

İPF TANISINDA RADYOLOJİK BULGULARIN ÖNEMİ

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ÖZET:

- **İPF'de radyolojik bulgular**

- Tanı

- Ayırıcı tanı

- Biyopsi yeri belirleme

- Mortaliteyi etkileyen faktörleri belirleme

- İPF patogenezinde ?

} amaçlı

ATS 2018

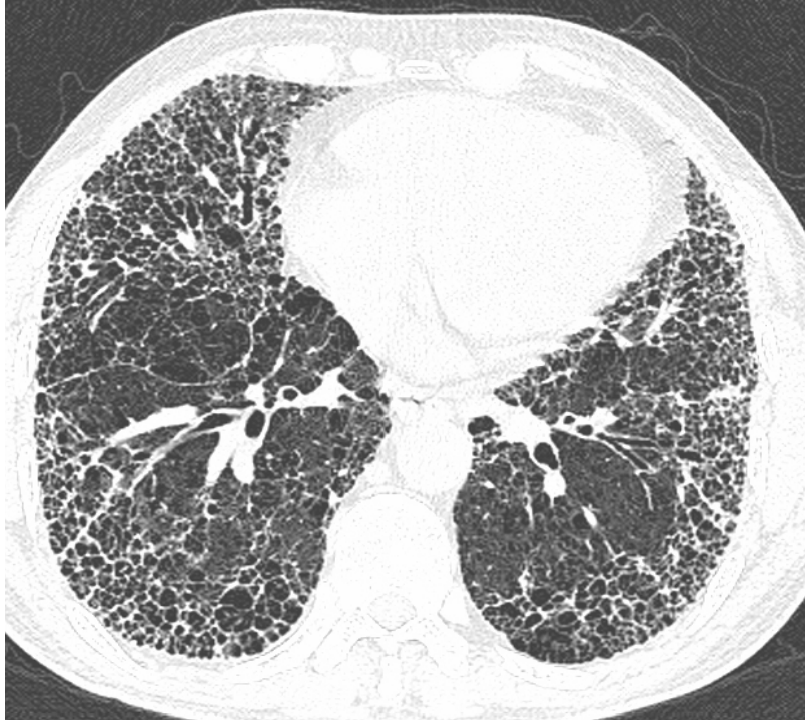
İPF KOMİTESİ: ATS, ERS, JRS, ALAT

- UIP
- Olası UIP
- Belirsiz
- Alternatif

	UIP	Olası (probable)UIP	Belirsiz	Alternatif tanı
Dağılım	<ul style="list-style-type: none"> • Bazal ve subplevral tutulum • Heterojen yerleşim • Balpeteği 	<ul style="list-style-type: none"> • Bazal ve subplevral diffüz tutulum • Heterojen yerleşim • Traksiyon bronşektazisi ile birlikte retikülasyon • İlimli buzlu cam 	<ul style="list-style-type: none"> • Subplevral ve bazal dağılım • İnce retikülasyon • BT özellikleri diğer spesifik fibrozis paternini düşündürmüyor olmalı • Erken UIP paterni 	<ul style="list-style-type: none"> • Dağılım -Üst-orta zon -Peribronşioler -Perilenfatik • Plevral plaklar • Dilate özefagus • Distal klavikuler erozyon • Yoğun LAP kümeleri • Plevral efüzyon ve kalınlaşma
-	Retiküler patern İlimli buz cam Parankimal ossifikasyon Dağılım varyasyonları: -Asimetrik -Diffuz			<ul style="list-style-type: none"> • Kistler • Mozaik atenüasyon • Nodüller • Konsolidasyon • Baskın buzlu cam

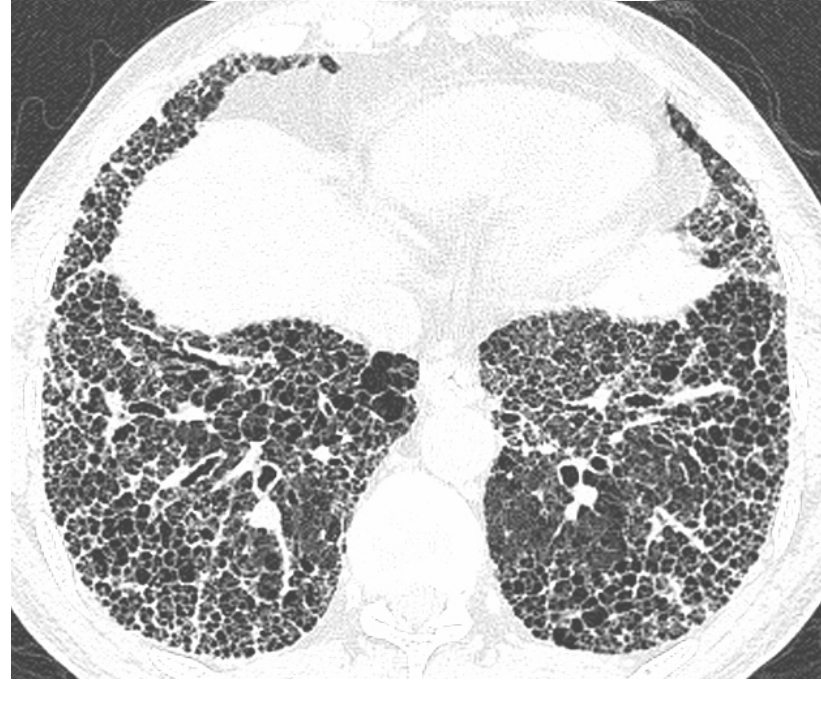
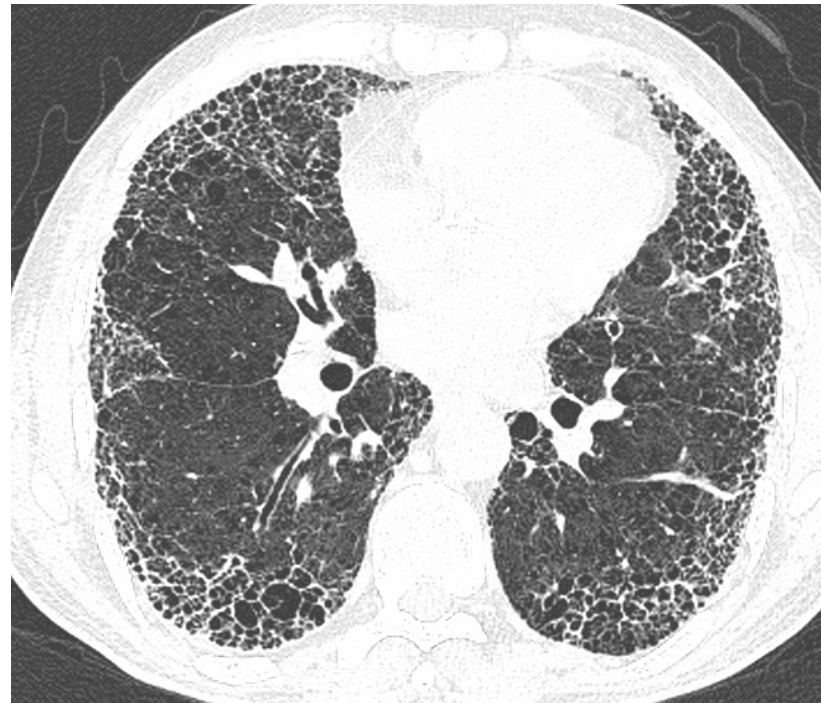
UIP PATERNİ

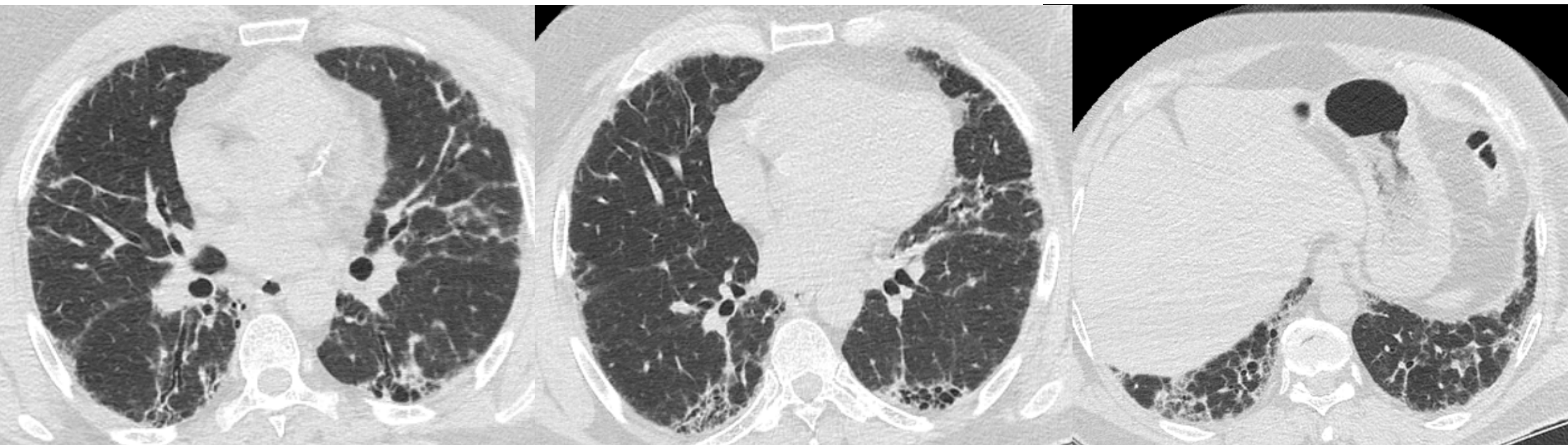
1. Subplevral /bazal ağırlıklı yerleşim
2. Retiküler patern
3. **Balpeteği görünümü** (\pm traksiyon bronşektazisi)
4. Alternatif tanıları düşündüren bulgu yok



Balpeteği
Subplevral
Bazal

UIP

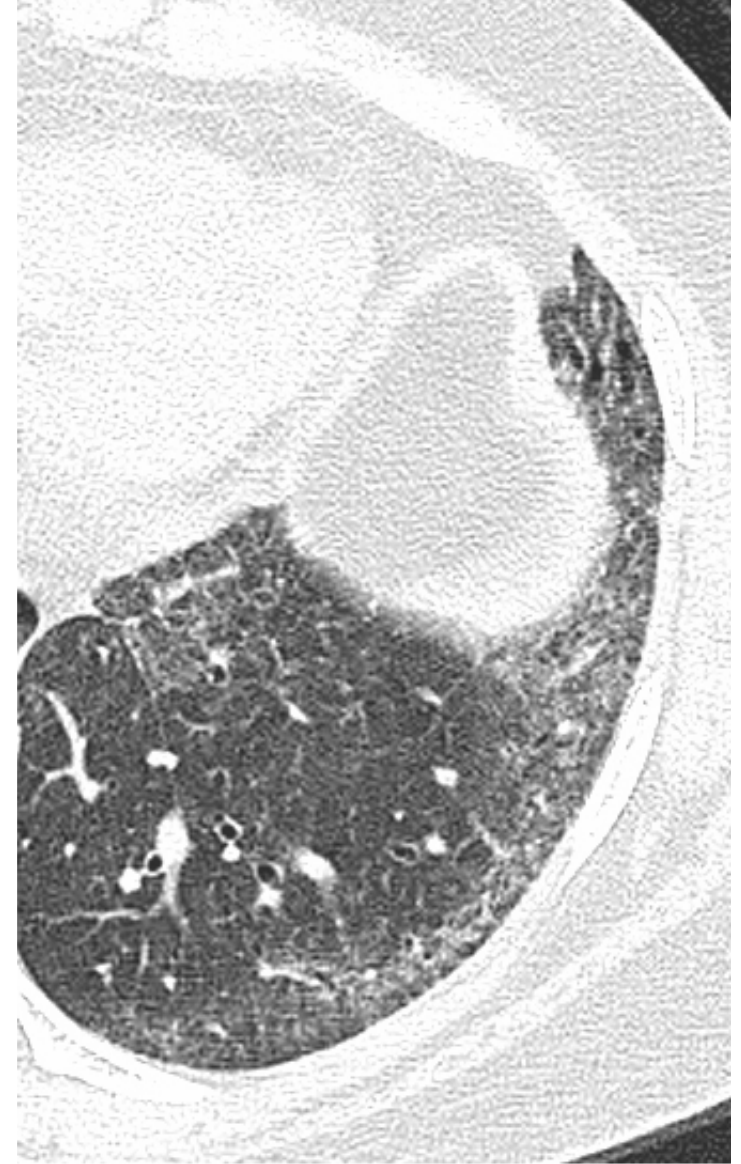




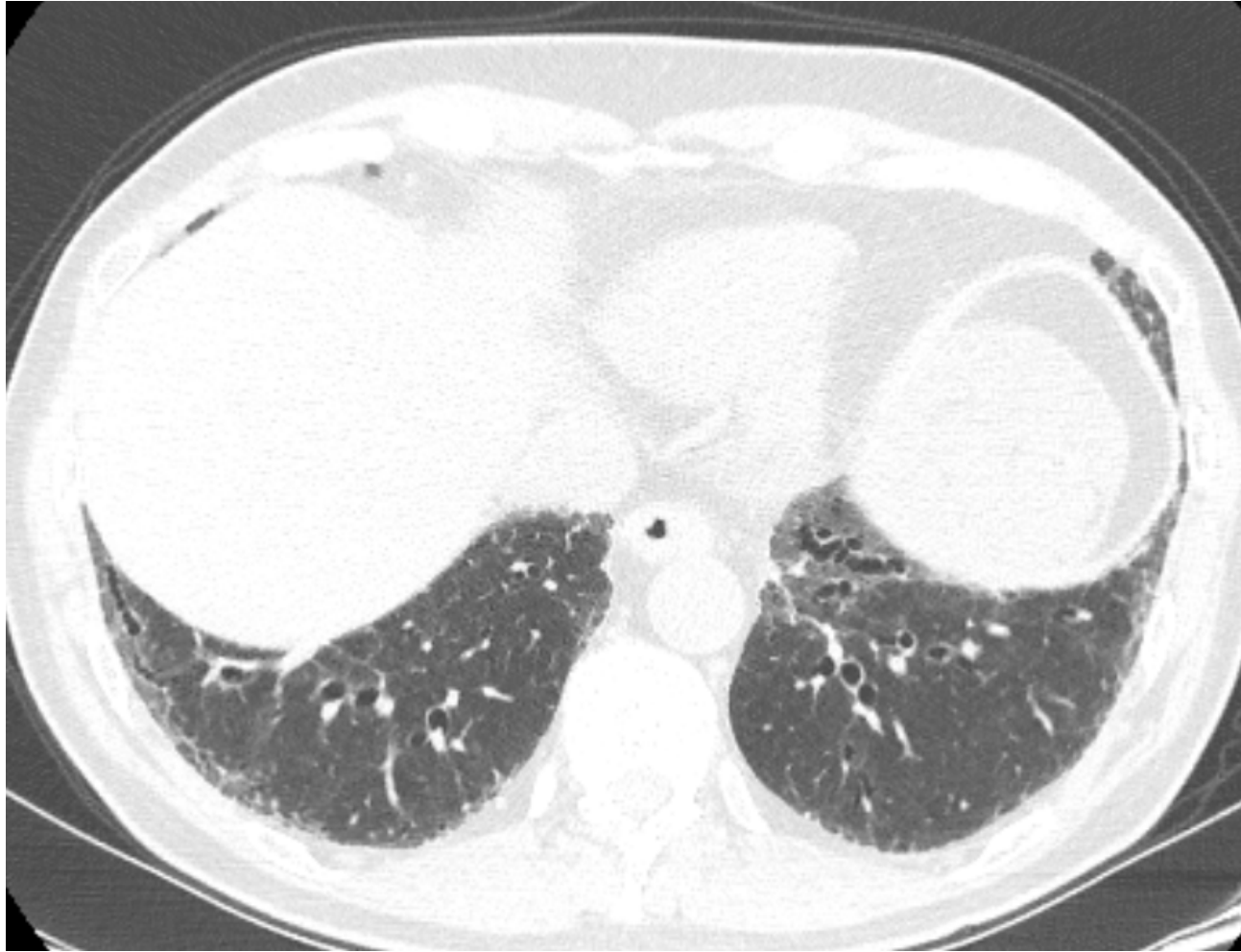
Balpeteği, subplevral, bazal : **UIP**

OLASI UIP PATERNİ:

1. Subplevral ve bazal ağırlıklı yerleşim
2. **Traksiyon bronşektazisi ve retiküler görünüm**
3. Alternatif tanıları düşündüren bulgu yok



Retikülasyon ve traksiyon bronşektazisi : **OLASI UIP**

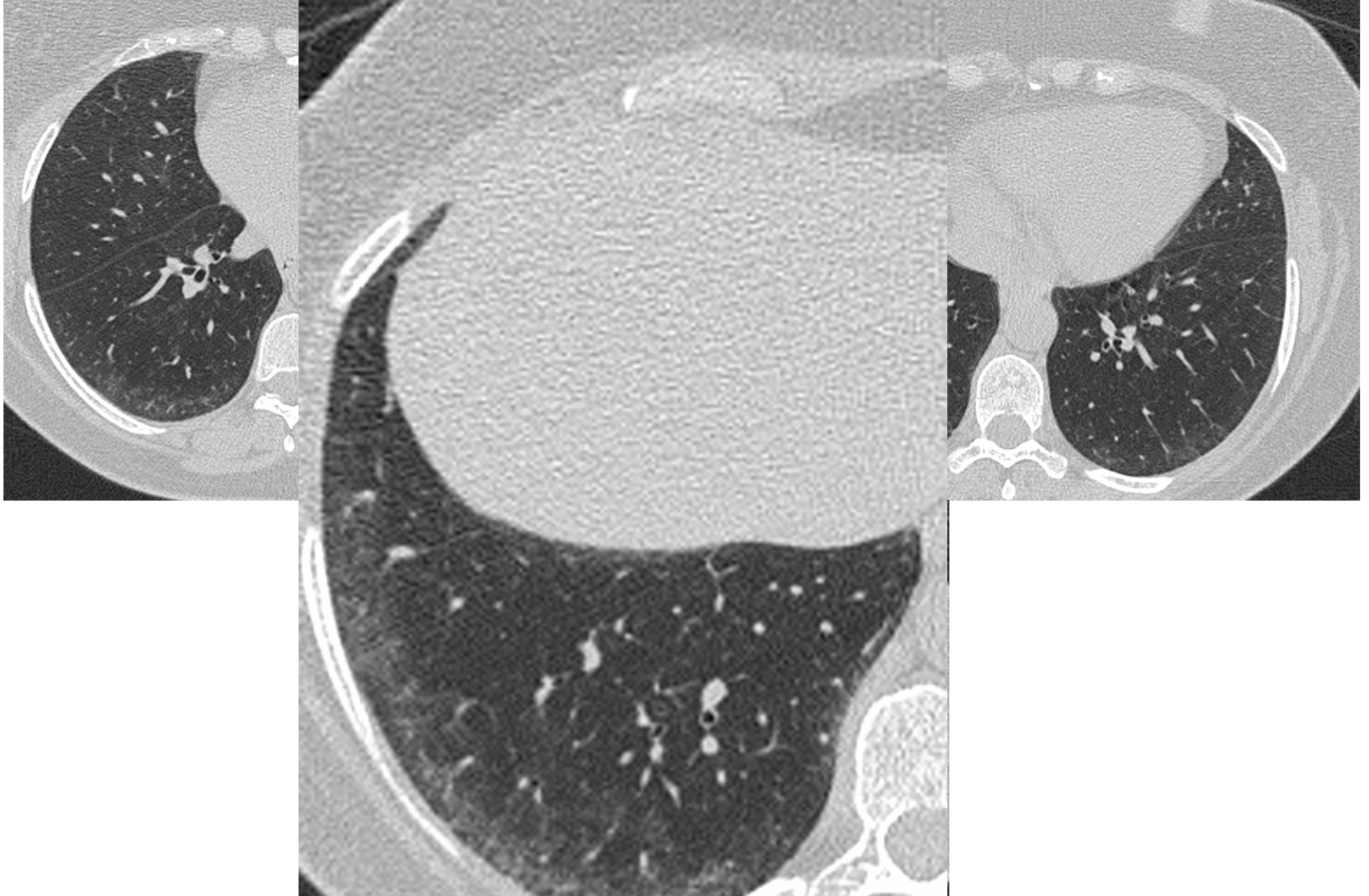


Retikülasyon ve traksiyon bronşektazisi

OLASI UIP

UIP İÇİN BELİRSİZ PATERN

1. Subplevral ve bazal dağılım
2. İnce retikülasyon (Erken UIP paterni)
3. BT özellikleri diğer spesifik fibrozis paternini düşündürtmüyor olmalı



