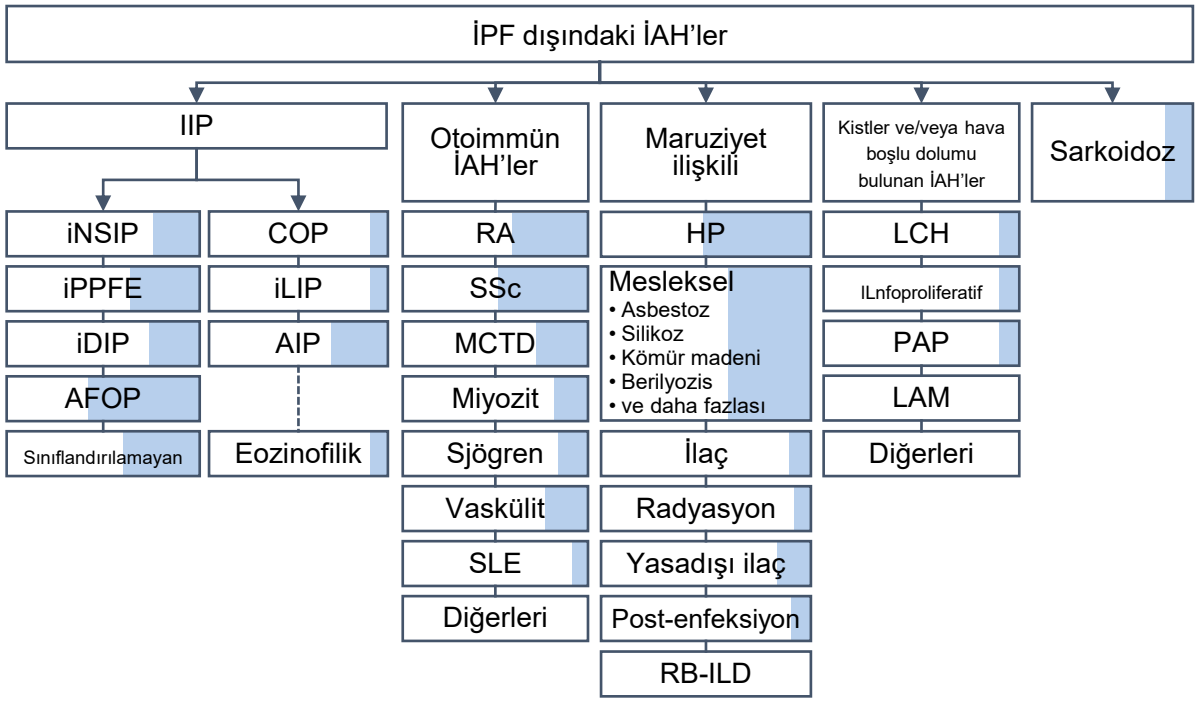
d. Retiküler anormalliğin yaygınlığında veya kalınlığında artış

e. Yeni veya artmış bal peteği görünümü

f. Lober hacim kaybında artış

Raghu G *et al. Am J Respir Crit Care Med* 2022;205:e18–47

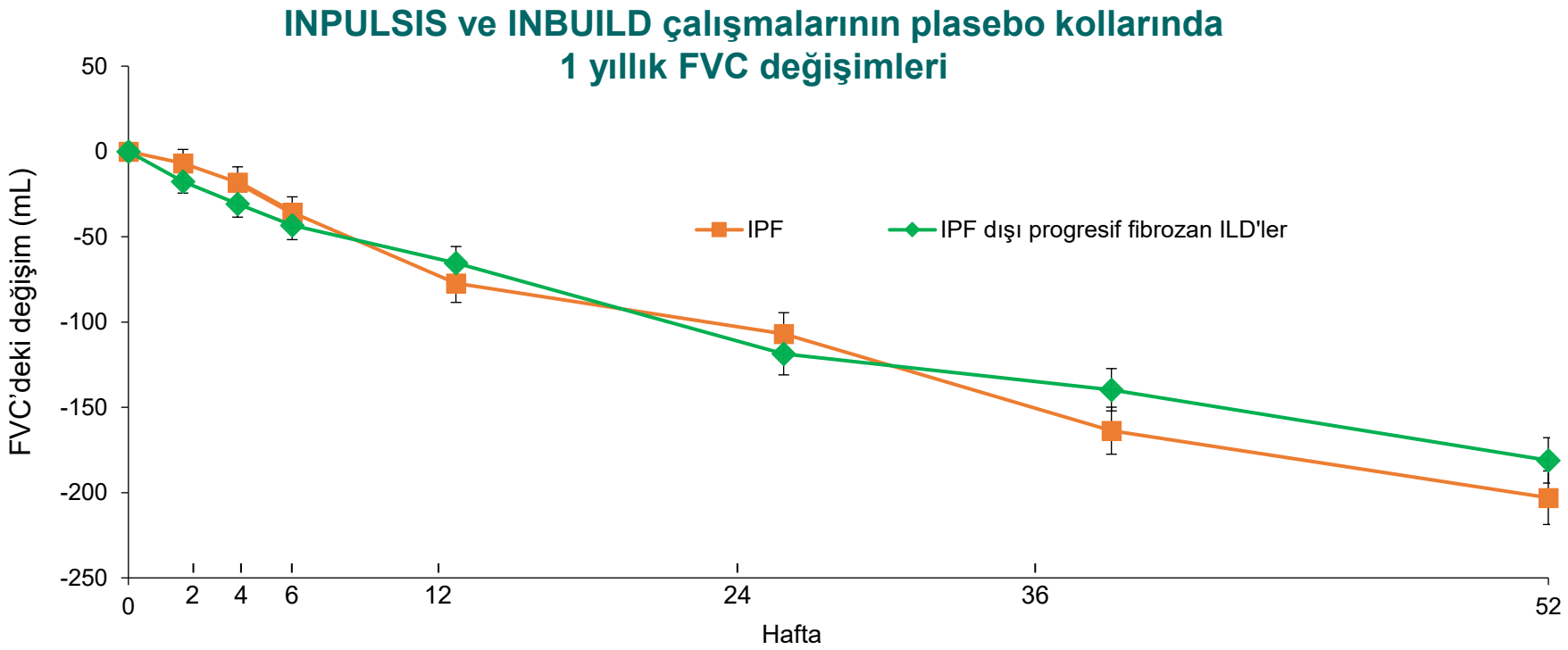
PPF saptanan ILD'li hastaların oranının, uluslararası komitenin konsensüsüne göre aşağıdaki gibi olduğu tahmin edilmiştir



- Taralı alanlar, uzman görüşüne dayalı olarak PPF ortaya çıkan hastaların tahmini oranını temsil etmektedir
- Bu tahminleri destekleyen veriler mevcut değildir

Raghu G *et al. Am J Respir Crit Care Med* 2022;205:e18–47

Progresif pulmoner fibrozisli hastalar IPF hastalarına benzer bir seyir göstermektedir



Brown KK et al. Eur Respir J 2020;55:2000085.

