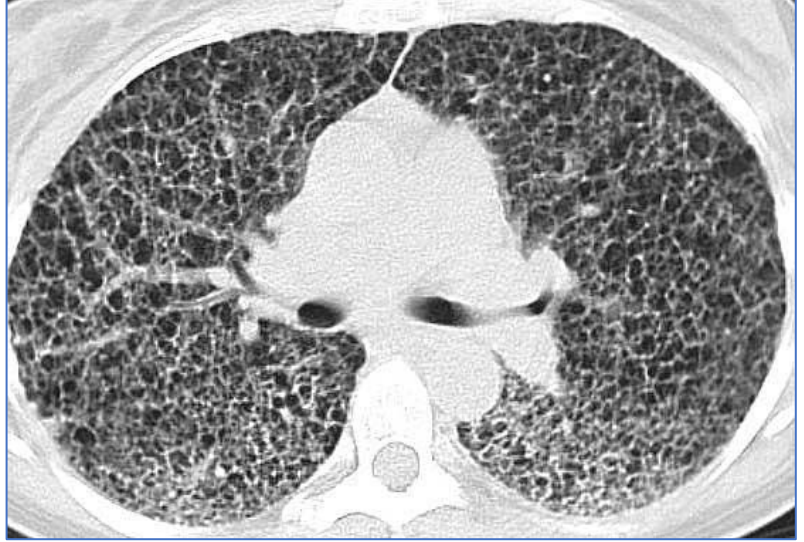


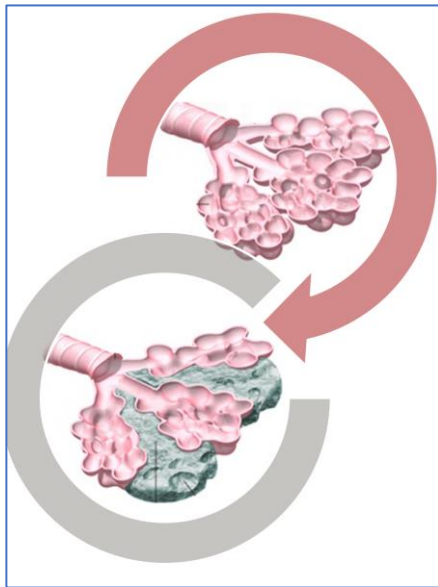
Prof. Dr. Tefvik ÖZLÜ



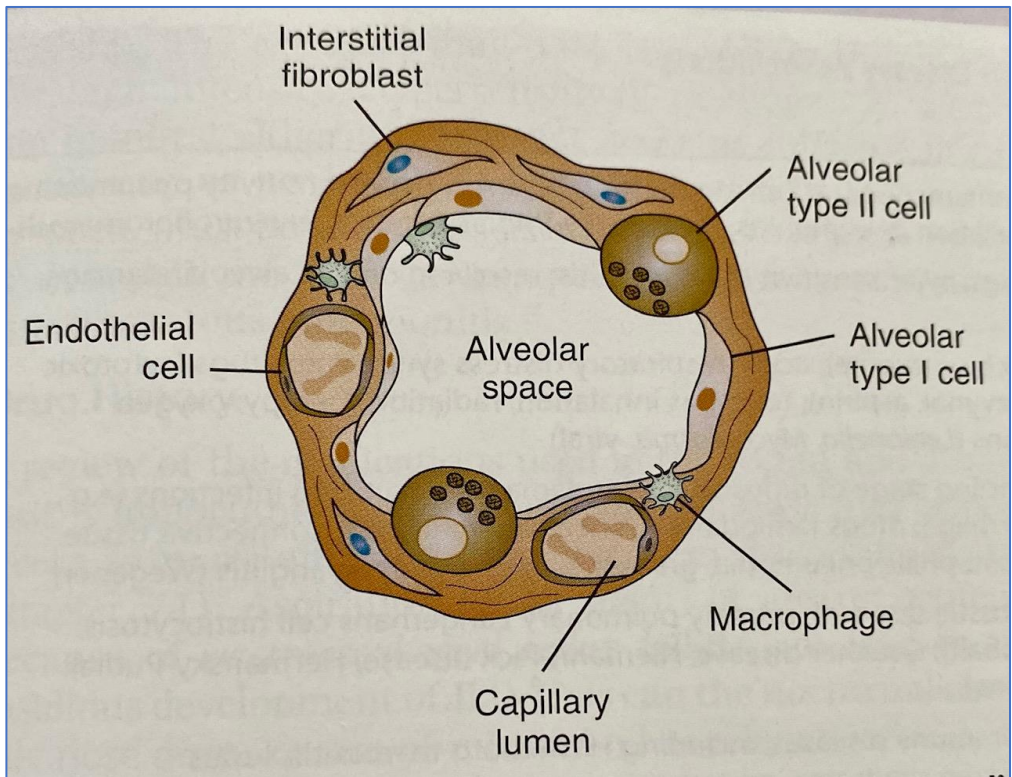
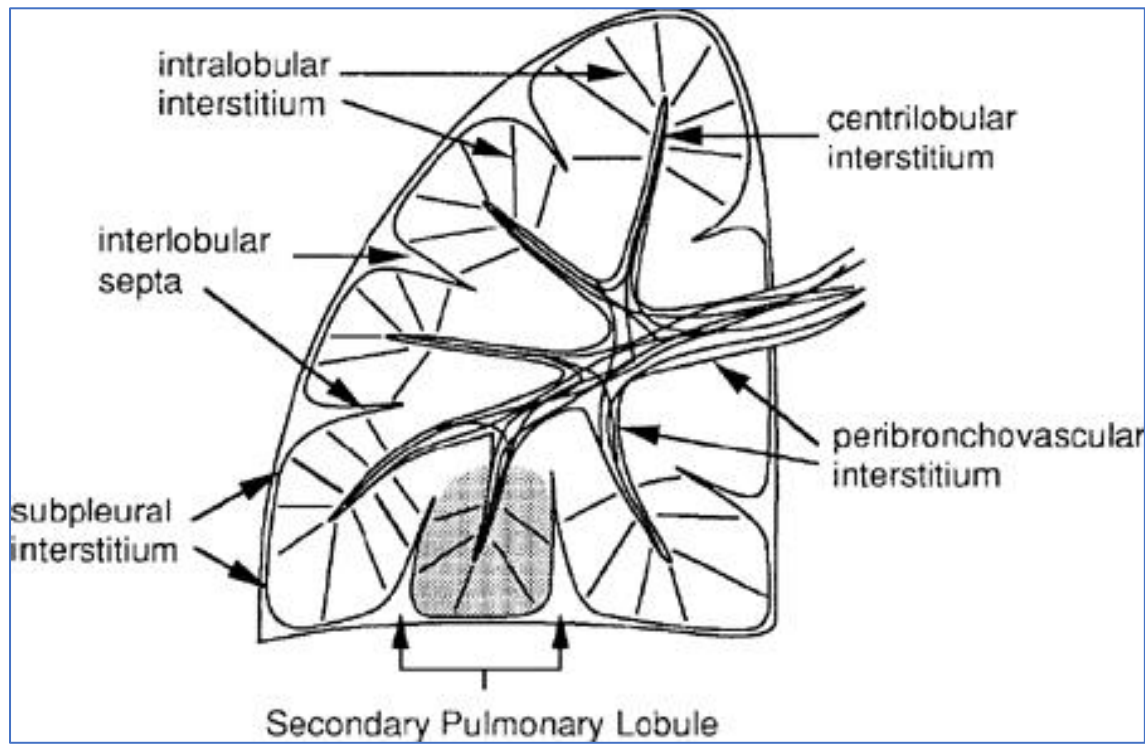
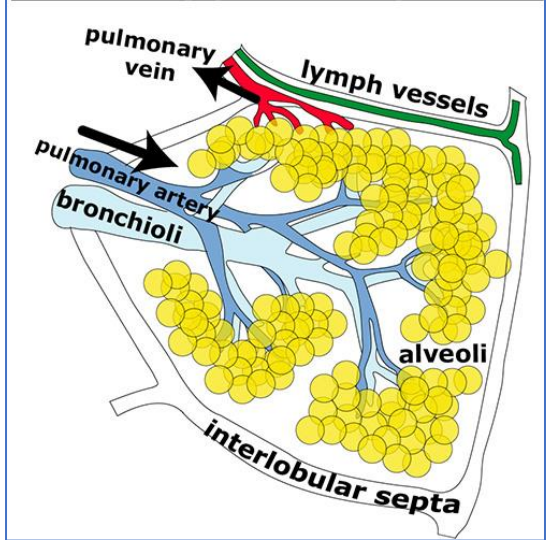
Birinci basamaktan özelleşmiş merkezlere
interstisyel akciğer hastaların yönetimi

Tanım

- İnterstisyel akciğer hastalığı
 - Diffüz parankimal akciğer hastalığı
 - Diffüz interstisyel akciğer hastalığı
 - İnterstisyel pnömoni/pnömonitis
- 
- Bilinen veya bilinmeyen çok sayıda faktöre bağlı olarak gelişen akciğer parankiminde enflamasyon ve/veya fibrozisle seyreden **200 kadar** hastalık
- **Benzer klinik, radyolojik, fizyolojik** ve **patolojik** görünümleri olan heterojen bir grup hastalık
 - Asıl olarak olay **intestisyumda** yerleşik olmasına karşın; **alveoller, havayolları, damarlar** ve **plevra** da kısmen tutulabilir



secondary pulmonary lobule



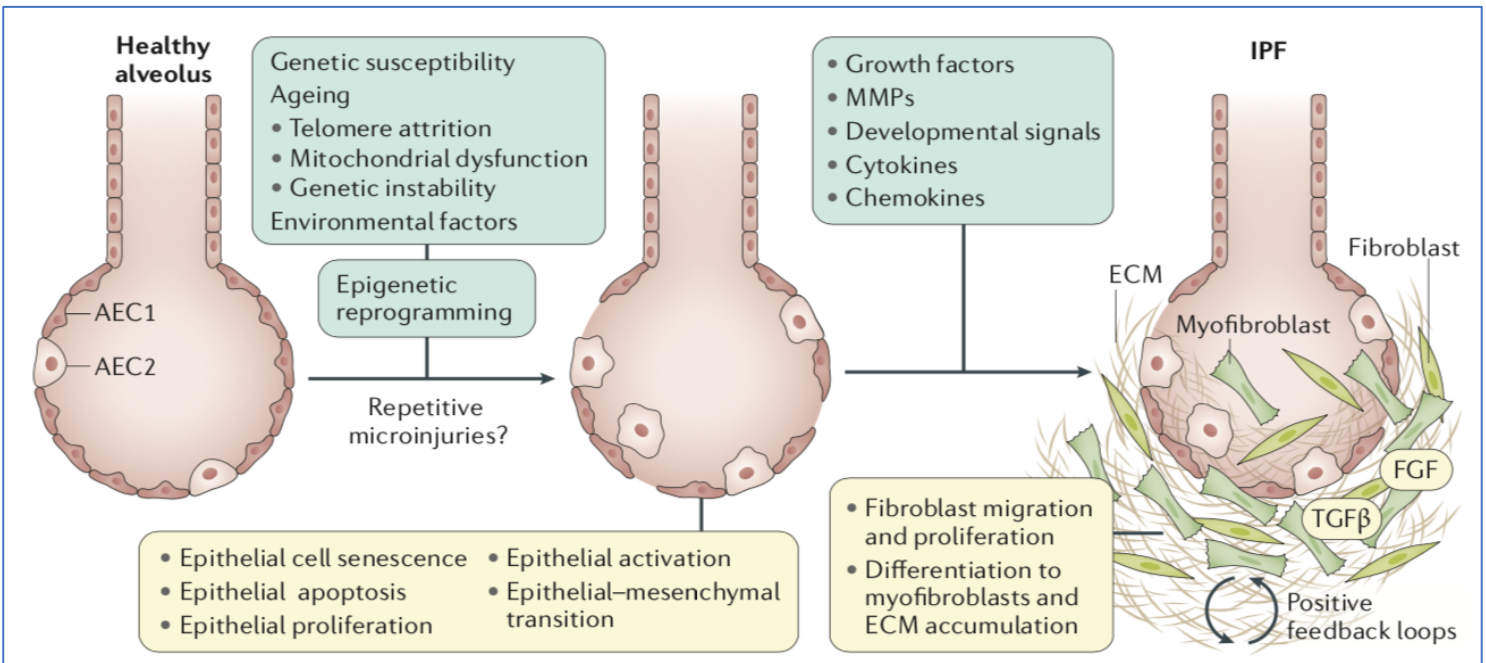
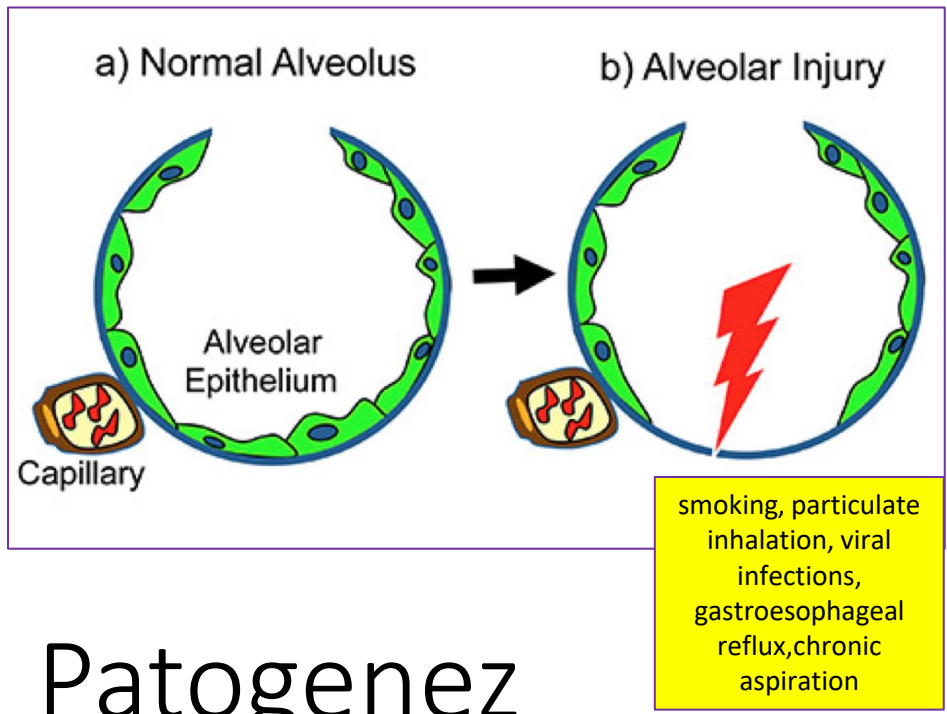
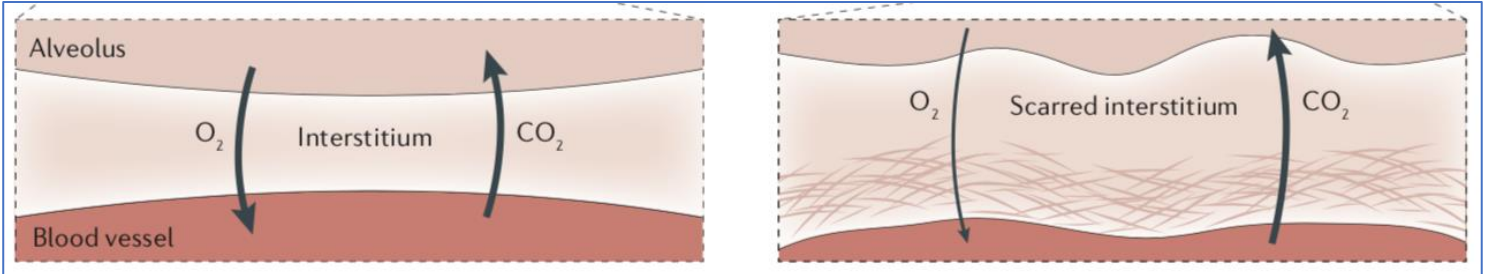
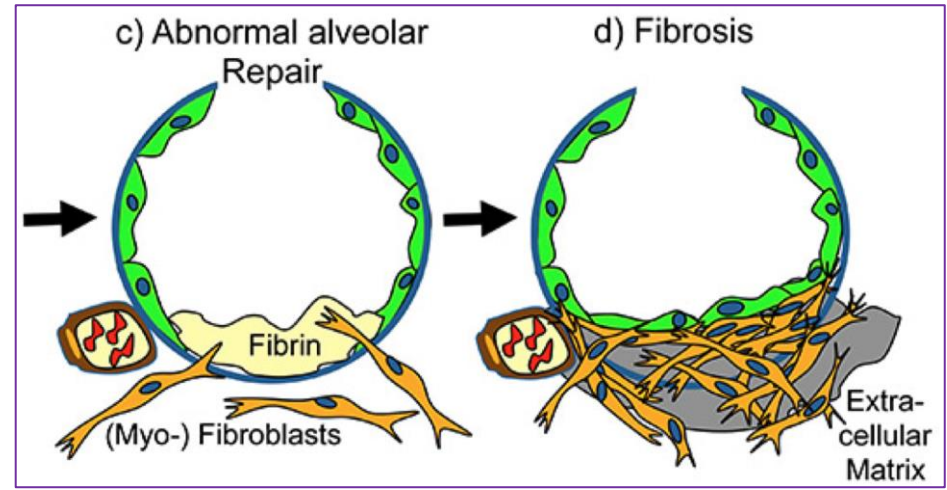


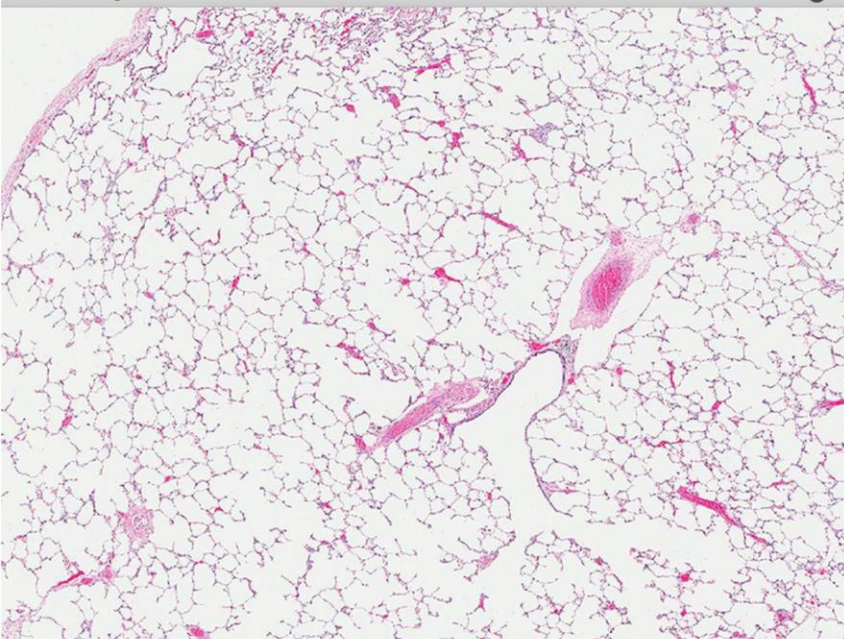
Figure 2 | **A proposed pathogenetic model of idiopathic pulmonary fibrosis.** The convergence of three elements — a genetic architecture affecting epithelial cell integrity, environmental factors and accelerated ageing-associated changes — results in a complex epigenetic reprogramming that promotes aberrant epithelial cell activation in idiopathic pulmonary fibrosis (IPF), which might or might not be exacerbated by injury. The activated epithelium secretes a plethora of mediators that induce migration, proliferation and activation of fibroblasts and myofibroblasts, which are resistant to apoptosis and persistently secrete extracellular matrix (ECM) components. The ECM is also a reservoir of growth factors that can be released as soluble ligands upon degradation. Rather than being linear, the sequence is characterized by a number of positive feedback loops, such as between matrix stiffness and fibroblast activation. Additionally, signalling crosstalk (for example, fibroblast growth factor (FGF) and transforming growth factor-β (TGFβ) signalling) adds to the complexity and likely leads to the inexorable progression of the disease. AEC1, alveolar epithelial type 1 cell; MMP, matrix metalloproteinase.



Patogeneez

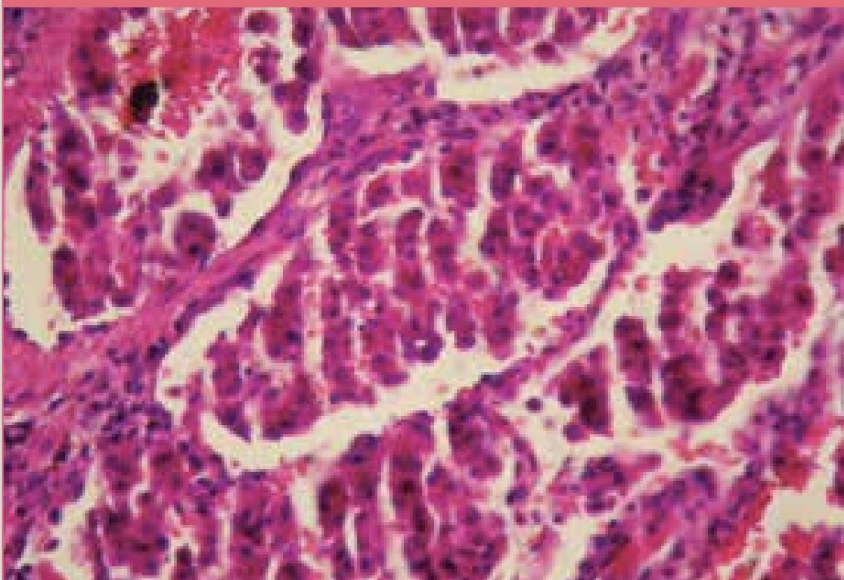
Martinez FJ, et al. NATURE REVIEWS, 2017.



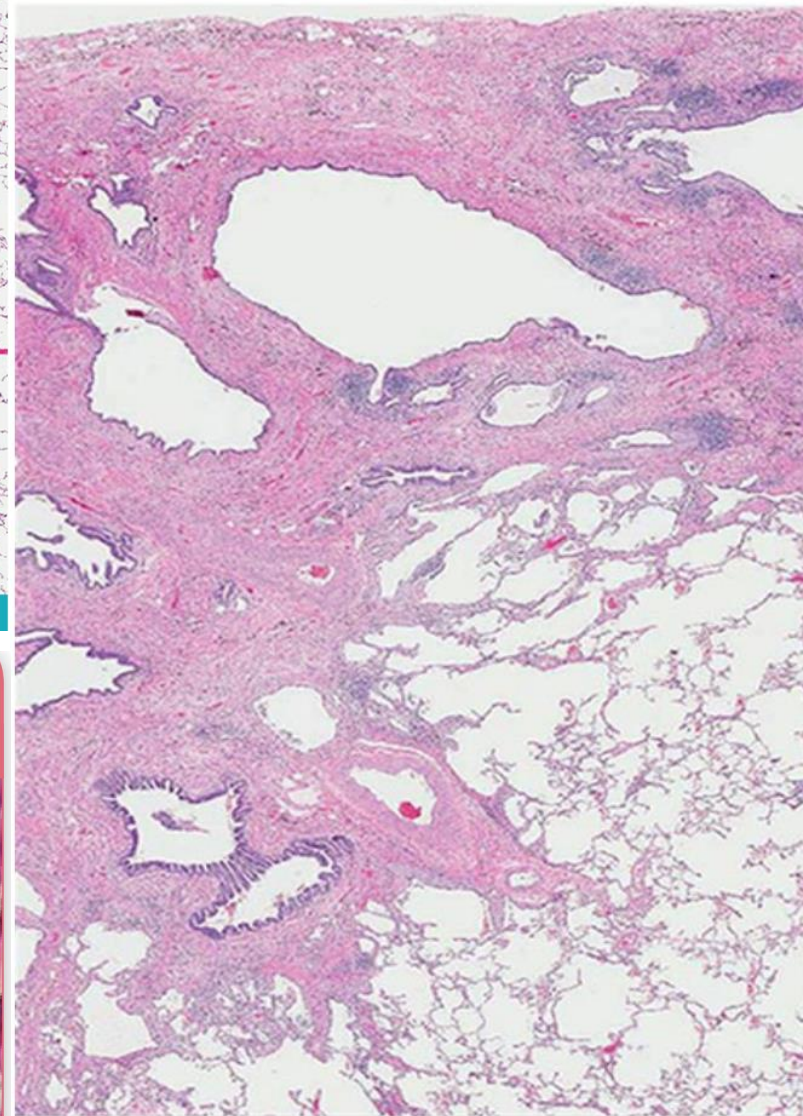


Low magnification photomicrograph showing the fine alveolar structure of normal lung tissue.

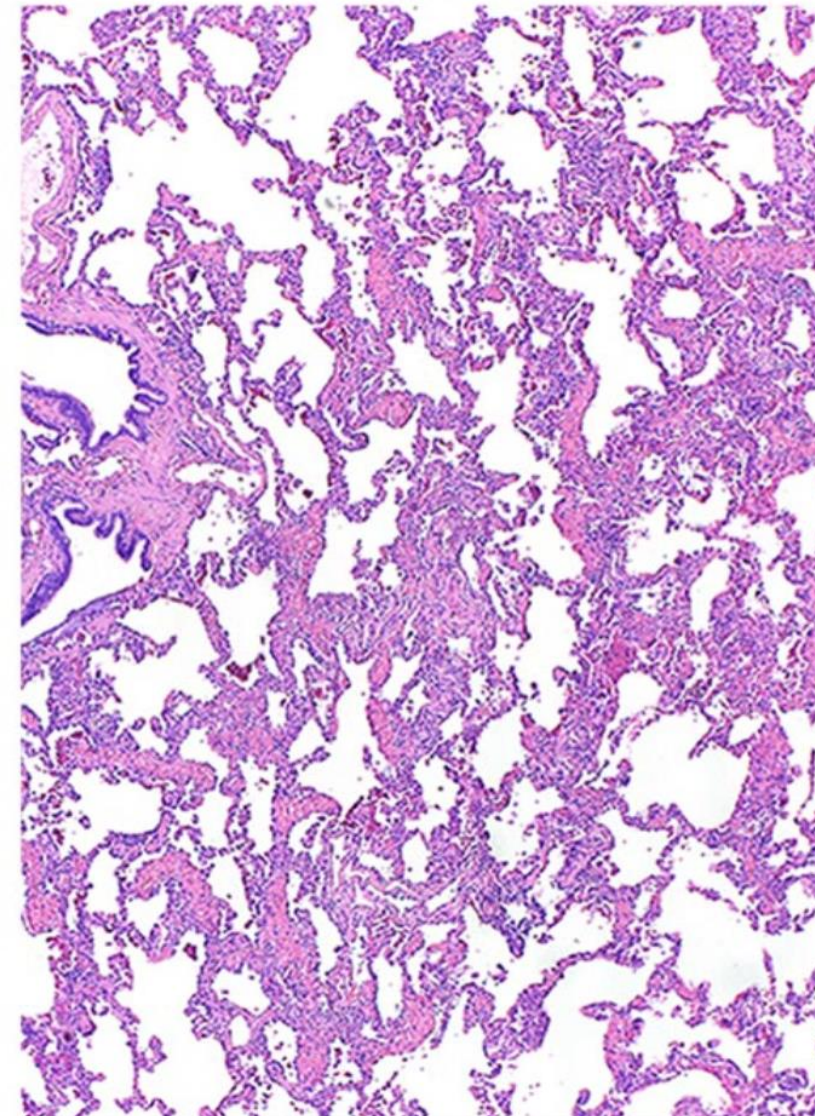
Resim 10. Deskumatif interstisyel pnömonide intraalveoler makrofaj depolanması.