İnce retikülasyon : **BELİRSİZ** **ERKEN UIP**



UIP İÇİN BELİRSİZ PATERN
OTO- İMMUN ÖZELLİKLİ İNTERSTİYEL PNÖMONİ
(IPAF)

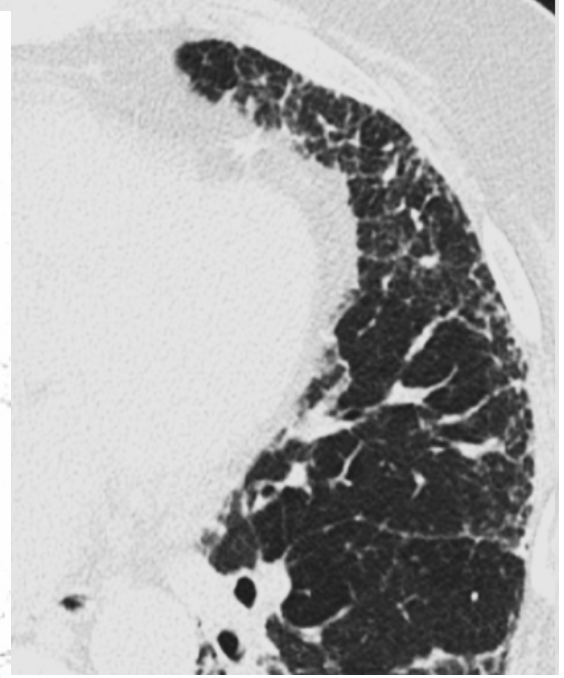
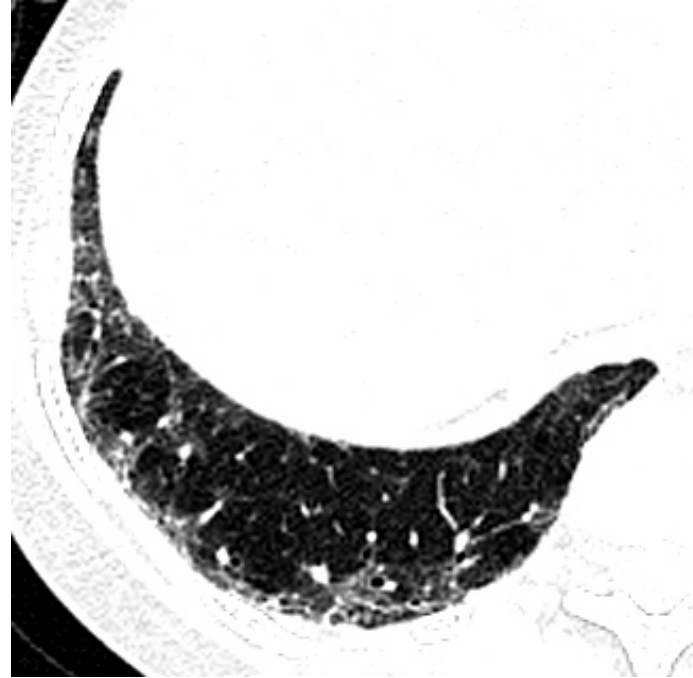


iPF' DE BT BULGULARI

RETİKÜLER GÖRÜNÜM

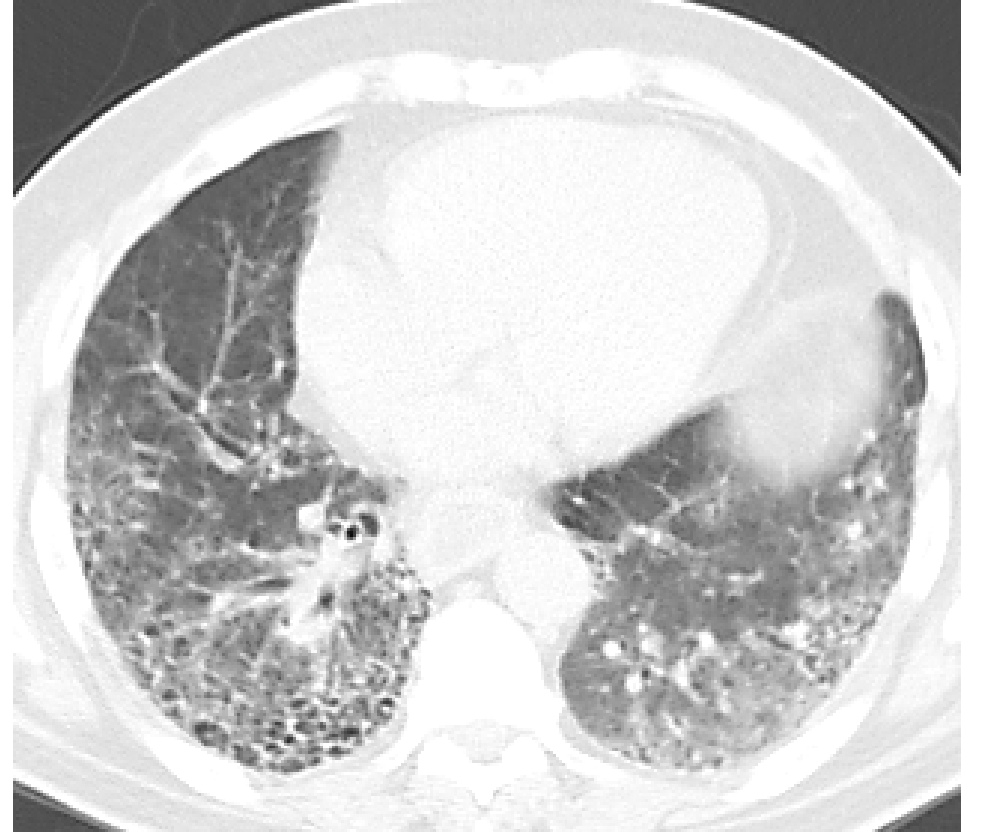
- İnce ağ şeklinde çizgilerden oluşan patern/ inter- ve intra septal kalınlaşmalar nedeniyle

- UIP' de: Kalın ve düzensiz aralıklarla
- NSIP'de: Düzenli ve homojen



BUZLU CAM GÖRÜNÜMÜ

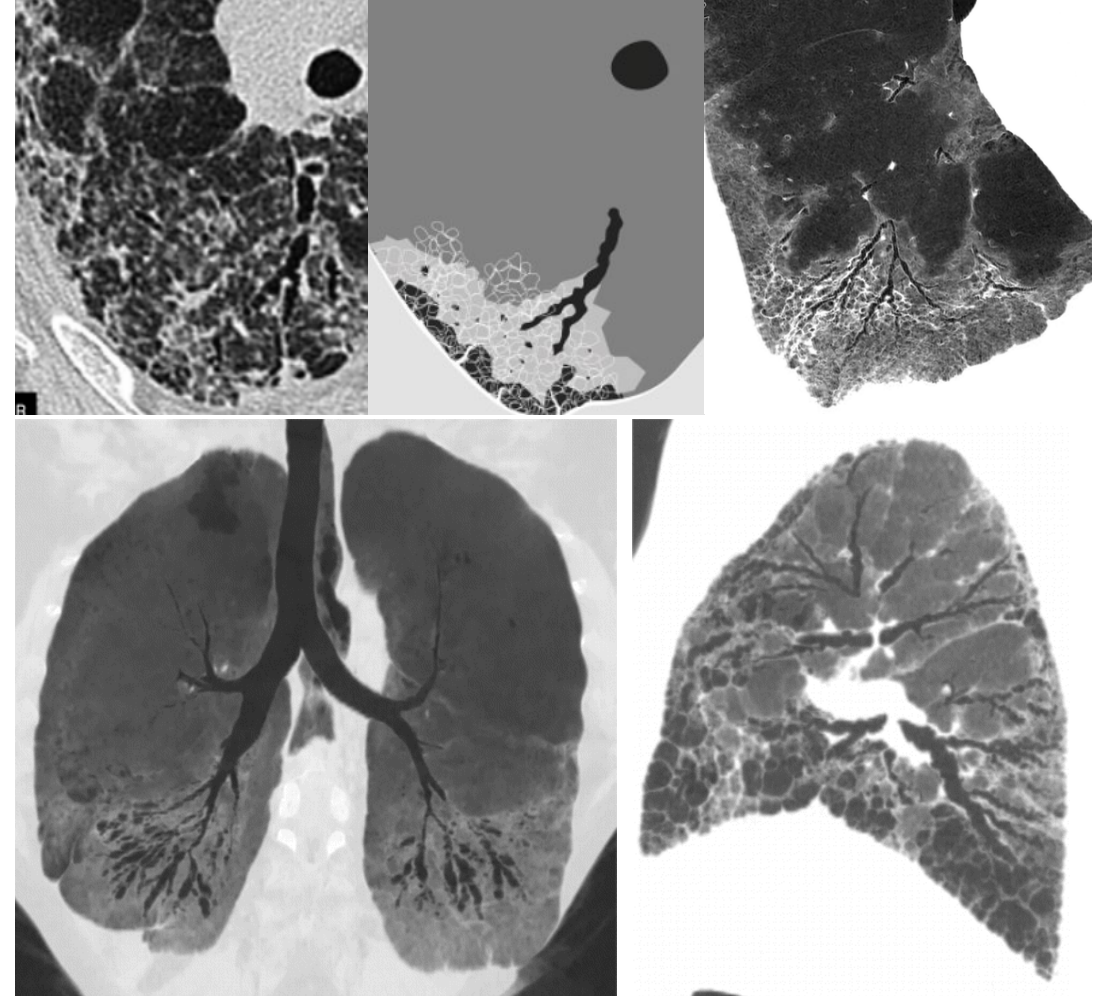
- Retikülasyon ve traksiyon bronşektazisi alanlarına komşu alanda görülür ve aslında fibrozisi temsil eder.
- Yeni çıkan buzlu cam ise alevlenme veya infeksiyon lehine



TRAKSİYON BRONŞEKTAZİSİ

Çevredeki retraktil fibrotik dokunun oluşturduğu bronşial dilatasyon

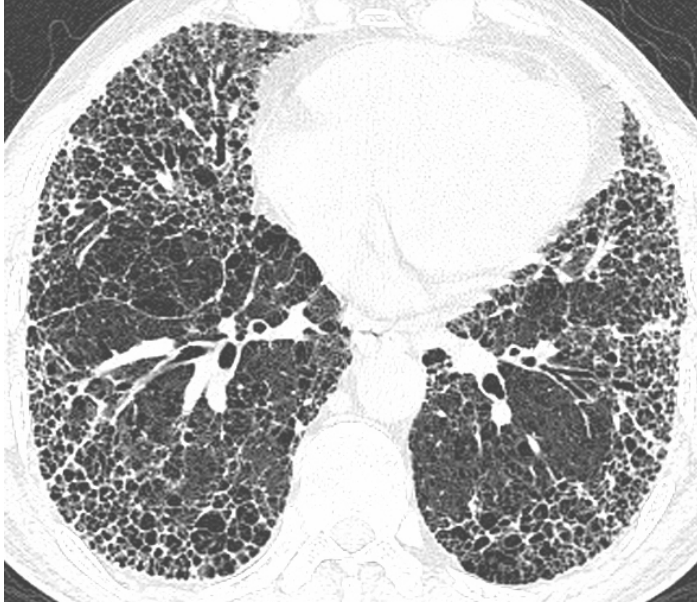
- UIP'de : Akciğerin periferinde, irregüler ve variköz
- NSIP'de: Santralde dilate bronşlar şeklinde



BAL PETEĞİ GÖRÜNÜMÜ

- Makroskopik

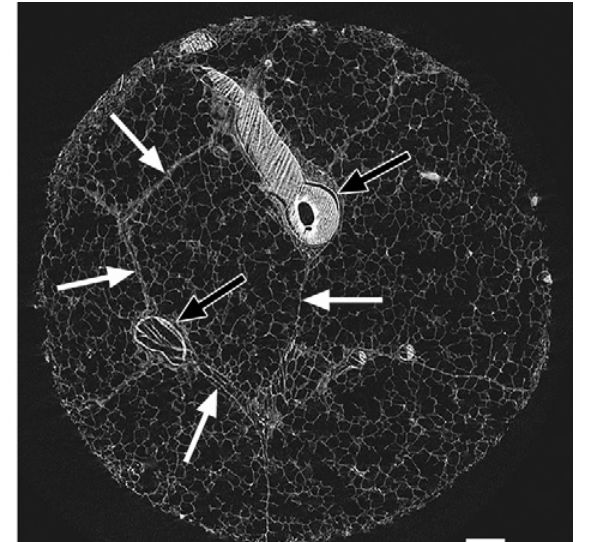
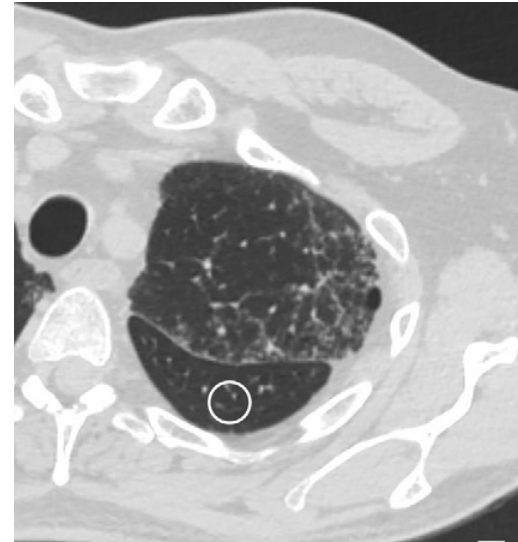
- Genelde 3-5 mm boyutta



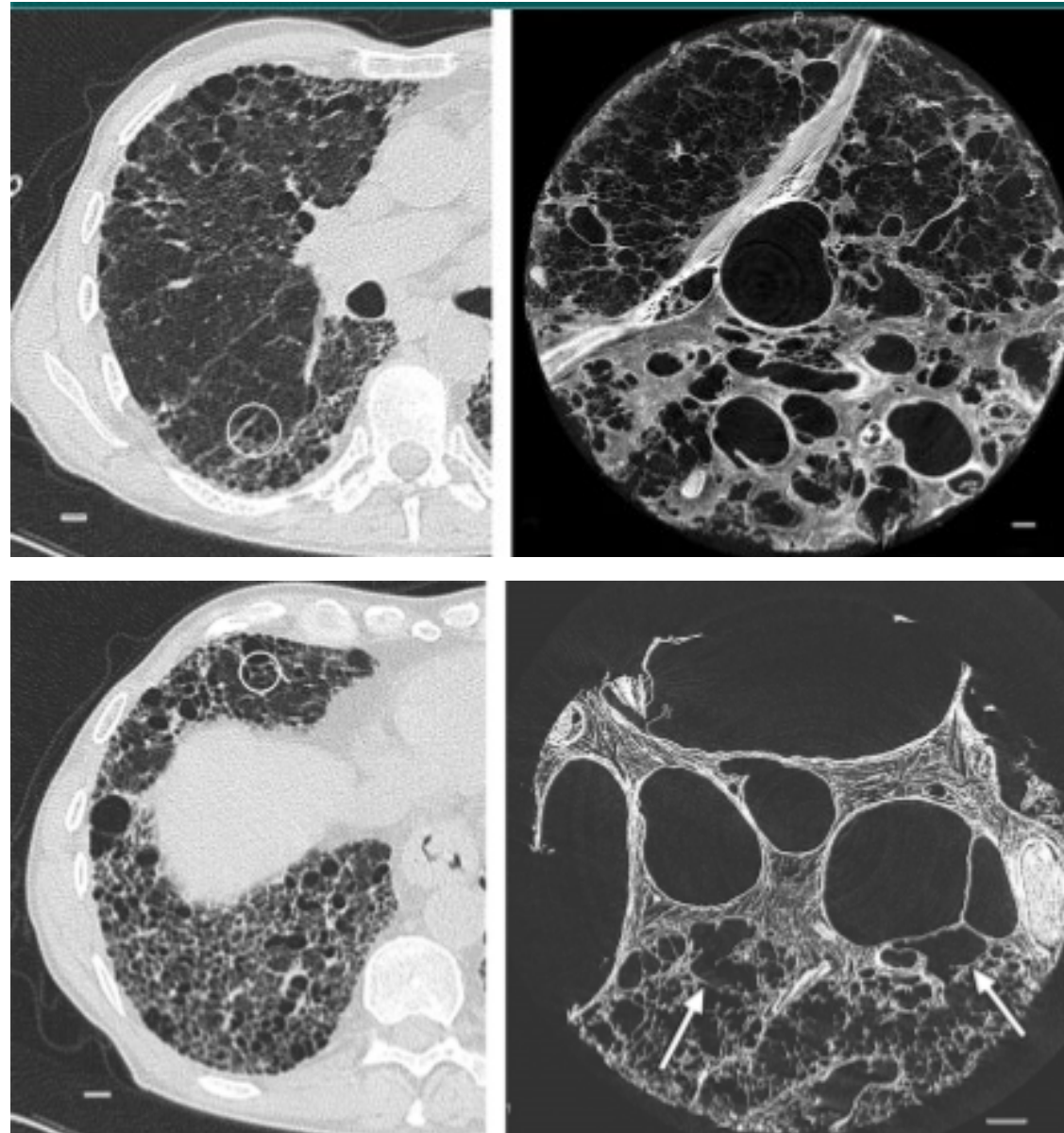
- Mikroskopik

- BT'de görülmeyen kistler

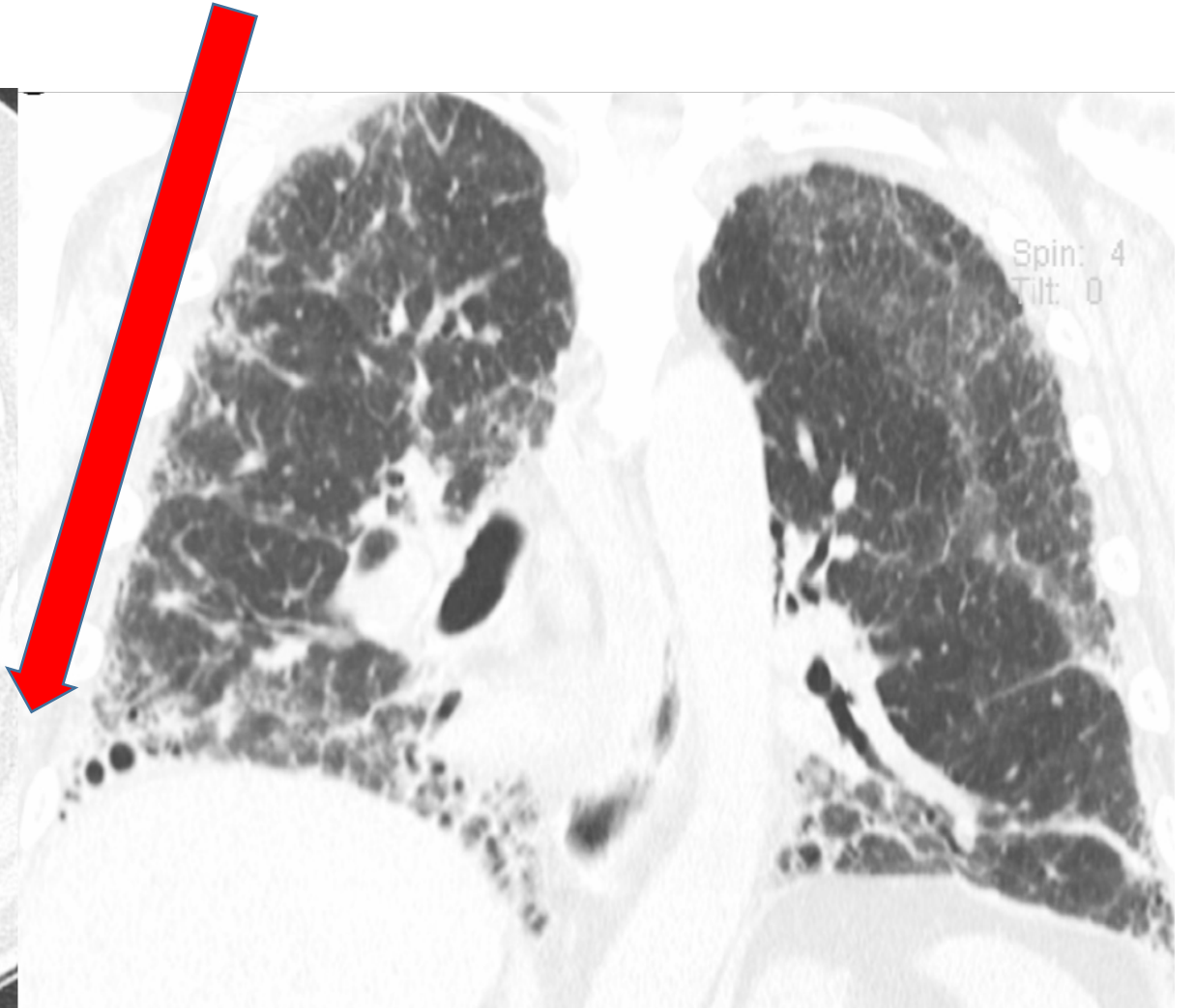
- MikroBT ile görülebilir.