Olgu-4

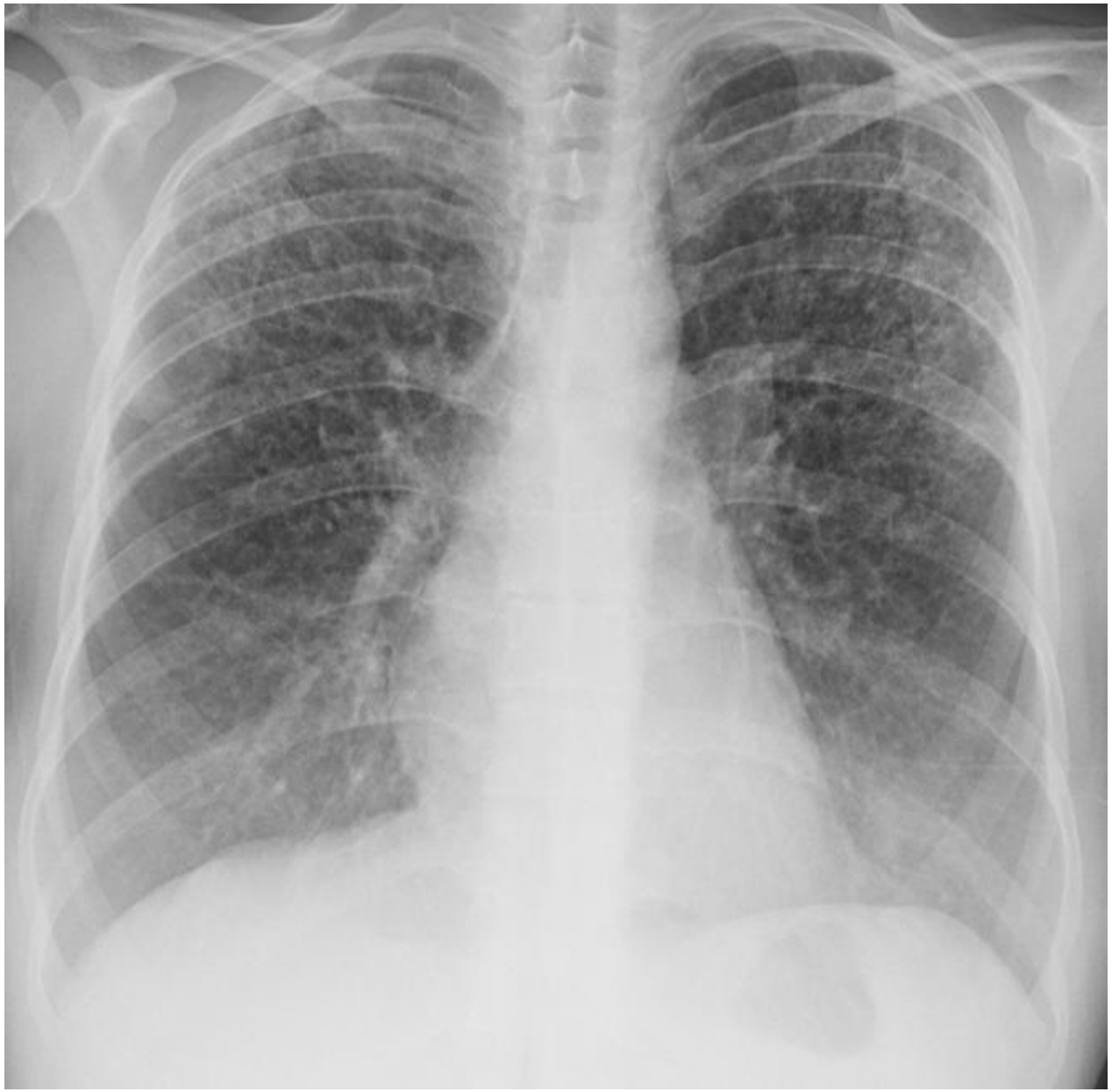
43 yaş, kadın hasta, ev hanımı.

8 yıldır öksürük, efor dispnesi. Sigara 25 paket yılı, ex-smoker. Çevresel maruziyeti yok.

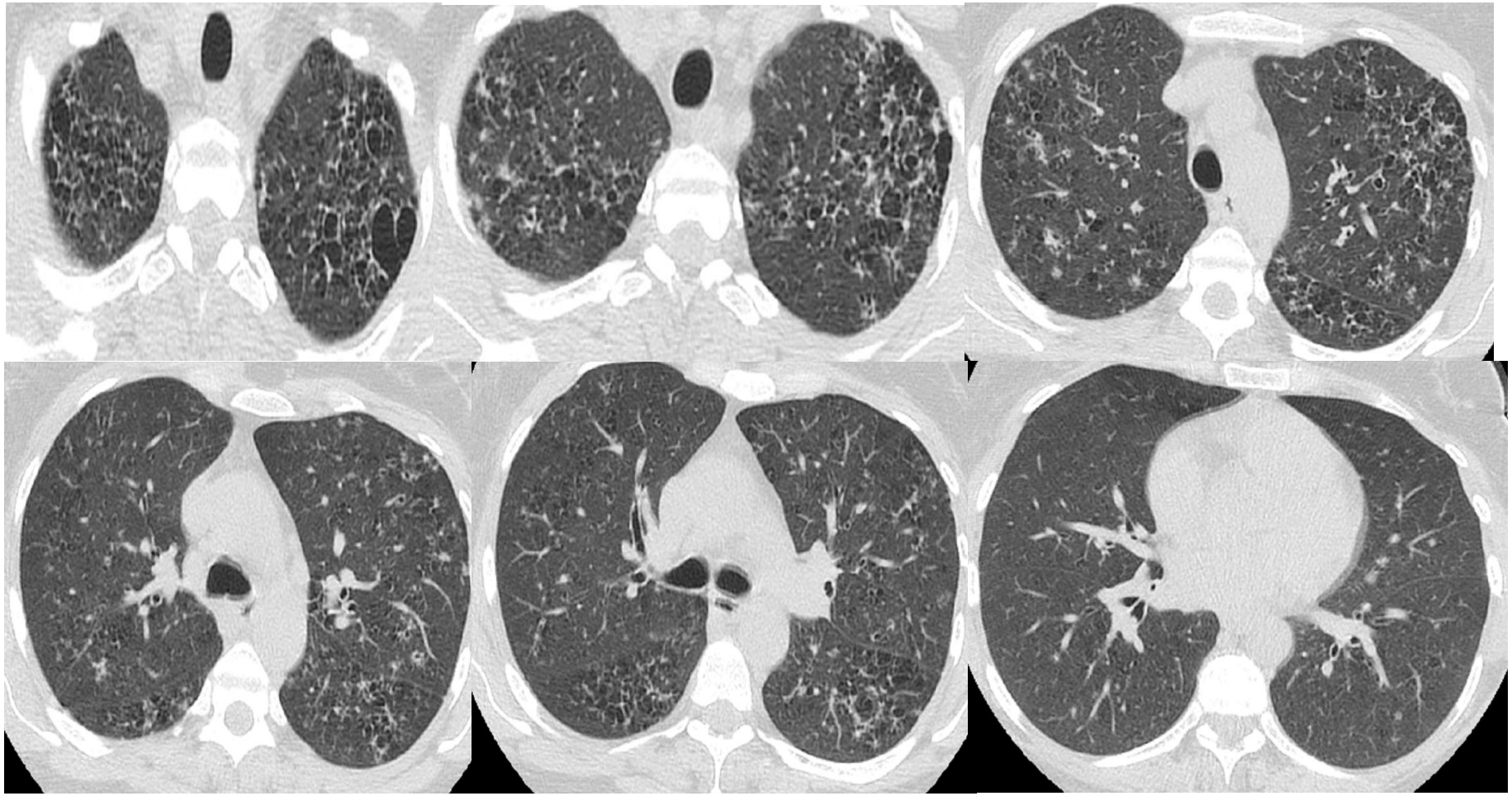
SpO2:%96. Solunum sesleri normal.

Hemogram ve rutin biyokimya normal, romatolojik biyobelirteçler ve HIV negatif.

FVC. % 80, FEV1: % 69, FEV1/FVC :74 TLCO .% 32.



Olgu-4: Toraks BT



Olgu-4

- BAL'da %90 alveoler makrofaj, %2 lenfosit,%3 eozinofil, %5 nötrofil lökosit, CD1a >%5, S-100 pozitif.