UIP



NSIP



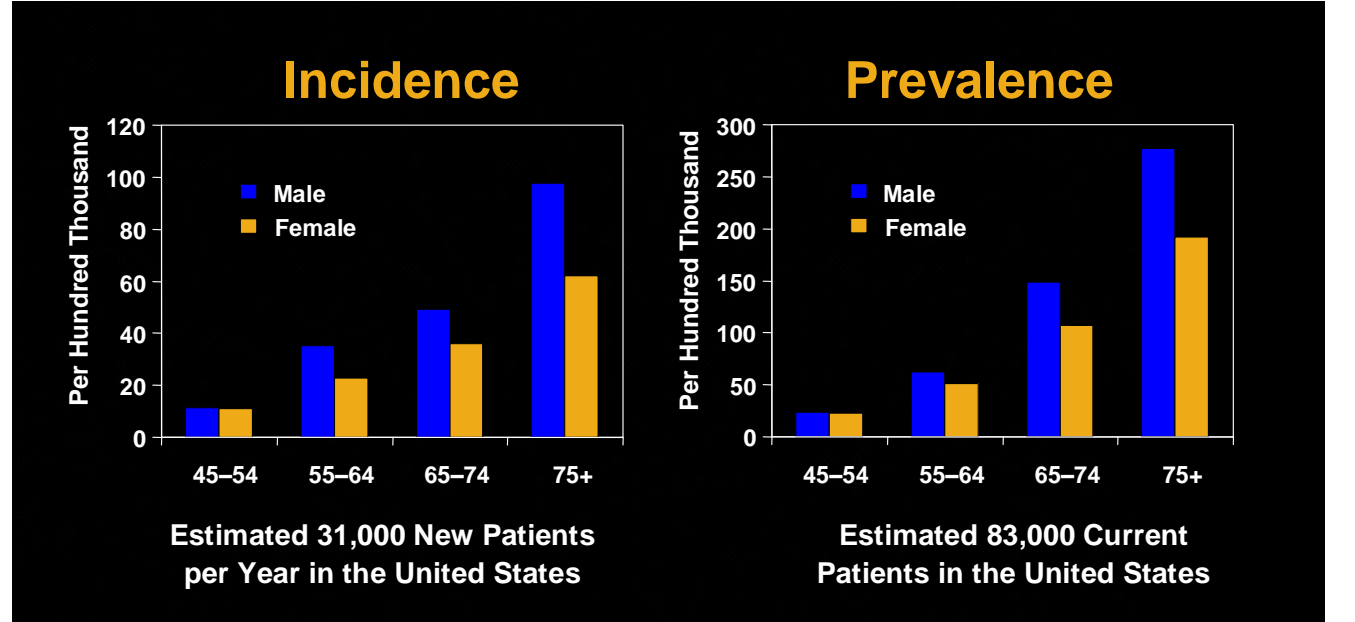
Side-by-side comparison of photomicrographs from patients with usual interstitial pneumonia (UIP, left) and nonspecific interstitial pneumonia (NSIP, right)). Note the similarity in the abnormality but the difference in distribution. UIP is a heterogeneous process, interspersed with normal lung, whereas NSIP is more homogeneous, affecting the entire lung.

Sınıflama

- Bilinen nedenlerle ilişkili olan
 - İlaçlar, radyasyon, KDH, enflamatuvar, infeksiyöz, malign hastalıklar, mesleki ve çevresel faktörler
- İdiyopatik
 - MAJOR: IPF, NSIP, RBILD, DIP, COP, AIP
 - NADİR: LIP, PPFE
- Granülomatöz
 - Sarkoidoz
- Diğer
 - LAM, LHH, PAP, EP

Sıklık

- Prevalans
 - Erkeklerde: 81 / 100 000
 - Kadınlarda: 67 / 100 000
 - Yaşlılarda (>75 yaş): 160 / 100 000
- İnsidens
 - Erkeklerde : 32 / 100 000
 - Kadınlarda : 26 / 100 000
- Mortalite:
 - İngiltere'de 3000 / yıl (%50'si İPF)



Weycker D, et al. Paper presented at: CHEST 2002, November 2-7, 2002; San Diego, CA.

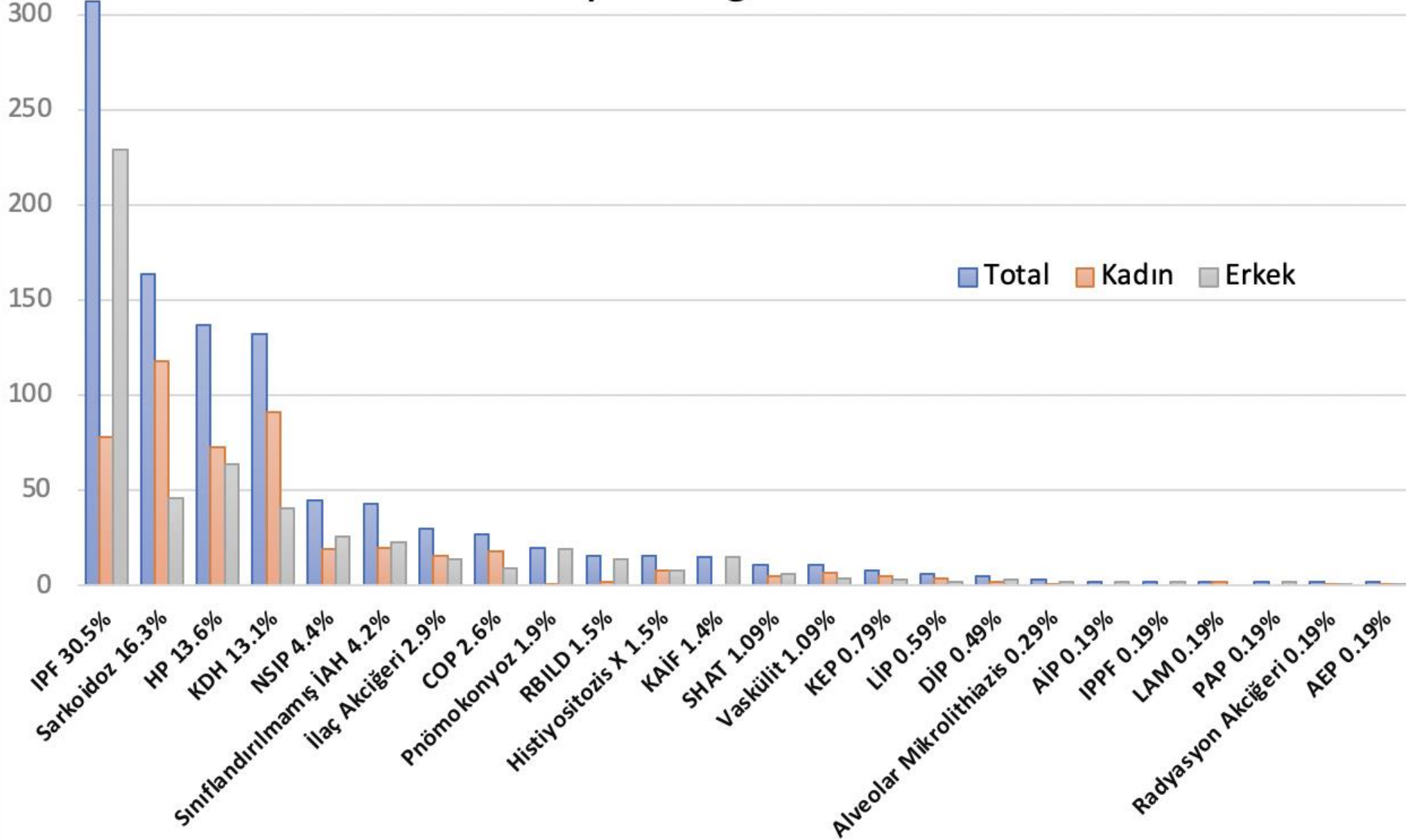
TABLE 1 | Incidence and prevalence of interstitial lung disease subtypes.

		Time Period	ILD (All Subtypes)	IPF	CTD	Sarcoid	HP	Drug	Occupational	Unclassifiable
North America										
New Mexico, USA	Incidence	1988–1990	Male 31.5 Female 26.1	Male 10.7 Female 7.4	Male 2.1 Female 3.0	Male 0.9 Female 3.6	–	Male 1.8 Female 1.1	Male 6.2 Female 0.8	–
New Mexico, USA	Prevalence	1988–1990	Male 80.9 Female 67.2	Male 20.2 Female 13.2	Male 7.1 Female 11.6	Male 8.3 Female 8.8	–	Male 1.2 Female 2.2	Male 20.8 Female 0.6	–
Europe										
Flanders (Belgium)	Incidence	1992–1996	1.0	0.22	0.07	0.26	0.12	0.05	0.07	0.10
Flanders (Belgium)	Prevalence	1992–1996	6.27	1.25	0.47	1.94	0.81	0.21	0.35	0.57
Greece	Incidence	2004	4.63	0.93	0.54	1.07	0.13	0.07	0.14	0.71
Greece	Prevalence	2004	17.3	3.38	2.14	5.89	0.45	0.30	0.36	1.46
Denmark	Incidence	2003–2009	4.1	1.3	–	–	–	–	–	–
Paris, France	Incidence	2012	18.3	2.8	3.3	4.9	0.9	1.2	0.8	1.8
Paris, France	Prevalence	2012	71.0	8.2	12.1	30.2	2.3	2.6	3.5	5.0
Turkey	Incidence	2007–2009	25.8	–	–	4.0	–	–	–	–

Incidence and prevalence defined as cases per 100,000.

ILD, interstitial lung disease; IPF, idiopathic pulmonary fibrosis; CTD, connective tissue disease; HP, hypersensitivity pneumonitis.

İnterstisyel Akciğer Hastalıkları



Sıklık

- Çok merkezli, prospektif, kesitsel, klinik gözlem çalışması
- Türkiye'nin 7 coğrafi bölge, 18 farklı il, 21 merkezden toplam 1070 hasta

İAH ne zaman akla gelmeli?

- Efor dispnesi, kronik öksürük
- Inspiratuvar raller
- Risk faktörlerinin varlığı
 - Meslek, çevresel maruziyet, ilaç kullanımı, KDH tanısı, aile öyküsü, sigara kullanımı, yaş
- Parankimal opasiteler
- Çomak parmak
- Hipoksemi, egzersiz desaturasyonu
- Spirometride restriksiyon

I. ve II. Basamak

- Aile hekimi
- Acil hekimi
- İşyeri hekimi

- Göğüs hastalıkları uzmanı
- Dahiliye uzmanı
- Kardiyoloji uzmanı

Özelleşmiş merkez

- Bilgili ve deneyimli

- Göğüs hastalıkları uzmanı
- Romatoloğ/immünoloğ
- Torasik radyoloğ
- Pulmoner patoloğ
- Uygun teknikle bilgisayarlı tomografi

- İnce kolimasyon, en kısa rotasyon zamanı ve en yüksek pitch, azaltılmış doz (1-3 mSv), hastaya uygun tüp potansiyeli ve tüp akımı (genellikle 120 kVp ve <240 mAs); lince hastalar için daha düşük tüp potansiyelleri (örneğin 100 kVp), supin pozisyon ve yeterli inspirasyon sonunda (volumetrik); supin pozisyon, ekspiryum sonunda (volumetrik veya sequential); prone, rekonstruksiyon

- Geniş laboratuvar menü

- Otoantikolar, vaskülit belirteçleri, genetik testler

Semptomlar

- **Asemptomatik**

- Sarkoidoz, erken dönem kronik İAH

- **Solunumsal**

- Efor dispnesi, kuru öksürük, siyanoz, hemoptizi, traksiyon bronşektazilerinde balgamlı öksürük, plöritik ağrı

- **Sistemik**

- Halsizlik, kırgınlık, yorgunluk, kilo kaybı, ateş, eklem ağrıları, güçsüzlük,

Cilt, eklem, böbrek, göz, kalp, GİS semptom ve bulguları

- Raynoud: SS, IPF
- Keratokonjonktivitis sicca: Sjogren
- Hepatosplenomegali: sarkoidoz, histiyositozis X, KDH, amiloidoz
- Diyabet insipidis: histiyositozis x, sarkoidoz
- Glomerülonefrit: KDH, Wegener, sarkoidoz, vaskülit
- Renal Kitle: LAM, tuberosklerozis
- Uveit: Sarkoidoz, Behçet
- Artrit: KDH, sarkoidoz, vaskülit
- Plevral hastalık: KDH, ilaçlar, asbestoz, tuberoskleroz, LAM ve histiyositozis X (pnömotoraks)
- Hipertansiyon: KDH, vaskülit
- Eritema nodosum: sarkoidoz, Behçet, KDH
- Heliotropik rash: dermatomyozit, polimyozit
- Makülopapüler rash: KDH, ilaçlar, amiloidoz
- Alopesi: KDH

Fizik muayene bulguları

- Velkro rall
 - IPF, fibrozis
- Squeaks (bronş gıcirtısı)
 - Bronşiolit, traksiyon bronşektazileri
- Çomak Parmak
 - IPF, DIP, asbestoz, RA
- Takipne, siyanoz, kor pulmonale
 - Terminal dönem IAH, kardiyak komplikasyon
- Ekstrapulmoner bulgular
 - Sistemik hastalık

Radyografi

- Normal PA grafi (% 10)
- Volüm azalması
 - Amfizem, Histiositozis-X, lenfanjiyoleiyomiyomatozis ve sarkoidoz, hariç
- Buzlu cam opasiteler
- Konsolidasyon
- Çizgisel, retiküler opasiteler
- Nodüller
- Plevral sıvı ve kalınlaşmalar
- Mediastinel LAP
- Kistik gölgeler, bal peteđi, kavite





Medscape www.medscape.com

Medscape www.medscape.com

Source: AIDS Read © 2002 Cllgott Publishing, Division of SCP Communications

Source: AIDS Read © 2002 Cllgott Publishing, Division of SCP Communications



Radyolojik paternler

KONSOLIDASYON

- Ödem
- Pnömoni
- Hemoraji
- Sarkoidoz
- Organize pnömoni
- Eozin. pnömoni
- PAP
- Bronkoalveoler ca
- Lenfoma

BUZLU CAM

- Ödem /ARDS
- Pnömoni/PCP, virus vb
- Hemoraji
- Hipersensit. pnöm
- Organize pnömoni
- Eozin. pnömoni
- PAP
- Bronkoalveoler ca
- Lenfoma
- NSİP
- RBİLD
- DİP

RETİKÜLER

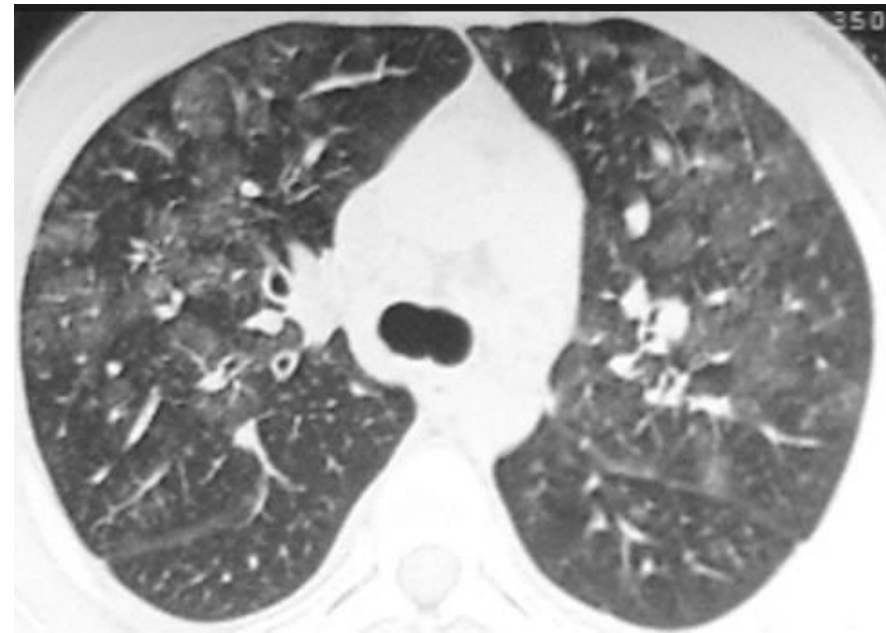
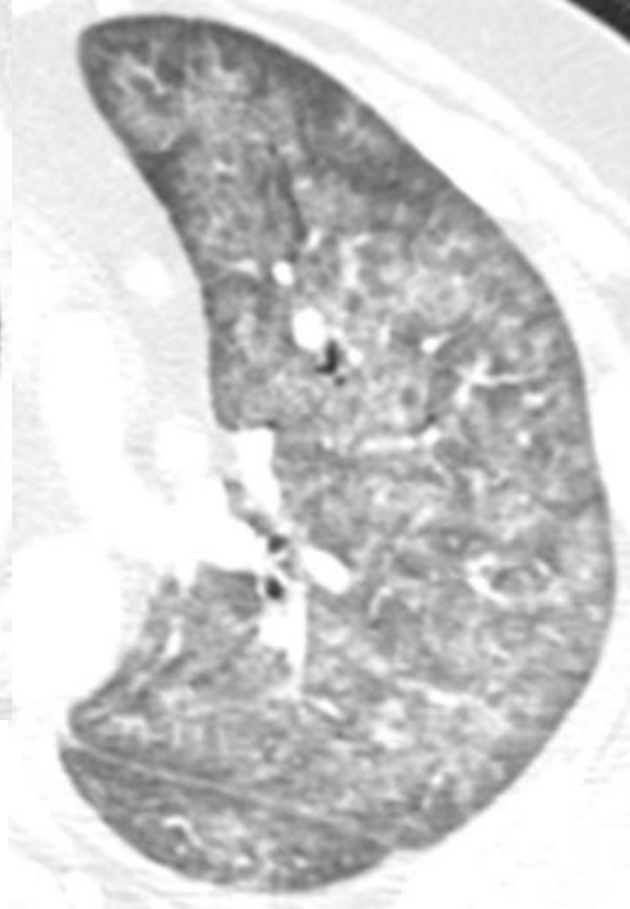
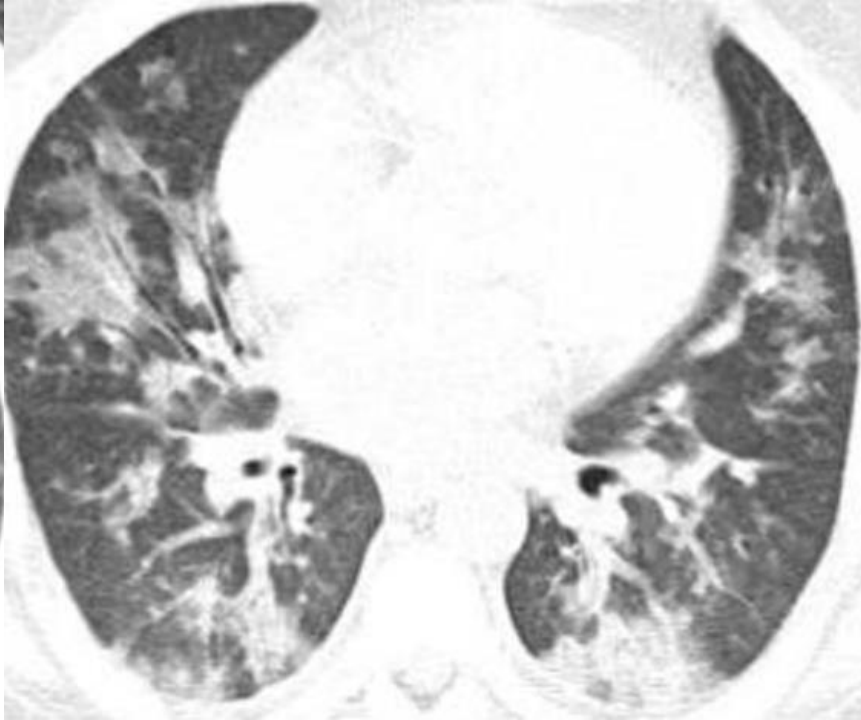
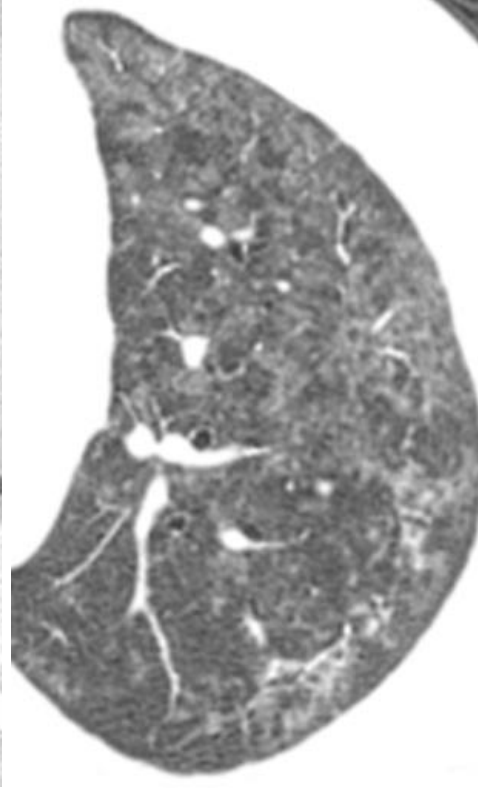
- Ödem
- Interst. pnömoni (virus, PCP)
- Limfanj. karsinom
- UIP (IPF, KDH vb)
- İlaçlar (Amiodaron vb)
- Kistik hastalıklar (LAM, LİP, Histiositozis)
- Sarkoidoz
- Metastaz
- Tbc

NODÜLER

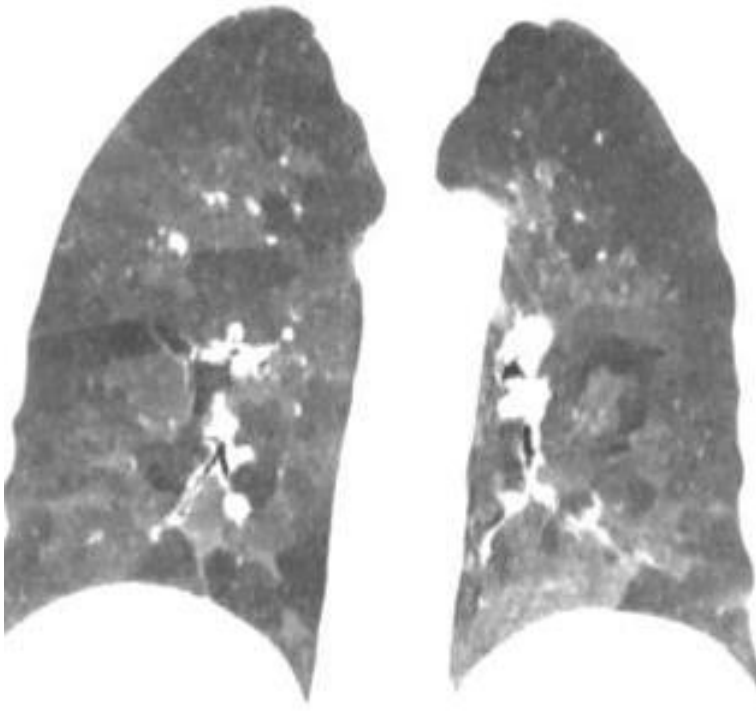
- Sarkoidoz
- Silikoz
- Limfanj. Karsinom
- Sarkoidoz
- Metastaz
- Tbc
- Histiositozis
- Hipersens. pnöm
- RBİLD
- BAC

KİSTİK

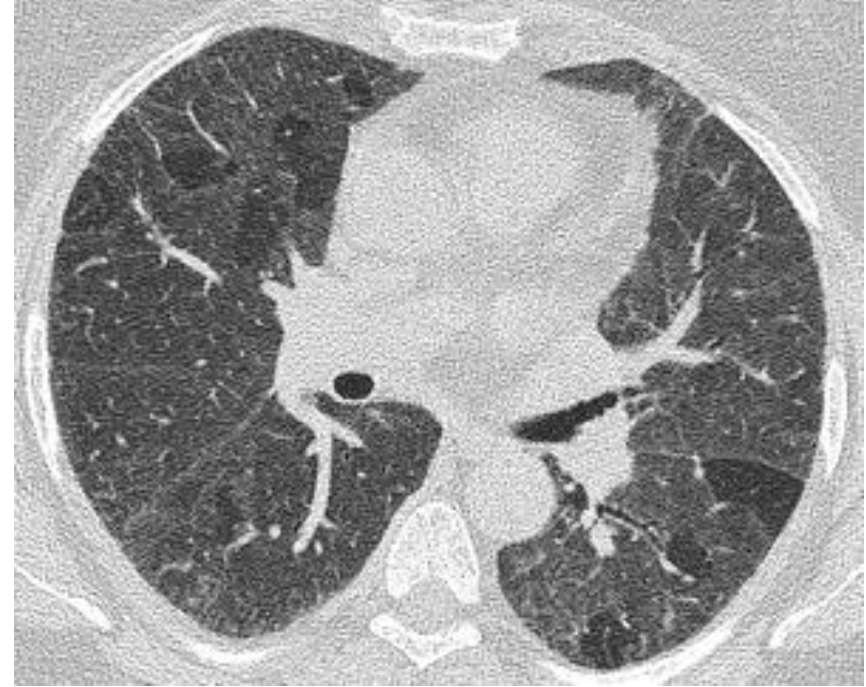
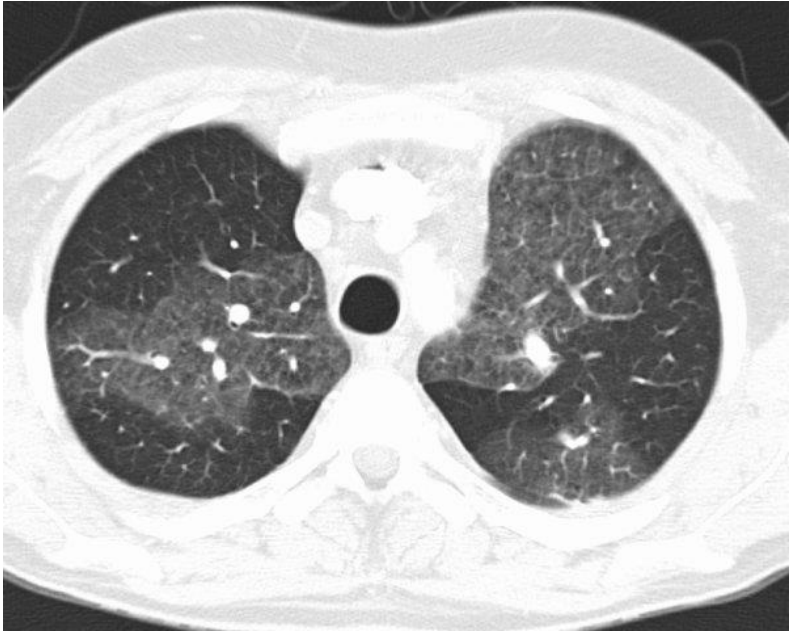
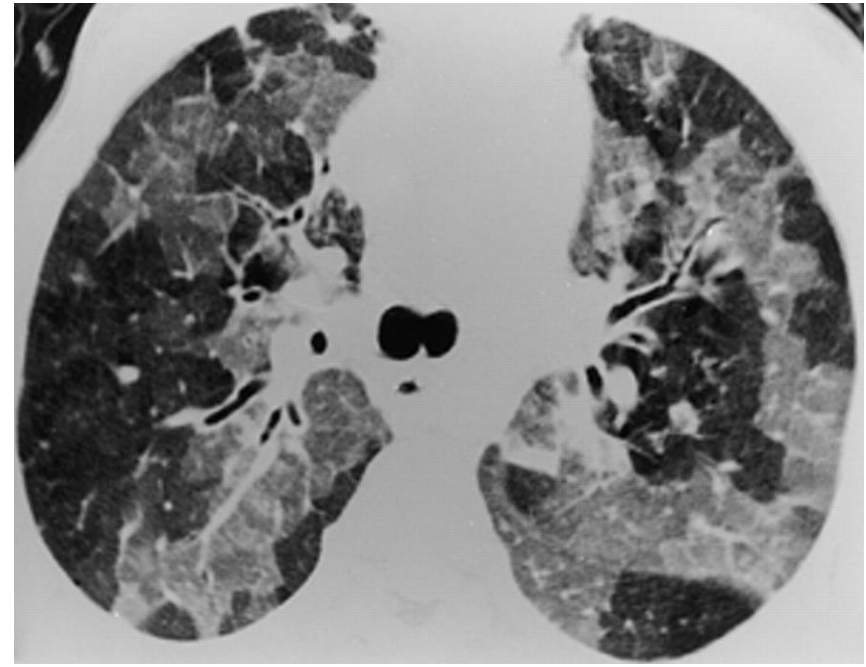
- Amfizem
- Balpeteği
- LAM
- Histiocytosis
- LİP
- Pulmonary metastases (squamous/adenocarcinoma)
- Birt Hogg Dubé syndrome
- Cystic mesenchymomas
- Light-chain disease