Thin-Section CT Features of Idiopathic Pulmonary Fibrosis
Correlated with Micro-CT and Histologic Analysis. *Radiology*:
Volume 283: Number 1—April 2017

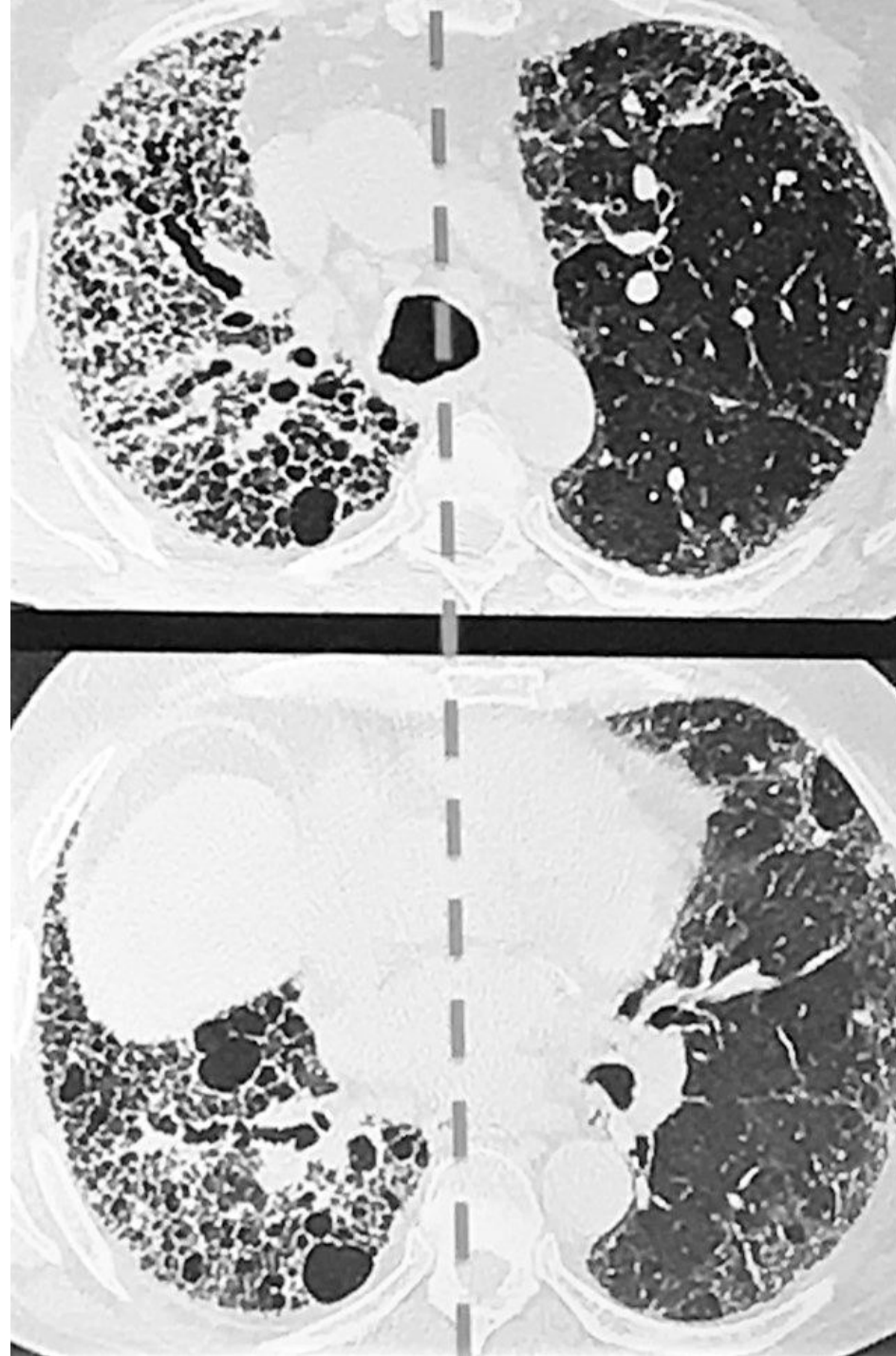


Thin-Section CT Features of Idiopathic Pulmonary Fibrosis Correlated with Micro-CT and Histologic Analysis. **Radiology**: Volume 283: Number 1—April 2017



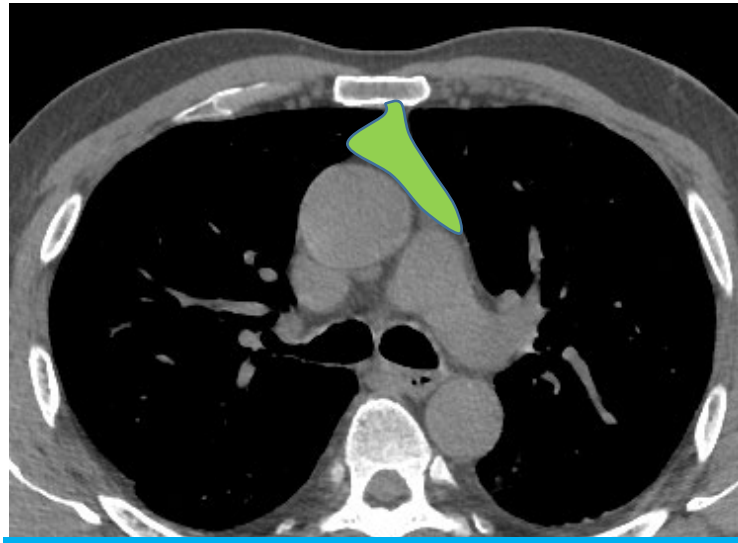
ESTİ2018, S Walsh

ASİMETRİ
SAĞ>SOL
İPF lehine

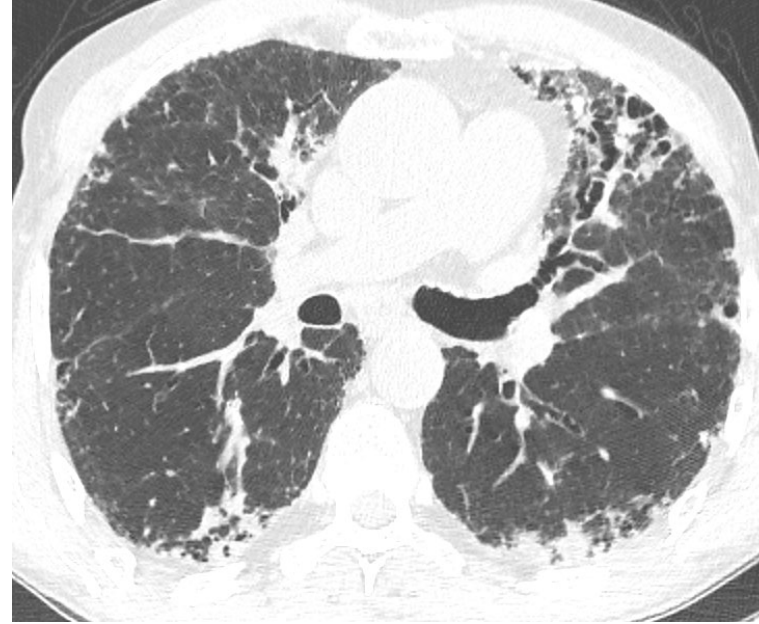
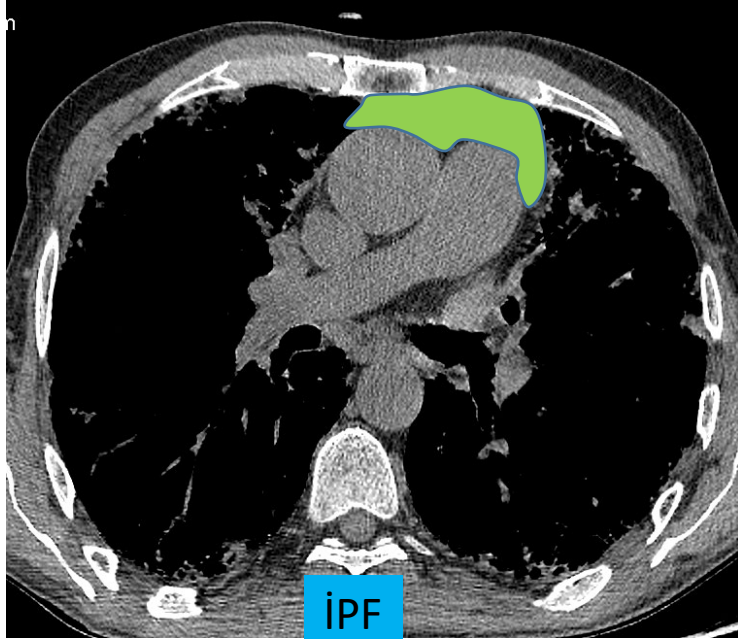


DİĞER BULGULAR

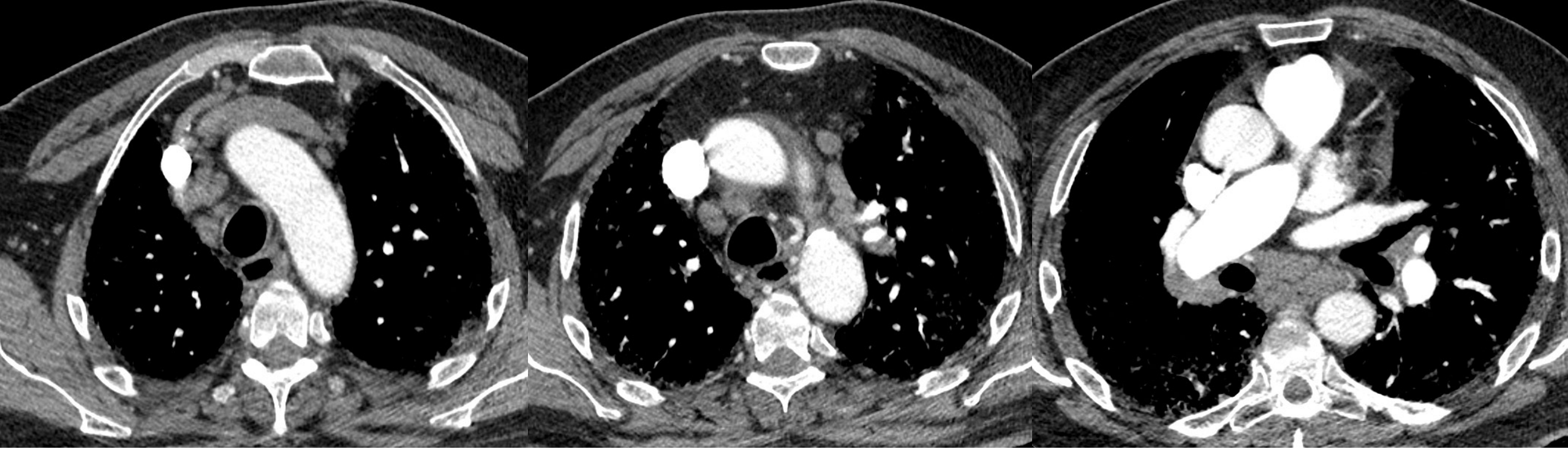
- Mediastinal yağ dokusu artışı
- Mediastinal lenf nodları (5-15 mm arası) % 70
- Hava hapsi (% 27-43)
- Ossifikasyon/kalsifikasyon (UIP'de % 28, diğer ILD % 8.3)
- Plevroparankimal fibroelastoz
- Amfizem



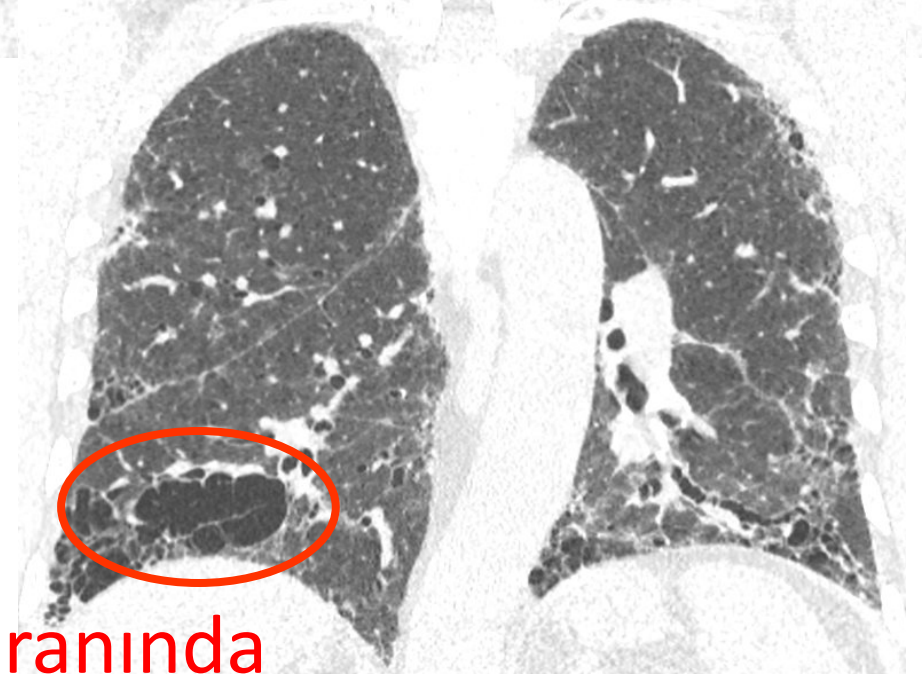
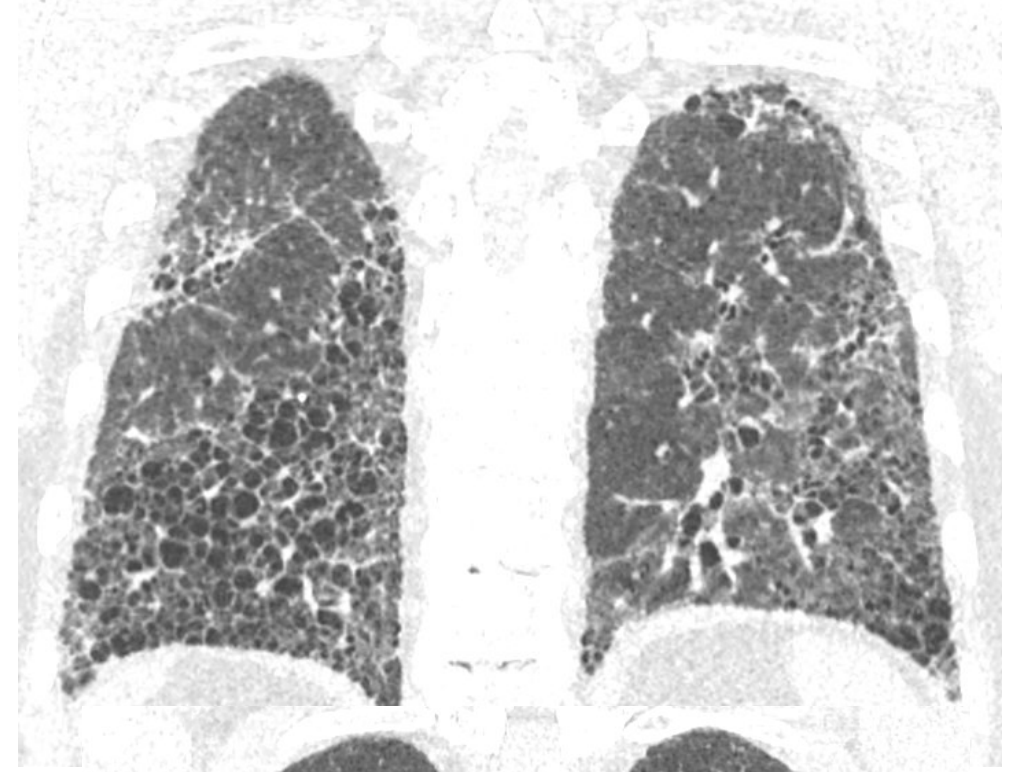
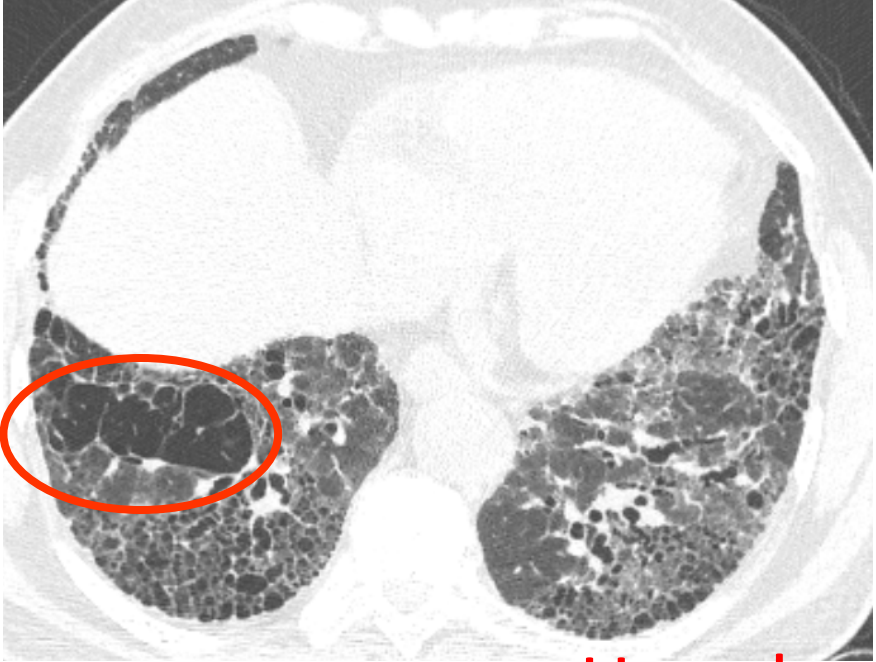
NORMAL mediasten genişliği ve yağı



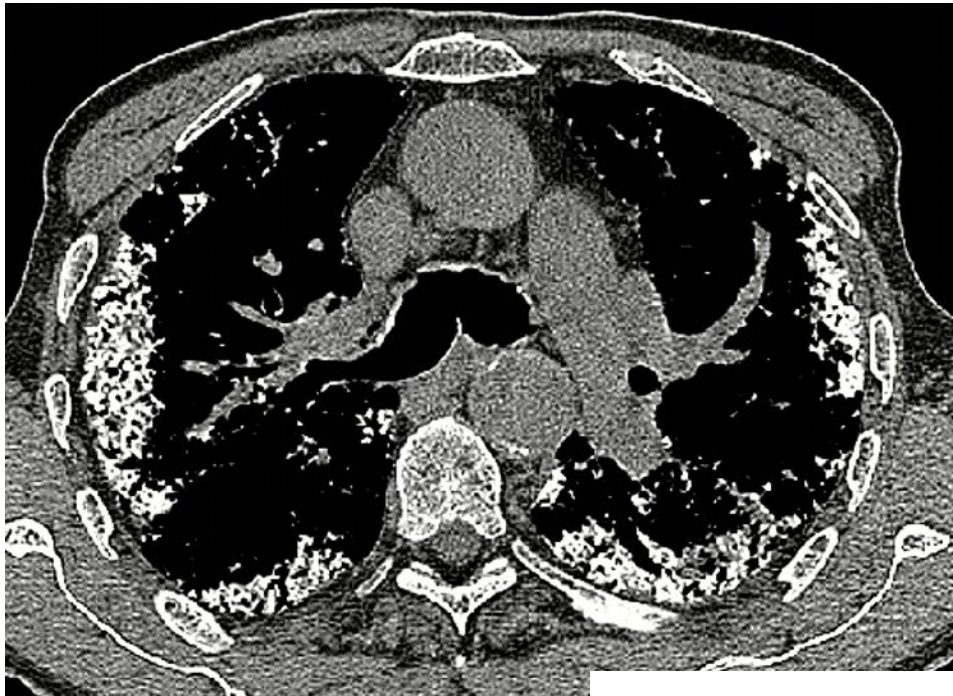
MEDIASTİNAL YAĞ DOKUSU ARTIŞI VE VOLÜM KAYBI