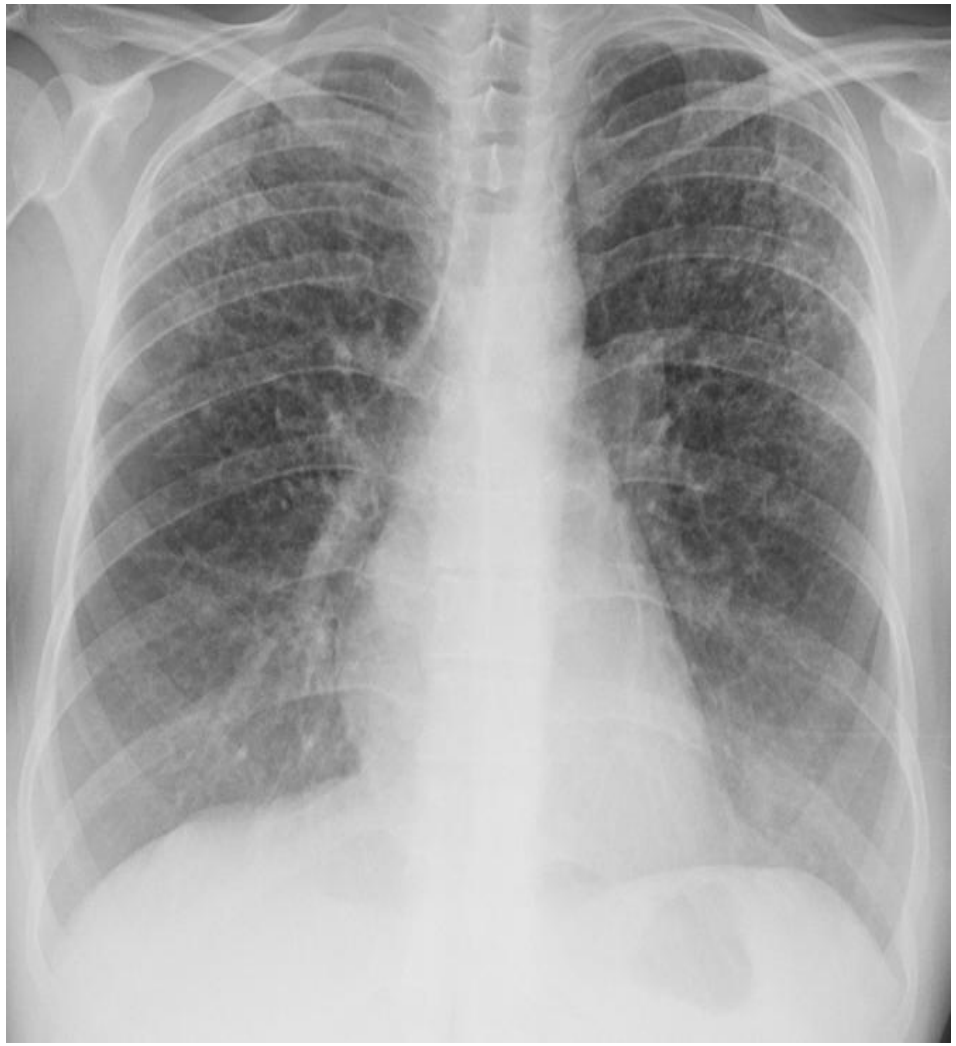
- Pulmoner langerhans hücreli histiositozis olarak değerlendirildi.

Extra organ tutulumu saptanmadı.

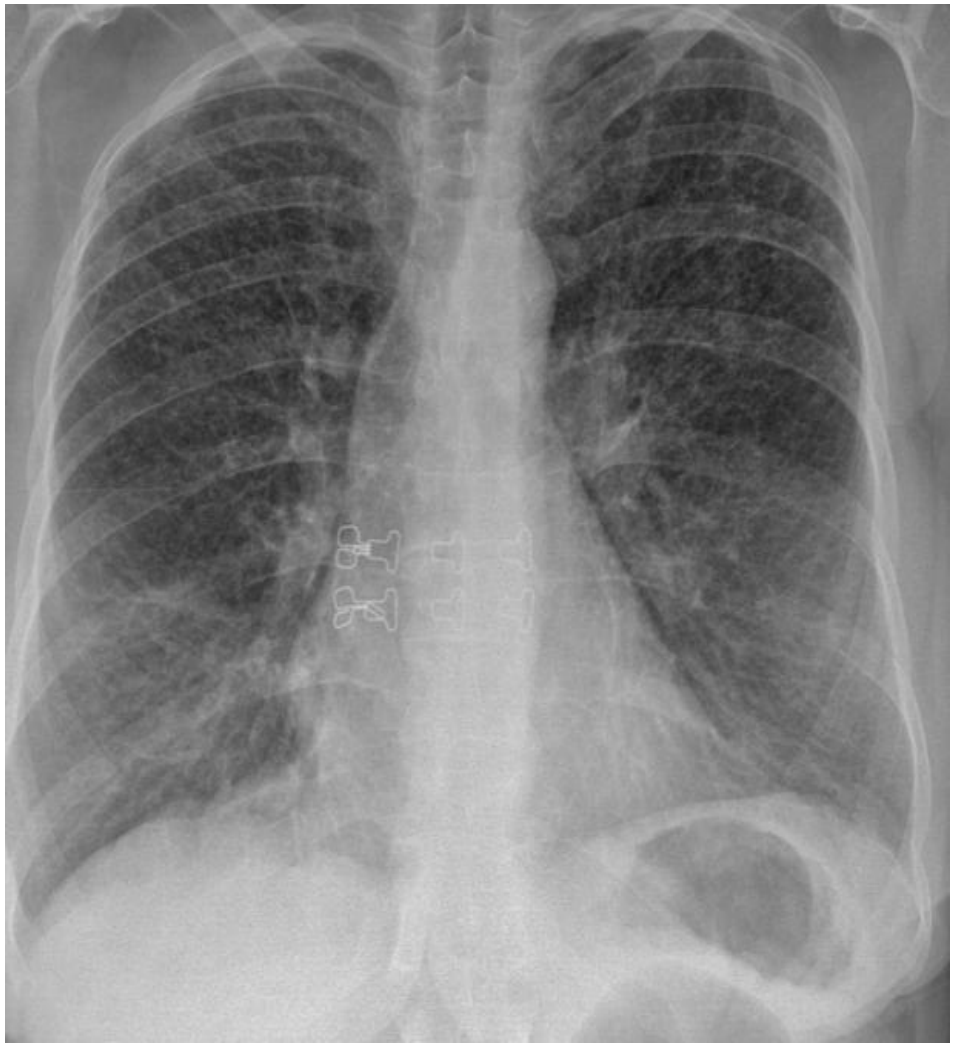
Sigarayı bırakması istendi 3-6 aylık takip önerildi.