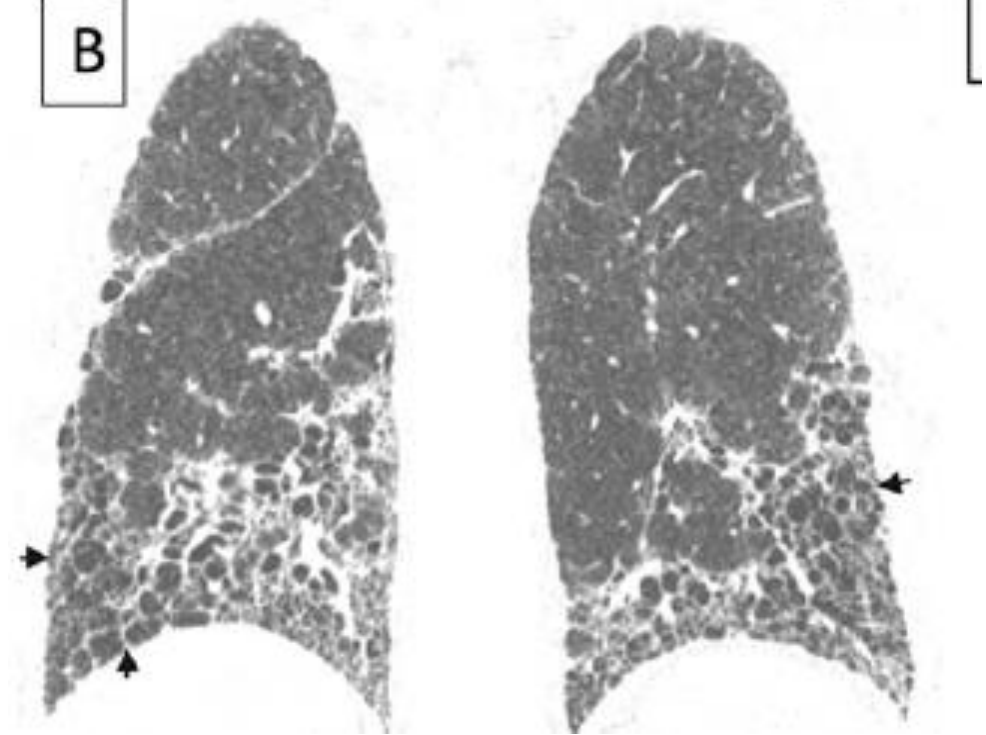
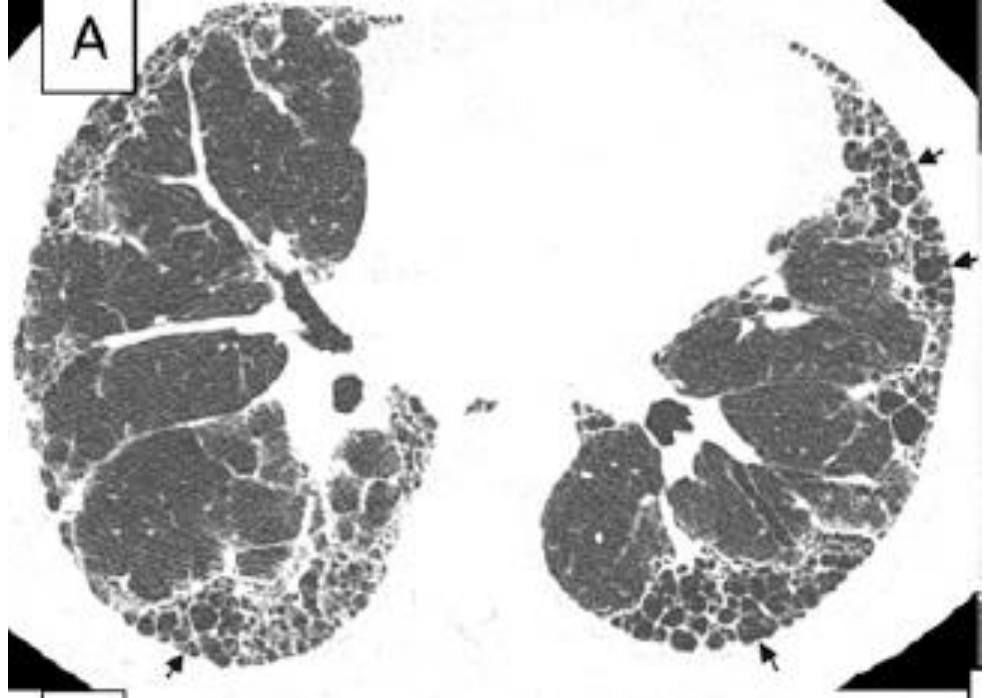


Buzlu
Cam

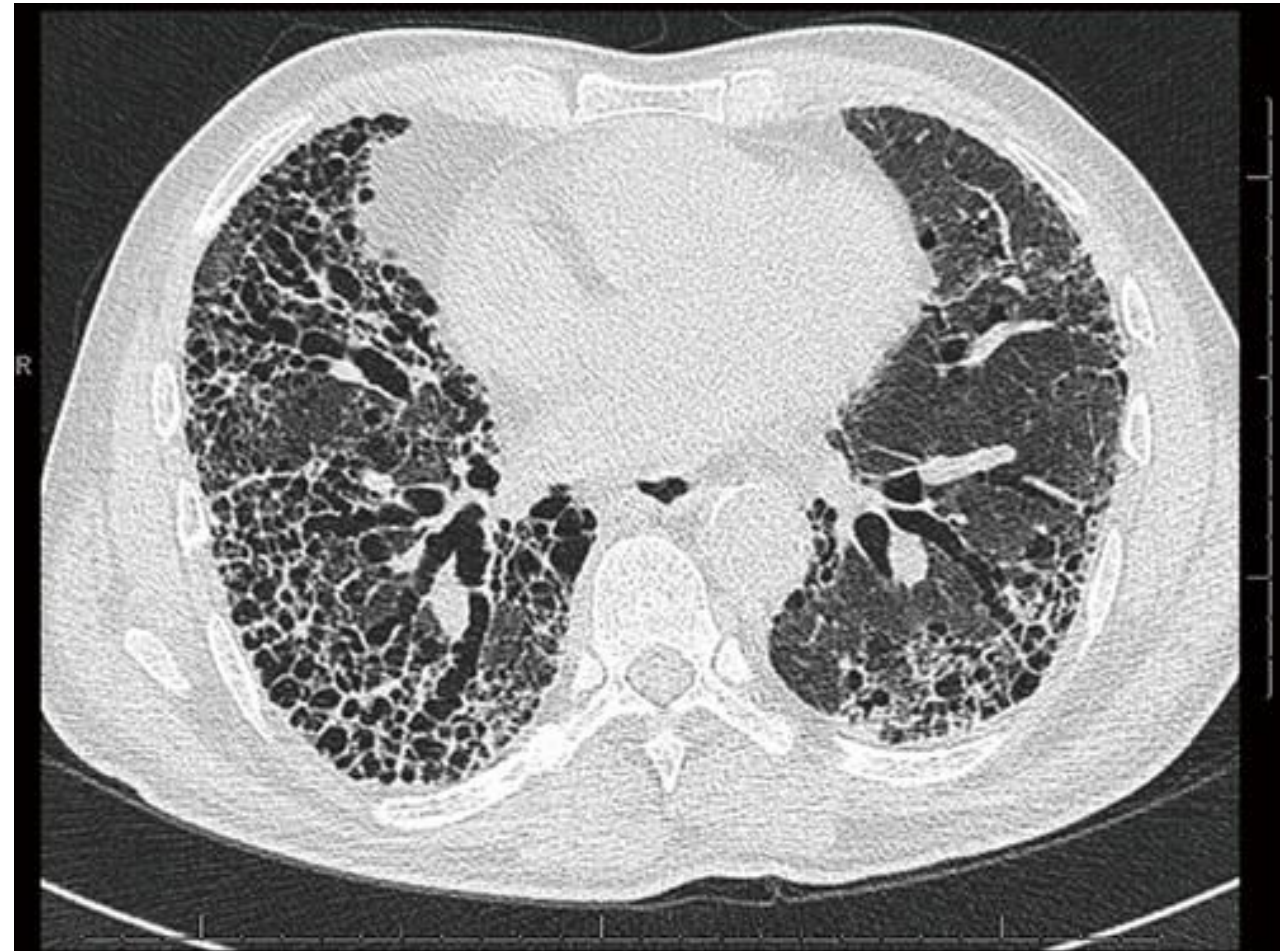


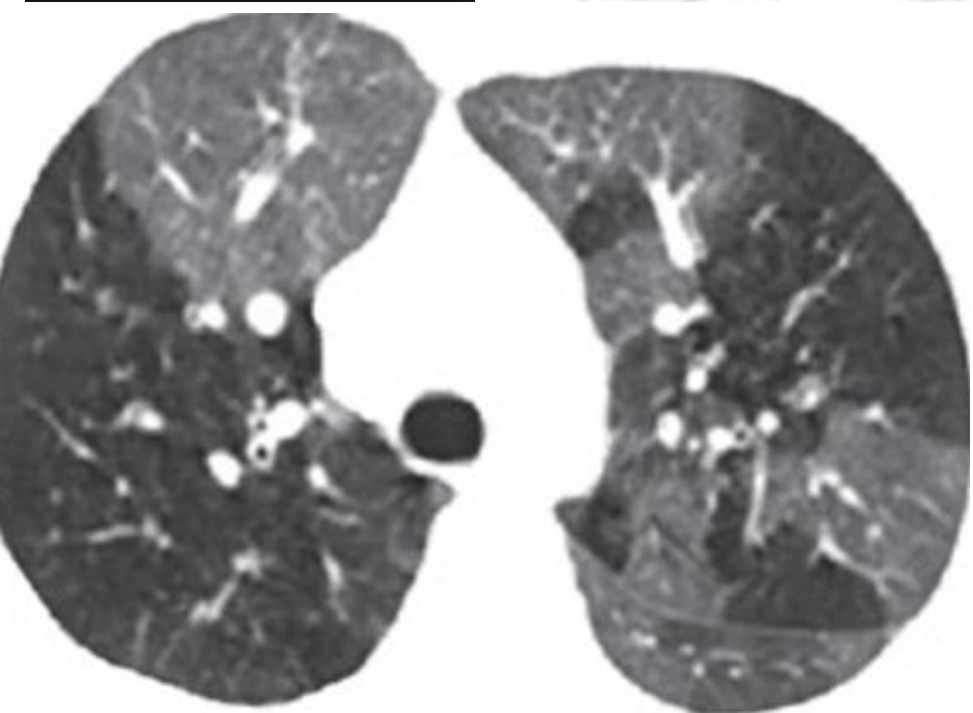
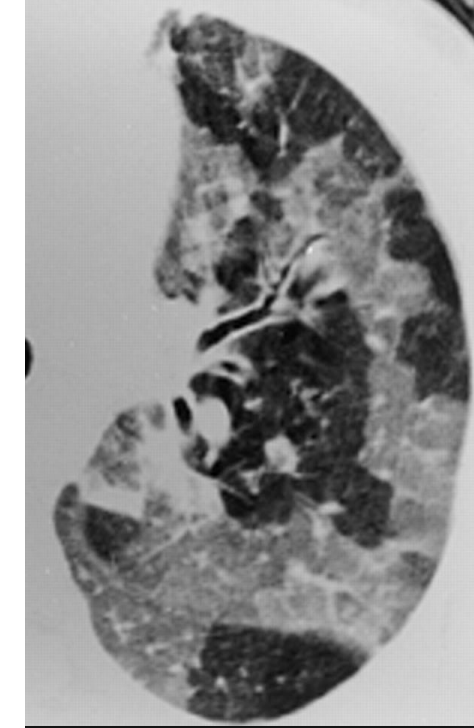
Buzlu cam
Mozaik
perfüzyon



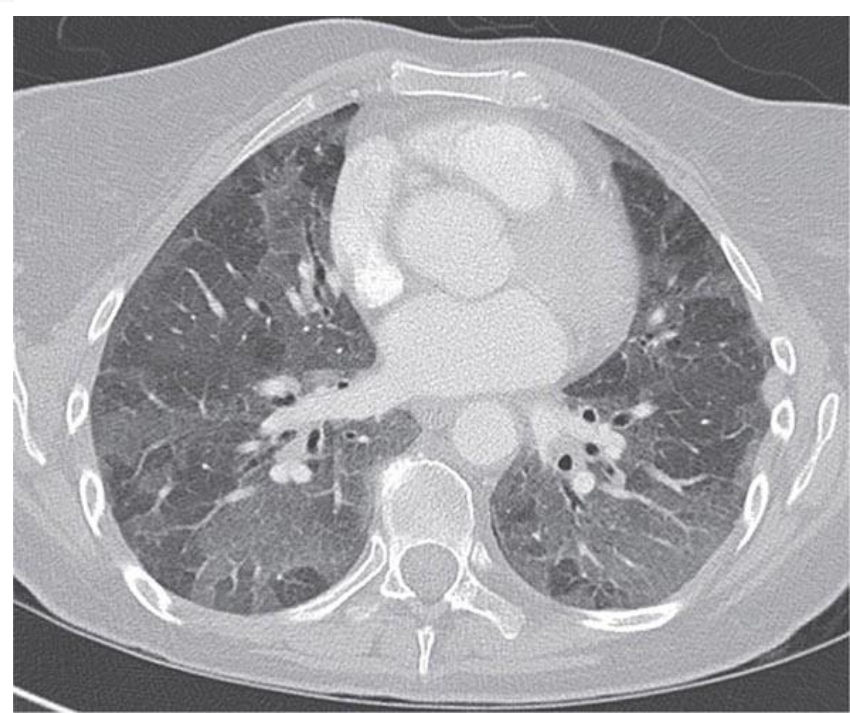


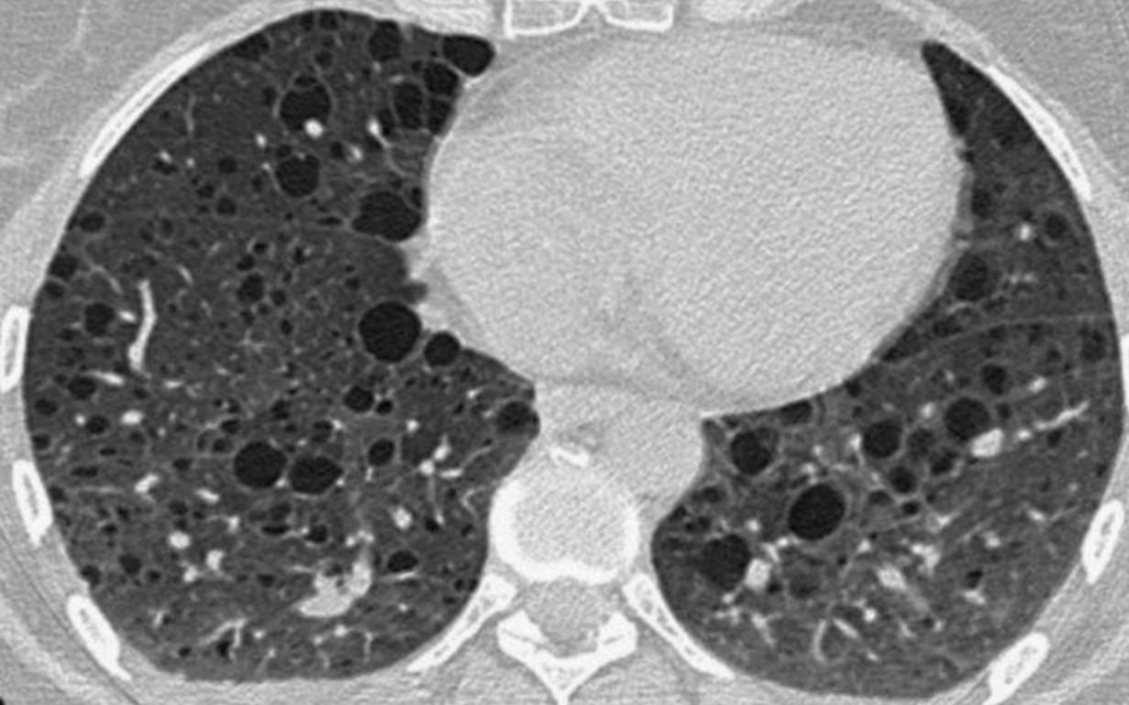
Balpeteği
Traksiyon
bronşektazisi



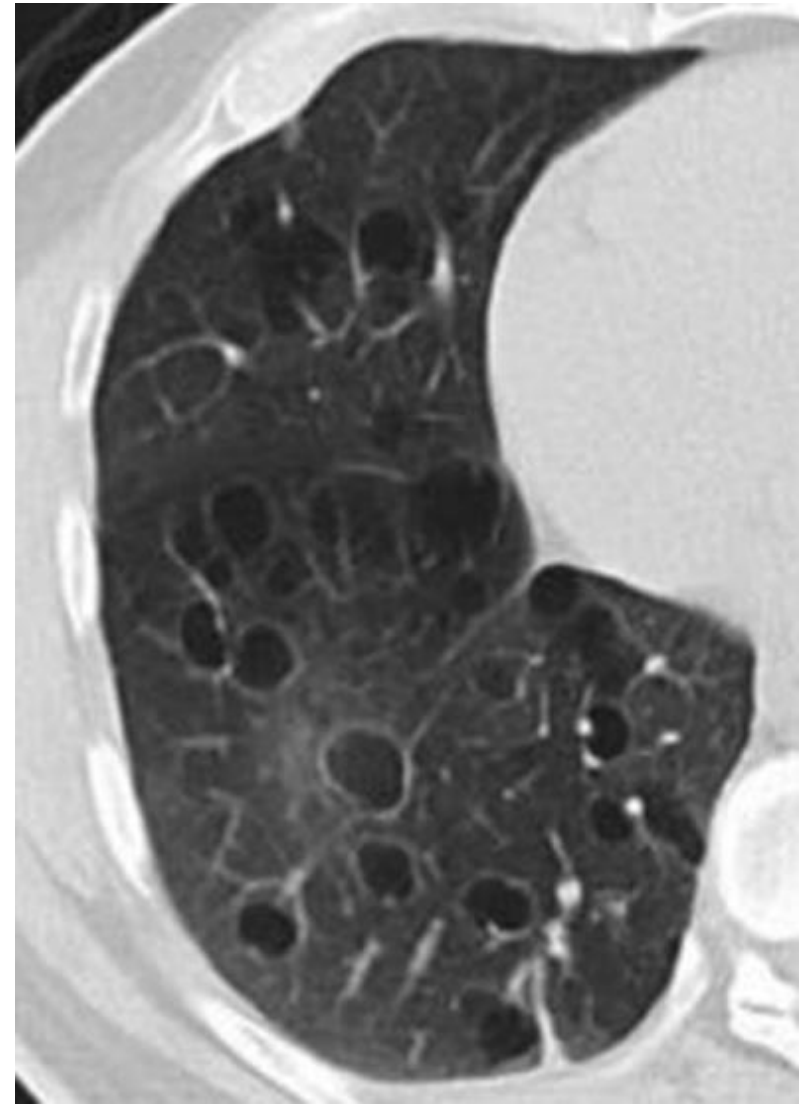
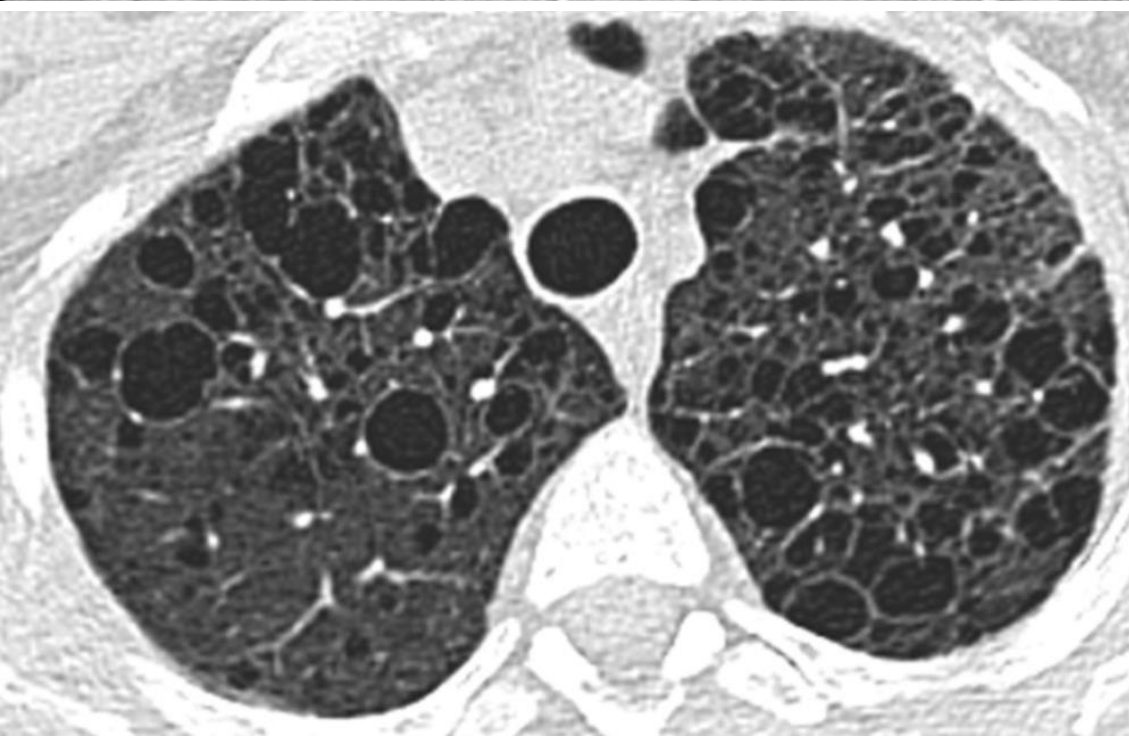


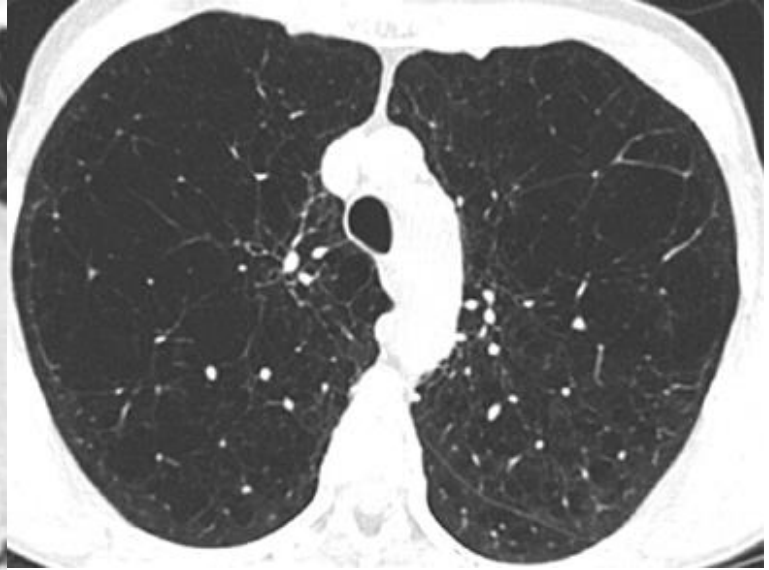
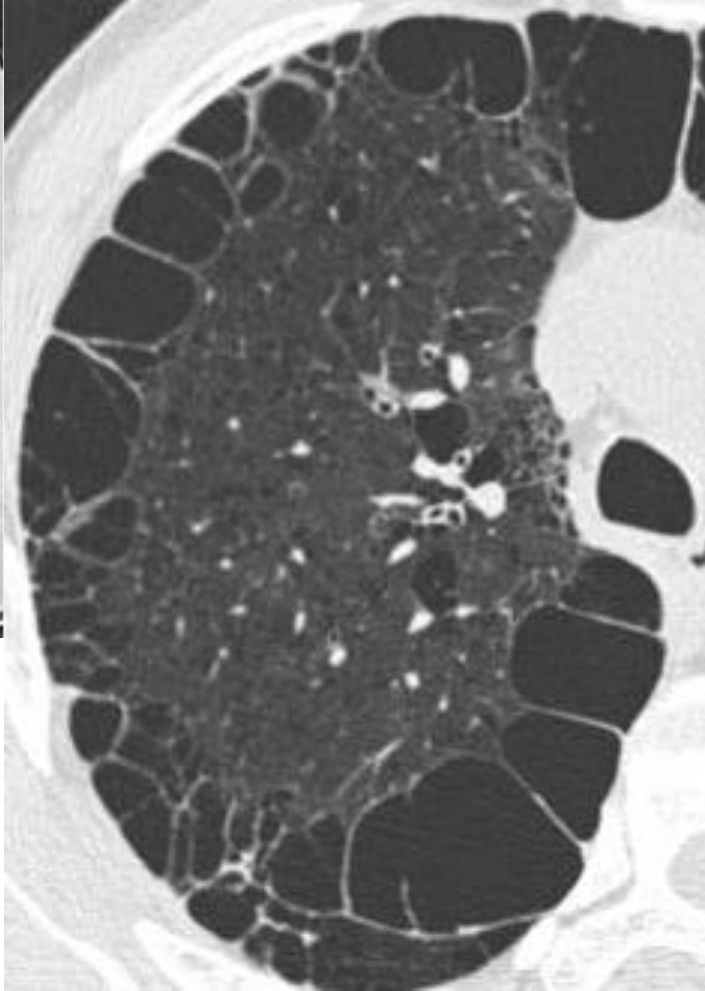
Mozaik
Patern



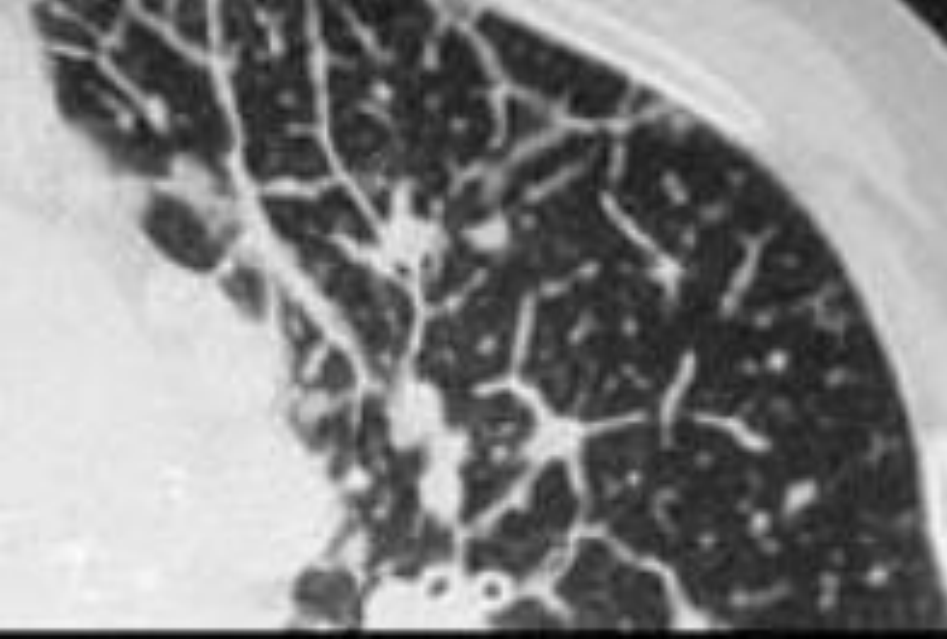
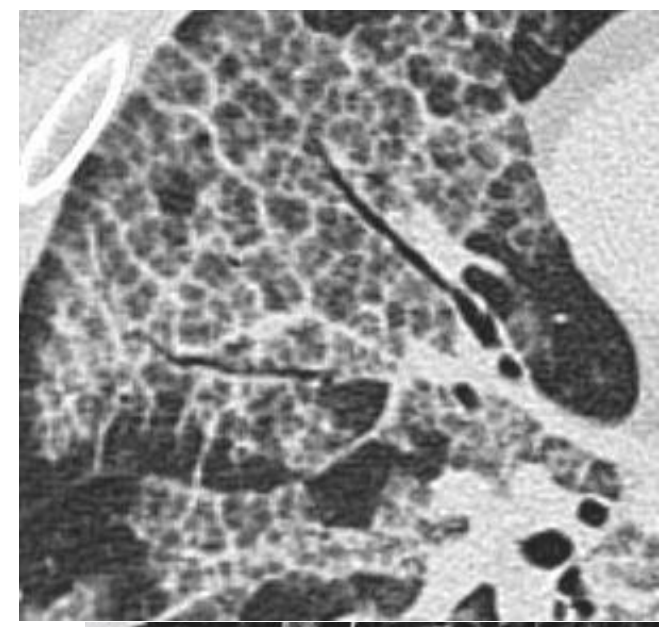
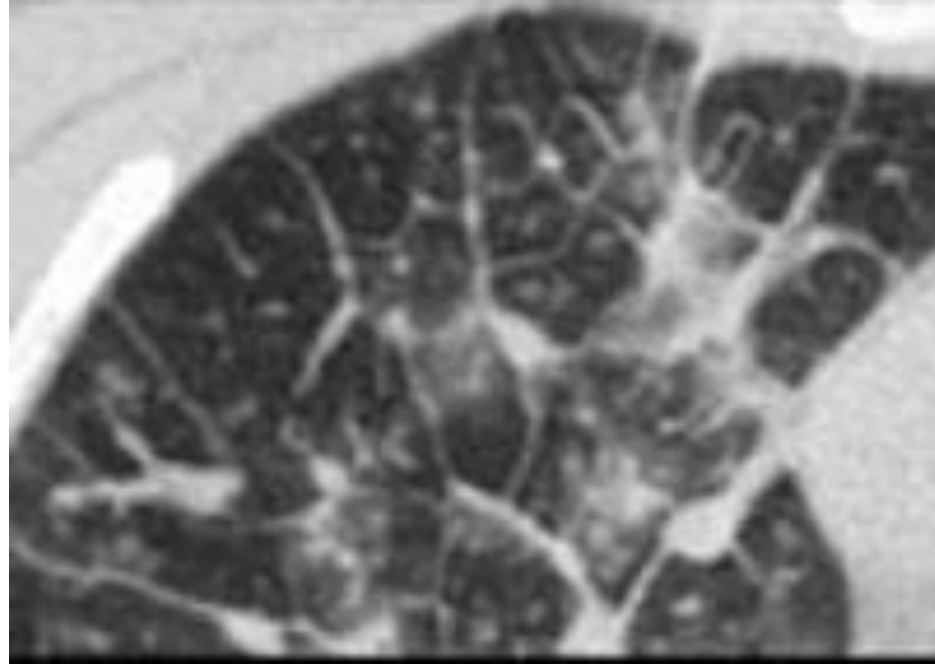
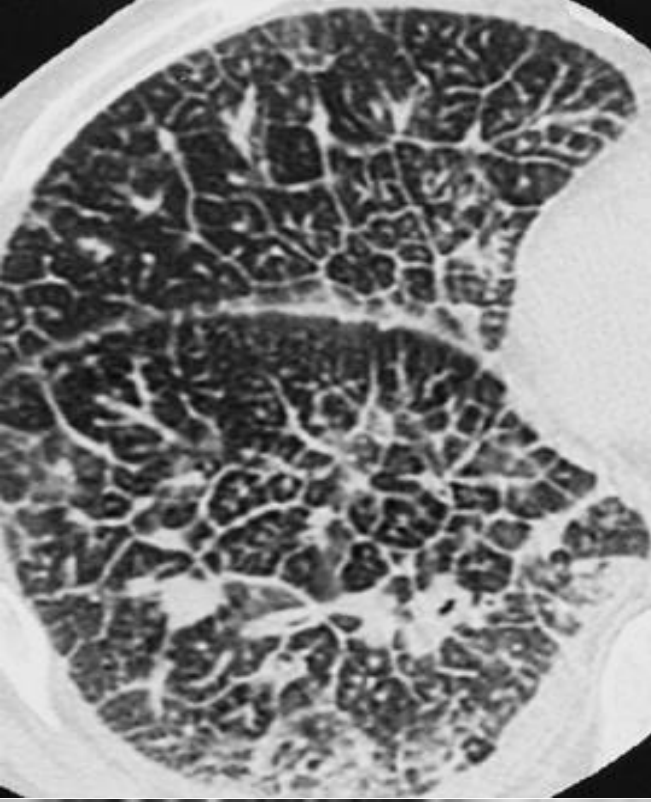