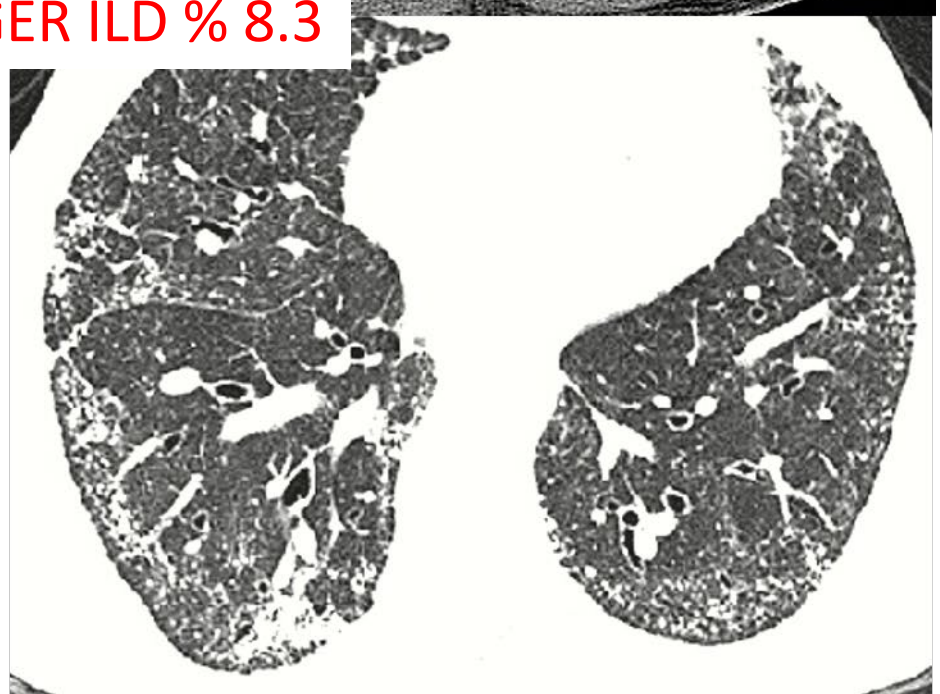
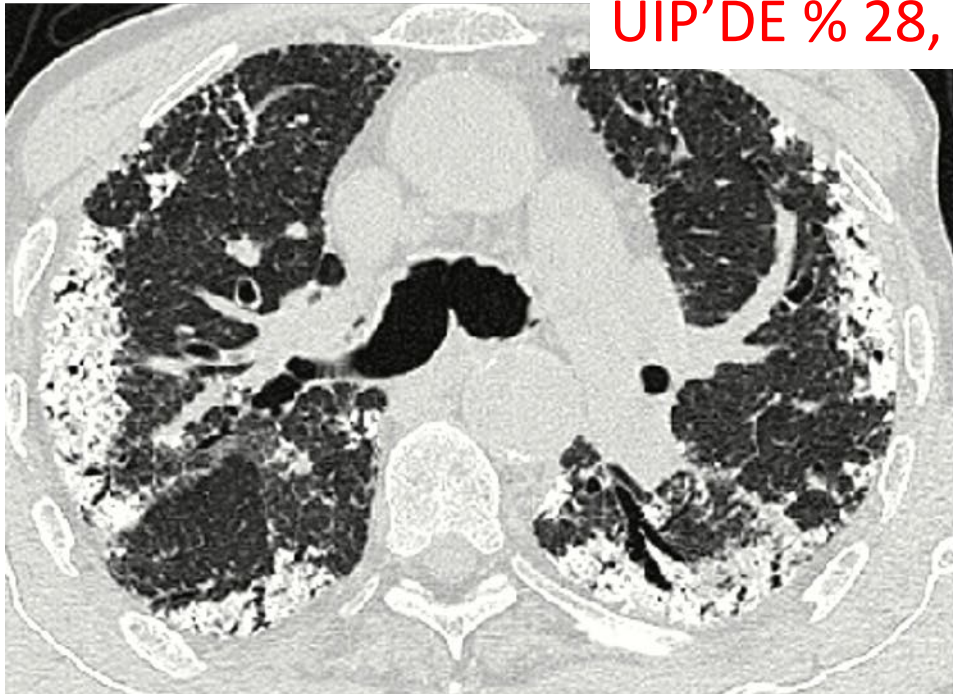
MEDIASTİNAL lenf nodları (5-15 mm arası) % 70

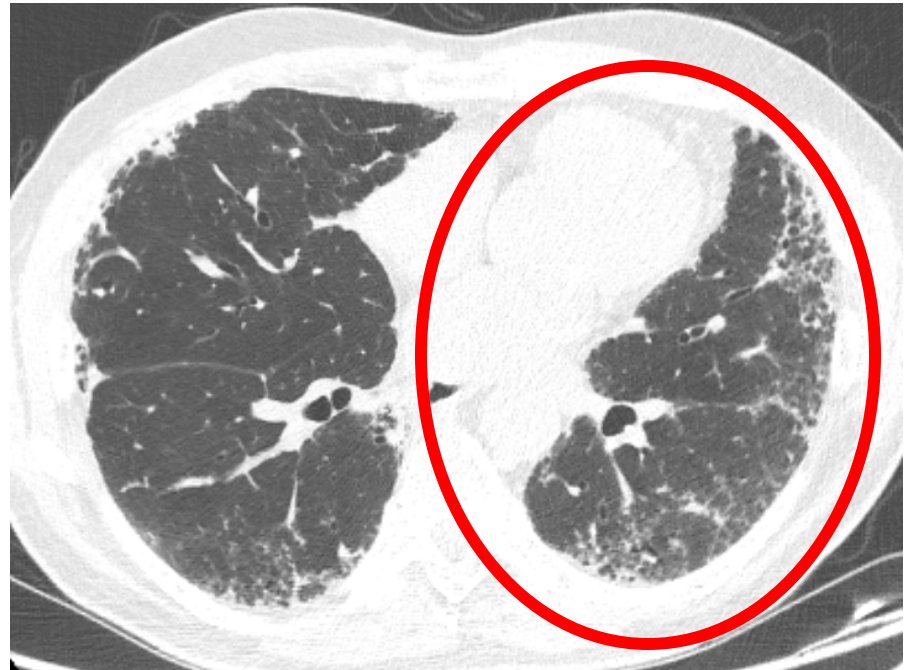
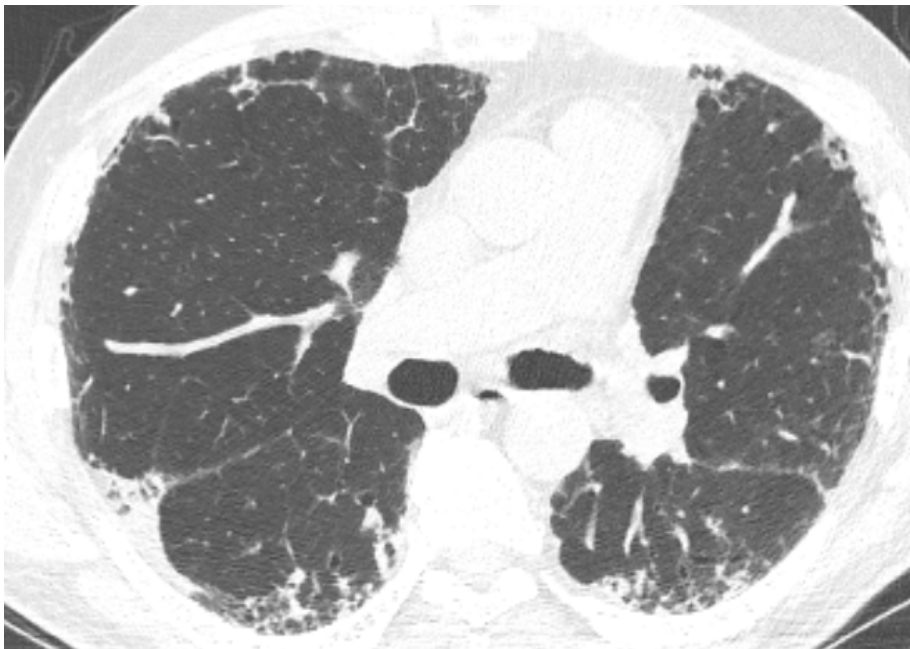
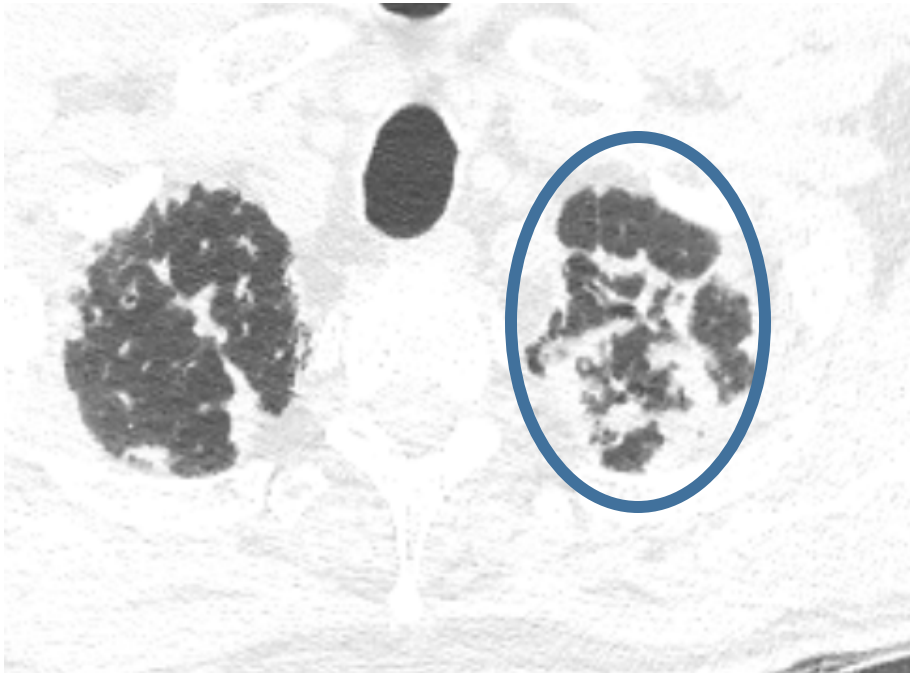


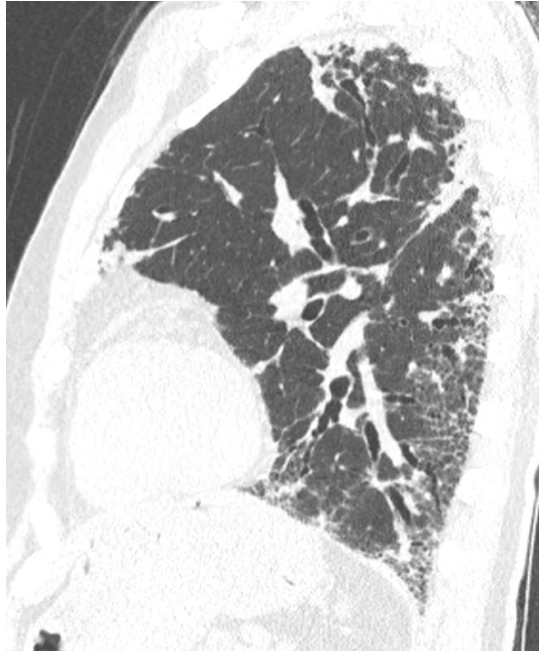
Hava hapsi % 27-43 oranında



UIP'DE % 28, DİĞER ILD % 8.3





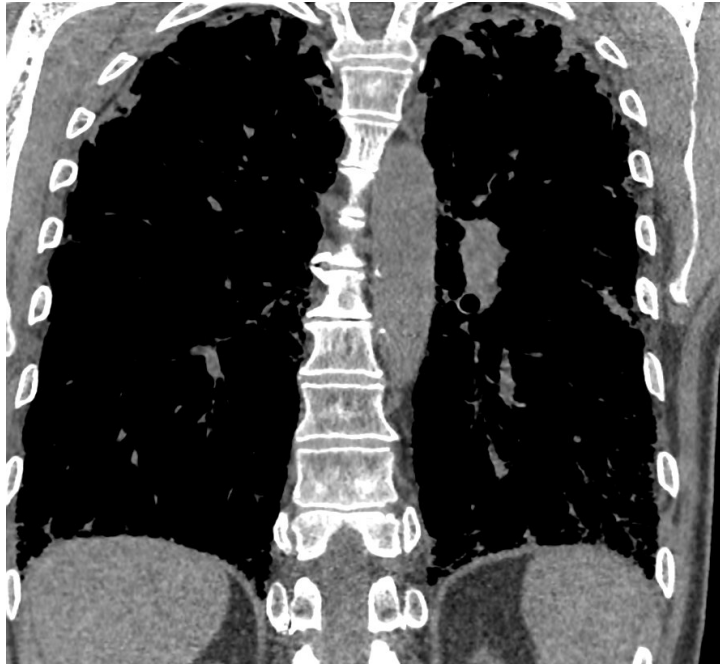
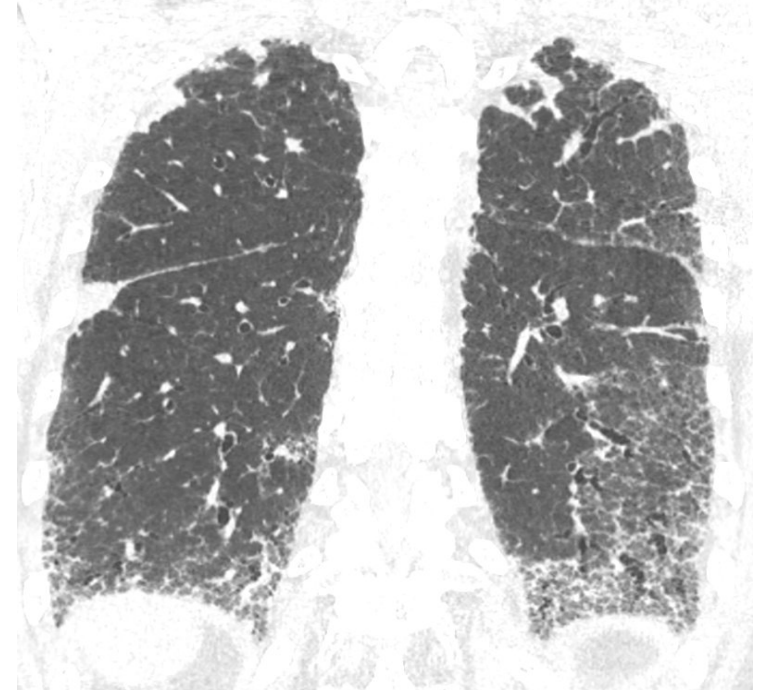


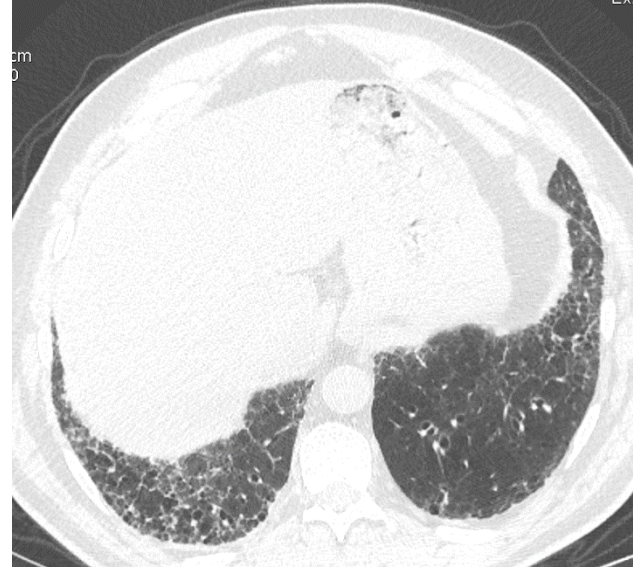
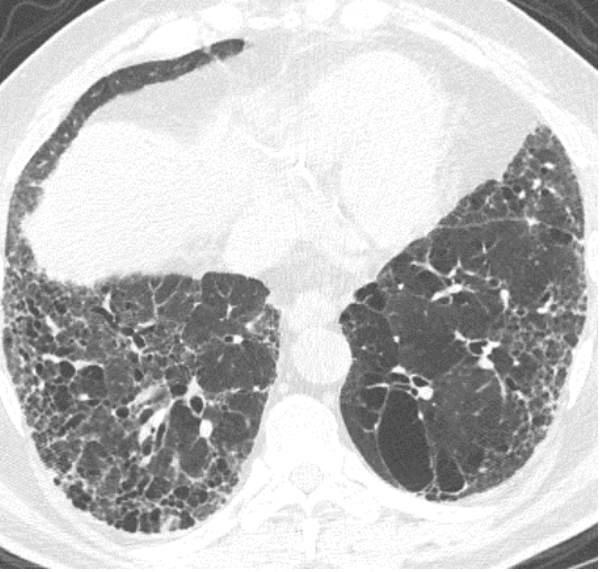
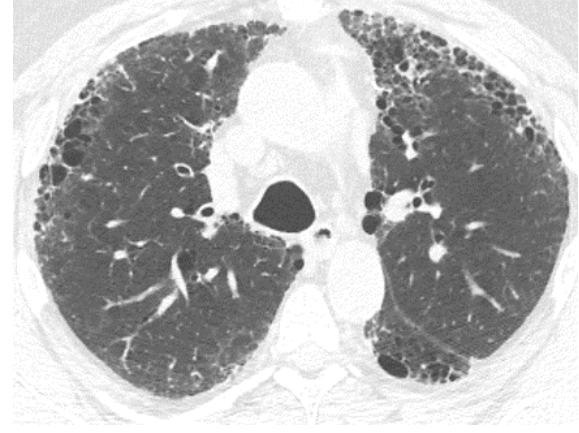
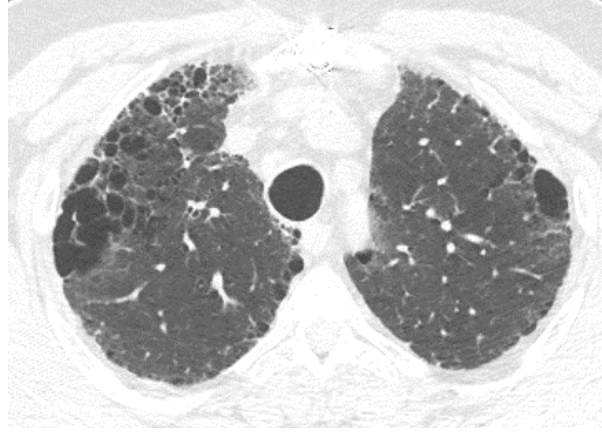
UIP VE PPFE

% 32 BİRLİKTELİK

ESTİ 2018, S Walsh

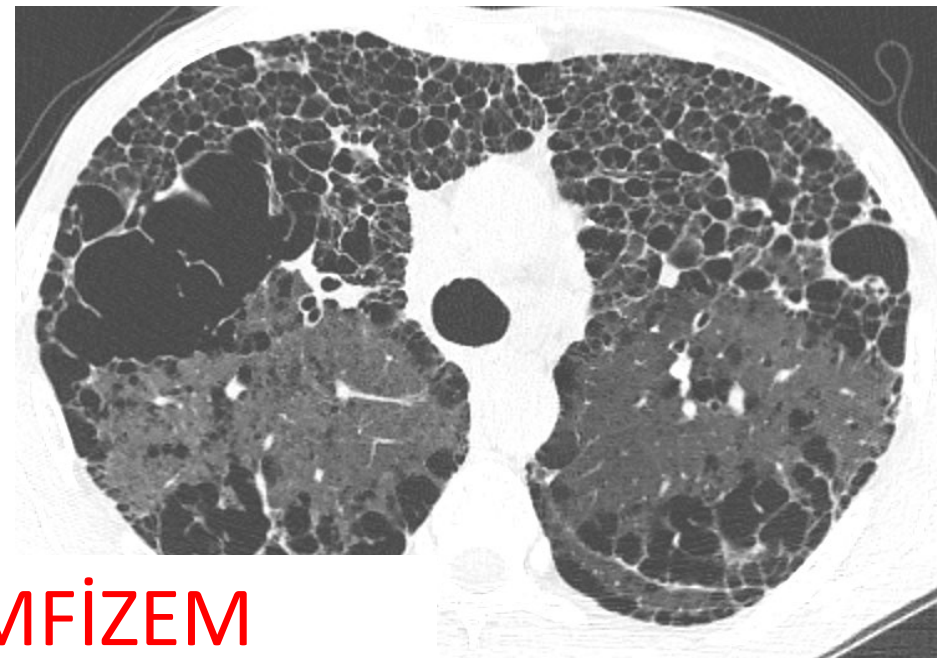
İPF tanımlamasına ters
Sadece akciğeri tutan ??