Olgu-4: PA akciğer grafileri

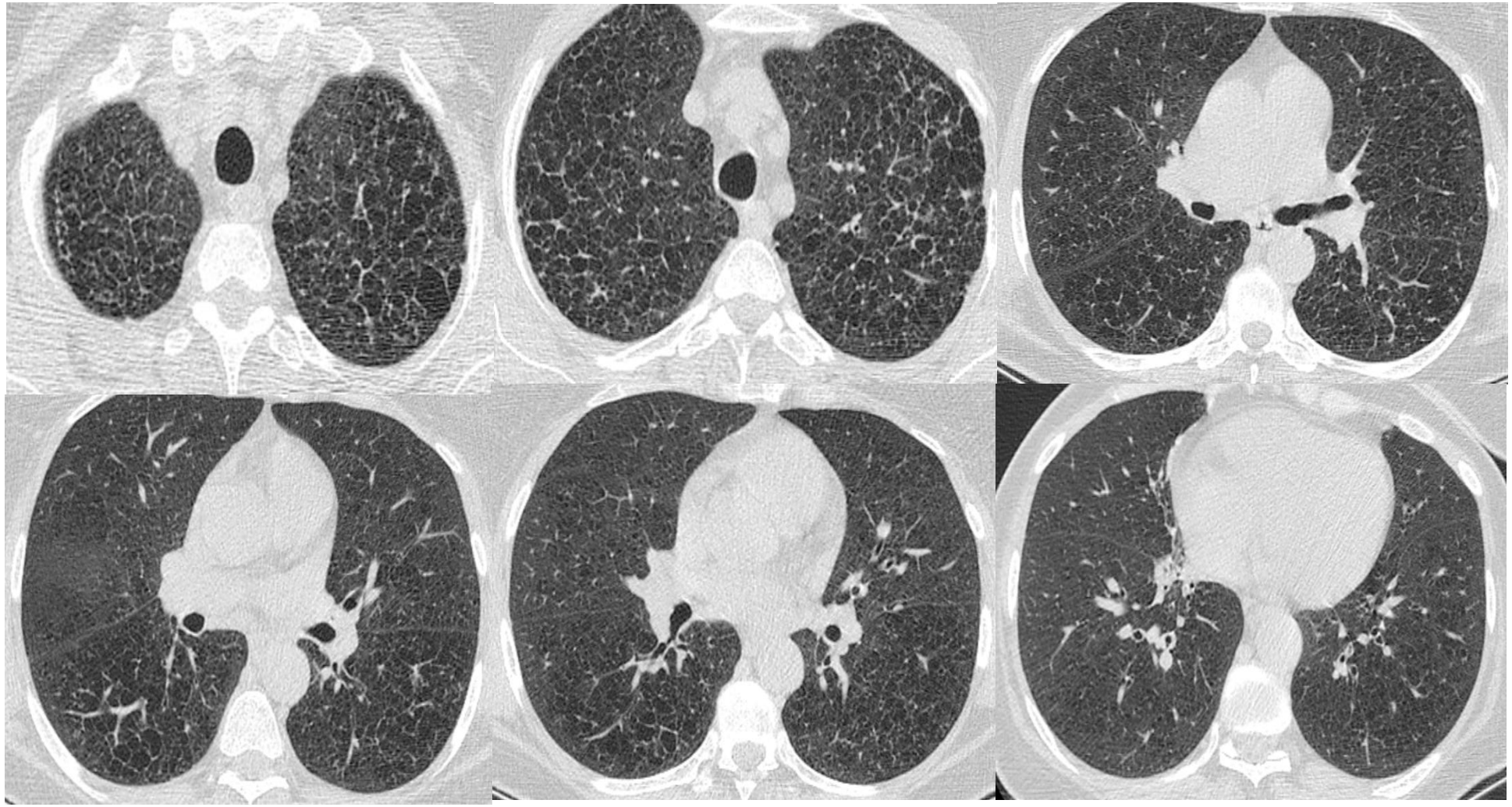
Başlangıçta



6 yıl sonra



Olgu-4: 6 yıl sonra



- Hasta takiplere gelmedi
- 6 yıl sonra dış merkeze başvurmuş. E-nabız bilgilerinden hastada solunum yetmezliği geliştiği öğrenildi.

Pulmoner langerhans hücreli histiositozis

*Difuz parankimal akciğer hastalıklarının %3-5'ini oluşturur

*%90'dan fazlası smoker veya ex-smoker

*20-40'lı yaşlarda sık

*İnflamasyon (alveoler makrofaj, eozinofil,nötrofil velenfosit içerir) bronşiyol duvarı ve akciğer parankiminde yıkıma neden olur

*Extrapulmoner tutulum %20

--Kemikte kistik lezyonlar

--Hipotalamus tutulumuna bağlı diyabetes insipidus

Pulmoner langerhans hücreli histiositoziste tedavi

- Sigaranın bıraktırılması
- Progresif ve semptomatik PLHH’de:

Kortikosteroidler (nodüler formda): 0.25-0.50 mg/kg/gün

Cladribin (kistik formda)

Cytarabine

Vinblastin (pediatrik LHH’de)

Characteristic features of pulmonary Langerhans cell histiocytosis

Clinical	▪ Typical age range 20 to 40 years ▪ Nearly all affected individuals are current or former cigarette smokers* ▪ May be asymptomatic, with abnormal chest radiograph, or may report dyspnea, constitutional symptoms ▪ History of pneumothorax ▪ Diabetes insipidus (<10%) ▪ Bone lesions (<15%)
Lung function tests	▪ Normal or reduced lung volumes and reduced diffusing capacity ▪ Airflow limitation and hyperinflation less common, typically in patients with more advanced, cystic disease
HRCT	▪ Mix of nodules (2 to 10 mm) and thick-walled cysts (early stages) or bizarrely shaped cysts varying in size and shape (advanced stages) ▪ Upper and mid-lung zone distribution with sparing of costophrenic angles ▪ Can show scattered cystic lesions without nodules ▪ Can show peribronchial nodules without cysts
Bronchoalveolar lavage	▪ BAL with ≥5% CD1a-positive cells is considered diagnostic, but frequently a lower percentage of CD1a cells is noted
Histopathology	▪ Peribronchial inflammatory lesions containing an admixture of Langerhans cells, eosinophils, lymphocytes, and neutrophils ▪ Langerhans-like cells express CD1a, langerin (CD207), and S100
Diagnosis based on clinical features and HRCT	▪ Classic HRCT features ▪ BAL with <5% CD1a-positive cells, but without lymphocytosis ▪ Improvement with smoking cessation
Diagnosis based on BAL or biopsy	▪ Compatible clinical features and one of the following: • BAL with ≥5% CD1a-positive cells • Transbronchial or surgical lung biopsy demonstrating diagnostic histological features • Compatible HRCT and extrapulmonary Langerhans cell histiocytosis confirmed by bone or skin biopsy

HRCT: high resolution computed tomography; BAL: bronchoalveolar lavage.

* Other cystic lung diseases can occur in smokers, but pulmonary Langerhans cell histiocytosis is almost unheard of in those without cigarette smoke exposure.

Reference: Meyer KC, Raghu G, Baughman RP, et al. An official American Thoracic Society clinical practice guideline: the clinical utility of bronchoalveolar lavage cellular analysis in interstitial lung disease. Am J Respir Crit Care Med 2012; 185:1004.