Hava kisti

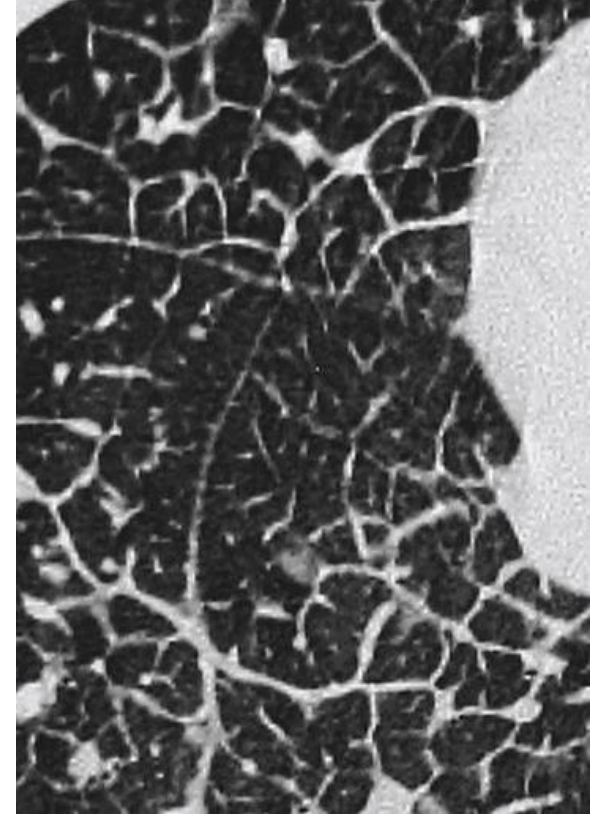


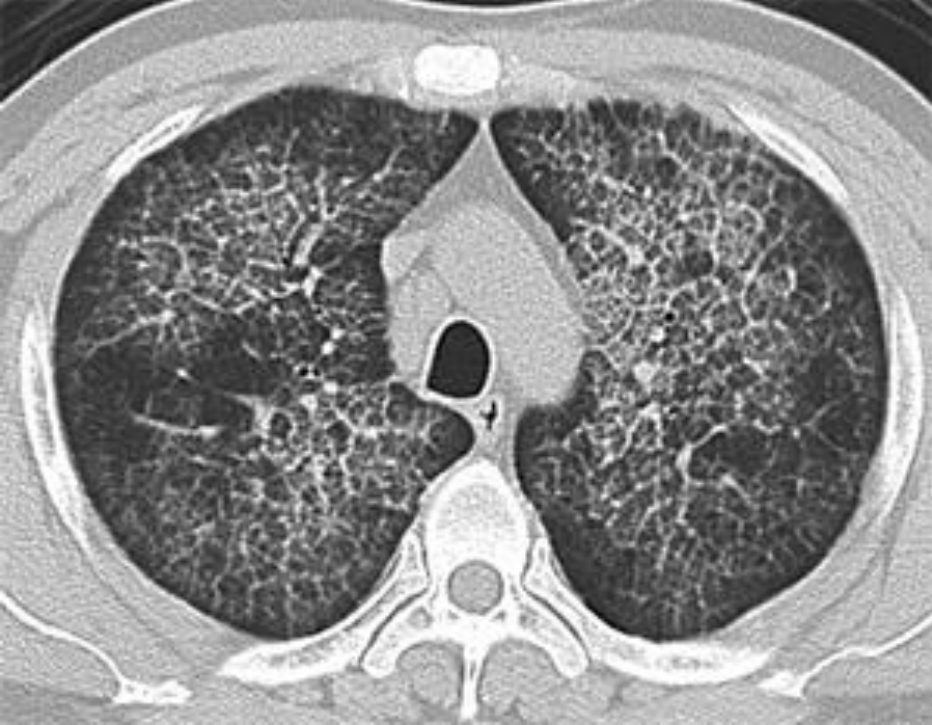


Amfizem

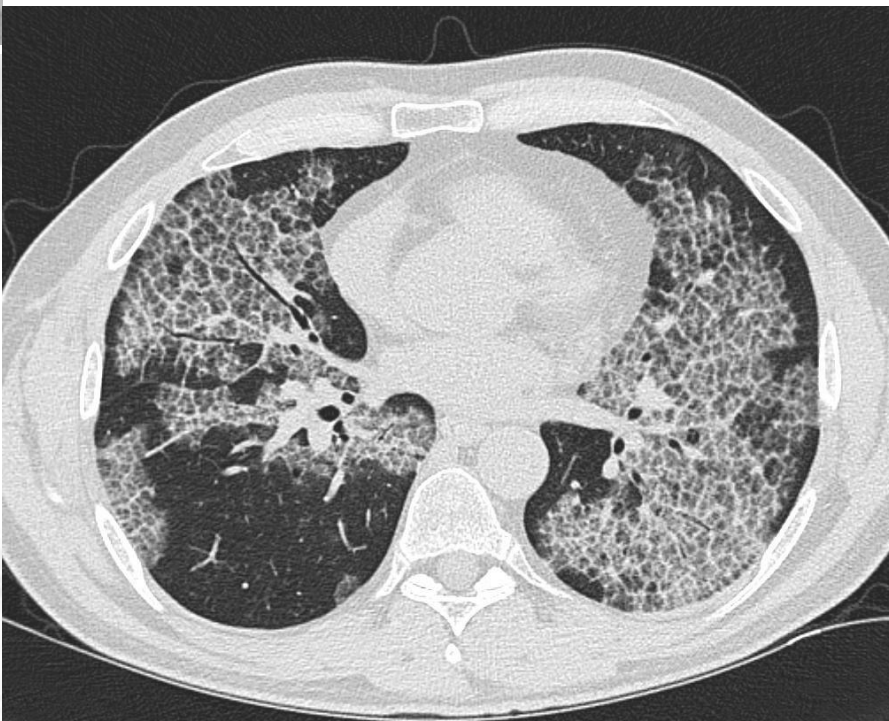


İnterlobüler Septal
Kalınlaşmalar



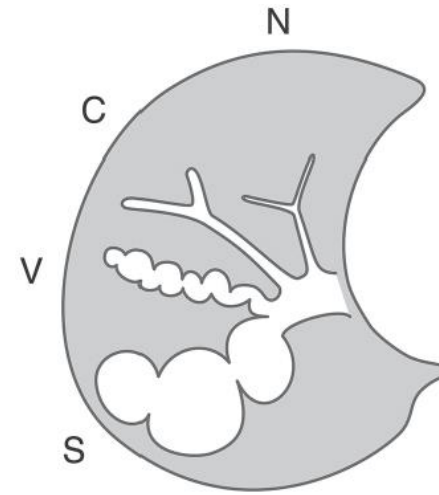
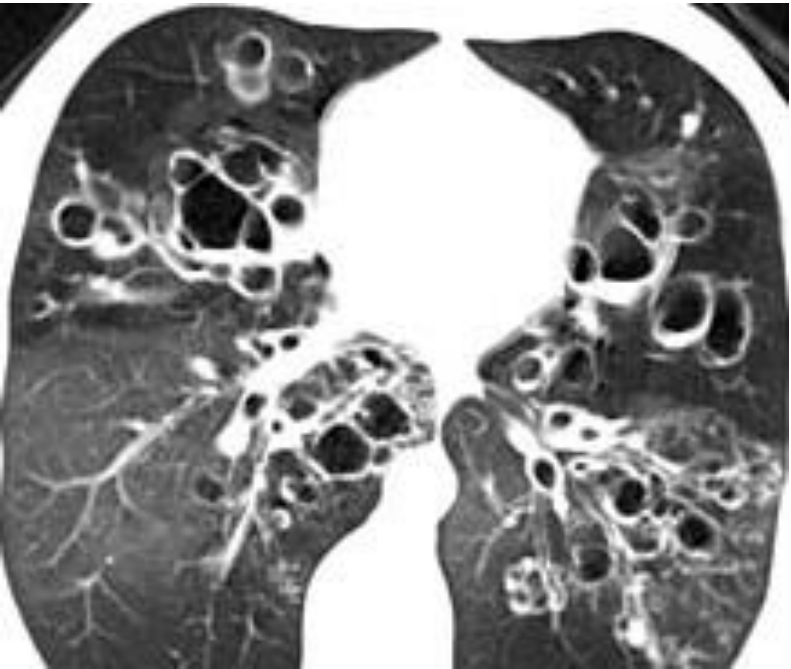
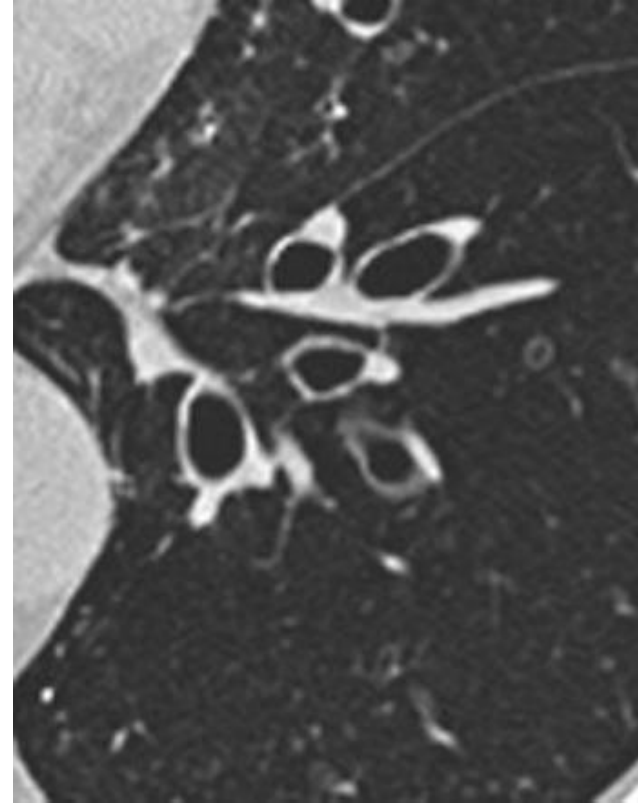


Kaldırım Taşı
Crazy Paving



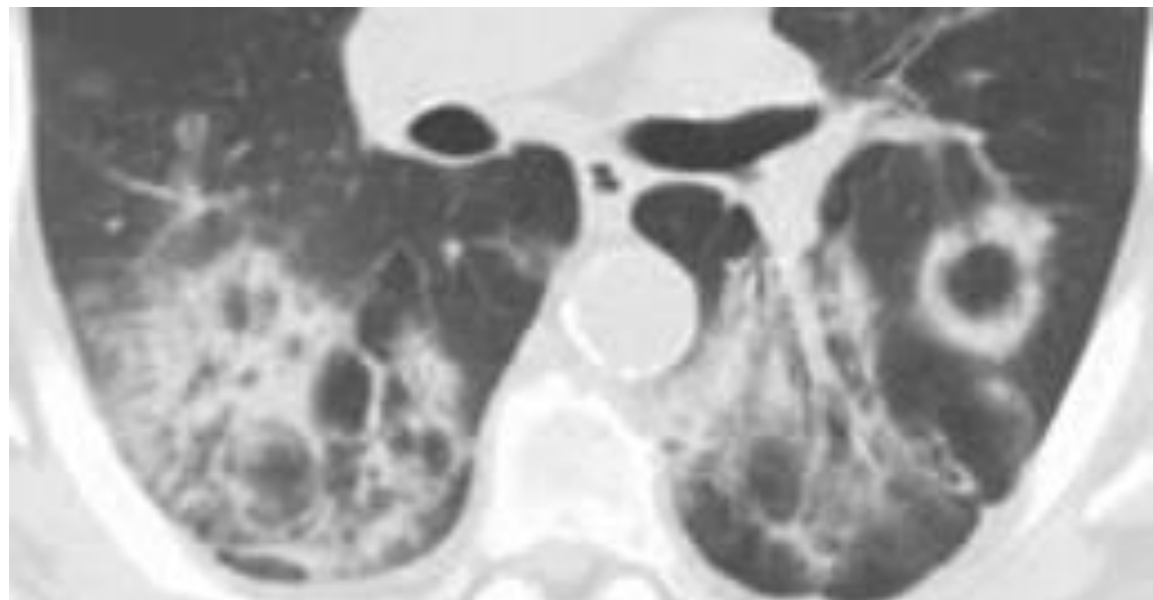
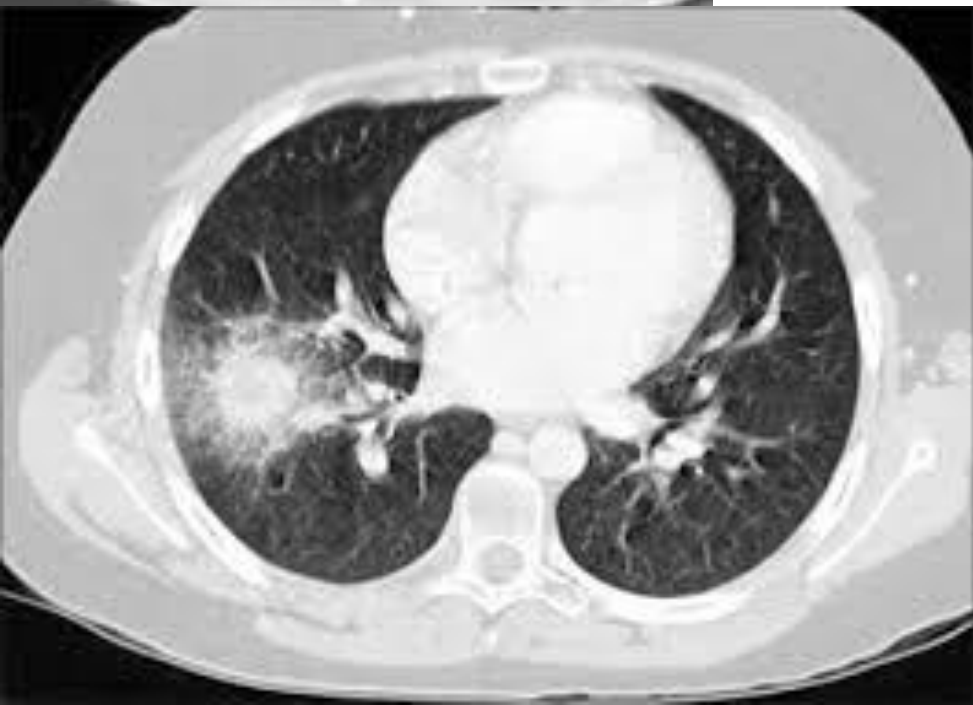
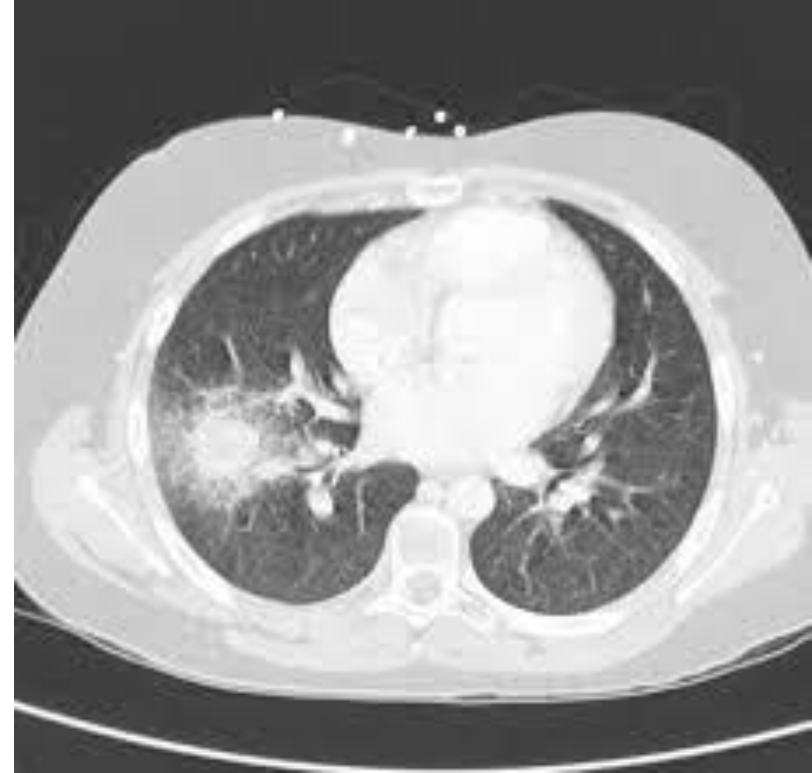


Bronşektazi

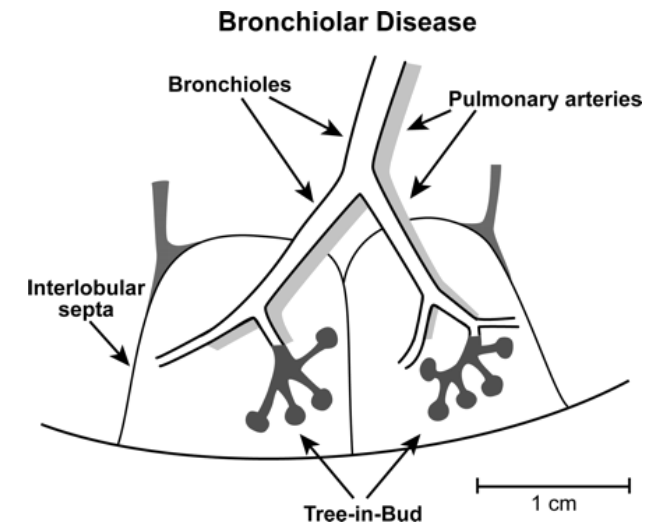
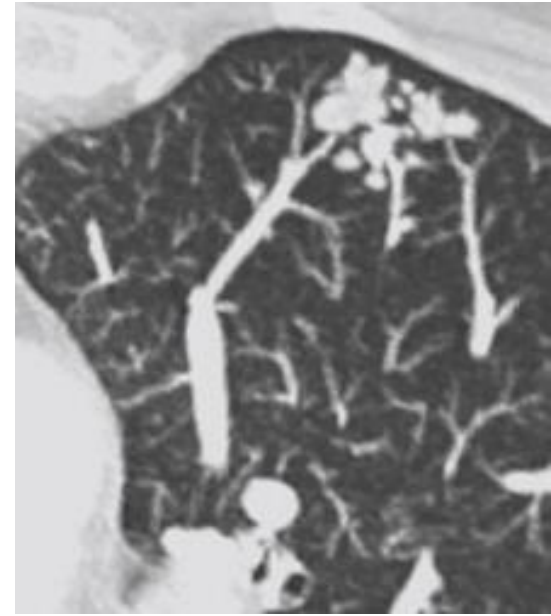
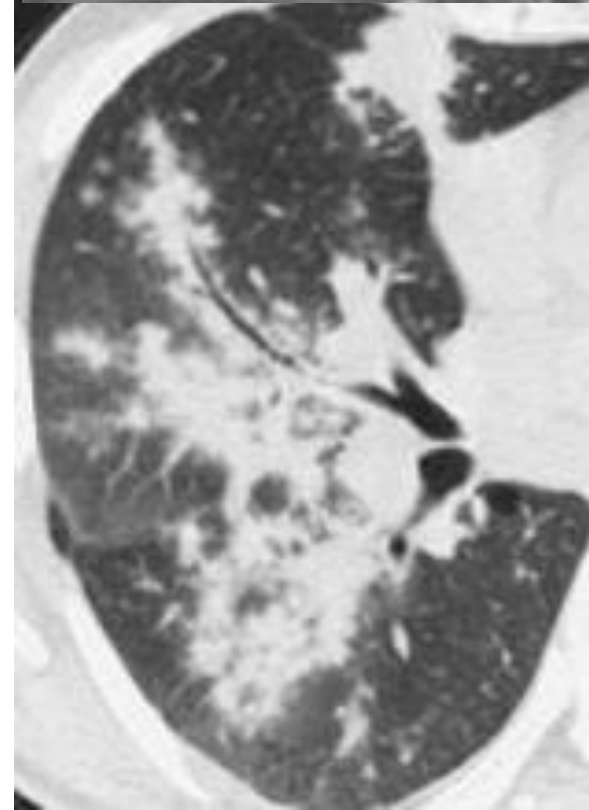
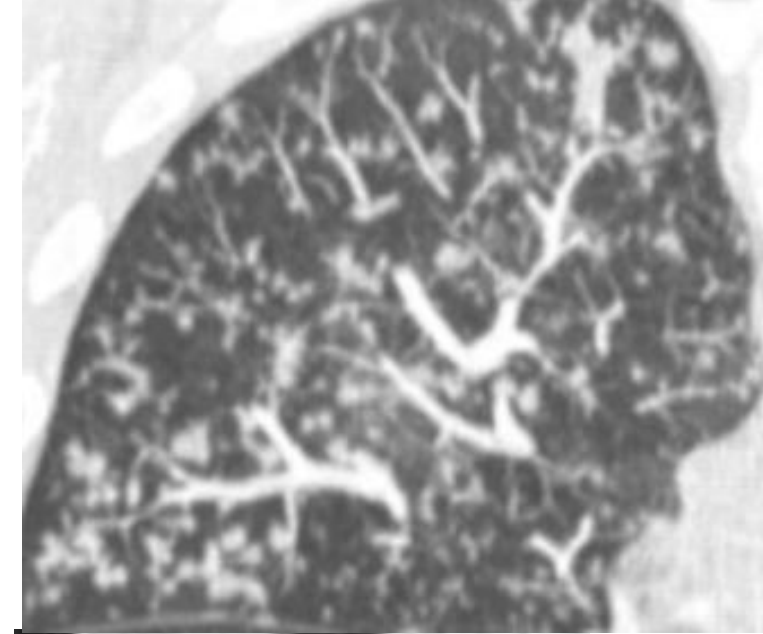