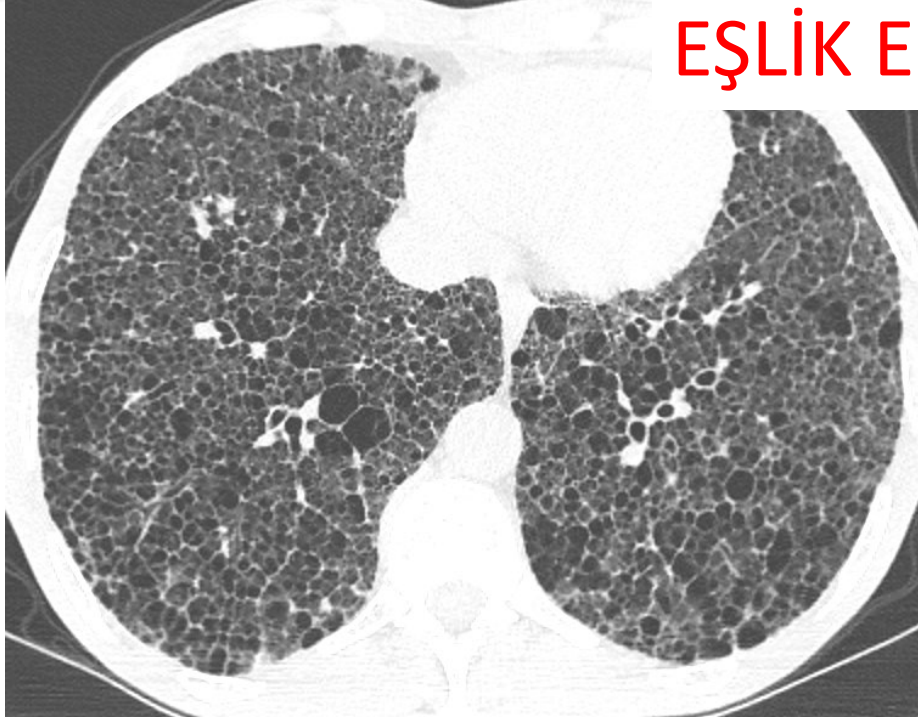


KOMBİNE PULMONER FİBROZİS VE AMFİZEM SENDROMU

Pulmoner HT riski yüksek???



**ŞİDDETLİ AMFİZEM
EŞLİK EDEN FİBROZİS ??**



ALTERNATİF TANILAR

- Üst veya orta zon tutulumu
- Peribronkovasküler dağılım
- Subplevral korunma
- Belirgin konsolidasyon
- Saf buzlu cam (akut alevlenme hariç)
- Ekspiryumda belirgin lobuler hava hapsi ile yaygın mozaik atenuasyon
- Diffüz nodül veya kist

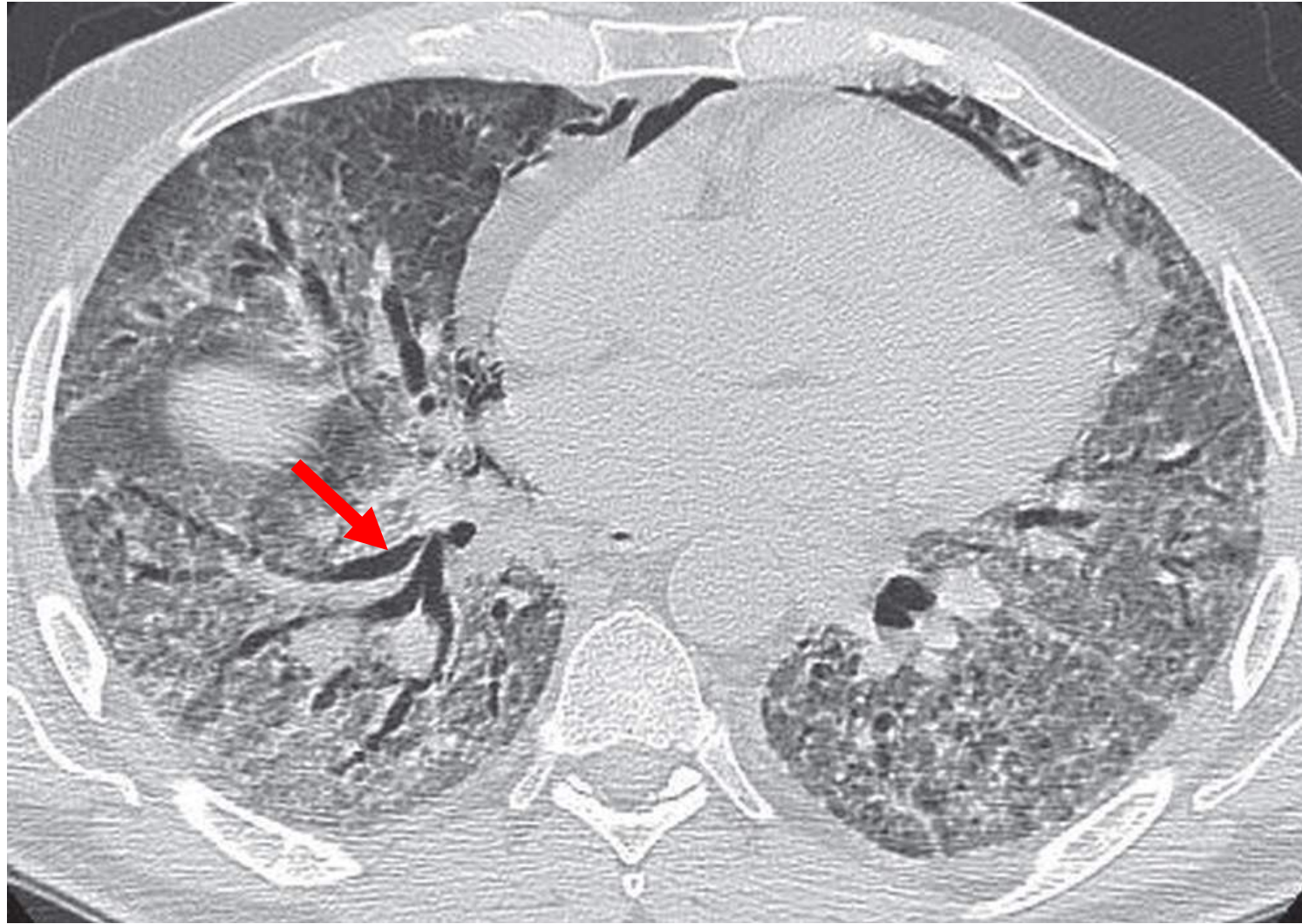
AYIRICI TANI :

- Nonspesifik interstisyel pnömoni (NSIP)
- Kronik hipersensitivite pnömonisi
- Otoimmün ilişkili interstisyel pnömoni (İPAF)
- İlaç reaksiyonlarına bağlı fibrozis
- Asbestozis
- Sarkoidozis
- İLA



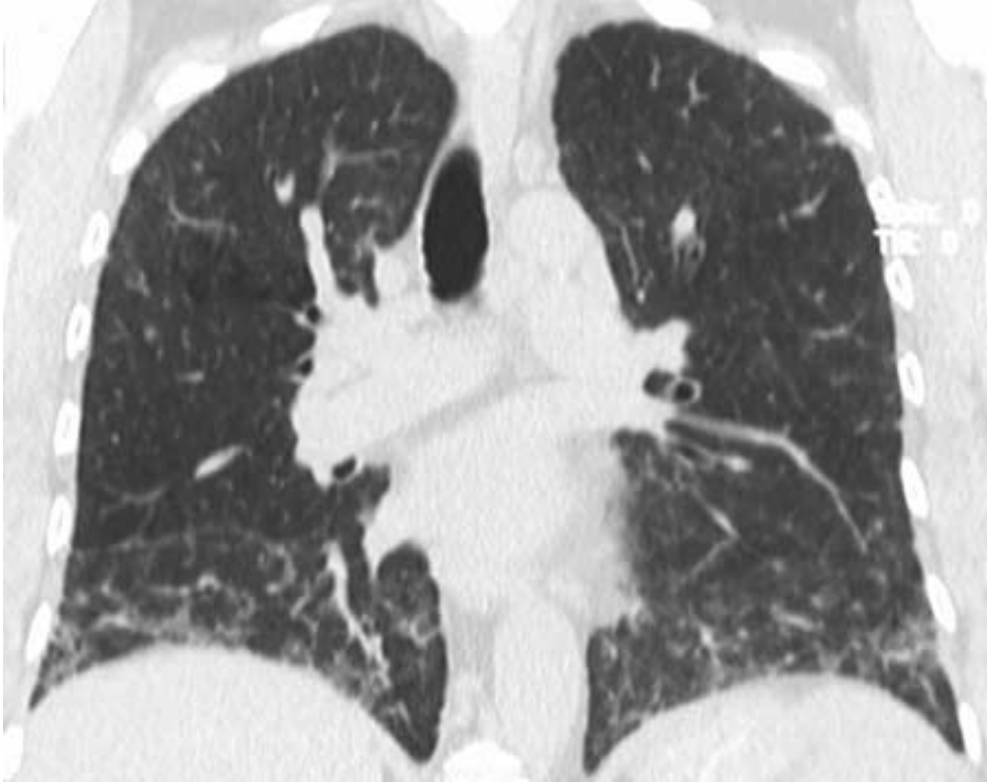
NSIP (sellüler form)

Yaygın peribronkovasküler dağılıma uyan buzlu cam alanları

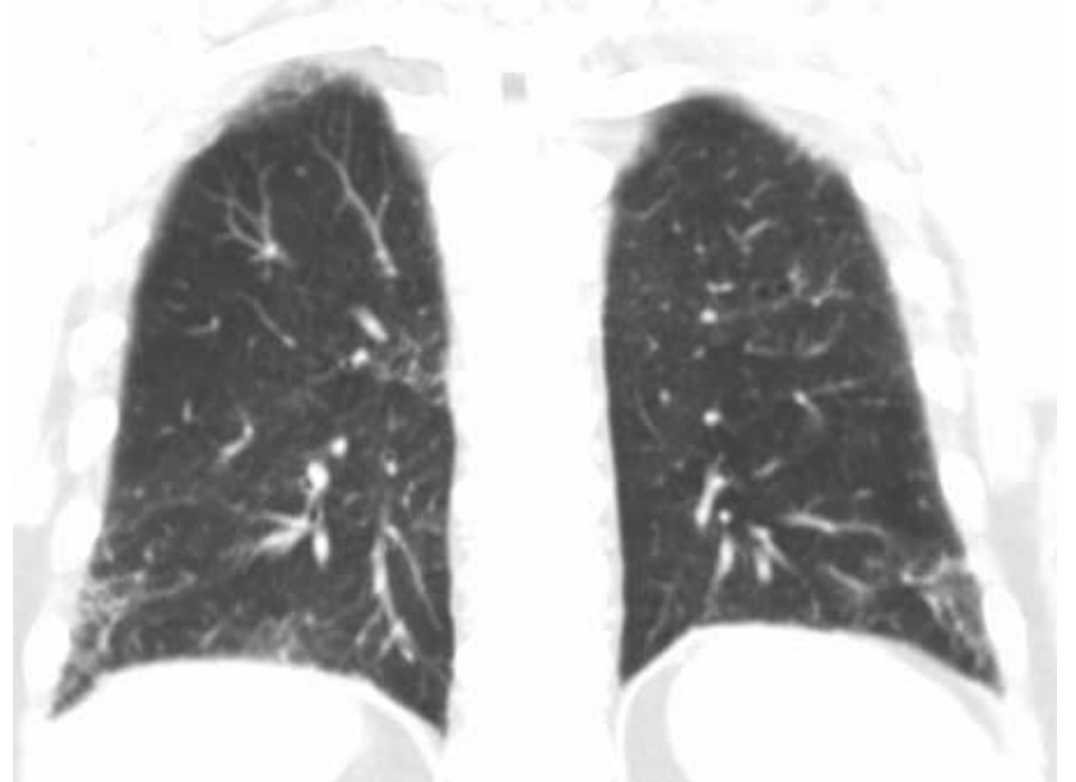


NSIP

Straight Edge sign – Düz kenar işareti



NSIP – SES var



UIP –SES yok

Differentiating Usual Interstitial Pneumonia From Nonspecific Interstitial Pneumonia Using High-resolution Computed Tomography The “Straight-edge Sign”. J Thorac Imaging 2018:266-270



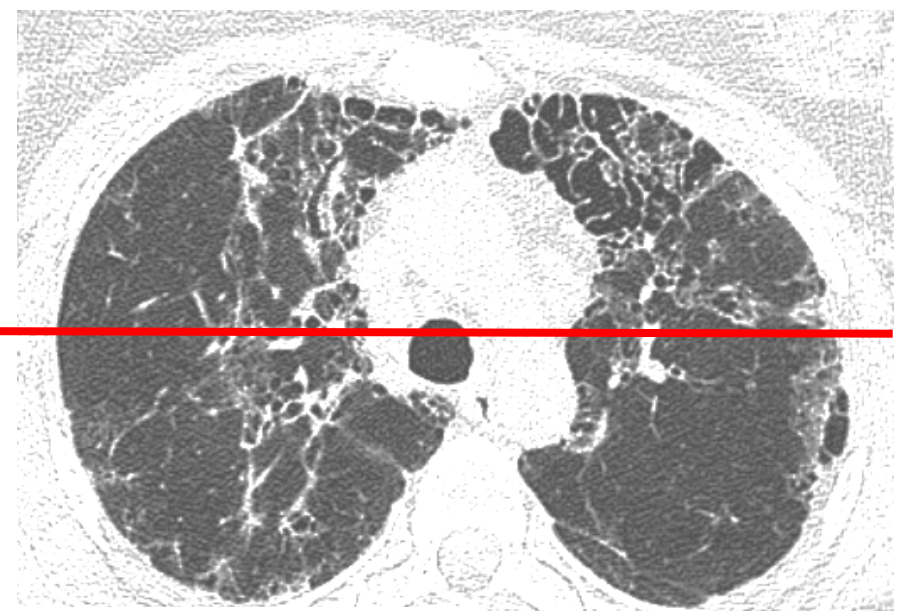
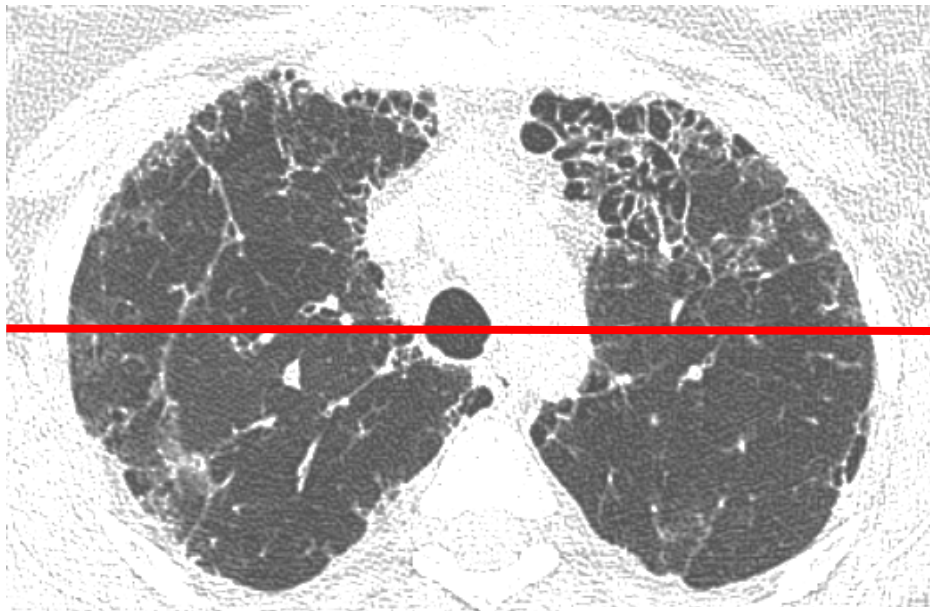
ASBESTOZIS



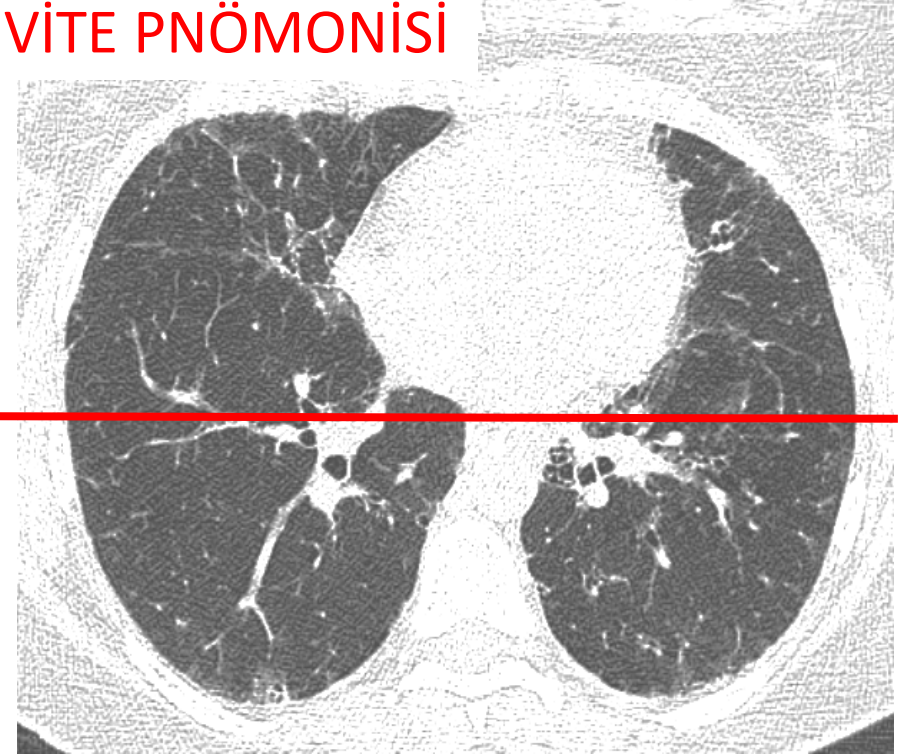
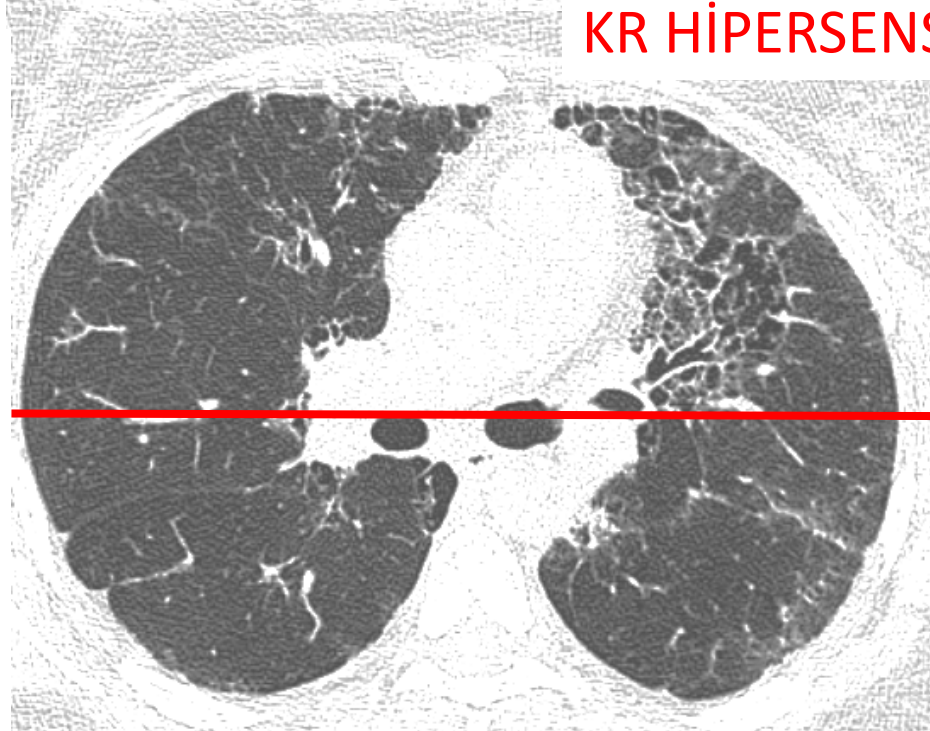
IPF=UIP 'dir.