Olgu-5

32 yaş , kadın hasta,

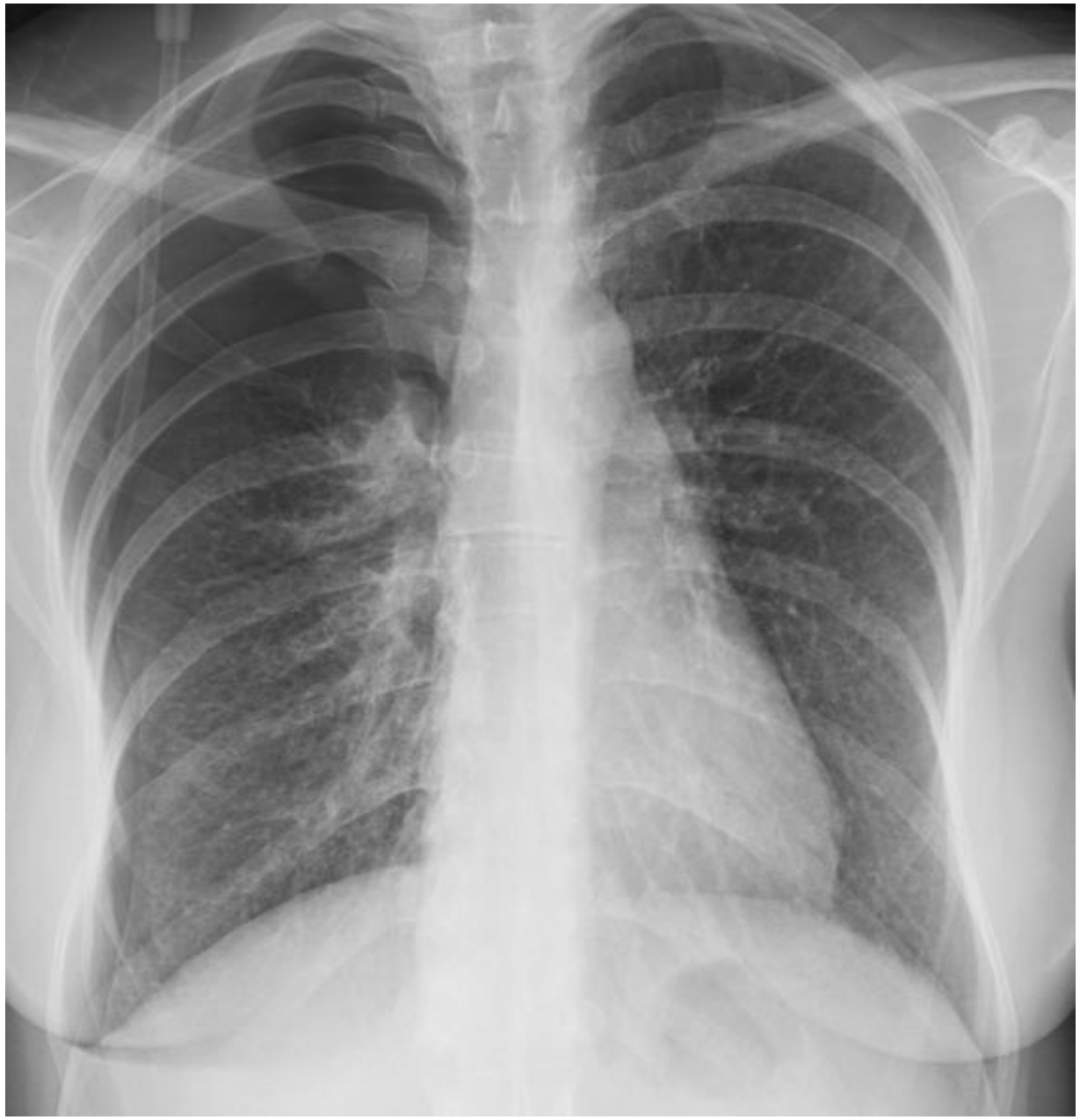
3 gündür göğüs ağrısı, ani başlayan nefes darlığı yakınmaları ile acil servise başvurmuş. Pnömotoraks saptanan hastaya KSAD uygulanmış.

Non-smoker, çevresel maruziyeti yok

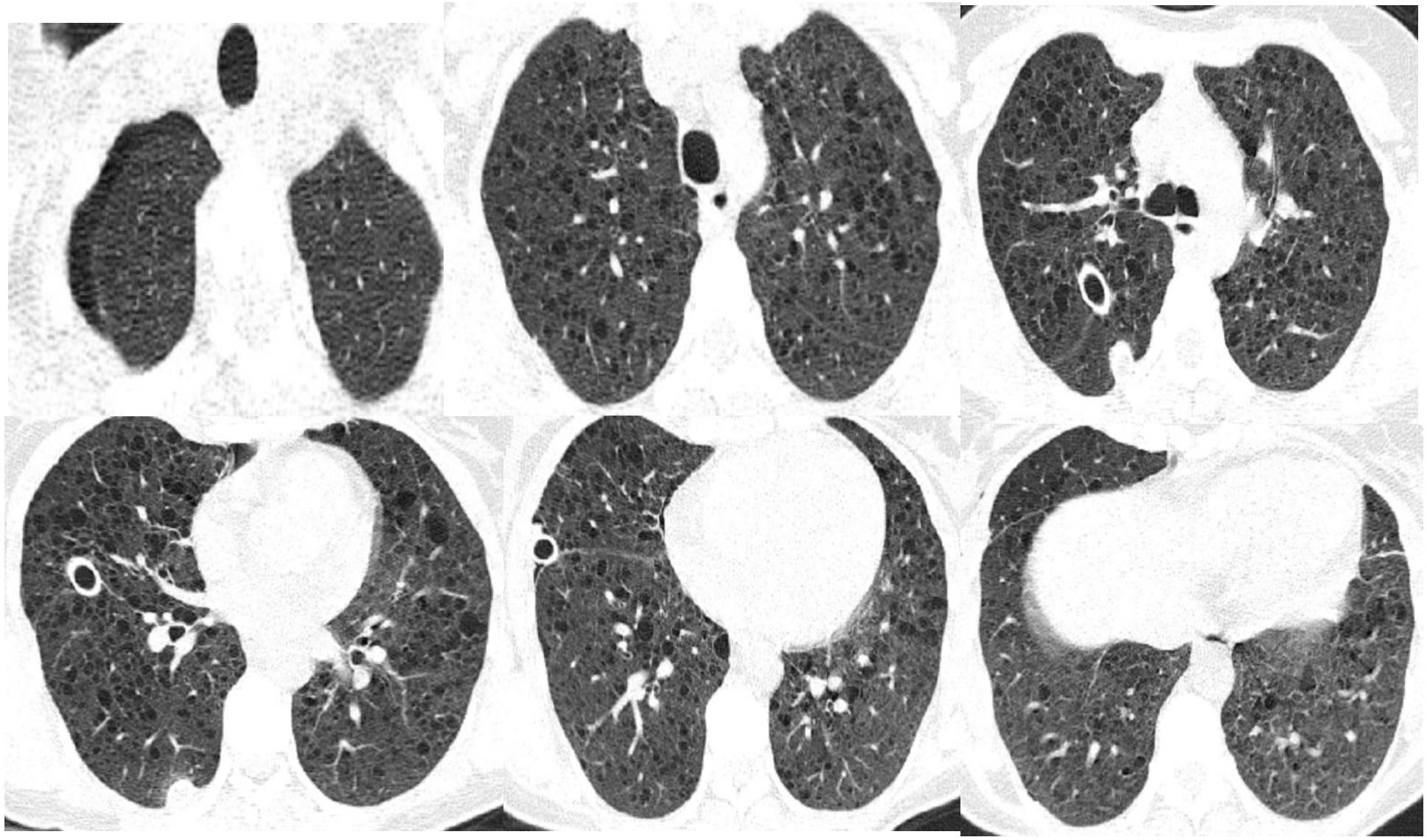
Soygeçmişinde özellik yok.

Solunum sesleri sağda alınmıyor.

Rutin hemogram ve biyokimya olağan, romatolojik biyobelirteçler ve HIV negatif.



Olgu-5: Toraks BT



Olgu-5:

Konsey kararı ile hastaya VATS ile cerrahi biyopsi yapıldı

- **LENFANGİOLEİOMYOMATOZİS**; SAĞ ÜST ve ORTA LOB WEDGE REZEKSİYON MATERYALLERİ
- NONSPESİFİK PLÖRİT; PARİETAL PLEVRA, BİYOPSİ MATERYALİ
- EPİKRİZ:
- Üst ve orta lobdan alınan örneklerde; alveoler alanda kistik dilatasyon oluşturan yapılar ve kistik yapıların duvarında iğsi nukleuslu hücre popülasyonu izlenmektedir. Bu hücrelerde İHK ile SMA, ER, PR ve HMB45 pozitif, EMA negatif saptanmıştır.