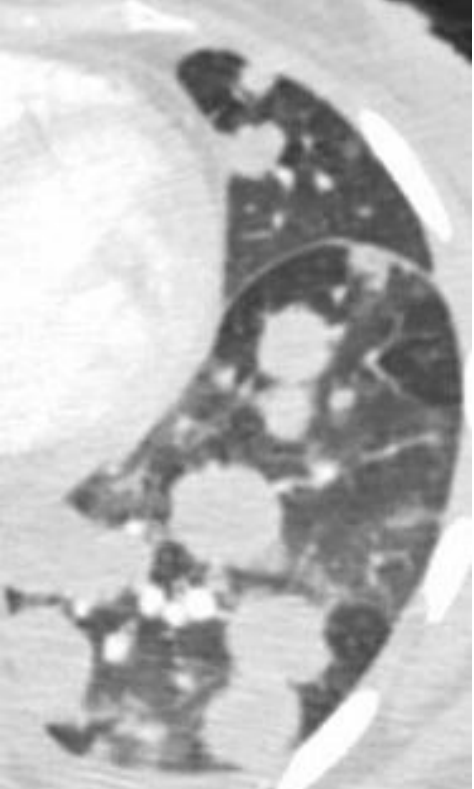


Halo
Ters Halo

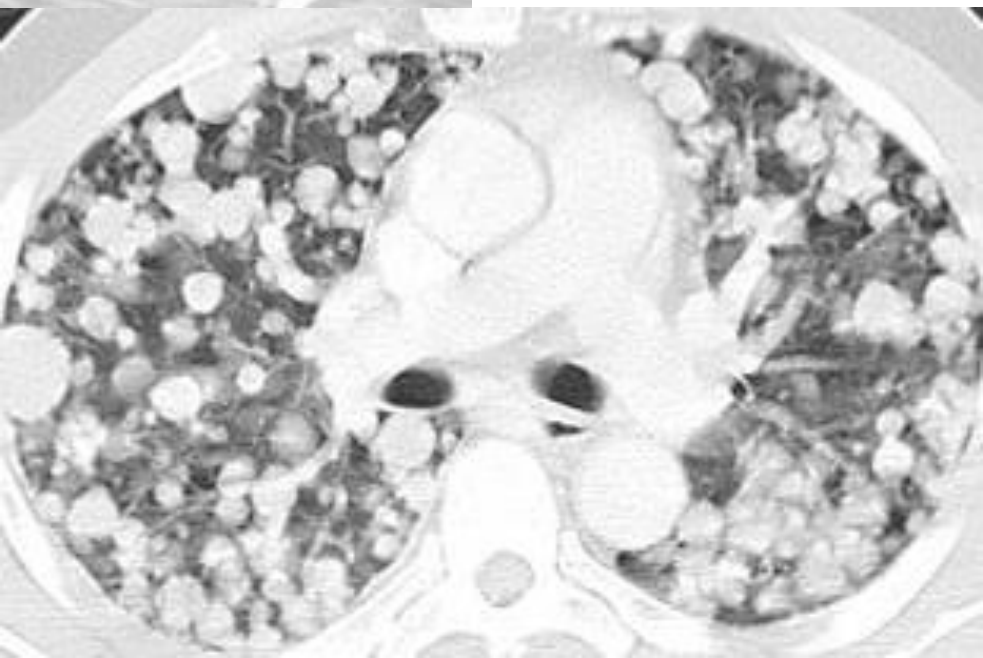


Tree in bud

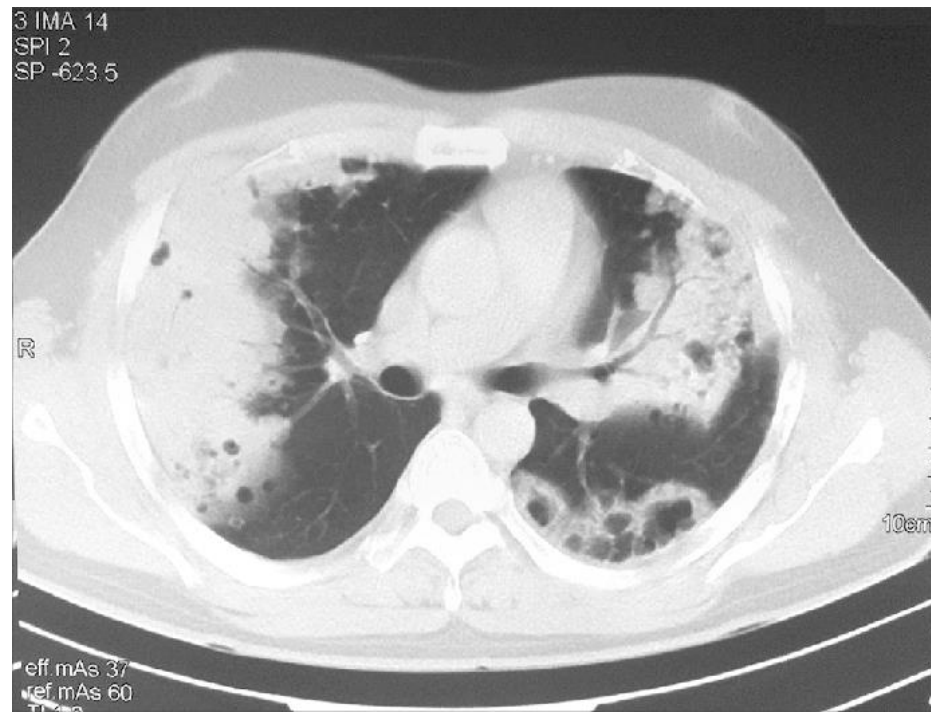
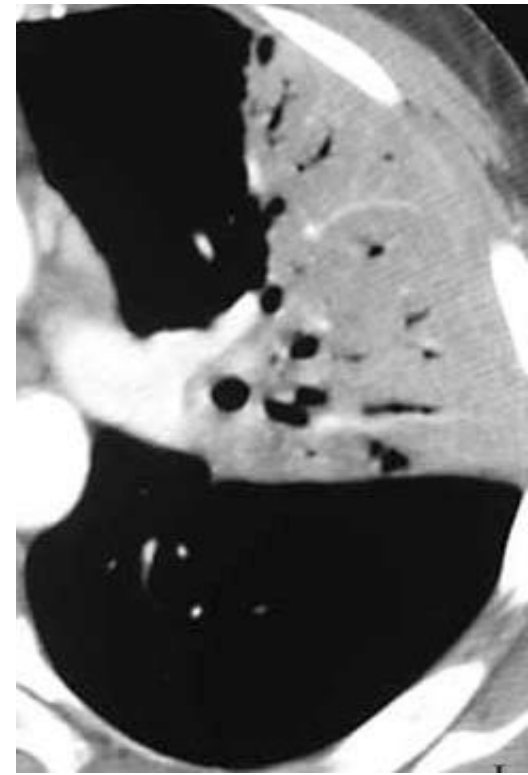
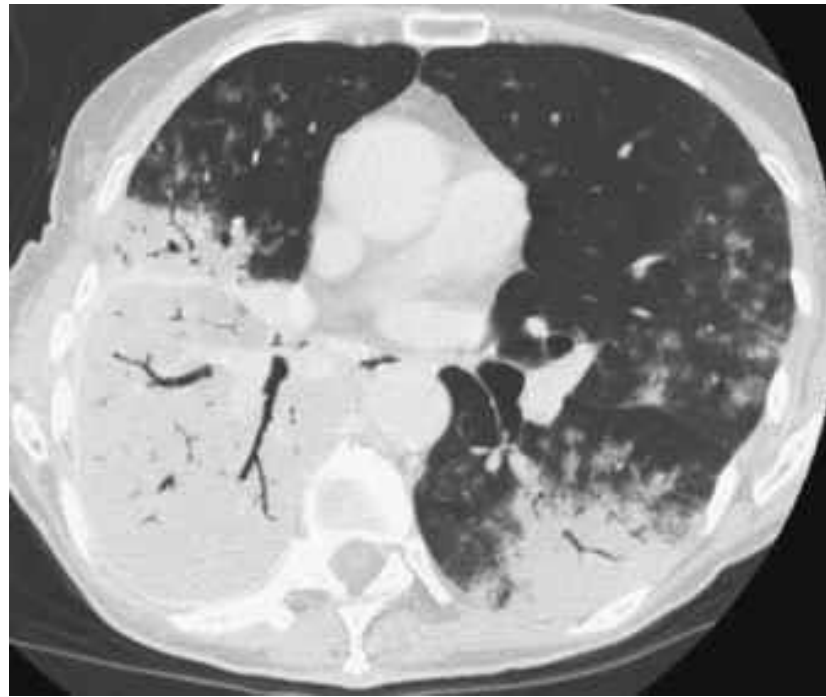
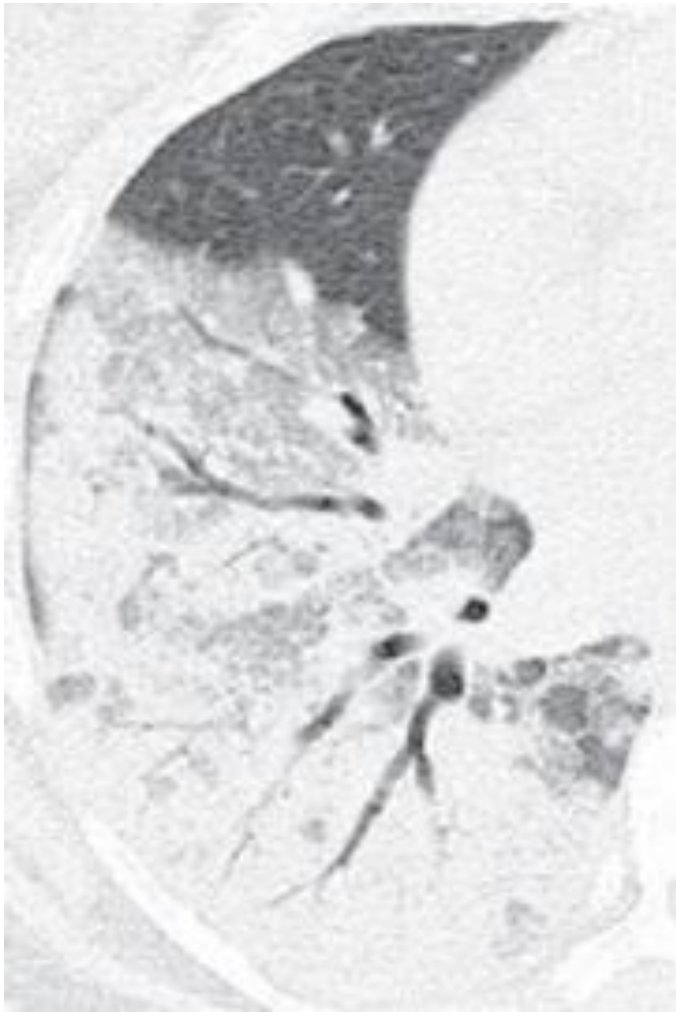


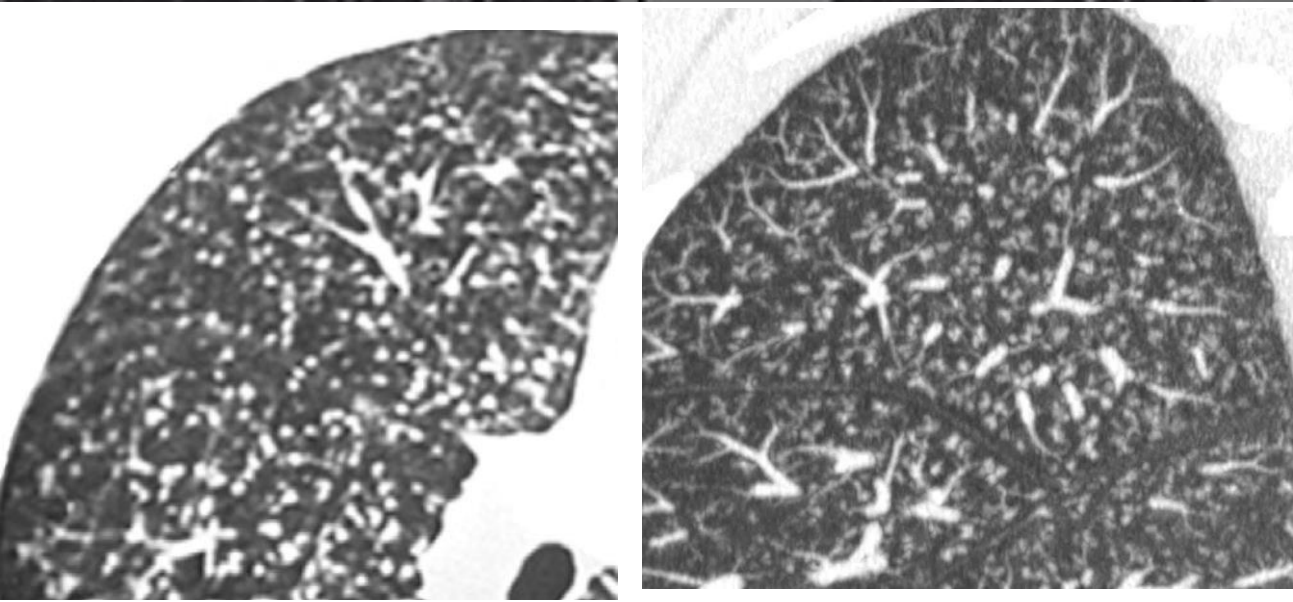
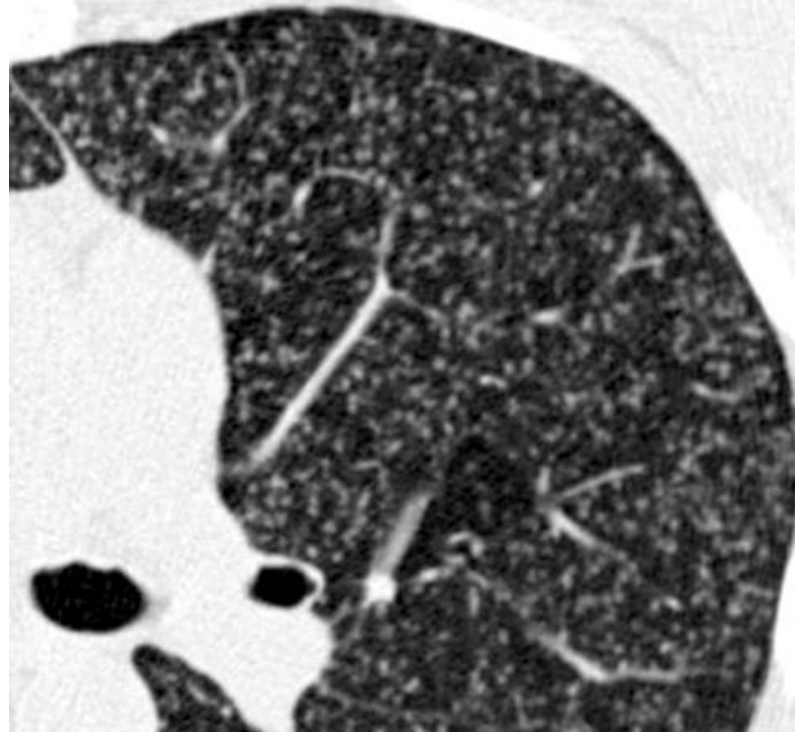
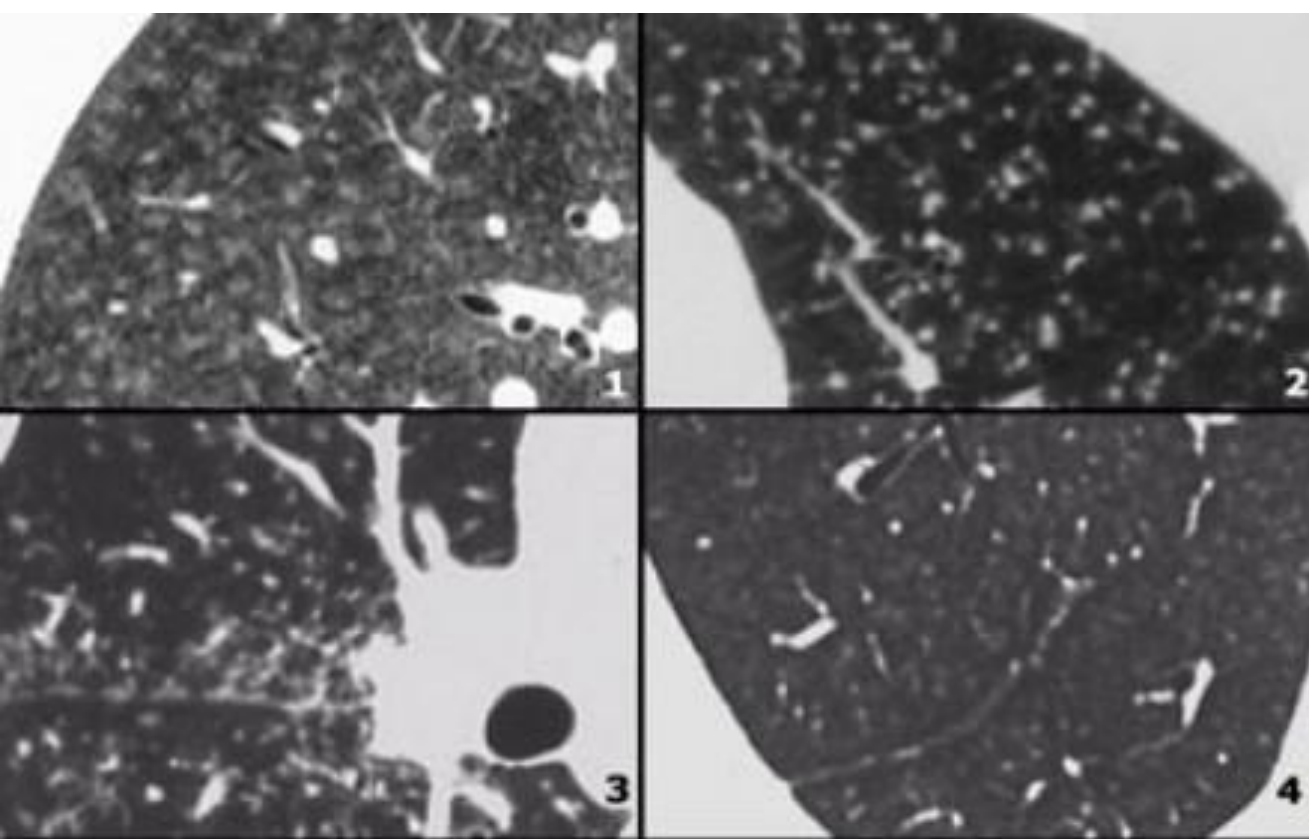


Cannonball

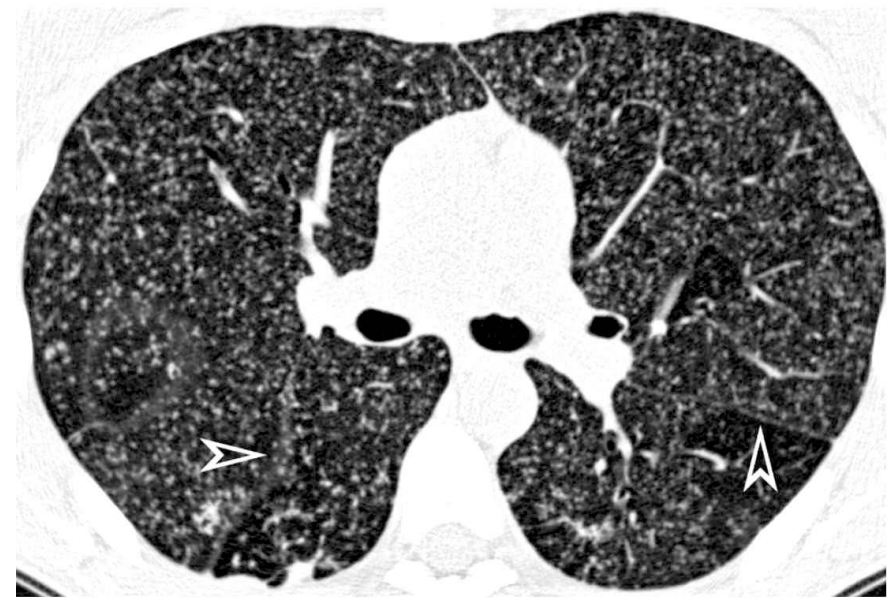


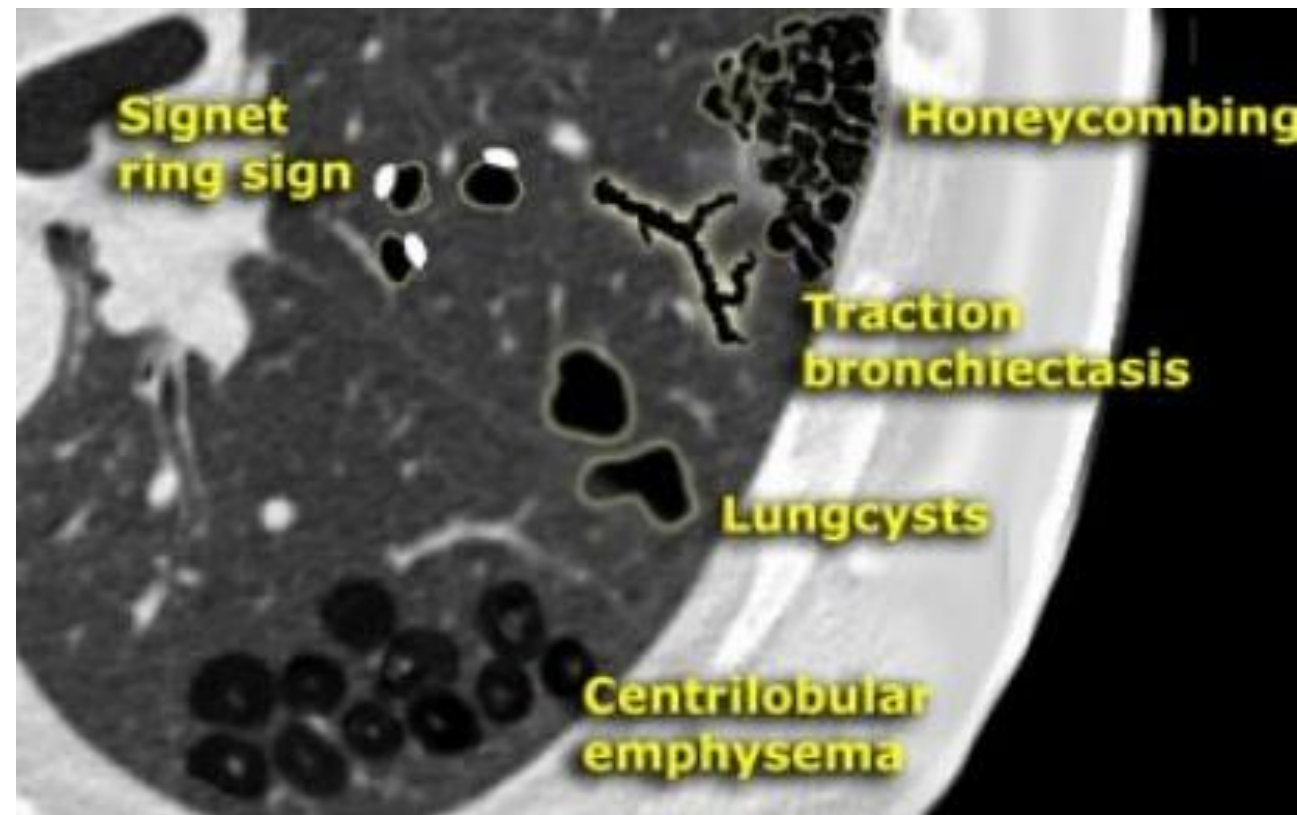
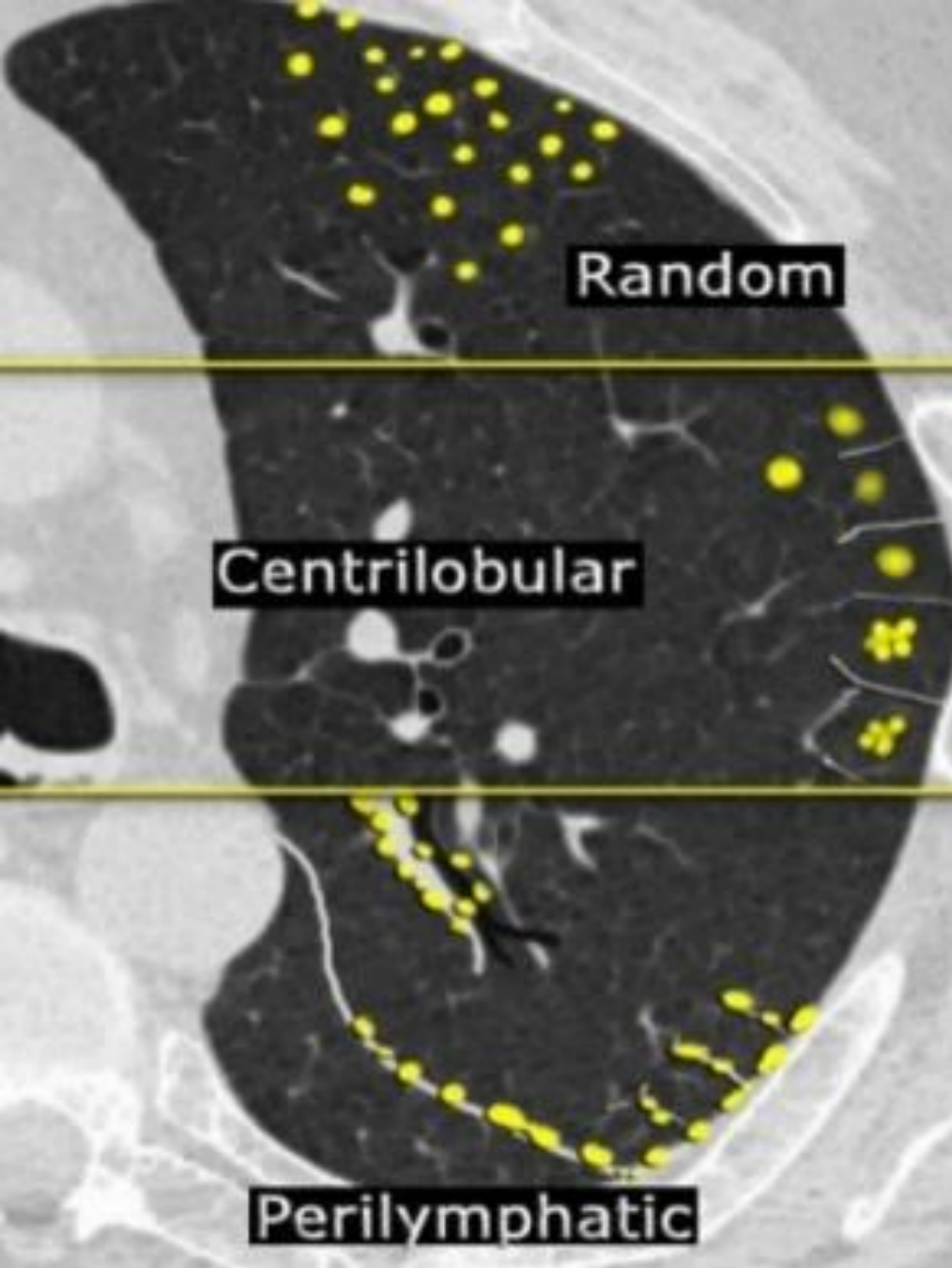
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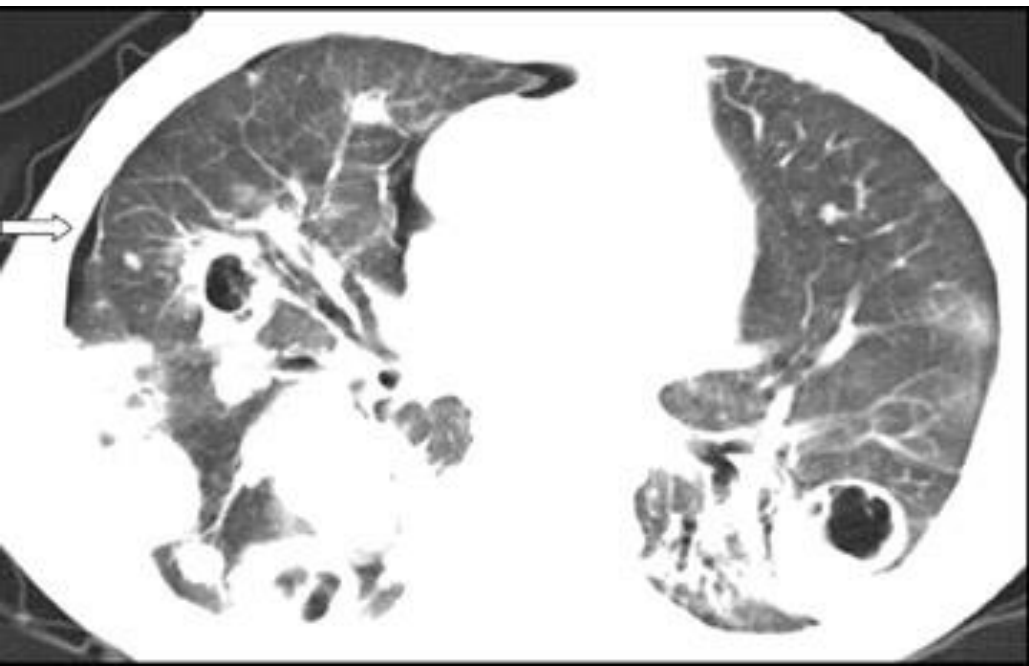