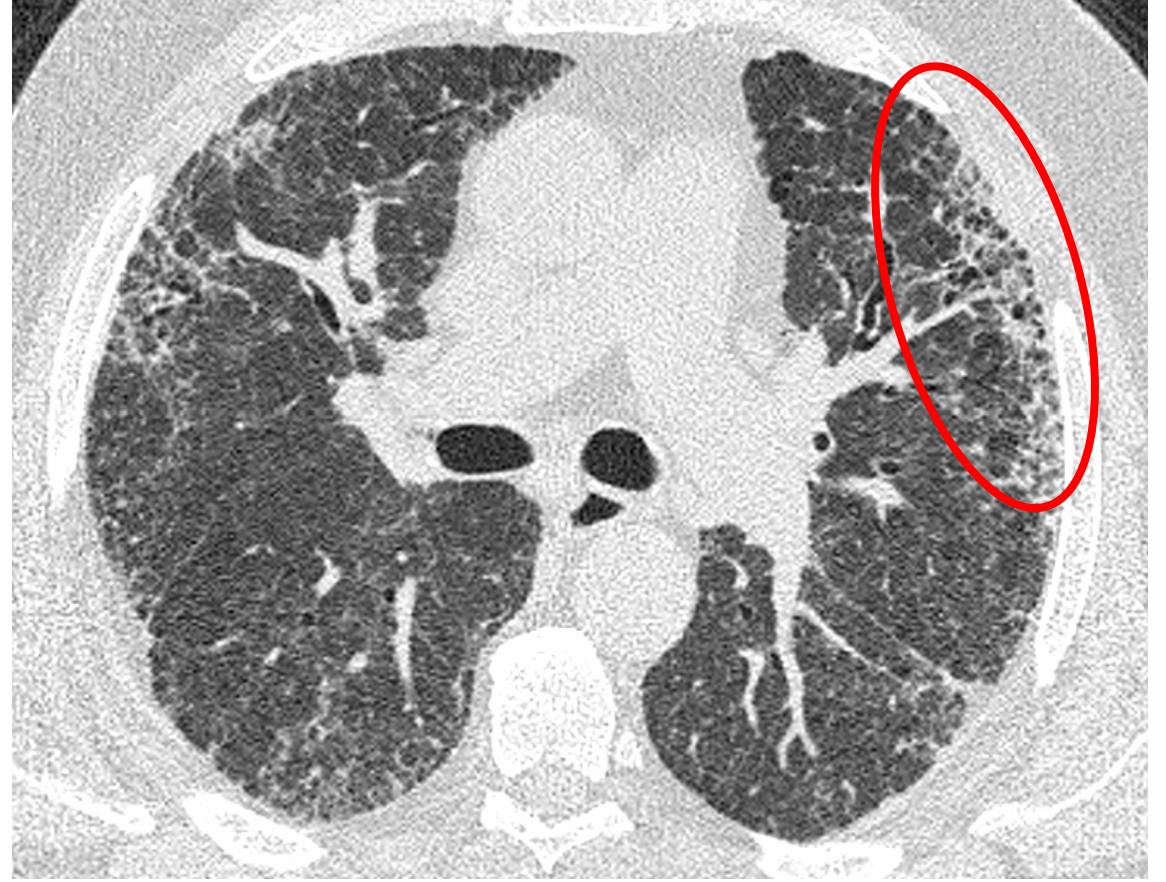
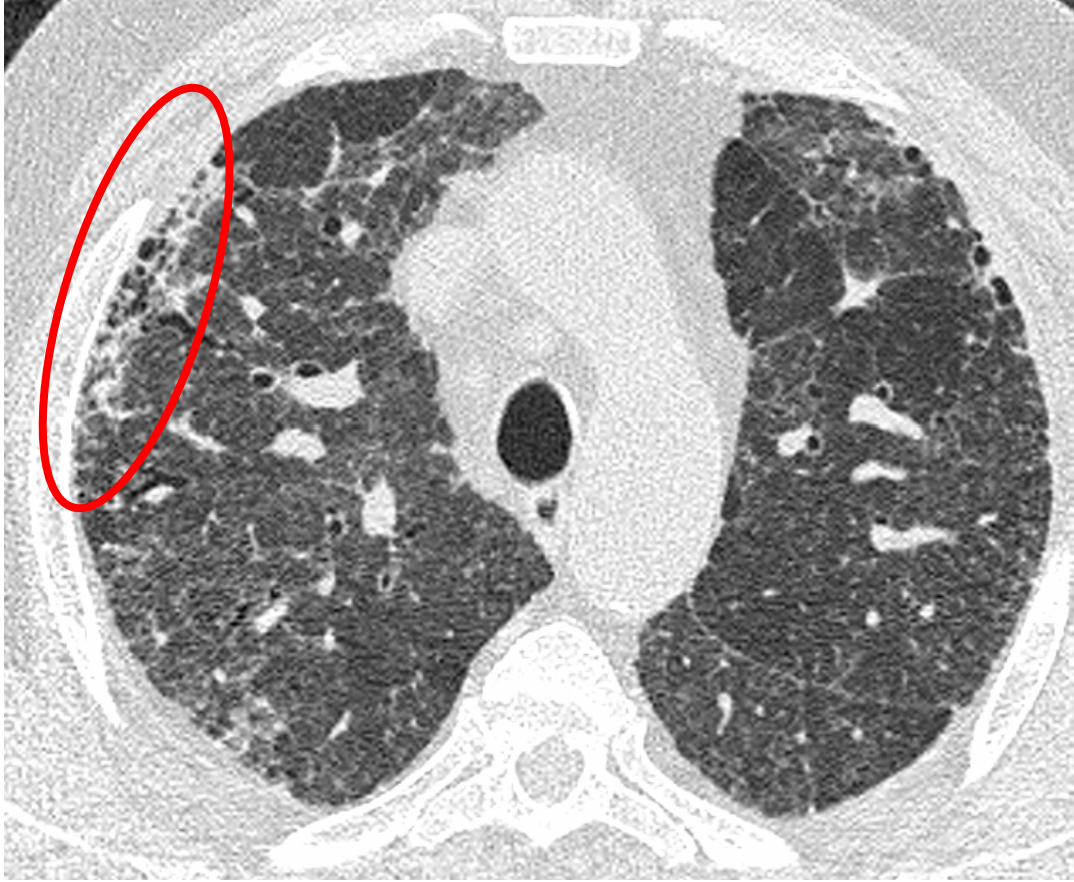
Ancak UIP her zaman İPF değildir

- Patolojik olarak UIP tanısı konmuş ise hasta:
 - IPF
 - Kronik Hipersensitivite Pnömonisi
 - Bağ dokusu hastalıkları
 - Sarkoidozis
 - Otoimmün özellikli interstisyel pnömoni
 - İlaç toksisitesi
 - Asbestozis
 - Kronik aspirasyon pnömonisi
 - Hermansky-Pudlak sendromuolabilir.



KR HİPERSENSİTİVİTE PNÖMONİSİ



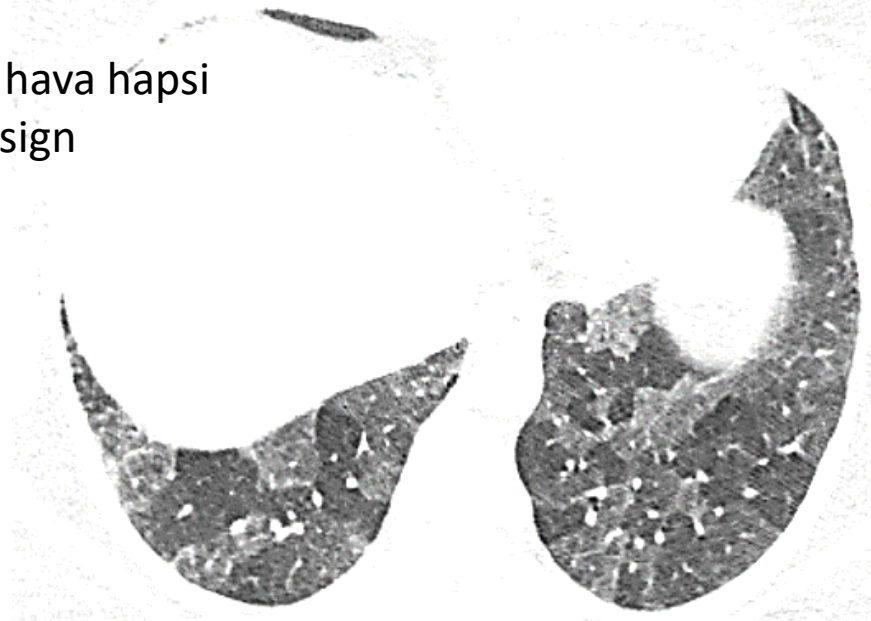
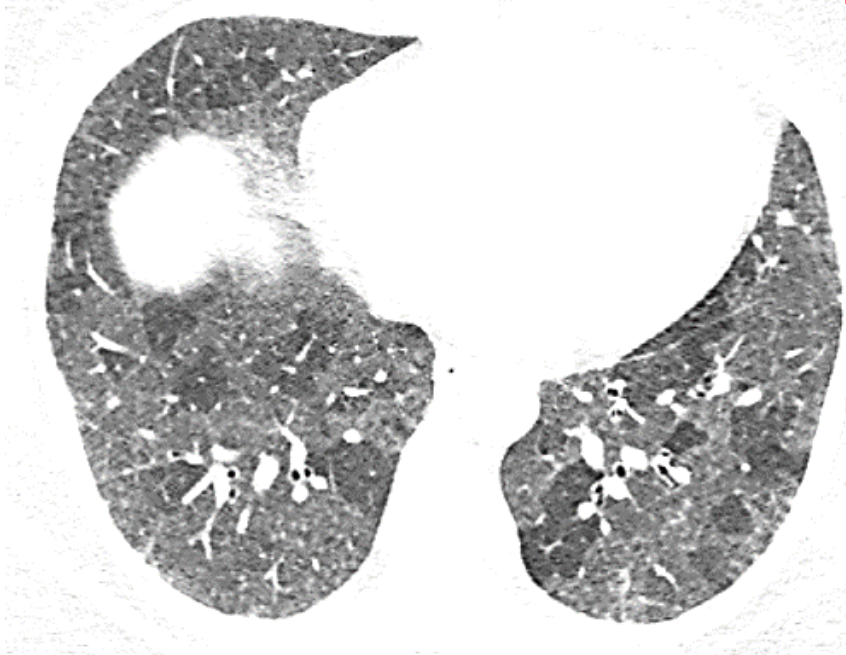


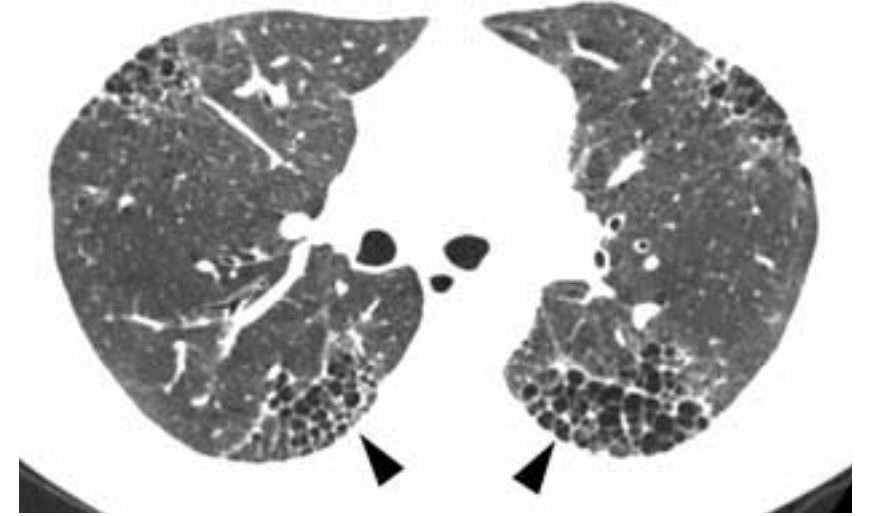
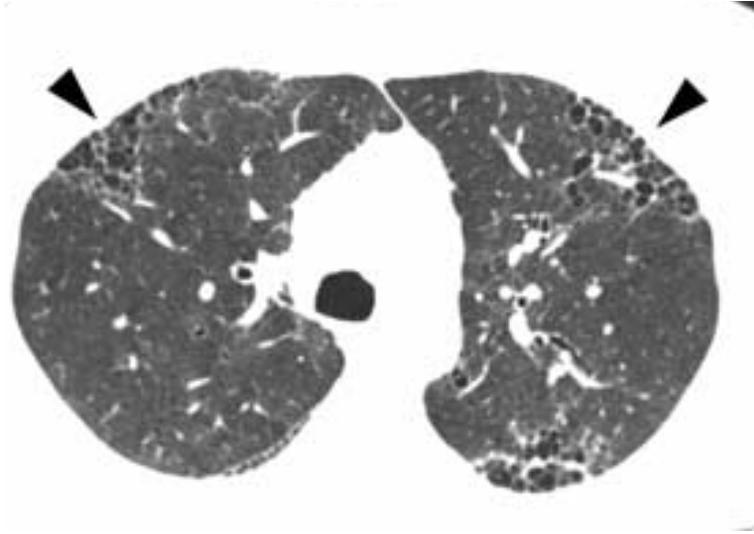
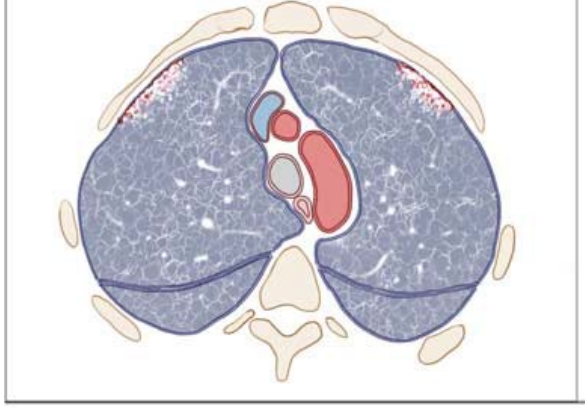
RİTUKSİMABA BAĞLI FİBROZİS



KR HİPERSENSİTİVİTE PNÖMONİSİ

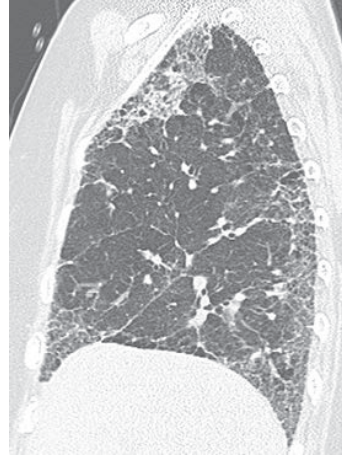
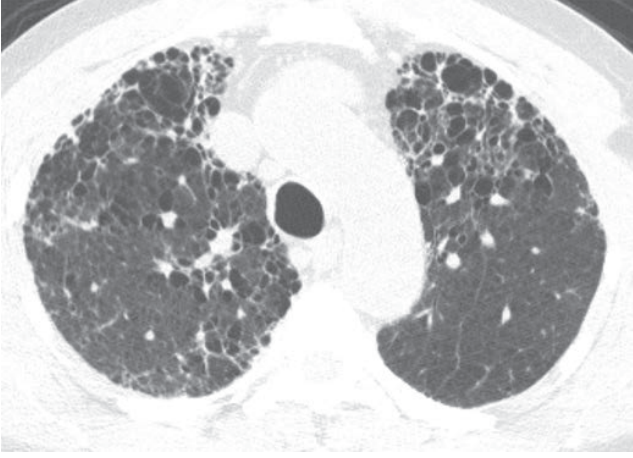
Buzlu cam ve hava hapsi
Head cheese sign



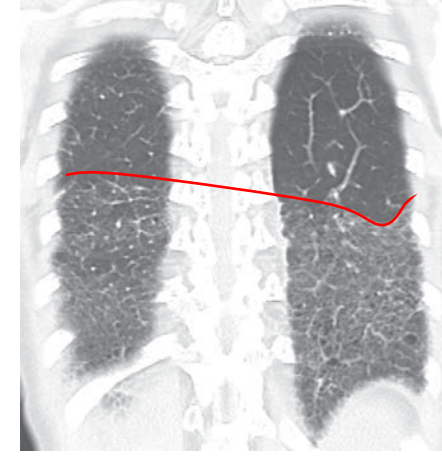


Dört Köşe işareti SKLERODERMA

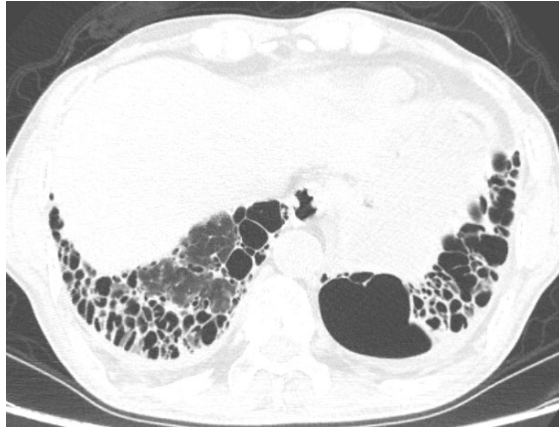
The Four Corners Sign A Specific Imaging Feature in Differentiating Systemic Sclerosis-related Interstitial Lung Disease From Idiopathic Pulmonary Fibrosis. Walkoff L et al J Thorac Imaging 2018 Jan 16.