Lenfanjiyoleyomyomatozis

- Çoğunlukla genç kadınları etkileyen nadir bir multisistem hastalık
- Düşük grade'li, metastaz yapan bir neoplazm
- Akciğer parankiminin anormal düz kas benzeri hücrelerle infiltrasyonuyla karakterizedir
- tüberoz skleroz kompleksi (TSC) genlerinde mutasyonlar
- Kistler uniform
- %50 hastada anjiyomyolipomabirlikteliği

-İzole LAM

-Tuberoskleros ile birlikte (TSC-LAM)

1/3 olguda pnömotoraks

¼ olguda plörezi

%10-30 olguda şilöz plevral sıvı veya assit

Tedavide:

-Semptomatik

-rapamisinin mekanistik target hedefi (mTOR) inhibitörleri=

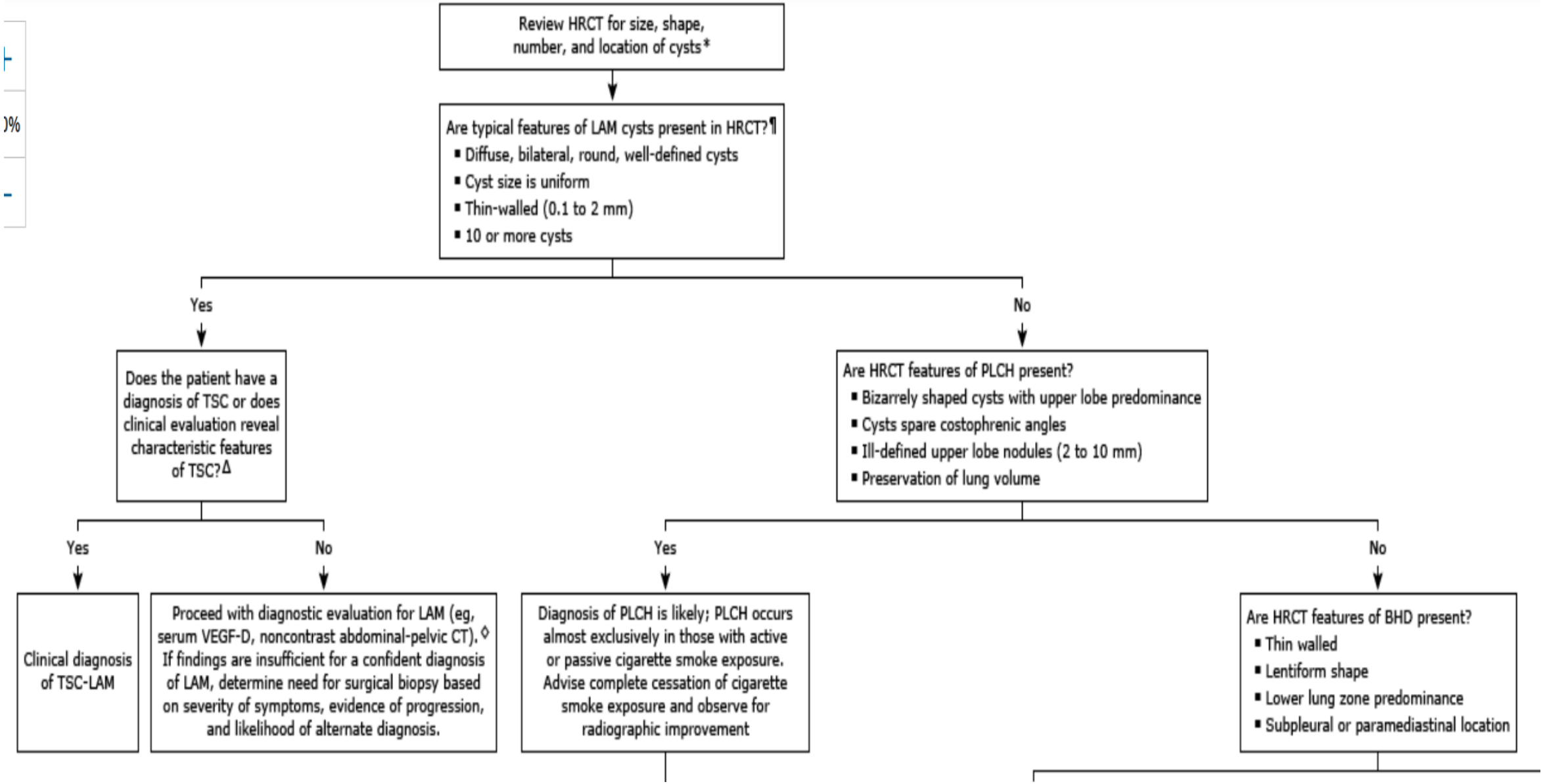
Sirolimus

everolimus

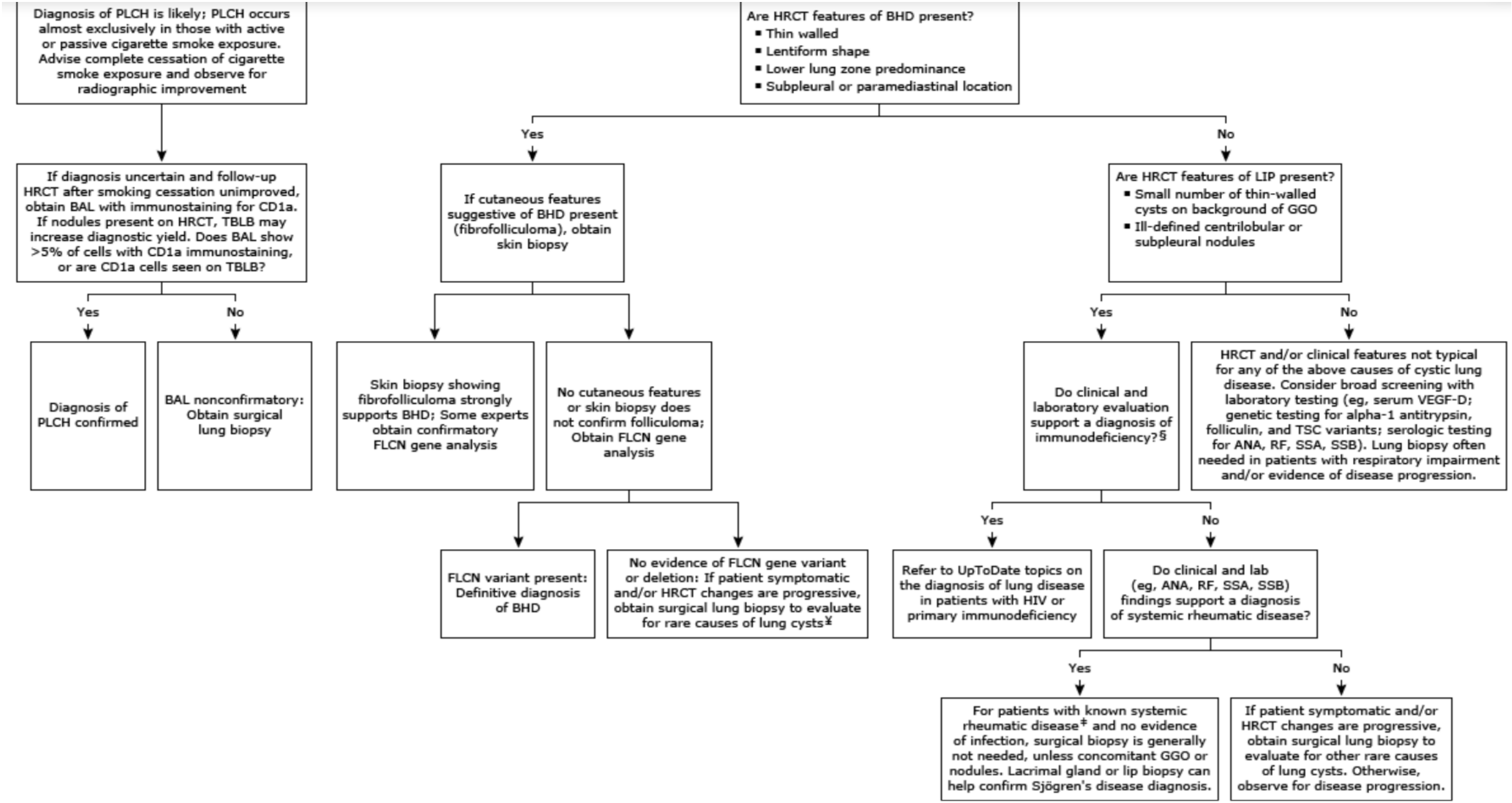
Cystic lung disease: Clinical, laboratory, and radiographic features