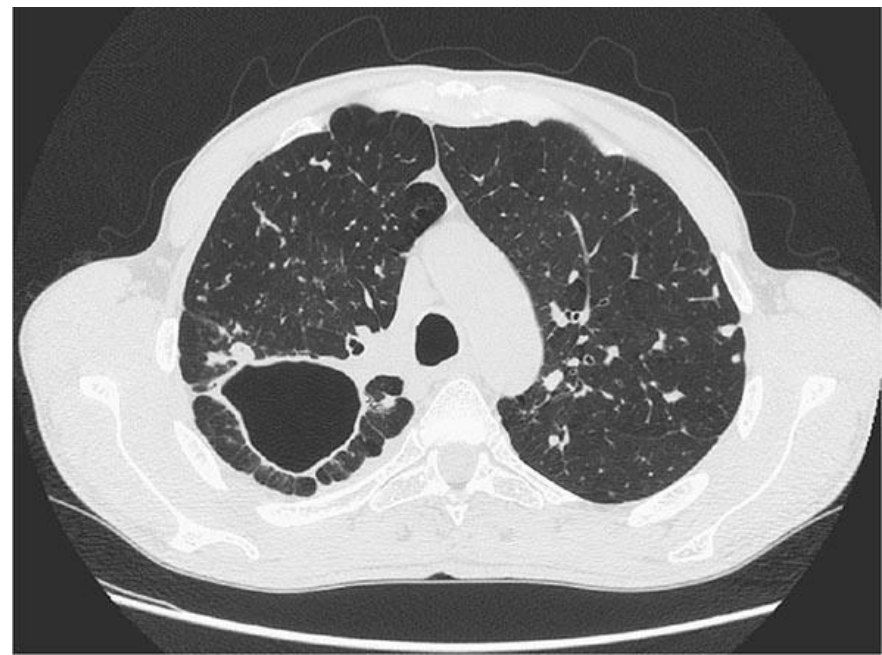
Nodül

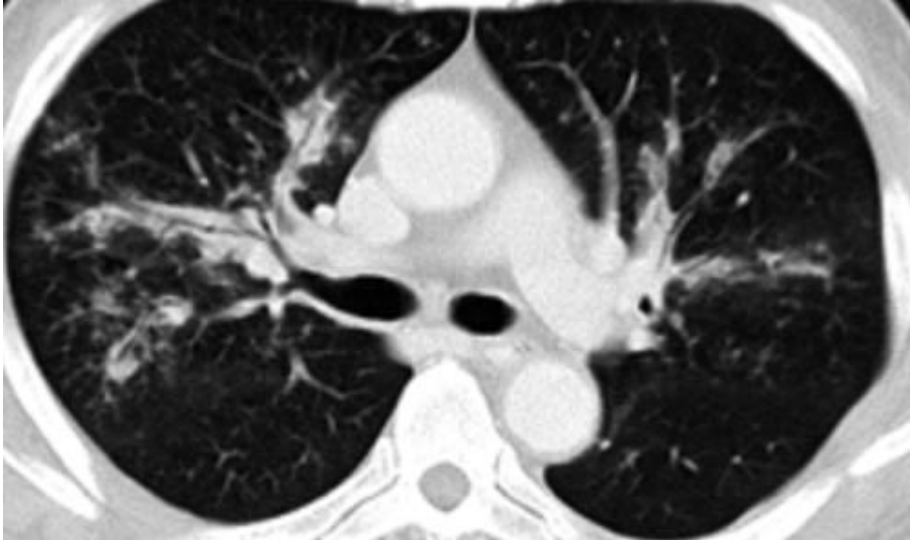




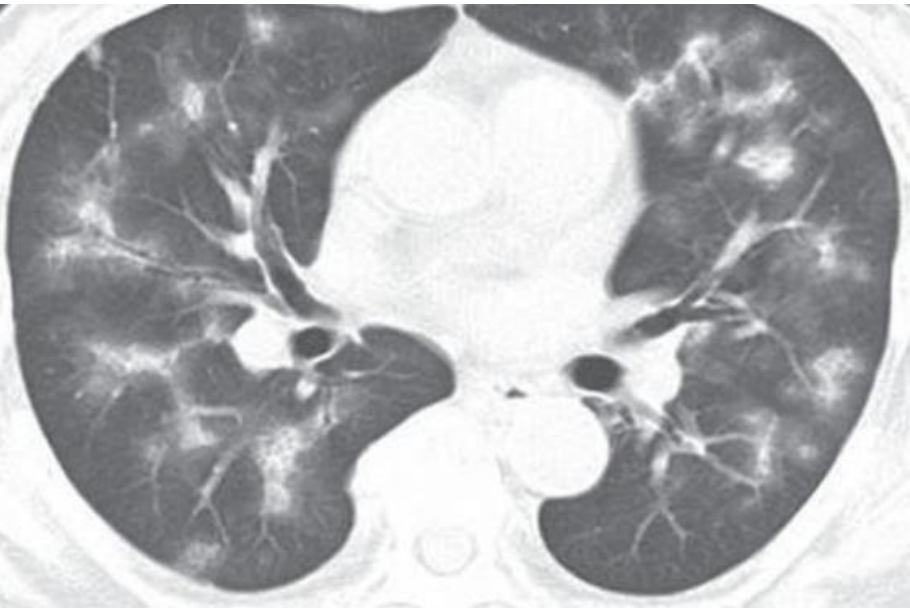
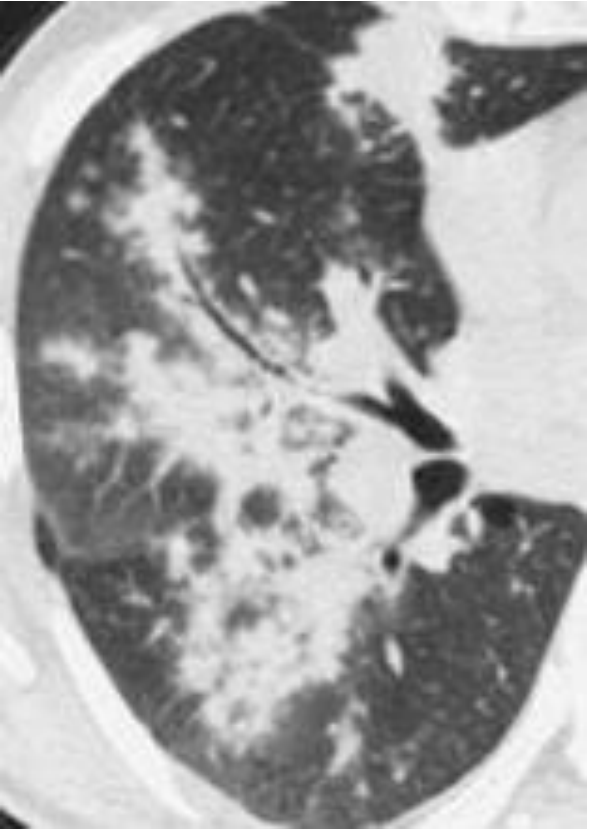
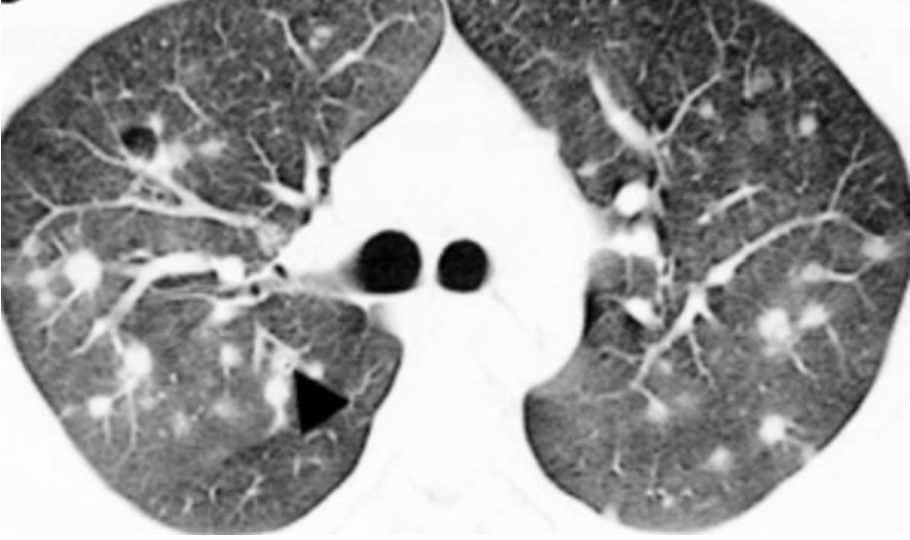


Kavite

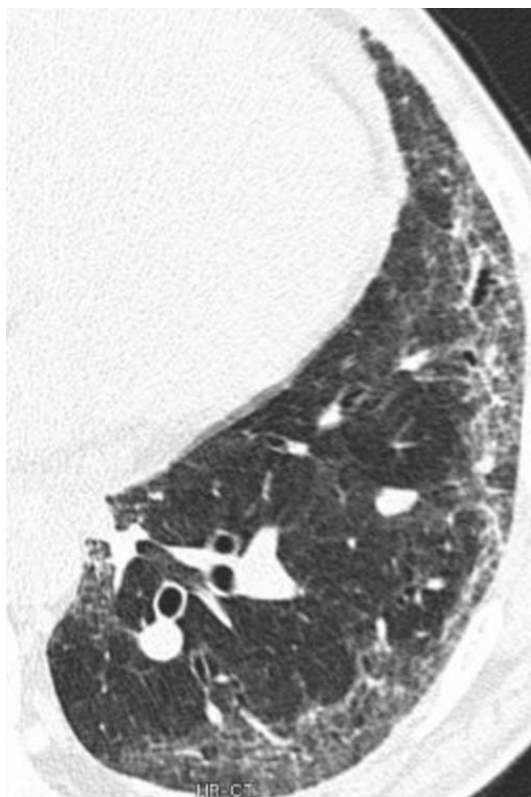
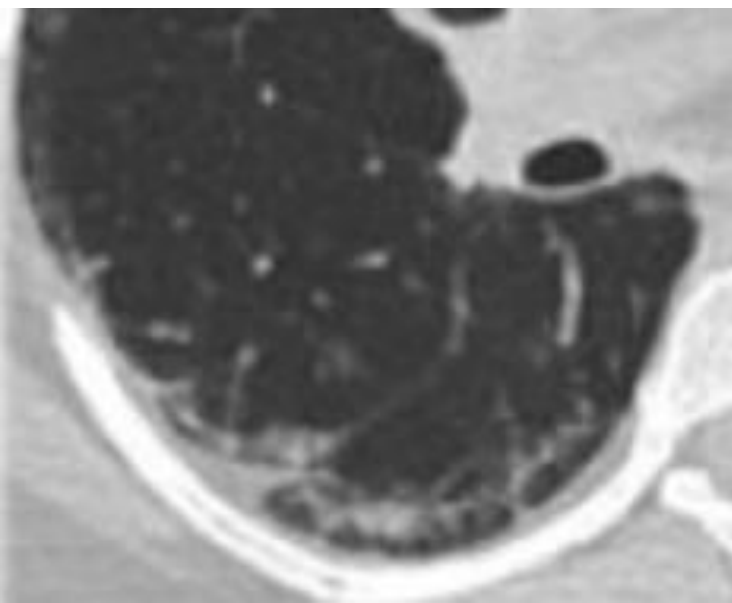
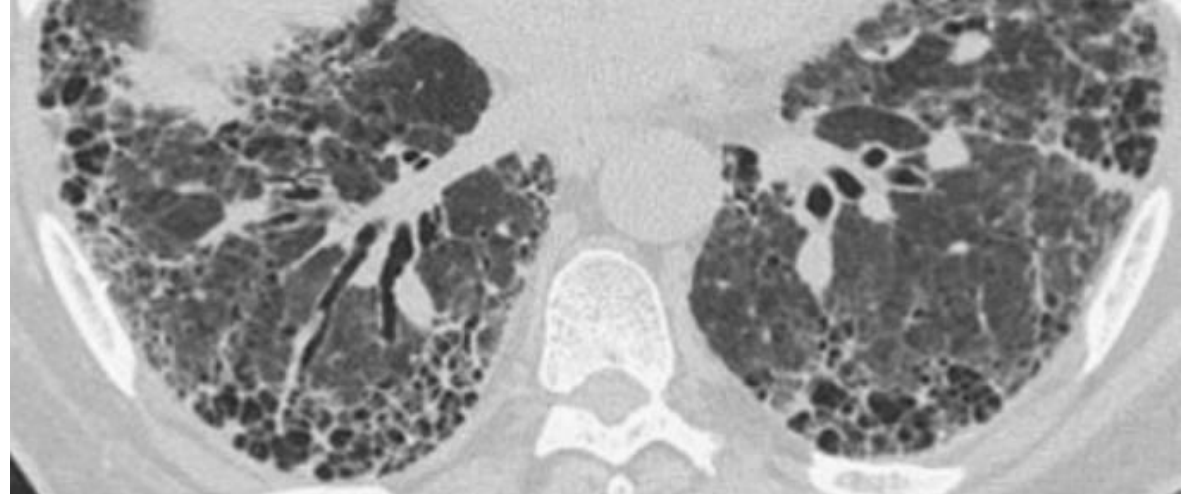
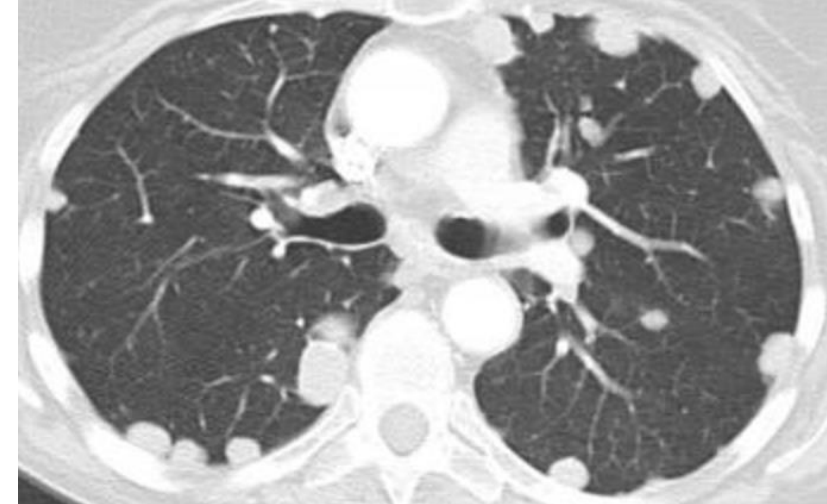


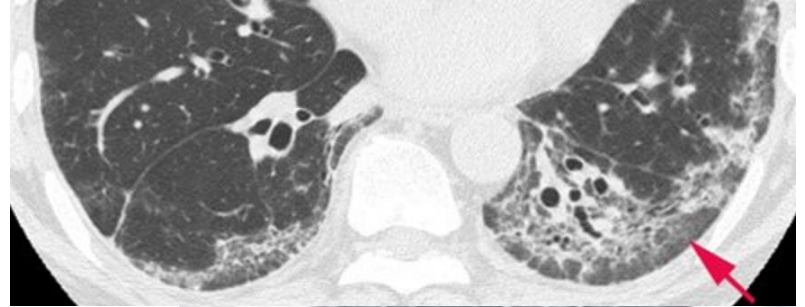
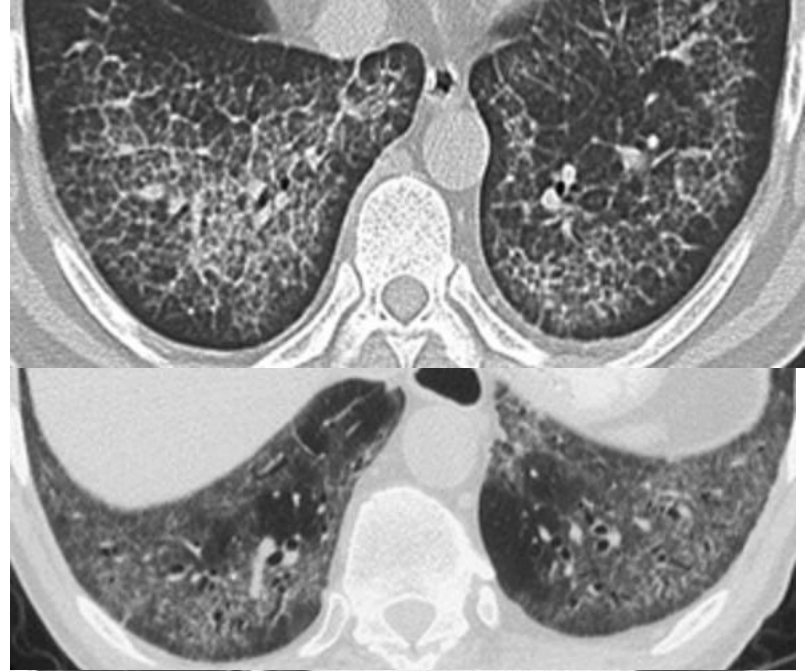
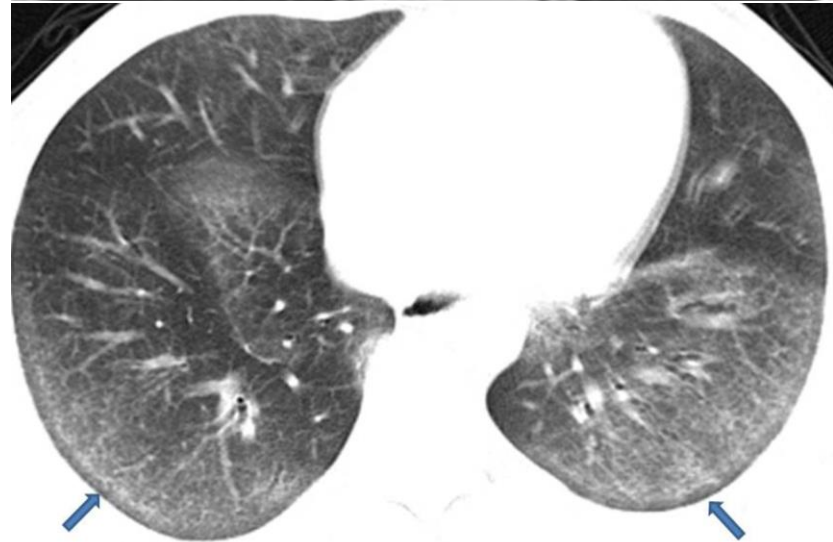


Peribronşiyoler tutulum

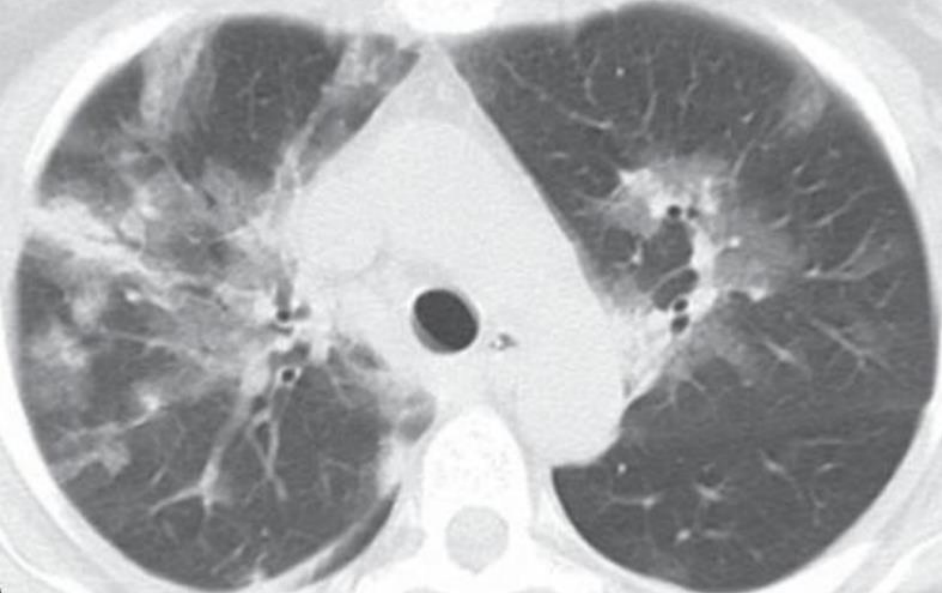


Subplevral
Periferik
tutululum

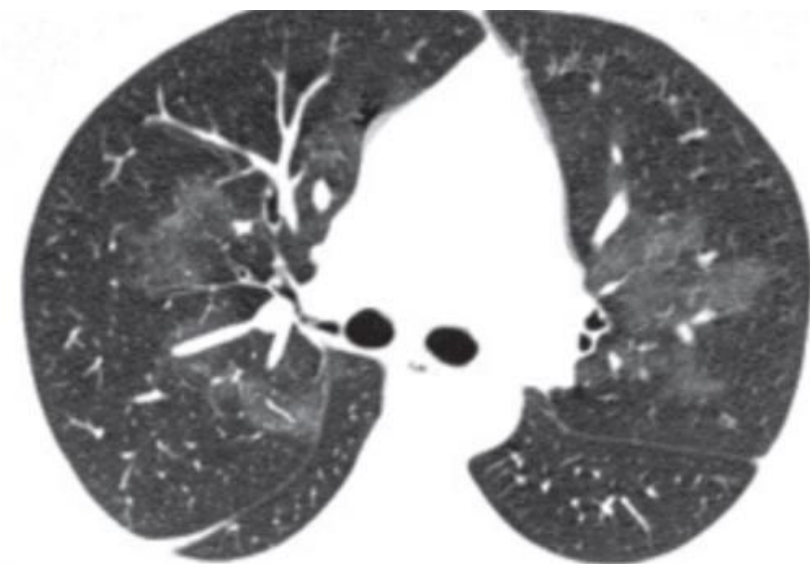
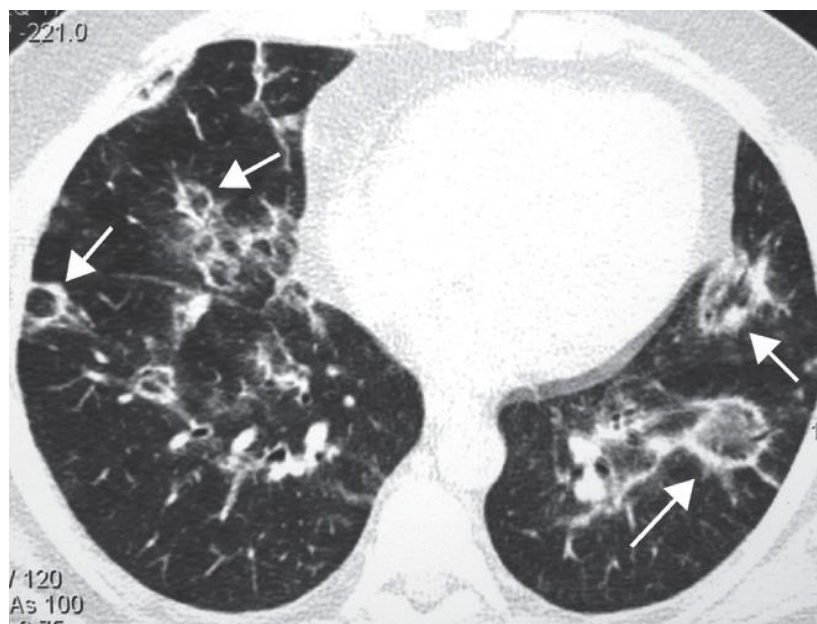
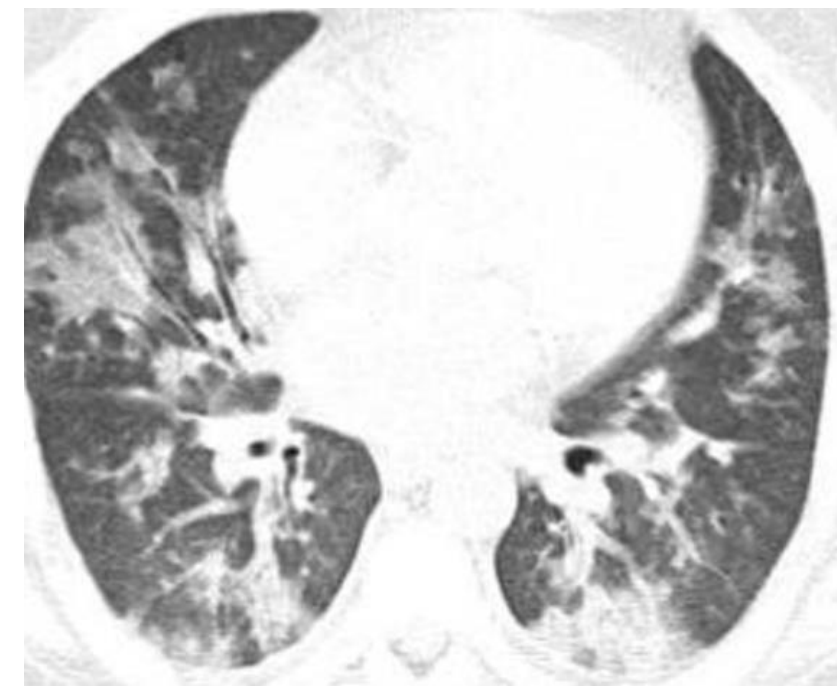
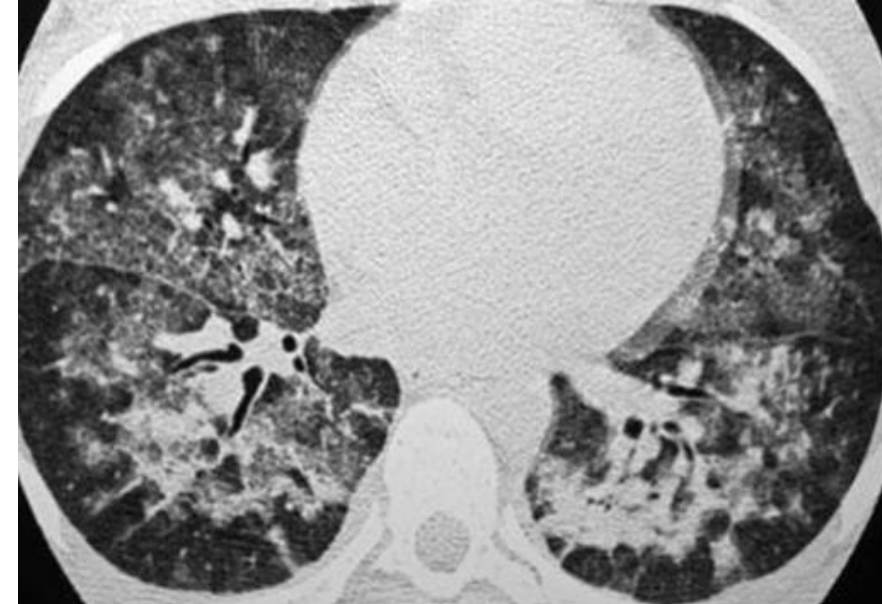




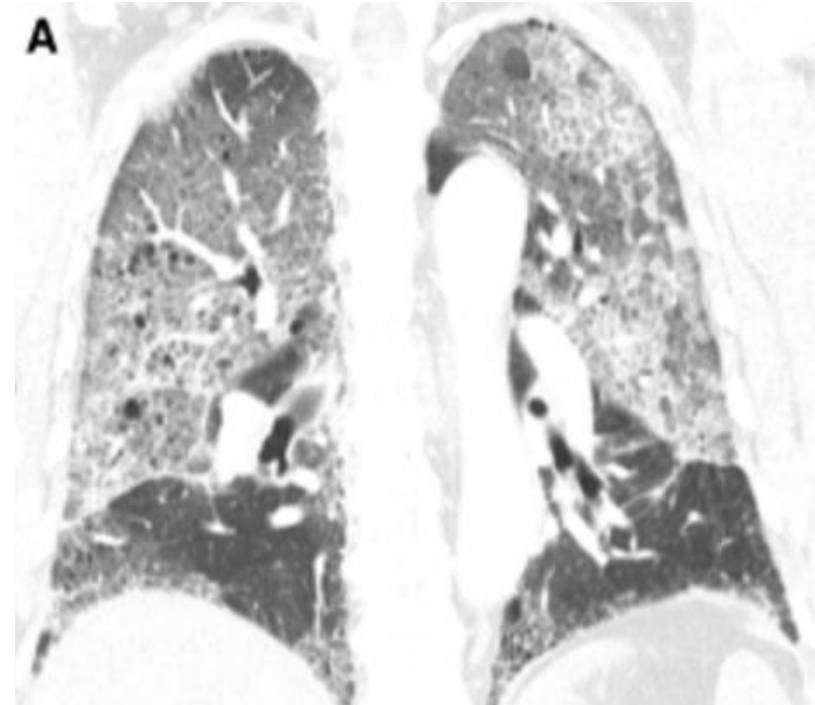
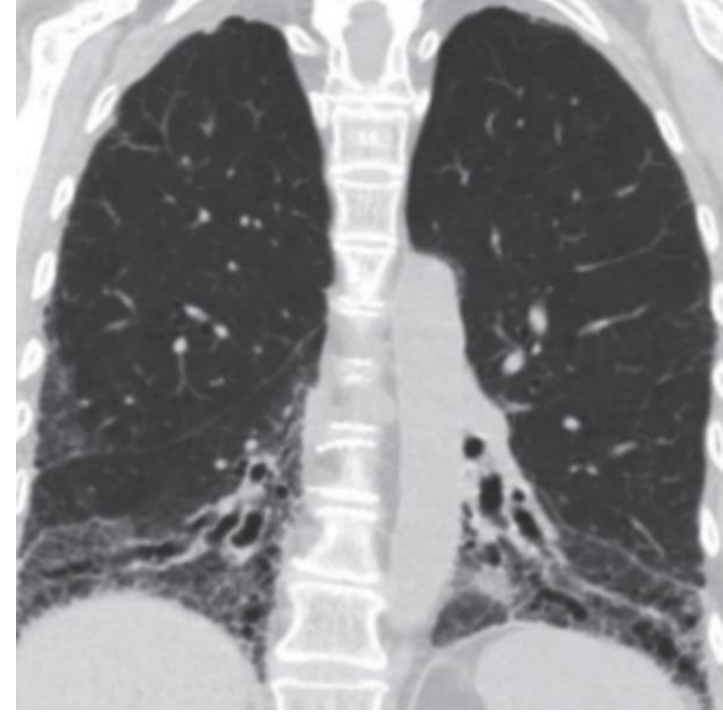
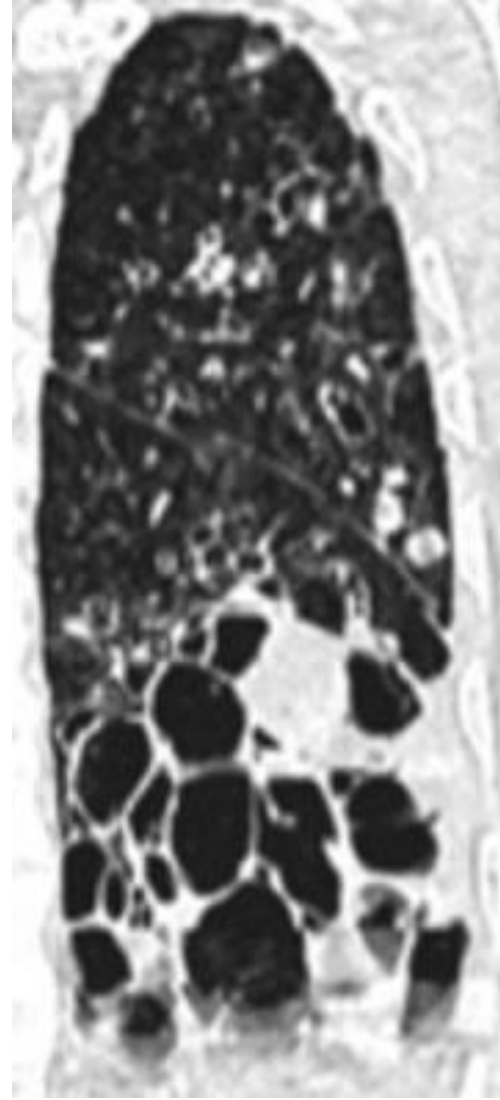
Subpleural sparing