Ön üst lob işareti



Düz kenar işareti



Abartılı bal peteği işareti

CT Features of the Usual Interstitial Pneumonia Pattern: Differentiating Connective Tissue Disease Associated Interstitial Lung Disease From Idiopathic Pulmonary Fibrosis. *AJR* 2018; 210:307–313





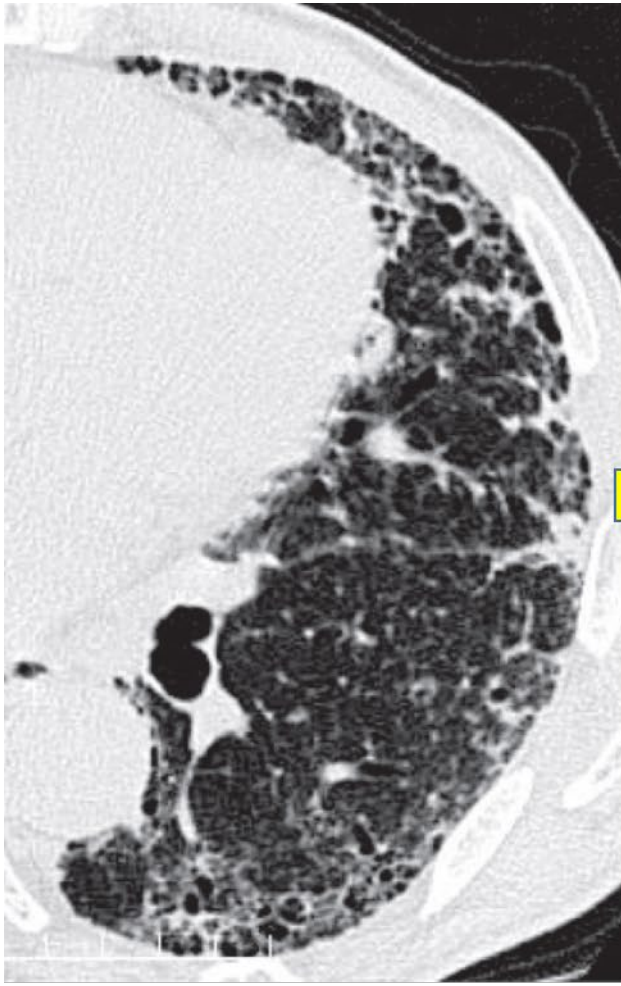
Düz kenar işareti

ROMATOİD ARTRİT

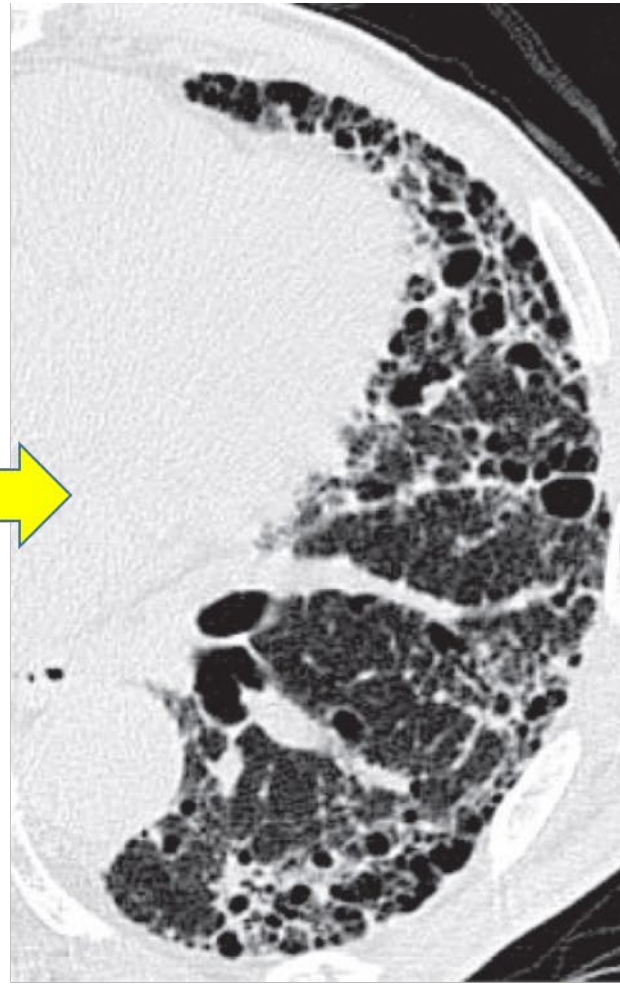
İPF'DE MORTALİTEYİ ETKİLEYEN DURUMLAR

RADYOLOJİK BULGULAR

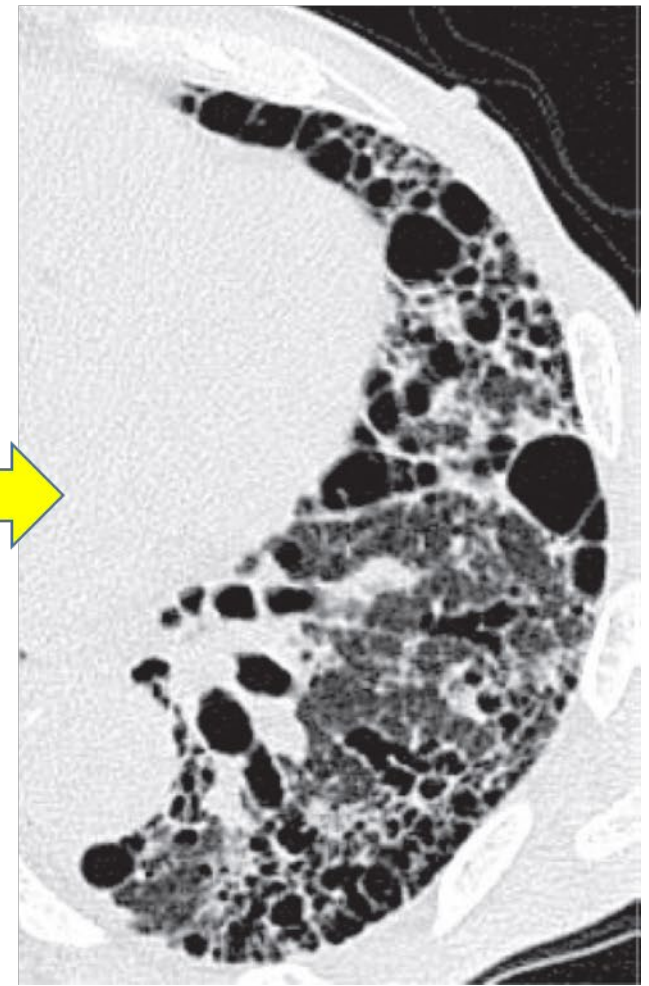
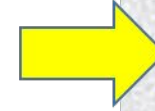
- Progresyon
- Akut alevlenme
- Pulmoner hipertansiyon
- Akciğer kanseri
- Pulmoner vasküler volüm (PVV) artışı (CALIPER vb bilgisayar programları ile görülüp tespit edilebilmekte)



Başlangıç



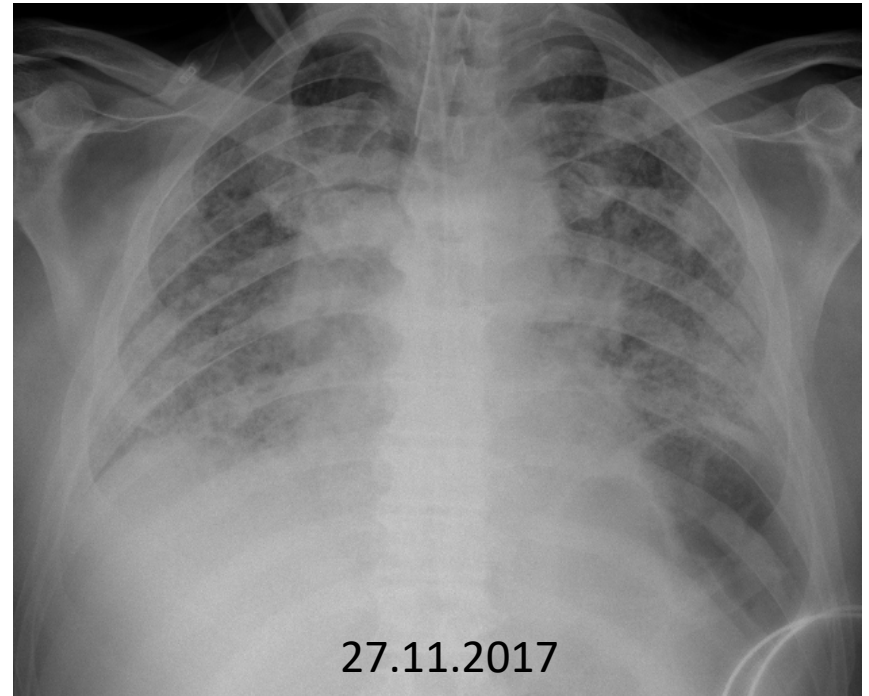
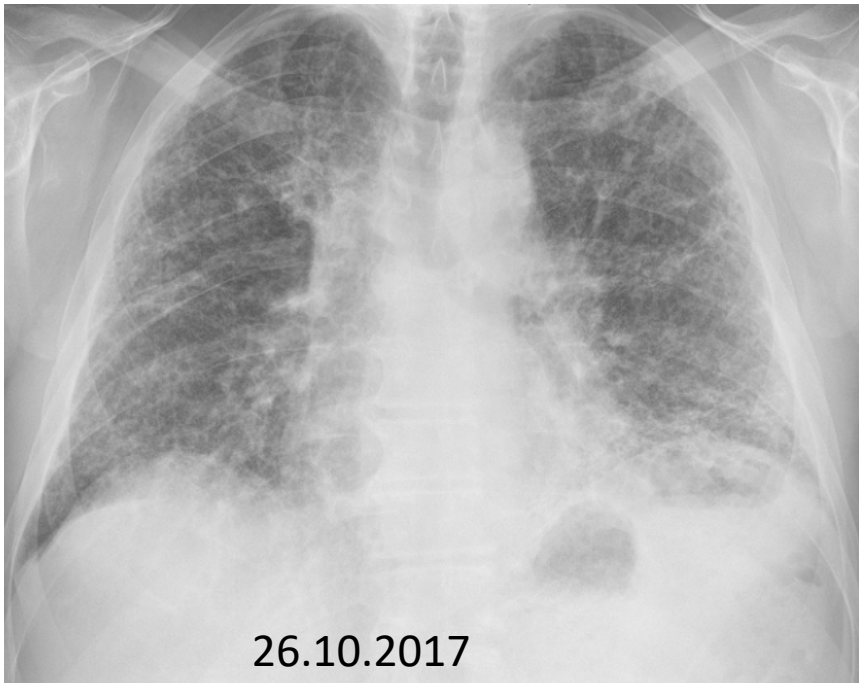
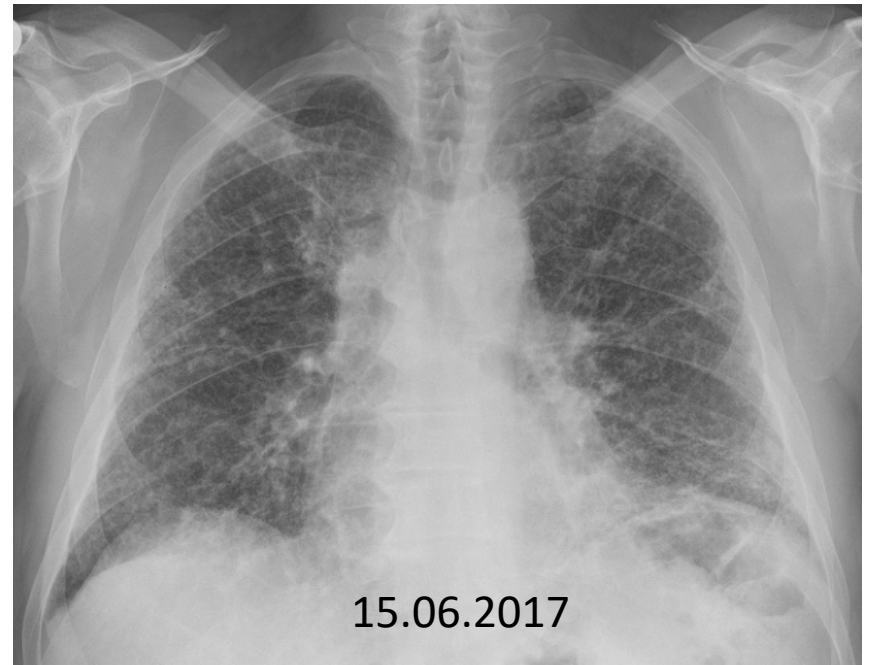
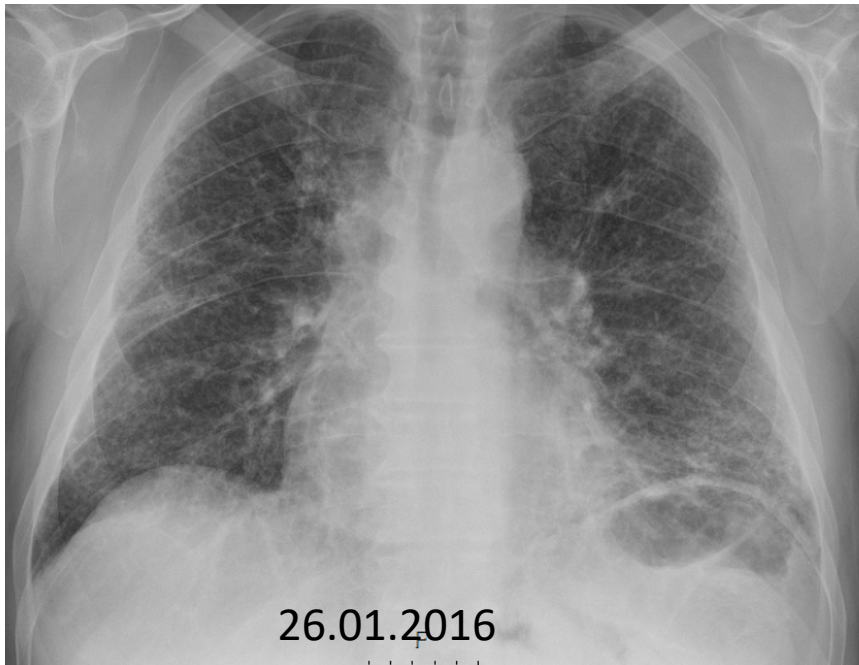
6 ay sonra

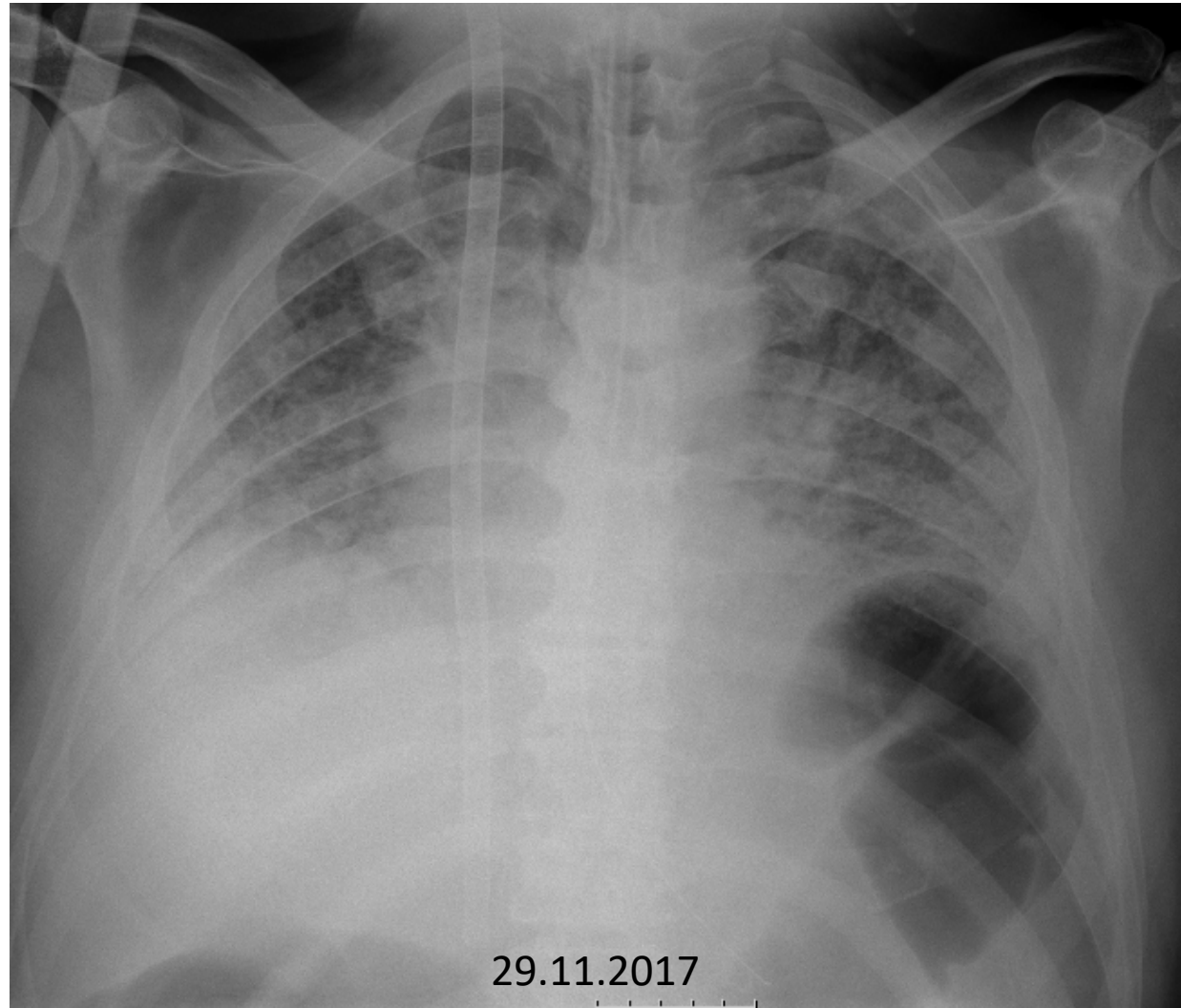


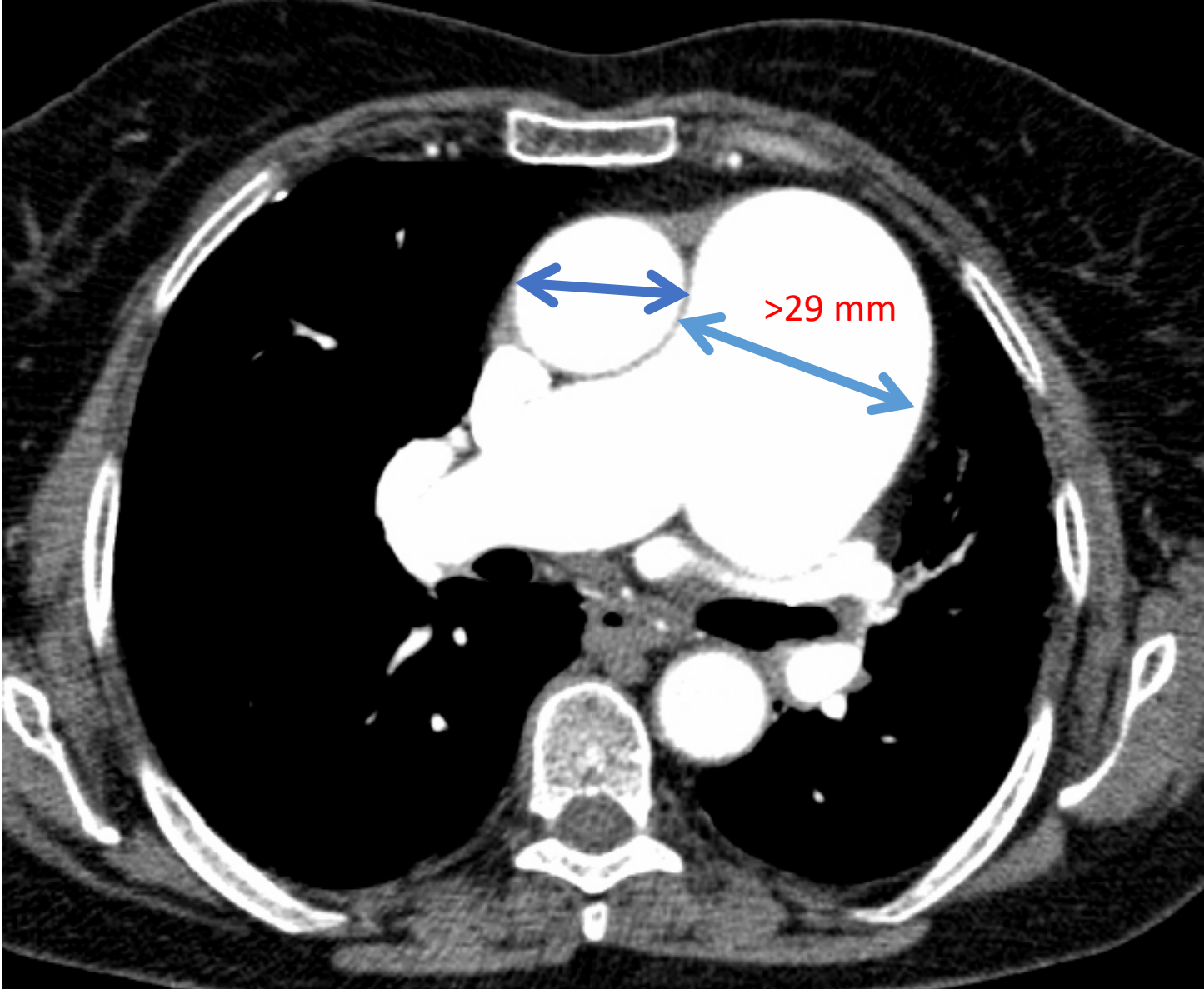
12 ay sonra

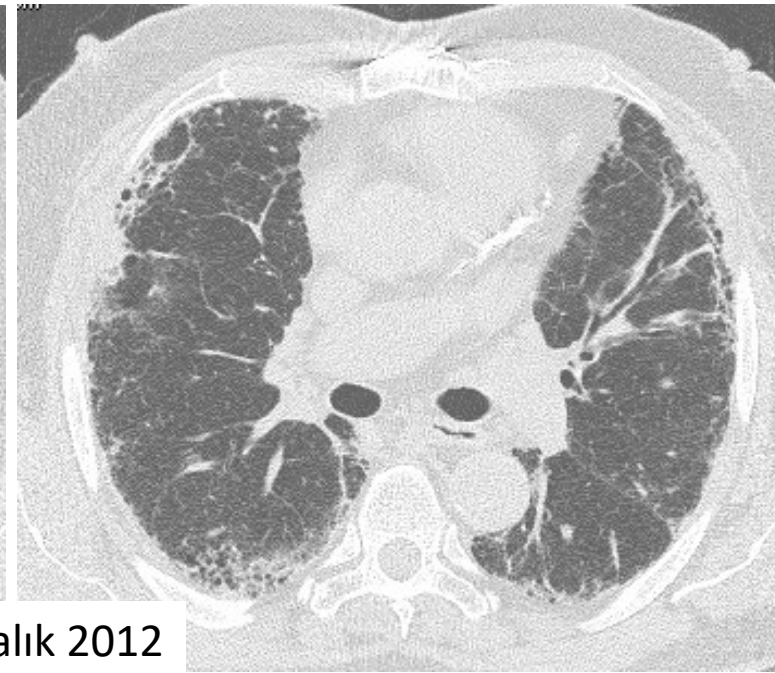
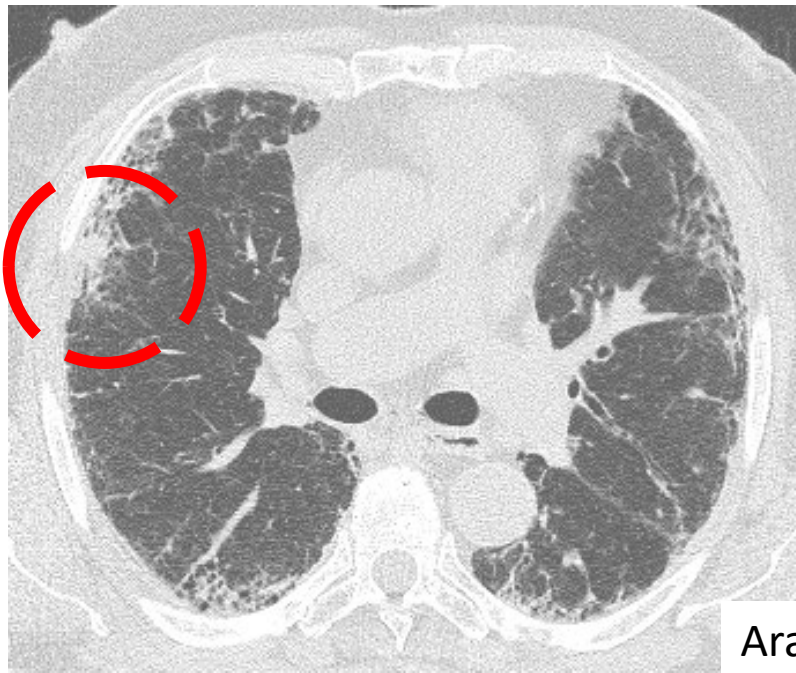
78 yaşında erkek