Cystic lung disease	Distinctive clinical features	Typical CT features*	Confirmation of diagnosis (in association with compatible chest CT)
LAM	▪ Occurs almost exclusively in females (occasionally in males with TSC) ▪ Renal angiomyolipomas in up to 50% of cases ▪ Retroperitoneal or pelvic lymphangioleiomyomas ▪ Chylous effusion, chylous ascites ▪ May be accompanied by clinical features of TSC	▪ Small, thin-walled, round cysts varying little in size or shape, uniformly distributed throughout the lungs	One of the following: ▪ Noncontrast abdomen/pelvis CT or MRI demonstrating renal angiomyolipoma or cystic lymphangioleiomyoma ▪ Presence of chylous pleural effusion or ascites¶ ▪ Serum VEGF-D >800 pg/mL ▪ Presence of TSC ▪ Transbronchial or surgical lung biopsy demonstrating diagnostic histological features△ (if all of the above are absent)
PLCH	▪ Nearly all affected individuals are current or former cigarette smokers◇ ▪ Diabetes insipidus ▪ Bone lesions	▪ Mix of nodules and thick-walled cysts (early stages) or bizarrely shaped cysts varying in size and shape (advanced stages) ▪ Upper and mid-lung zone distribution with sparing of costophrenic angles	One of the following: ▪ BAL with ≥5% CD1a-positive cells ▪ Transbronchial or surgical lung biopsy demonstrating diagnostic histological features
BHD	▪ Fibrofolliculomas ▪ Renal neoplasms, typically multifocal hybrid oncocytic tumors or chromophobe renal cell carcinomas ▪ Family history of pneumothorax or renal neoplasms	▪ Bilateral thin-walled round or lentiform cysts of variable sizes, often subpleural and/or abutting the mediastinum, with a lower lung zone predominance	One of the following: ▪ Biopsy-proven skin fibrofolliculoma ▪ Bilateral, multifocal hybrid oncocytic renal tumors or chromophobe renal cell carcinomas ▪ Genetic testing positive for *FLCN* mutation **NOTE:** Lung biopsy NOT helpful
LIP	▪ Underlying autoimmune or immunodeficiency state in 80% of cases ▪ Sjögren syndrome is most common underlying disorder	▪ Thin-walled cysts, typically few in number, associated with ground glass opacities and centrilobular nodules	▪ Serologic testing for HIV and systemic rheumatic disease (eg, antinuclear antibody, anti-Ro/SSA, anti-La/SSB, rheumatoid factor) adds support to clinical diagnosis ▪ Lacrimal gland or lip biopsy ▪ Surgical lung biopsy, when a definitive diagnosis is needed to guide therapy **NOTE:** Transbronchial biopsy size too small to be definitive for LIP, so TBLB only helpful if an alternate diagnosis suspected (eg, LAM)

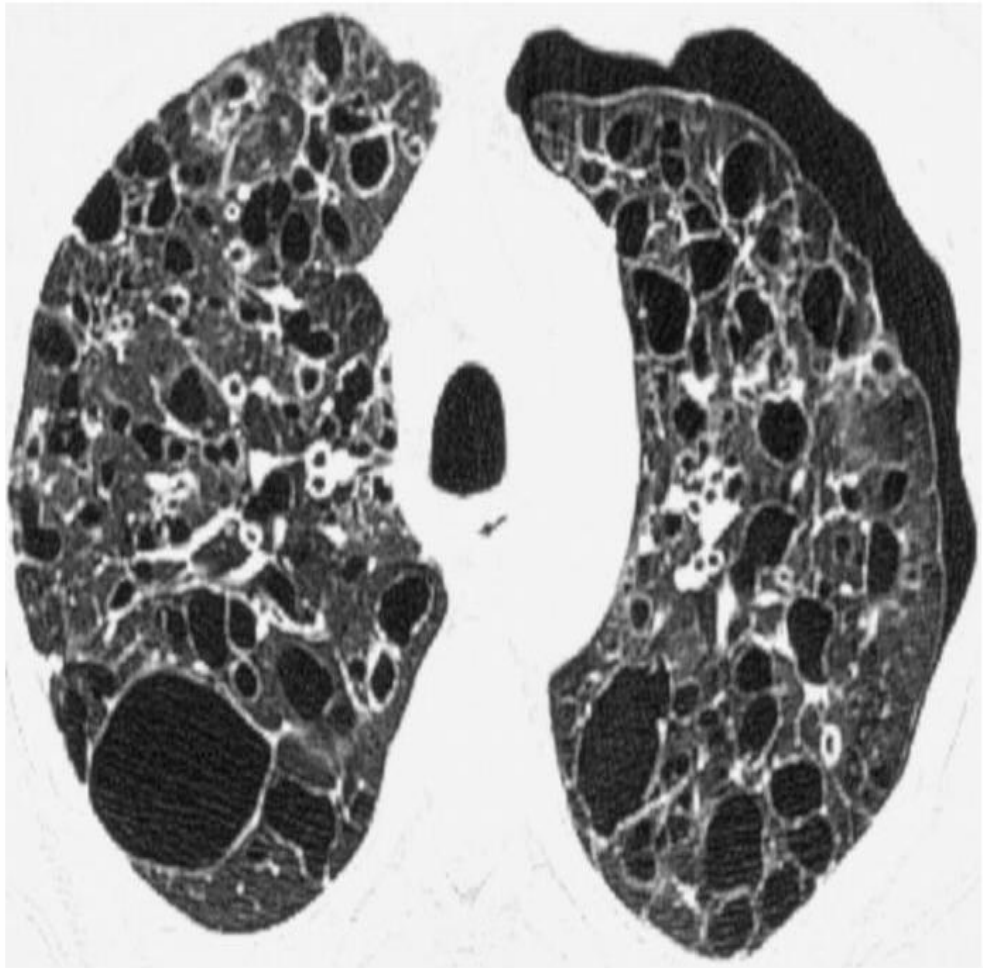
CT: computed tomography; LAM: lymphangioleiomyomatosis; TSC: tuberous sclerosis complex; MRI: magnetic resonance imaging; VEGF-D: vascular endothelial growth factor-D; PLCH: pulmonary Langerhans cell histiocytosis; BAL: bronchoalveolar


Review HRCT for size, shape, number, and location of cysts*
Are typical features of LAM cysts present in HRCT?¶
Diffuse, bilateral, round, well-defined cysts
Cyst size is uniform
Thin-walled (0.1 to 2 mm)
10 or more cysts
Yes
No
Does the patient have a diagnosis of TSC or does clinical evaluation reveal characteristic features of TSC?Δ
Are HRCT features of PLCH present?
Bizarrely shaped cysts with upper lobe predominance
Cysts spare costophrenic angles
Ill-defined upper lobe nodules (2 to 10 mm)
Preservation of lung volume
Yes
No
Yes
No
Clinical diagnosis of TSC-LAM
Proceed with diagnostic evaluation for LAM (eg, serum VEGF-D, noncontrast abdominal-pelvic CT).◊ If findings are insufficient for a confident diagnosis of LAM, determine need for surgical biopsy based on severity of symptoms, evidence of progression, and likelihood of alternate diagnosis.
Diagnosis of PLCH is likely; PLCH occurs almost exclusively in those with active or passive cigarette smoke exposure. Advise complete cessation of cigarette smoke exposure and observe for radiographic improvement
Are HRCT features of BHD present?
Thin walled
Lentiform shape
Lower lung zone predominance
Subpleural or paramediastinal location