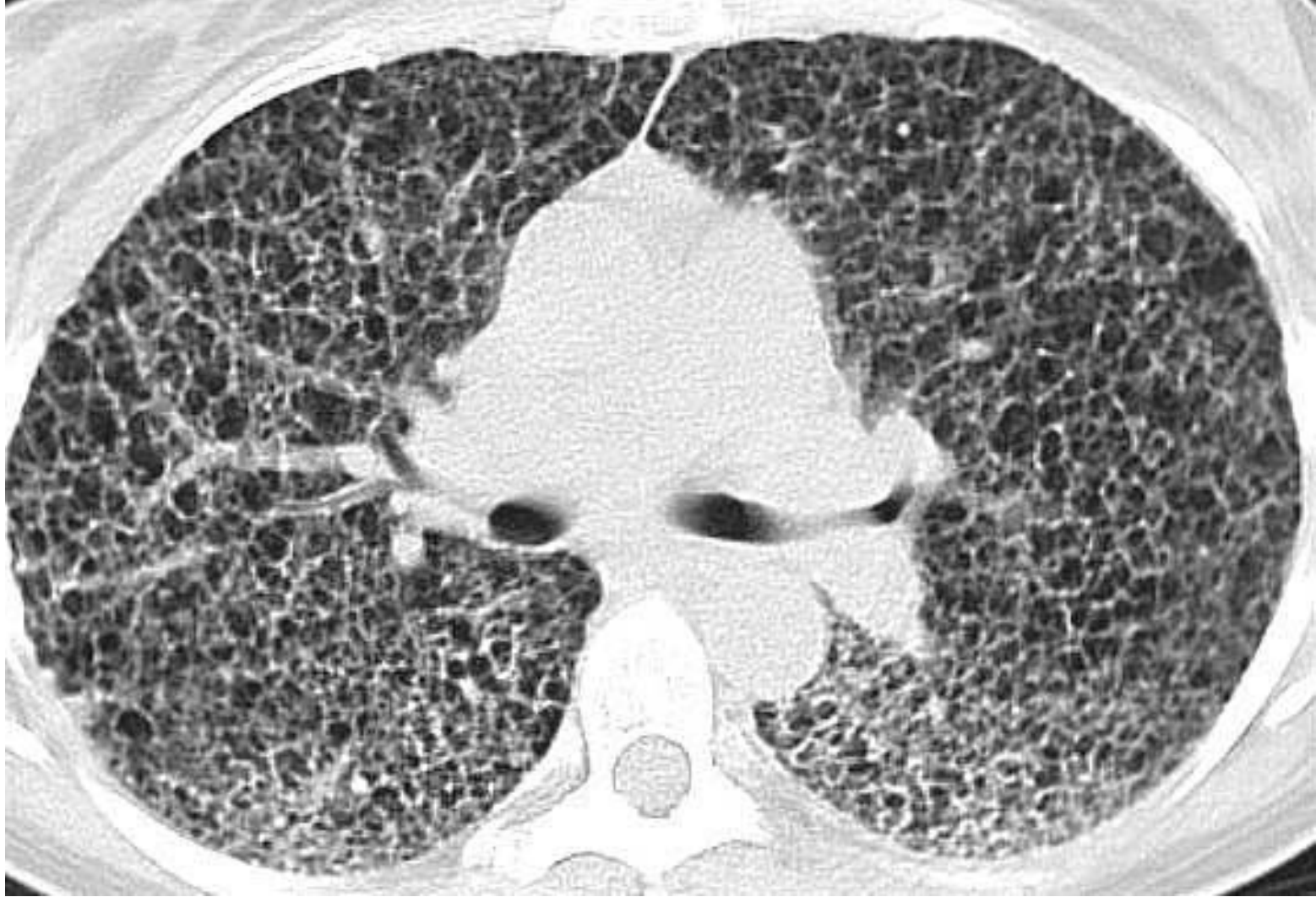


Santral Tutulum



Üst ve alt zon
tutulumu





Yüksek rezolüsyonlu bilgisayarlı tomografi (HRCT)

Laboratuvar testler

- Hemogram, Karaciğer testleri, LDH, Kreatinin, sedimentasyon
 - Eozinofili; KEP
 - Lökopeni; sarkoidoz, KVH, lenfoma
 - Karaciğer testleri; sarkoidoz, amiloidoz,
 - Renal testler; KVH, WG, Goodpasture
 - Fe eksikliği anemisi; DAH
 - Sedimentasyon: KDH, malignite
- İdrar sedimenti
 - KVH, Wegener, mikroskopik polianjitis, Goodpasture
- ANA, RF
 - IIP, KDH (IPF'de %20)
- EKG, EKO
 - Sağ kalp yüklenmesi, kor pulmonale

Laboratuvar testler

- ANCA C, WG
- ANCA P, Mikroskopik polianjitis
- Anti-GBM, Goodpasture
- Presipitan antikor, HSP
- RF (IgG), ANA, RA
- Scl-70, PM-Scl, PSS
- ds-DNA, ANA, Lupus antikoagulan, SLE
- U1-RNP, Miks KDH
- Jo-1, DM/PM
- R0 (SS-A), La (SS-B), RF(IgM), Sjögren
- ACE, sarkoidoz

Solunum fonksiyon testleri

- **Restriktif bozukluk**

- FVC, FEV1, TLC, RV azalır

- **Obstruktif patern**

- FEV1/FVC, FEV1 azalır

- **Diffüzyon**

- DLCO azalır
- DLCO'da artabilir DAH (SLE)

- **Arter Kan Gazları**

- Hipoksemi, hipokarbi, P(A-a)O₂ artar, egzersizde belirgin

Egzersiz testleri

- **Max VO₂ (oksijen tüketimi)**
 - Cihaz gereksinimi
 - Randevu
 - Kompleks
 - Kardiyak/respiratuvar ayırımı yapabilir
- **6 dk yürüme testi**
 - Çok pratik

Tanı, ayırıcı tanı

- Klinik
 - Efor dispnesi, öksürük, çabuk yorulma
 - Çomak parmak, subskapuler inspiratuvar raller, siyanoz
- Risk Faktörleri
 - Sigara, çevresel ve mesleki maruziyet, ilaç kullanımı, radyoterapi, otoimmün hastalık, vaskülit, sarkoidoz, idiyopatik
- Görüntüleme
 - HRCT, PA grafi, kontrastlı BT
- SFT
 - FVC ve DLCO
- Laboratuvar
 - Otoantikorler, vaskülit belirteçleri
- Örnekleme, Histo-sitopatoloji
 - Bronkoskopi, torakoskopi
- Konsültasyon, MDK



Progressif Pulmoner Fibrozis

- Radyolojik olarak akciğer fibrozisi bulguları olan **İPF dışında İAH tanılı bir hastada**, başka bir nedenle açıklanamayan aşağıdaki üç bulgudan en az ikisinin son bir yıl içinde ortaya çıkmasıdır
 - **Solunumsal semptomlarda kötüleşme**
 - **Hastalığın fizyolojik olarak kötüleştiğini gösteren aşağıdakilerden biri**
 - FVC'de %5 veya daha fazla düşme
 - DLCO'da (Hb'ne göre düzeltilmiş) %10 veya daha fazla düşme
 - **Progresyonun radyolojik kanıtları**
 - Traksiyon bronsektazi/bronşiolektazilerin şiddet veya yaygınlığının artması
 - Traksiyon bronşektazileriyle beraber yeni buzlu cam opasiteler
 - Yeni ince retikülasyonlar
 - Retiküler anormalliklerin yaygınlığının veya kalınlığının artması
 - Yeni veya artmış balpetekleşme
 - Artmış lobar volüm kaybı



Kasım 2016



Nisan 2019

Progrese hastalık

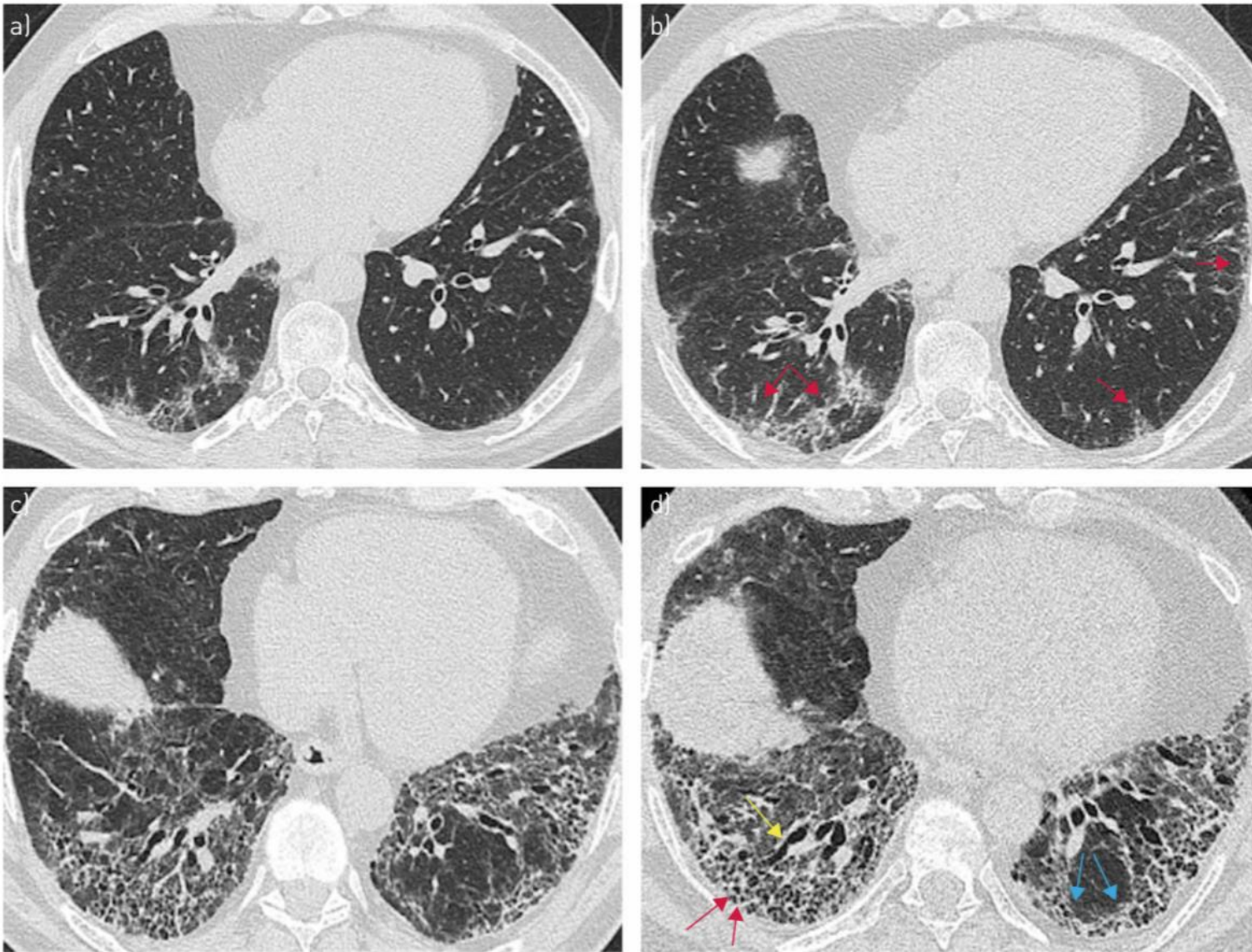


FIGURE 1 Axial computed tomography (CT) images a) at baseline and b) at a 12-month follow-up scan, in a patient with idiopathic pulmonary fibrosis (IPF). b) Note more progressive reticulation in the right lower lobe with new areas of subpleural reticulation in the left lower lobe (red arrows). Axial CT images in another patient with IPF c) at baseline and d) at a 12-month follow-up scan. d) Note the progressive honeycombing (red arrows), traction dilation of the airways (yellow arrow) and reticulation in both lobes (blue arrows).

Progressif hastalık

Robbie H, et al. Eur Respir Rev 2017.

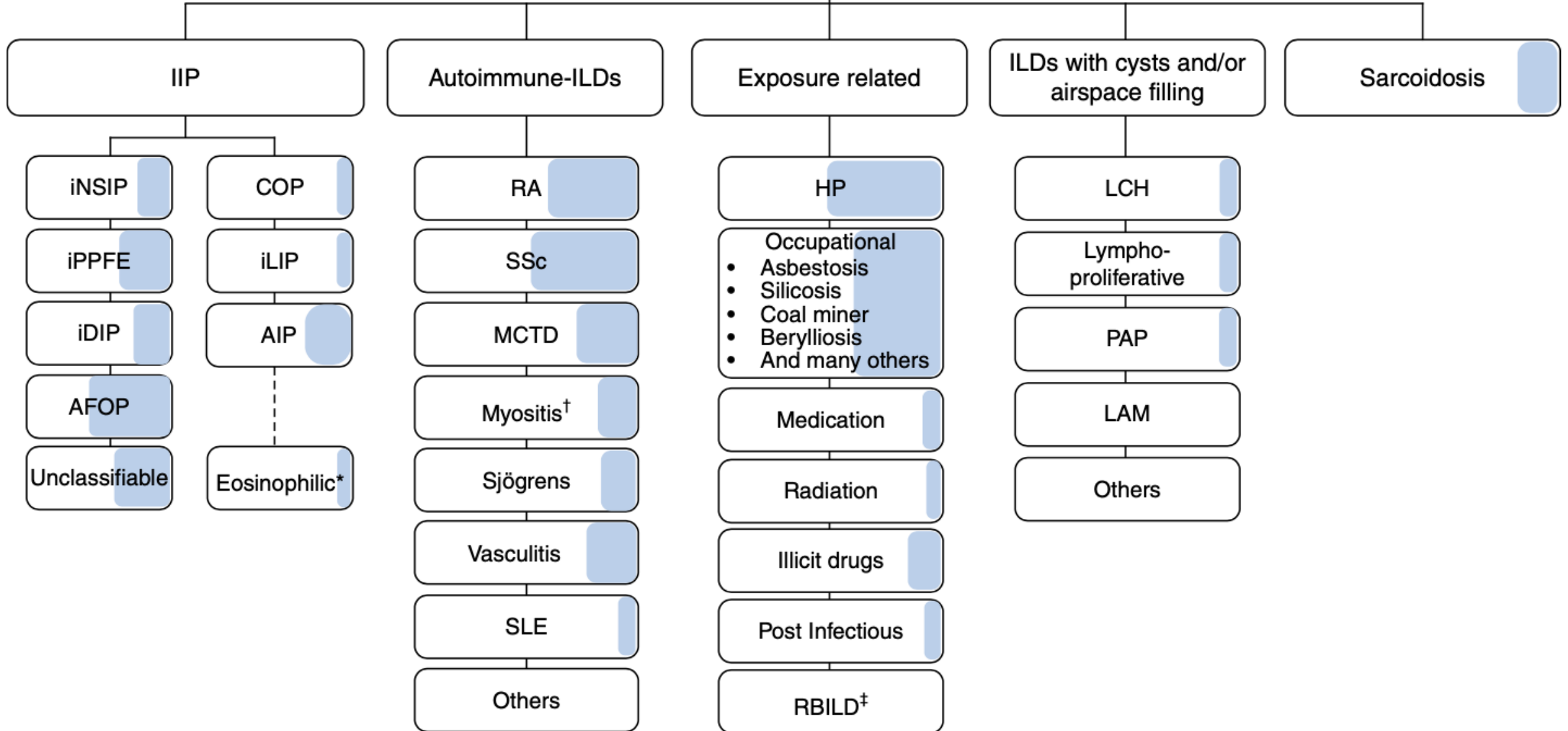
PPF'e ilerleyen fibrotik akciğer hastalıkları

Table 5. Selected Fibrotic Lung Diseases That Can Manifest Progressive Pulmonary Fibrosis

Potentially Fibrotic Interstitial Lung Diseases	Histologic Patterns
Idiopathic F-NSIP	<ul style="list-style-type: none"> • F-iNSIP (179)
PPFE	<ul style="list-style-type: none"> • IAFE (179) • May coexist with other patterns such as UIP in patients with other forms of concomitant ILD (e.g., IPF) (180)
FOP	<ul style="list-style-type: none"> • Cicatricial organizing pneumonia (181) • Organizing pneumonia with concomitant interstitial fibrosis (sometimes secondary to diffuse alveolar damage/acute interstitial pneumonia) (179, 182)
DIP	<ul style="list-style-type: none"> • DIP*
Fibrotic CTD-related ILD	<ul style="list-style-type: none"> • F-NSIP, FOP, UIP (use histopathological criteria for idiopathic diseases [179])
Fibrotic HP	<ul style="list-style-type: none"> • HP and probable HP (138) • Fibrotic element may be that of UIP, F-NSIP, or bronchiolocentric fibrosis
Fibrotic occupational ILD	<ul style="list-style-type: none"> • Dependent on occupational lung disease (asbestosis, fibrotic HP, silicosis, pneumoconiosis, or other) (183)
Fibrotic LCH	<ul style="list-style-type: none"> • F-LCH (184)
Fibrotic sarcoidosis	<ul style="list-style-type: none"> • Discrete nonnecrotizing granulomas with a lymphatic distribution with coexistent fibrosis (185)
Unclassified fibrotic ILD	<ul style="list-style-type: none"> • Cases should ideally be termed “unclassifiable” only after multidisciplinary discussion. Most cases represent combined or overlapping patterns of classifiable interstitial pneumonias, and these should be reported as such (179)
Other	<ul style="list-style-type: none"> • Fibrosis in association with inborn errors of metabolism, surfactant protein disorders, pulmonary involvement by systemic disorders, or others

Definition of abbreviations: CTD = connective tissue disease; DIP = desquamative interstitial pneumonia; F-LCH = fibrotic Langerhans cell histiocytosis; F-iNSIP = fibrotic idiopathic nonspecific interstitial pneumonia; F-NSIP = fibrotic nonspecific interstitial pneumonia; FOP = fibrosing organizing pneumonia; HP = hypersensitivity pneumonitis; IAFE = intraalveolar fibrosis and elastosis; ILD = interstitial lung disease; IPF = idiopathic pulmonary fibrosis; LCH = Langerhans cell histiocytosis; PPFE = pleuroparenchymal fibroelastosis; UIP = usual interstitial pneumonia.
*Terminology for fibrotic interstitial pneumonias with DIP-like features is controversial, this overlapping with F-NSIP.

Interstitial Lung Diseases (ILDs) other than Idiopathic Pulmonary Fibrosis (IPF)

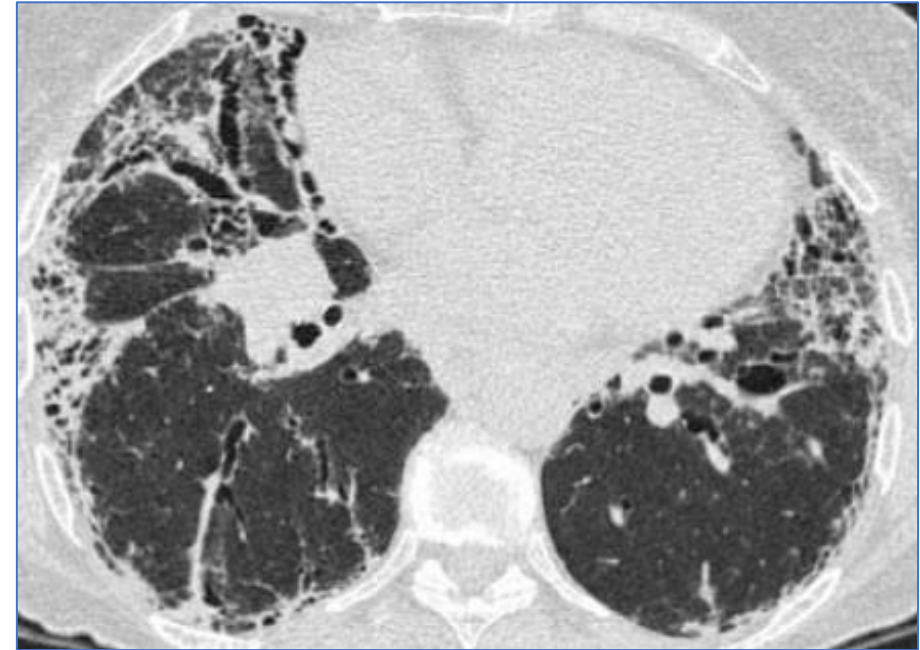
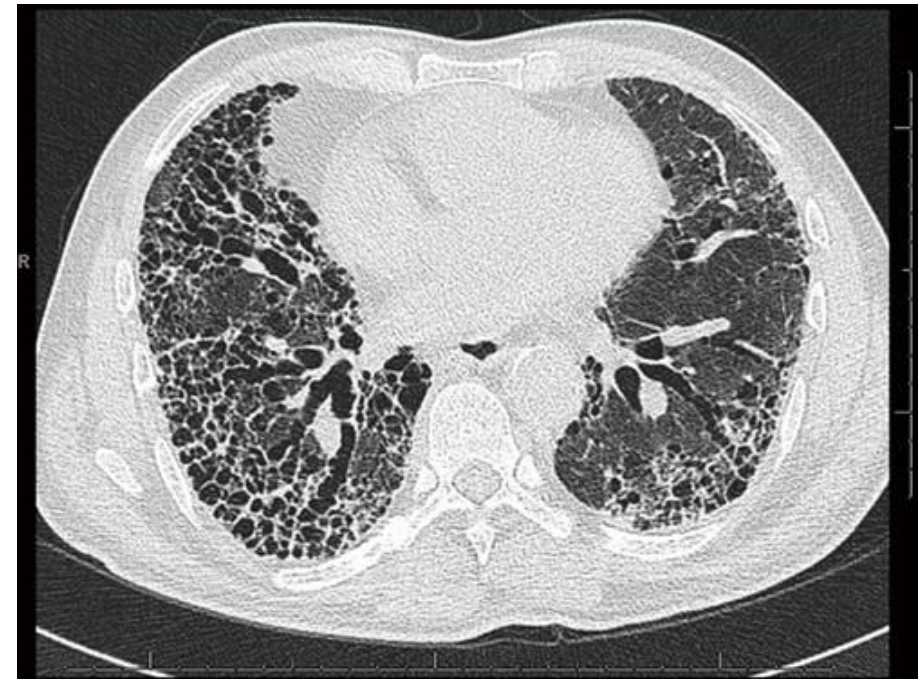
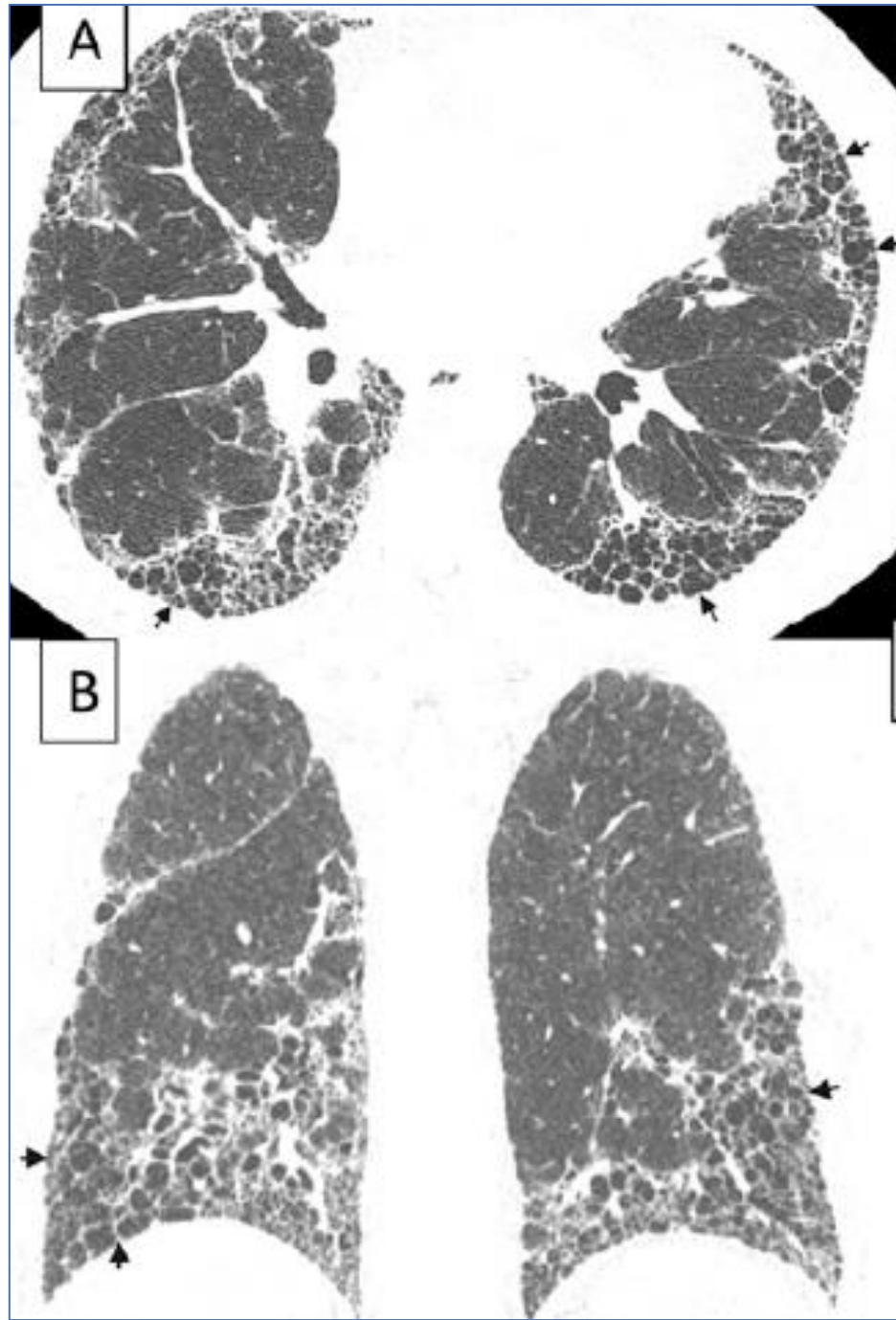