Approach to the diagnosis of cystic lung disease

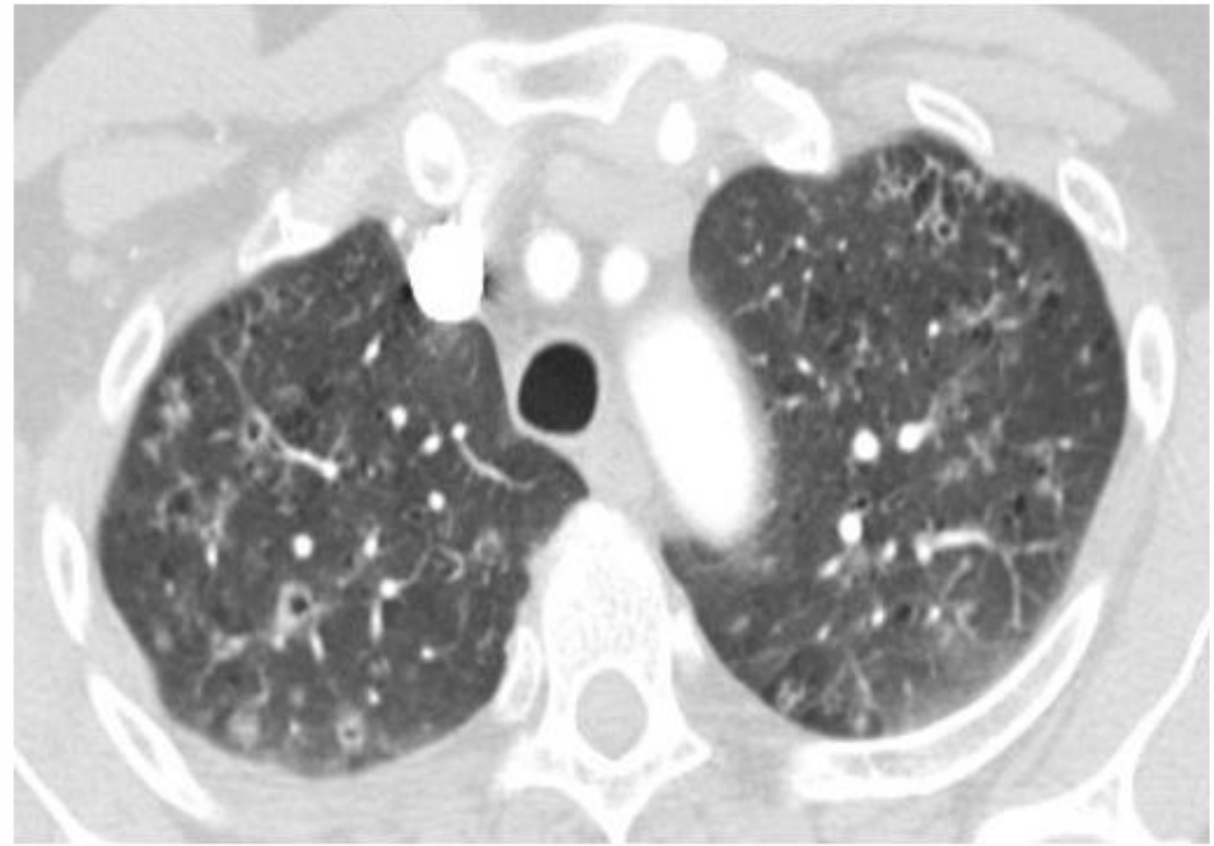


Computed tomography of pulmonary Langerhans cell histiocytosis



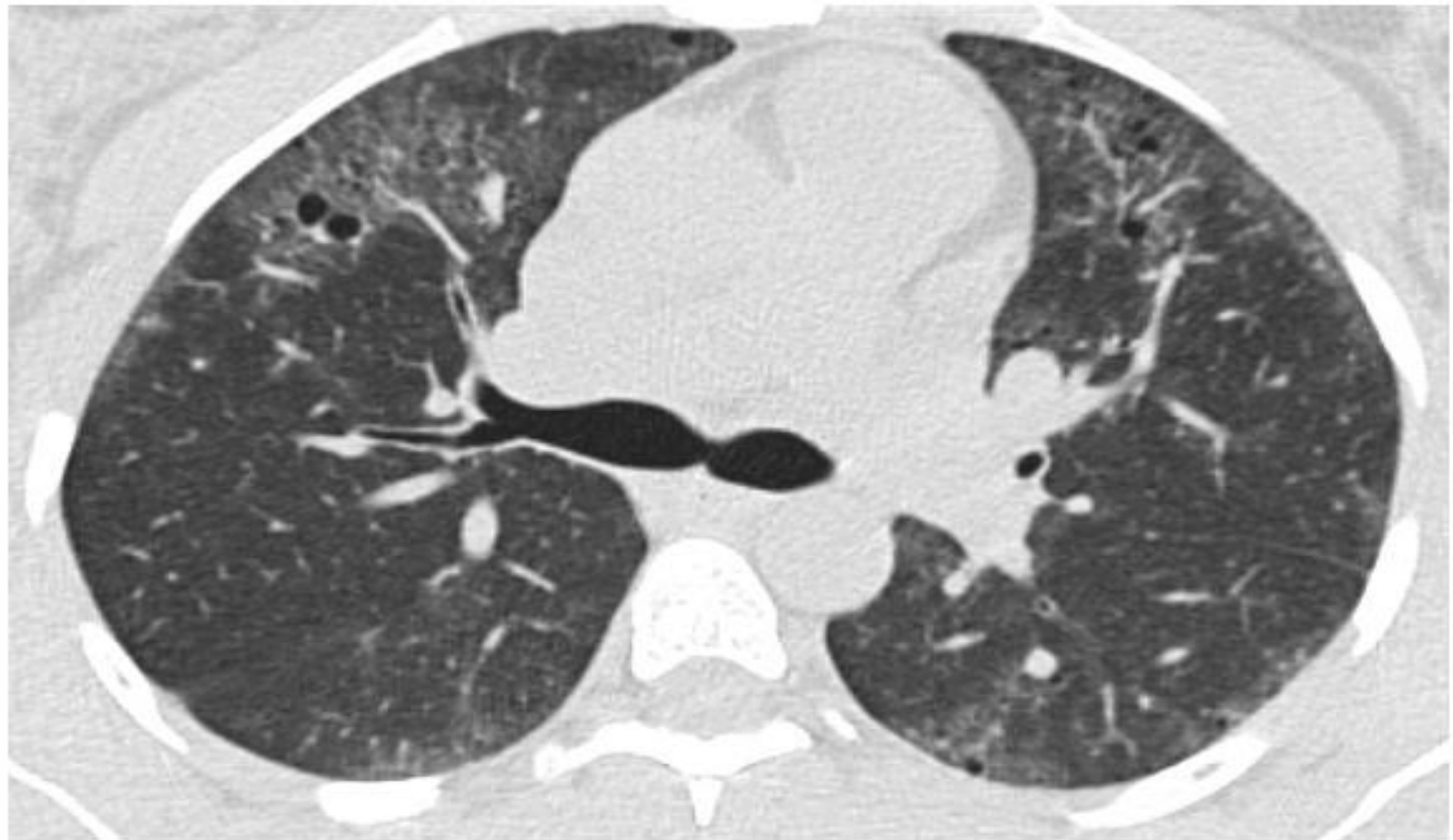
Axial computed tomography image of pulmonary Langerhans cell histiocytosis (PLCH) showing multiple irregular and bizarrely shaped cysts with prominent walls, characteristic of PLCH. Also present is a left pneumothorax.

Computed tomography of pulmonary Langerhans cell histiocytosis



Axial computed tomography image of upper lung zones in a patient with pulmonary Langerhans cell histiocytosis. Solid nodules, cavitary nodules, and thick-walled cysts are present in the apices of both lungs.

Computed tomography of lung cysts in lymphoid interstitial pneumonia



Axial computed tomography image from a patient with lymphoid interstitial pneumonia showing thin-walled lung cysts in areas of ground glass opacities.

Sabrınız için teşekkürler