Fibrozisin radyolojik paternleri ve nedenleri

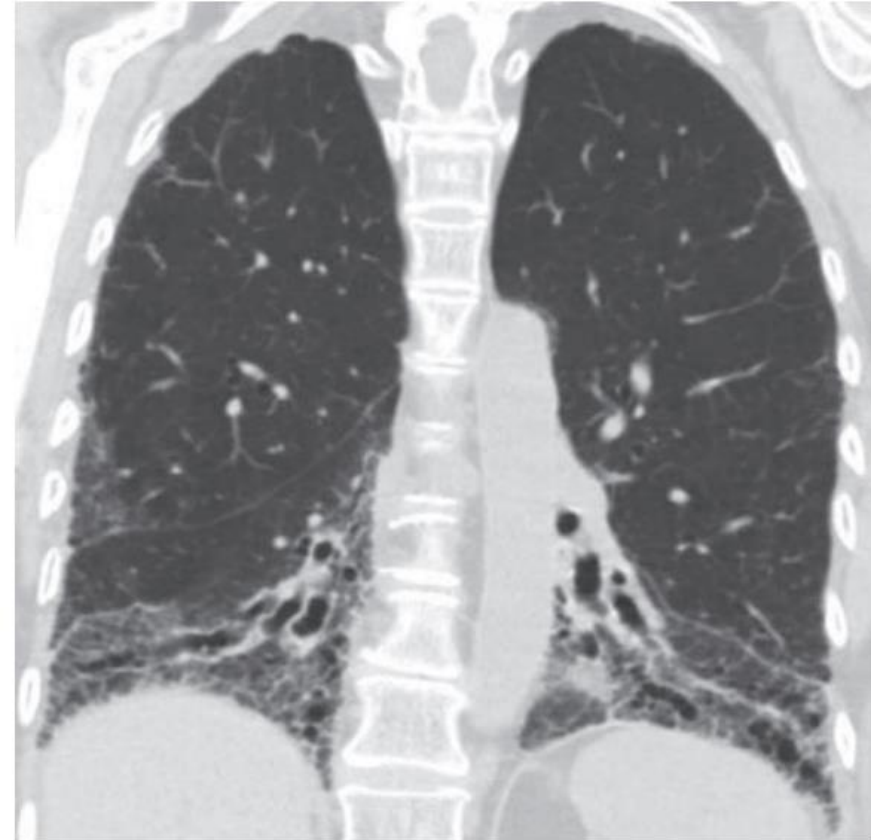
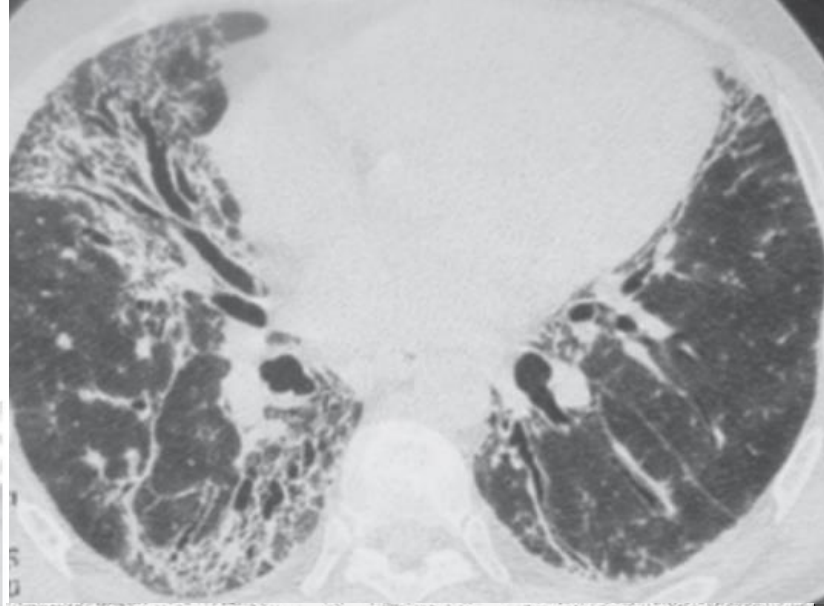
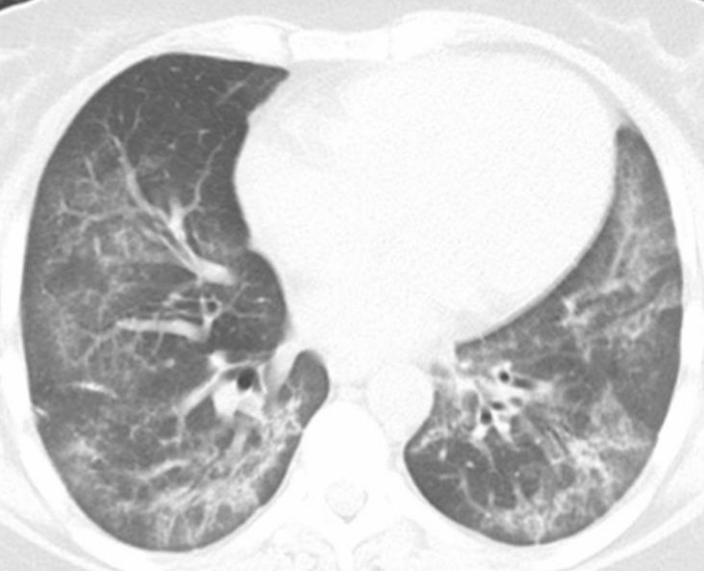
- UIP
 - IPF, RA, HSP, SS, Sarkoidozis, Asbestozis
- NSIP
 - KDH, SS, maruziyet ve tedaviyle ilgili fibrozis, iNSIP, HSP
- OP
 - COP, maruziyet ve tedaviyle ilişkili, KDH, dermatomyozit, HSP, vaskülit, malignite

UIP

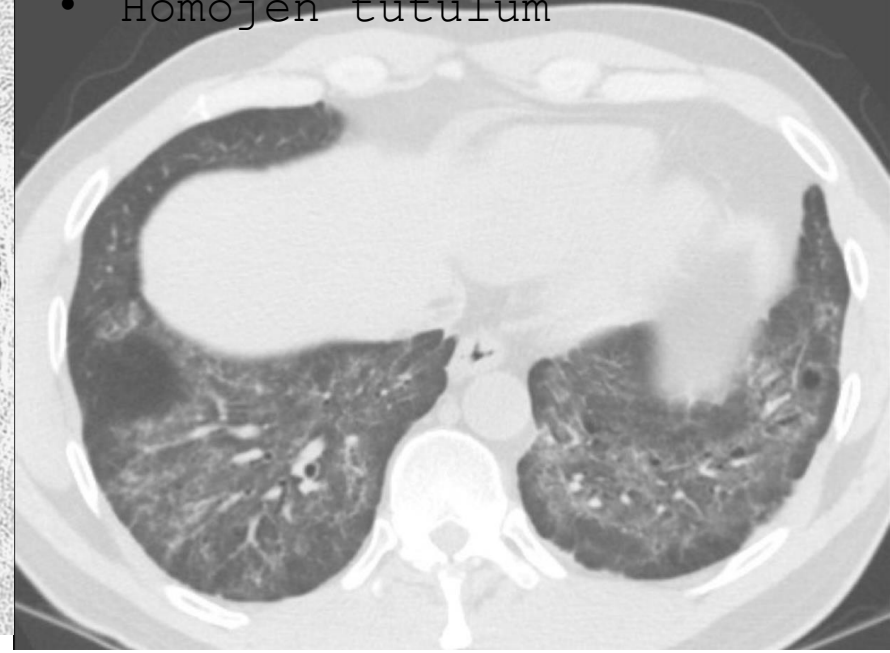
- Subplevral, bazal tutulum
- Retiküler opasiteler
- Bal peteđi \pm traksiyon bronşektazi
- Uyumsuz bulguların olmaması

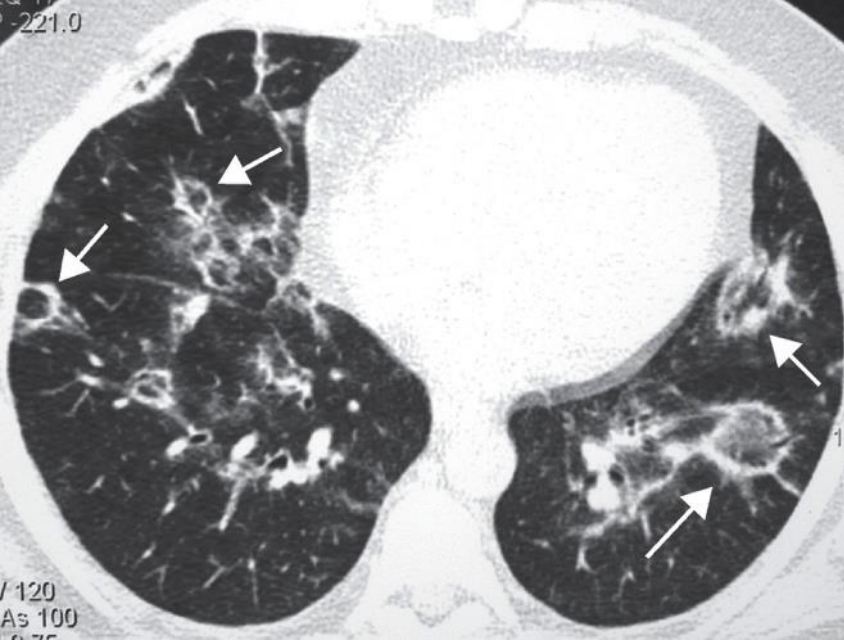


NSIP



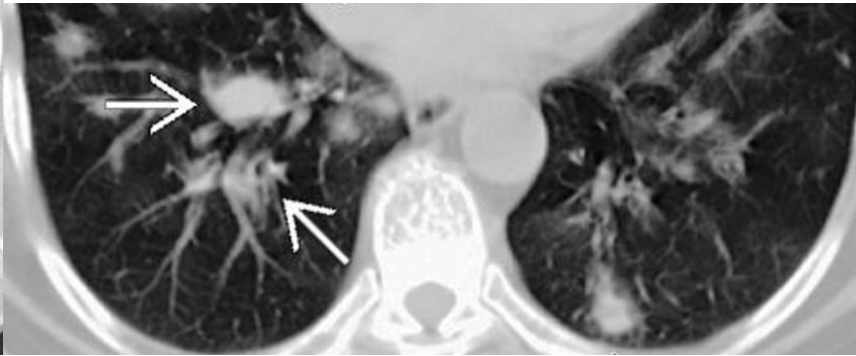
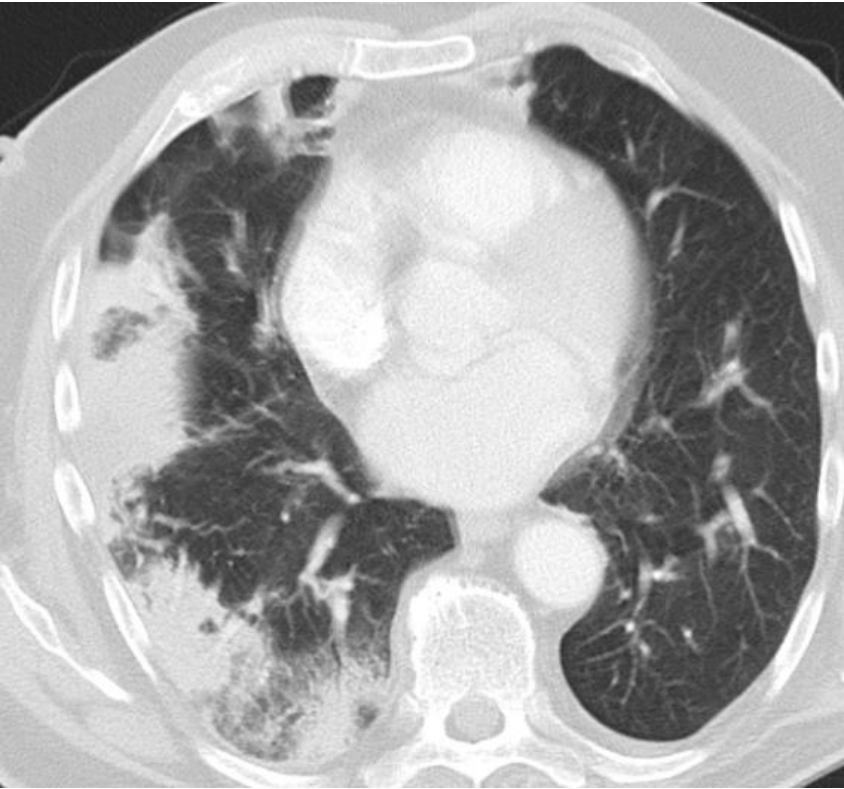
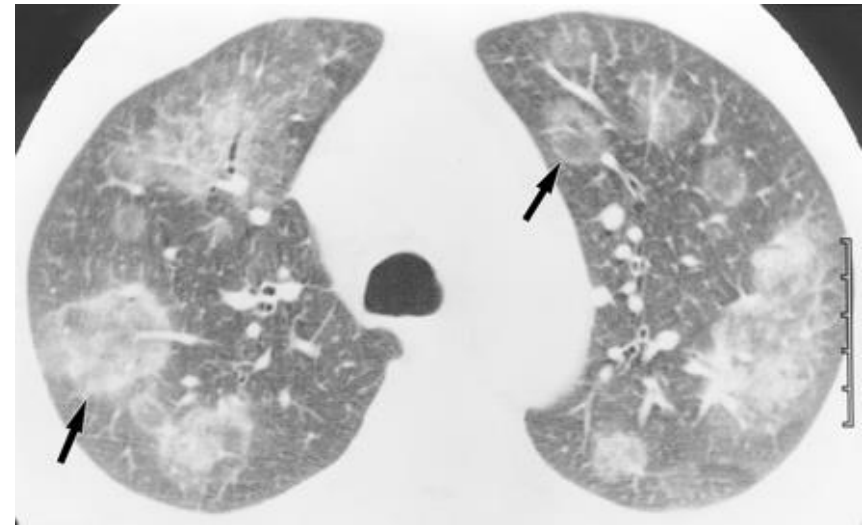
- Peribronkovasküler tutulum
- Yamalı buzlu cam alanlar
- Retiküler opasiteler
- Subplevral alanların korunması
- Bazal simetrik tutulum, volüm kaybı
- Mikrokistik balpeteği
- İntralobüler septal kalınlaşmalar
- Traksiyon bronşektazileri
- Homojen tutulum





OP

- Yamalı konsolidasyon alanları, bazen subplevral yerleşimli
- Spiküler, multipl, nodüller
- Ters halo belirtisi
- Retiküler opasiteler, fibrozis



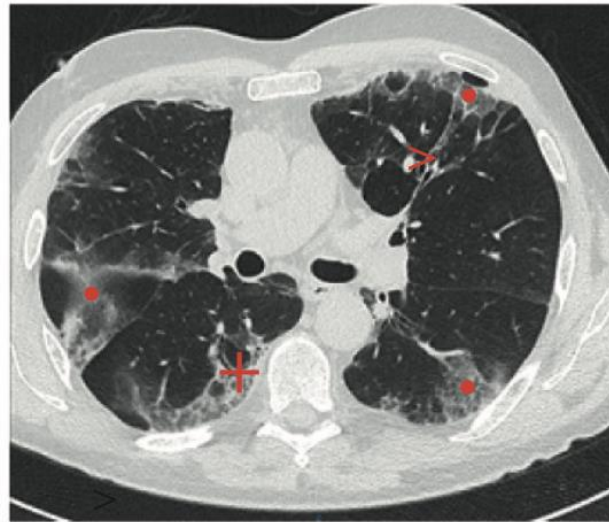
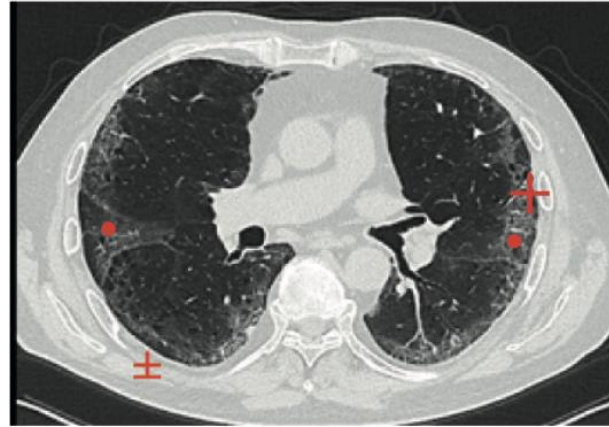
Usual interstitial pneumonia

Typical image



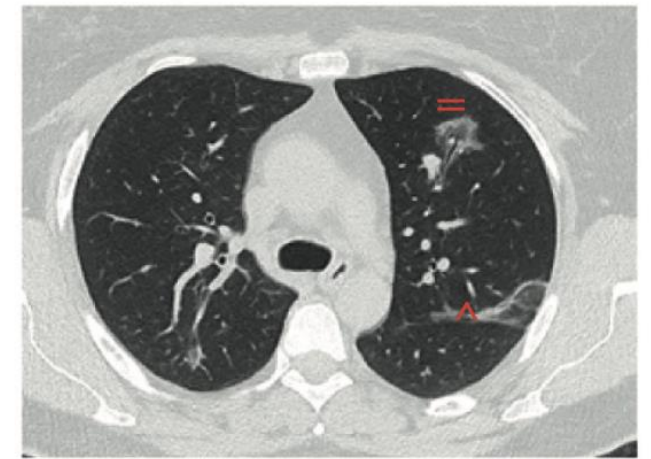
Honeycombing (*) with or without peripheral traction bronchiectasis (+), in a subpleural and basal predominant, often heterogeneous, distribution.

Non-specific interstitial pneumonia

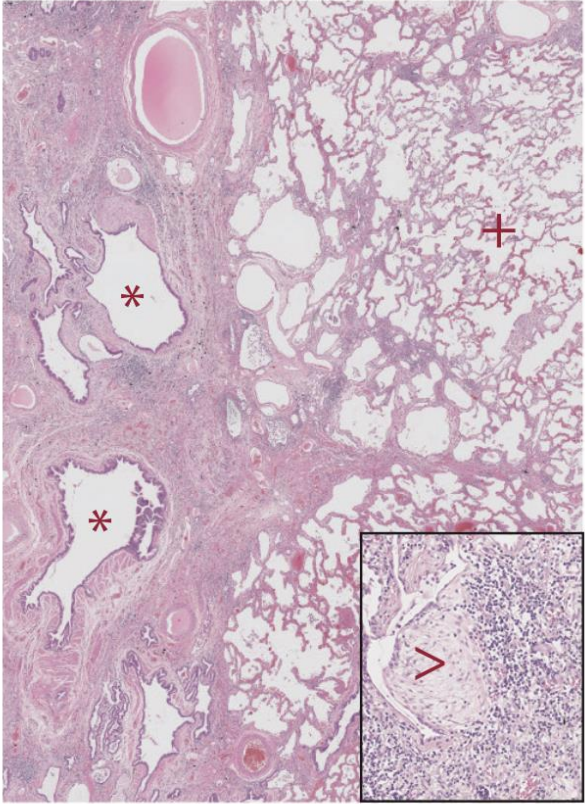
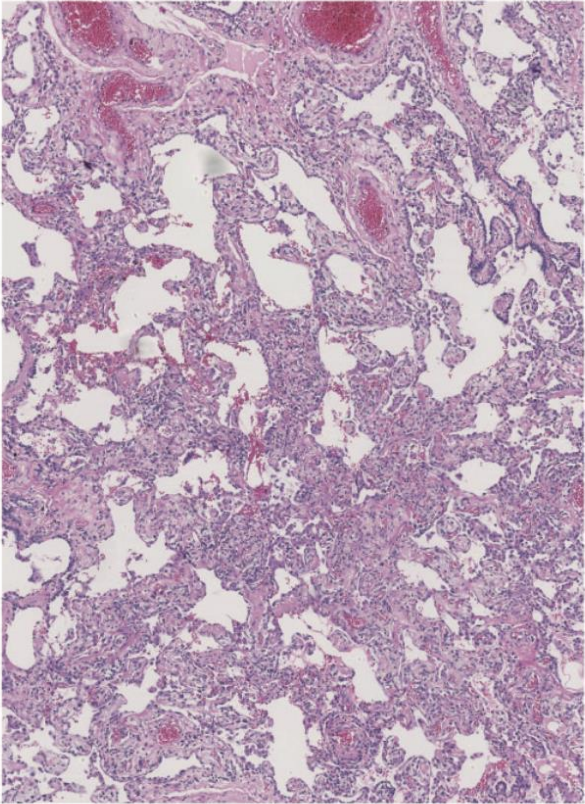
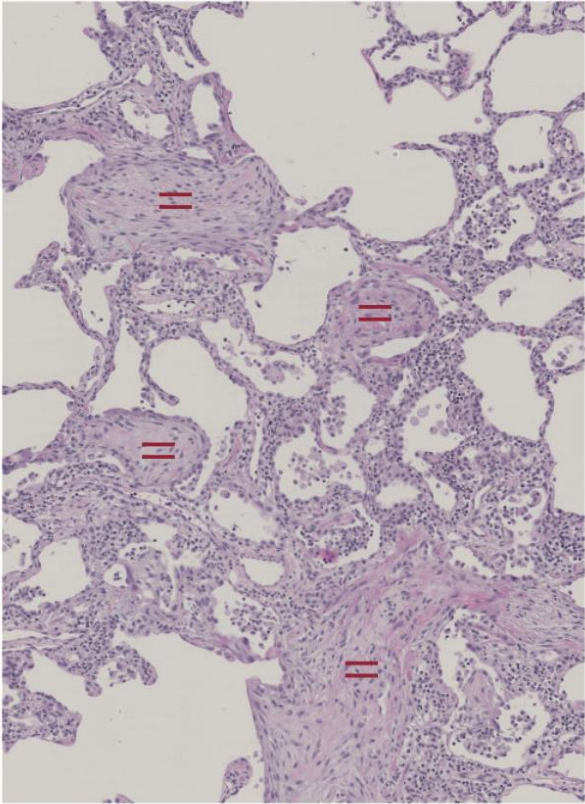


Ground-glass opacities (•) with traction bronchiectasis (+), often peribronchovascular (>) predominance with subpleural sparing (±).

Organising pneumonia



Peripheral consolidation with air bronchograms (<), bronchocentric distribution, a perilobular pattern, reversed halo sign (=), and band-like consolidations (^) can also be seen.

	Usual interstitial pneumonia	Non-specific interstitial pneumonia	Organising pneumonia
Typical pathology	 <p data-bbox="573 1110 1154 1368">Marked fibrosis, architectural distortion with or without honeycombing (*) in predominant subpleural or paraseptal distribution, presence of patchy involvement, and areas of preserved normal lung tissue (+). Presence of fibroblast foci (>) and absence of features suggesting an alternate diagnosis.</p>	 <p data-bbox="1205 1110 1786 1368">Diffuse alveolar wall thickening by uniform fibrosis (pale pink) with preservation of the alveolar architecture and mild interstitial inflammation (purple).</p>	 <p data-bbox="1837 1110 2418 1368">Patchy distribution, filling of the distal airways, and adjacent alveoli with fibromyxoid plugs (=) of granulation tissue with temporal uniformity. Relative preservation of the underlying pulmonary architecture. Mild to moderate interstitial inflammation can be present.</p>

Tedavi

- Düzenli izlem
- Risk kontrolü
- Bilinen etyolojilerin tedavisi
 - KDH, sarkoidoz, HSP
- Farmakolojik tedavi
 - Antifibrotik tedavi
- Tedaviye uyum
- Enfeksiyon kontrolü, aşılama
- Yan etkilerin yönetimi
 - DM, GIS...
- Komorbiditelerin yönetimi
 - KY, KAH, KBH, HT, DM, KOAH
- Komplikasyonların yönetimi
 - Akut alevlenme, PH,
- Semptom kontrolü
 - Dispne, öksürük..
- Destek tedavisi
 - Oksijen, NIMV, rehabilitasyon, beslenme, psikososyal
- Transplantasyon

Farmakolojik Tedavi

- **Kortikosteroidler**

- Prednizolon, metilprednizolon

- **Anti-enflamatuvar ve sitostatik ajanlar**

- Siklofosamid, azatioprin, metotreksat, mycophenolate, cellcept, sirolimus, leflunomide

- **Antifibrotik**

- Kolsişin, Pirfenidon, Nintedanip

- **İmmunmodülatör**

- Siklosporin, gama interferon, infliximab, etarnecept

- **Antioksidanlar**

- N-asetil sistein, glutation, niasin, taurine

- **Destek tedavi**

Time

Diagnosis of IPF

TREATMENT CONSIDERATIONS

PHARMACOLOGICAL

- Nintedanib
- Pirfenidone

NONPHARMACOLOGICAL

- Oxygen supplementation (if hypoxemic)
- Pulmonary rehabilitation

COMORBIDITIES

- Pulmonary hypertension
- Gastroesophageal reflux
- Obstructive sleep apnea
 - Lung cancer

SYMPTOM CONTROL

- Palliative care

If increased risk of mortality, evaluate for lung transplantation at diagnosis

MONITOR FOR DISEASE PROGRESSION

Consider pulmonary function testing and the 6-minute-walk test every 4–6 months or sooner if clinically indicated

Consider annual HRCT if there is clinical suspicion of worsening or risk of lung cancer

Consider an HRCT if there is concern for an acute exacerbation

Consider a CT pulmonary angiogram if there is a clinical concern for pulmonary embolism

ACUTE EXACERBATION

Corticosteroids

RESPIRATORY FAILURE DUE TO PROGRESSION OF IPF

Evaluate and list for lung transplantation

Palliative care

Patients should be made aware of available clinical trials for possible enrollment at all stages

Komplikasyonlar

Medical Complications



- Akut atak, akut solunum yetmezliđi
- Kronik solunum yetmezliđi
- Kardiyovasküler olaylar
- Pulmoner hipertansiyon
- Akciđer kanseri

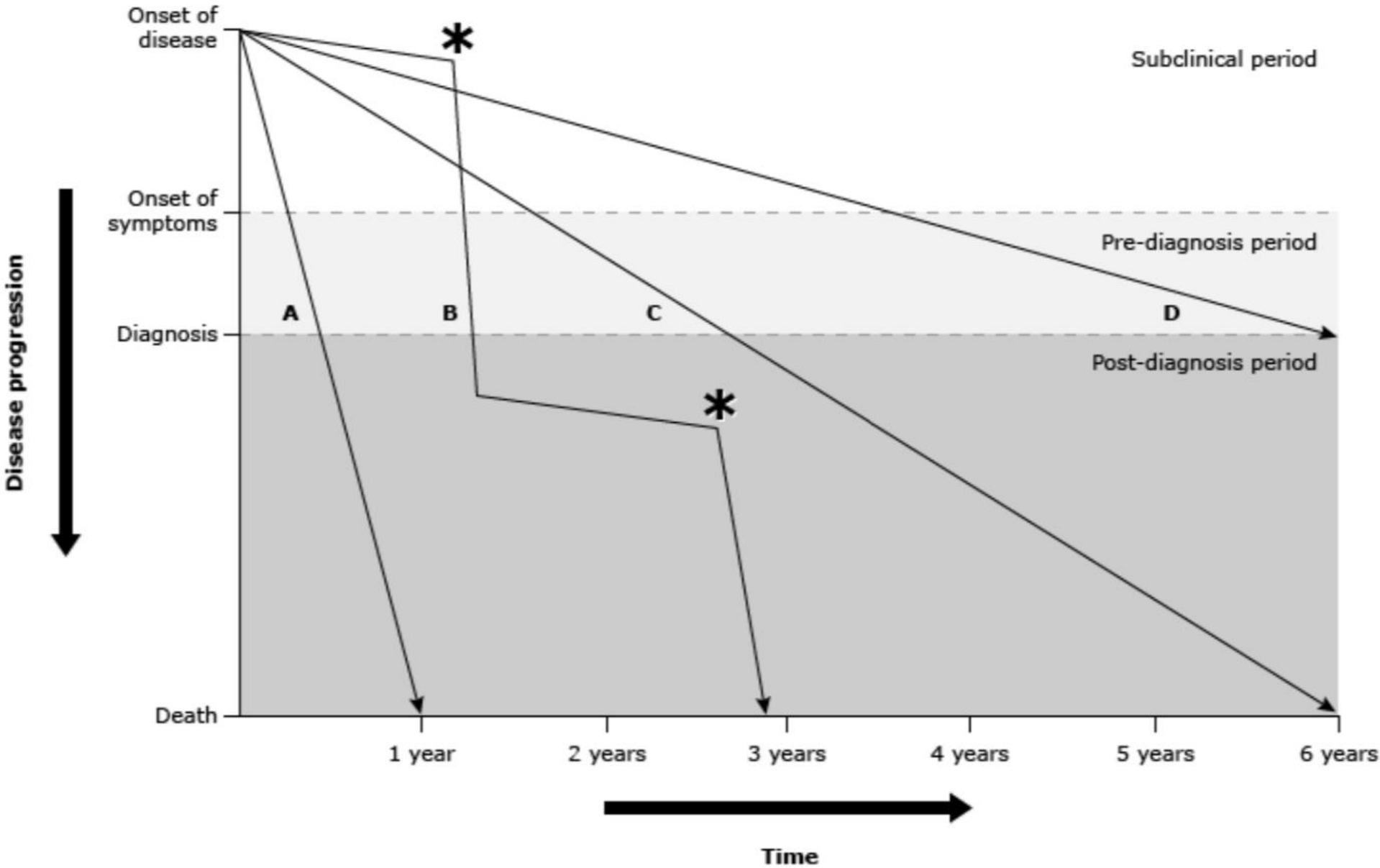
İzlem

- Semptomlarda artış, kötüleşme
- 3-6 aylık düzenli periyotlarla
- Progresyon
- Akut Alevlenme
- Komorbidite
- Komplikasyon
- İlaç yan etkisi
- Tedavi başlama
- Tedavide değişiklik
 - Ara verme
 - Doz azaltma
 - İlaç değiştirme
 - Kombinasyon
- Palyatif bakım
- Transplantasyon

İzlem Parametreleri

- Semptomlar
 - Dispne
 - Egzersiz kapasitesi
 - Öksürük
 - İlave semptomlar
- SFT
 - Spirometri, DLCO, 6DKYT
- Oksijenizasyon
 - Pulseoksimetri, AKG, Monitorizasyon
- Görüntüleme
 - Direkt Grafiler, HRCT

Schematic representation of potential clinical courses of idiopathic pulmonary fibrosis (IPF)





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