PROGRESYON

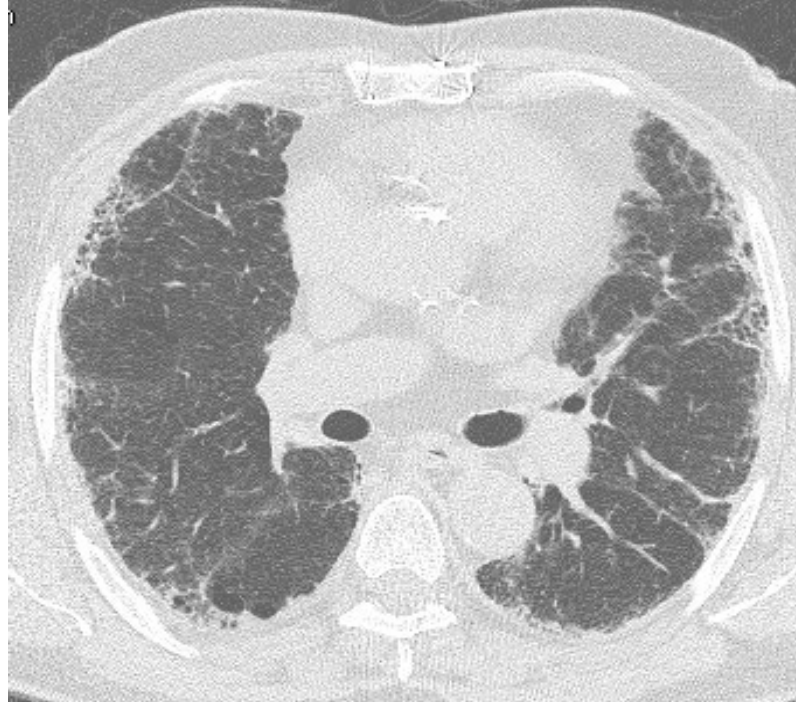


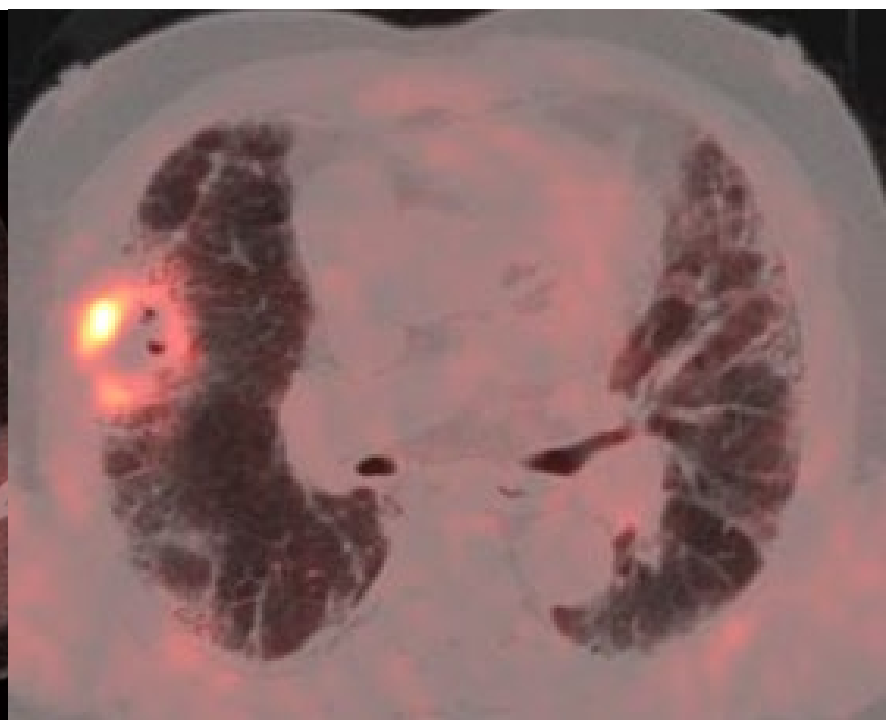
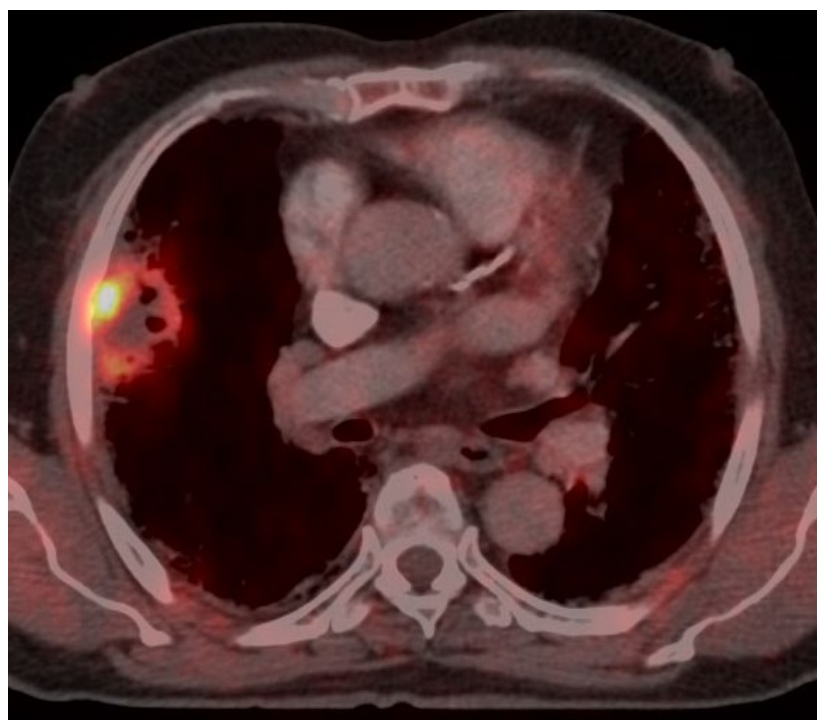






Aralık 2012





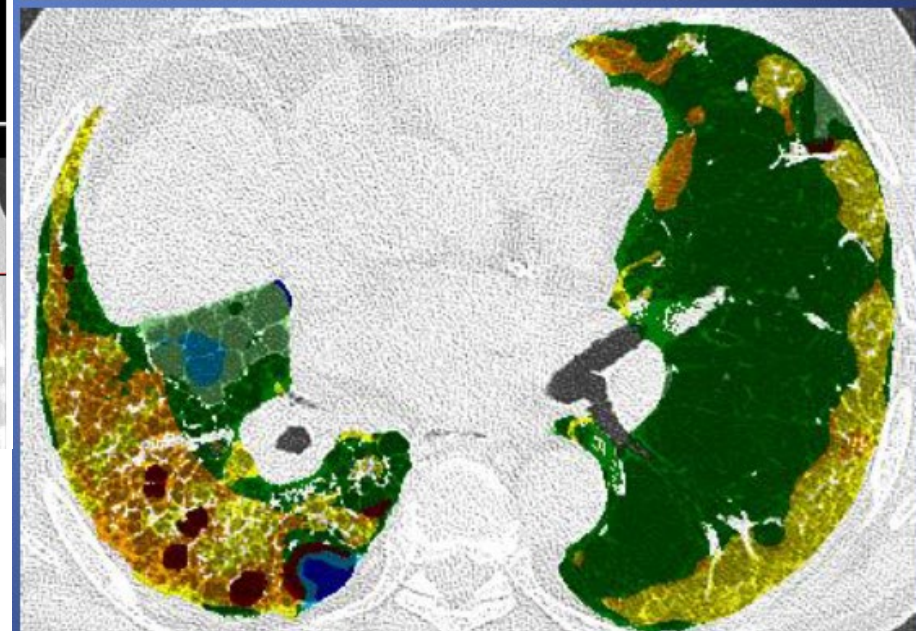
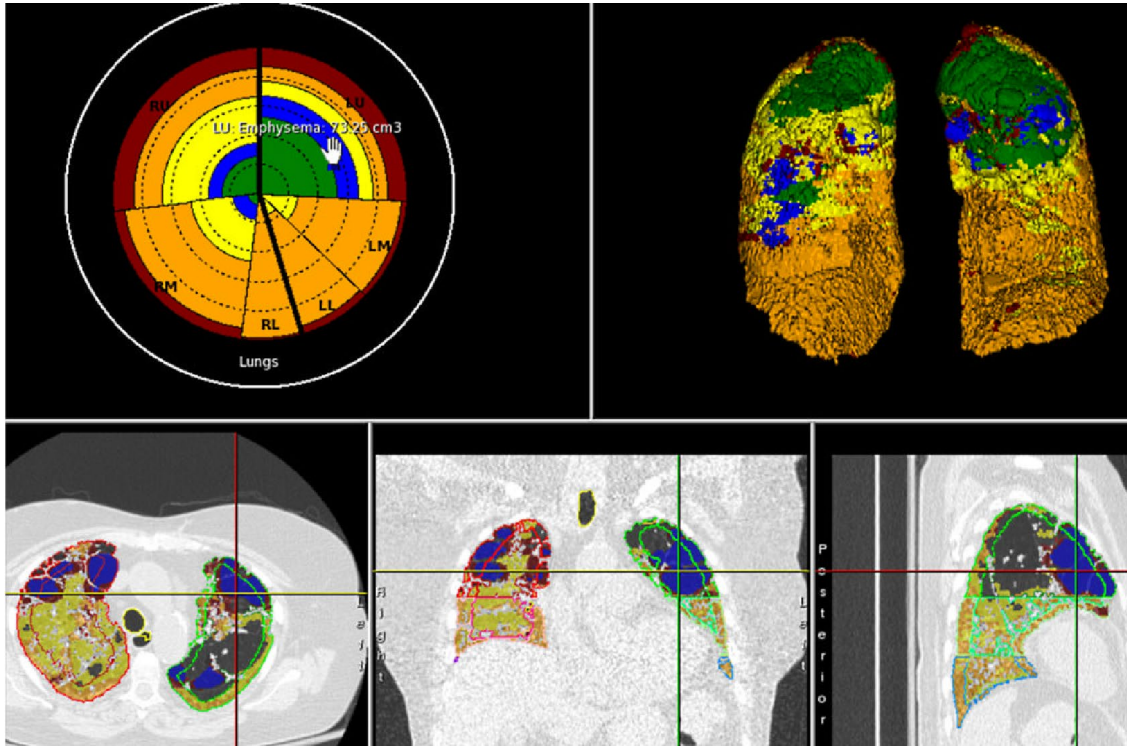
SqCC

Mayis 2014

İPF zemininde akciğer kanseri

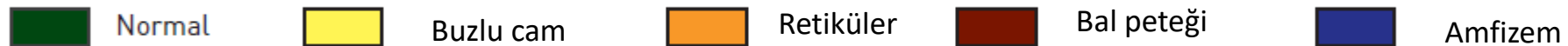
- İPF de akciğer kanseri riskinin artmış olabileceği söyleniyor
(fibrozisde atipik ve displastik epitelyal değişiklikler malignansi riskini artırıyor?)
- 5 yıllık takipte % 15 akciğer kanseri gelişmekte
- Kanserlerin çoğu periferik (SqCC ve adenoca)
- Geniş serilere ihtiyaç var

CALIPER analizi



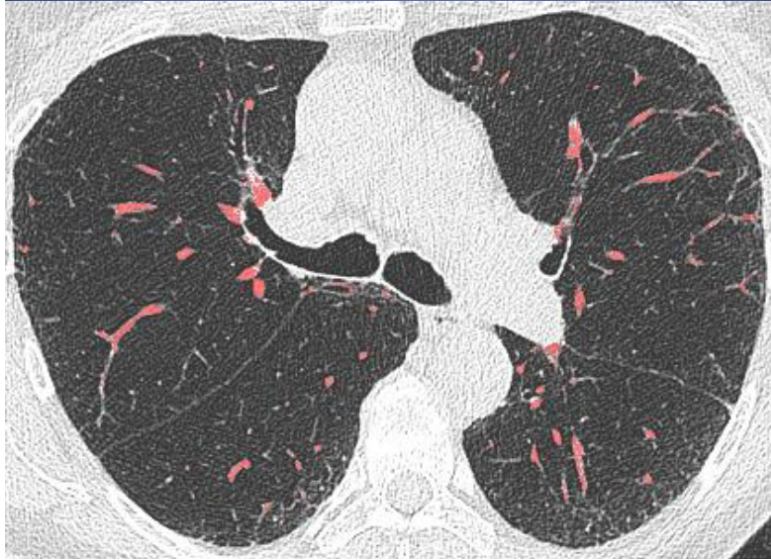
Automated quantification of radiological patterns predicts survival in idiopathic pulmonary fibrosis.

Maldonado F et al Eur Respir J 2014; 43: 204–212



PULMONER VASKÜLER VOLÜM

Minimal fibrozis

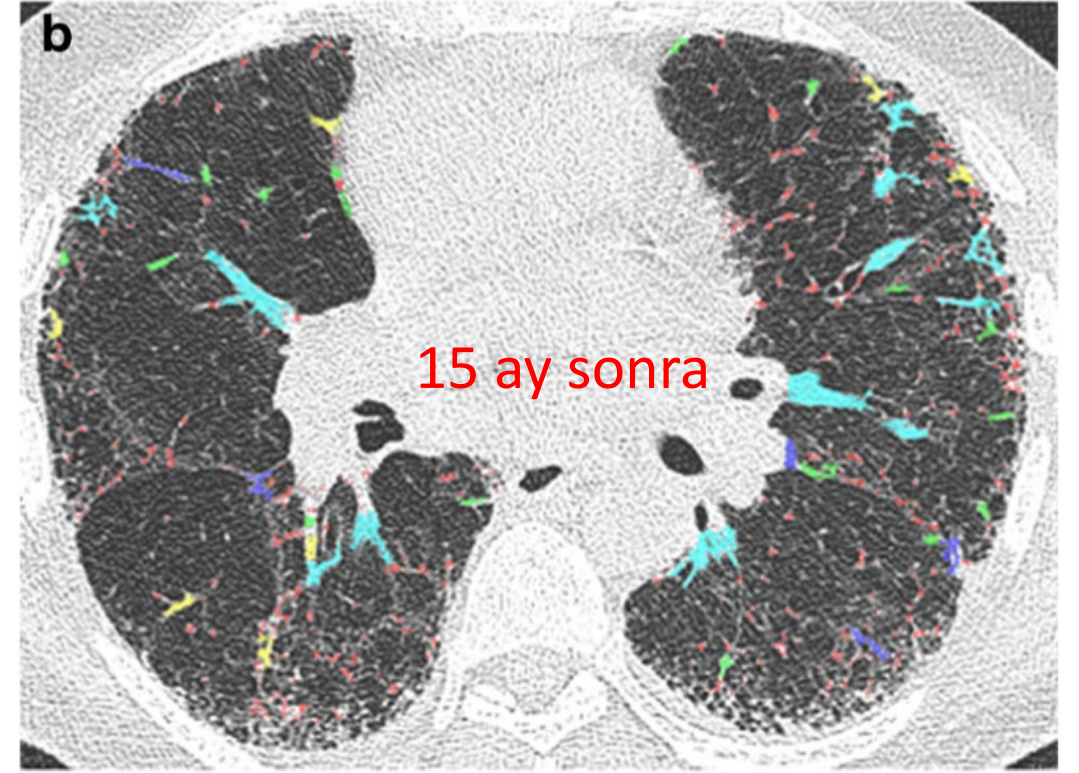
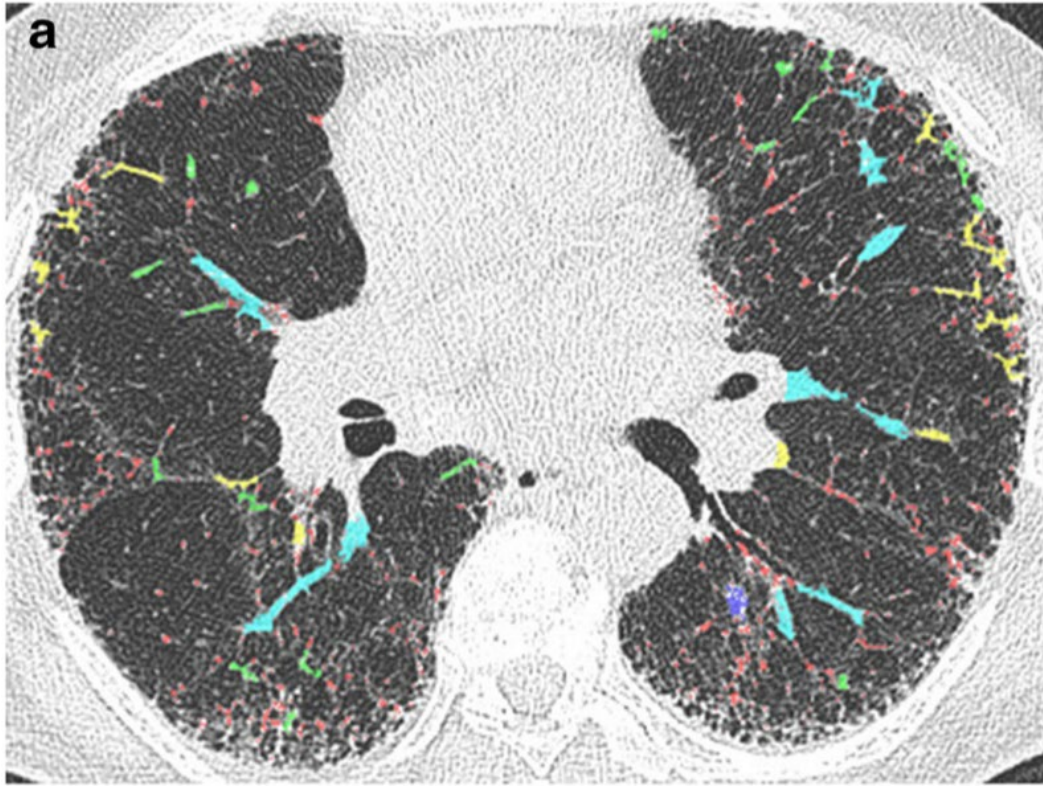


Orta fibrozis



Ciddi fibrozis





CALIPER (Computer Aided Lung Informatics for Pathology Evaluation and Rating) Vessels are colour coded according to size: $<5\text{mm}^2$ =red, $5-10\text{mm}^2$ =green, $10-15\text{mm}^2$ =yellow, $15-20\text{mm}^2$ =purple, $>20\text{mm}^2$ =blue

Pulmoner vasküler volüm % 4.6 dan % 4.9 a (retikülasyon gibi görülmekte)
İnterstisyel değişiklikler de % 13.7 den % 16.3 e çıkmış.
Göz ile ayırt etmek mümkün değil.

Serial automated quantitative CT analysis in idiopathic pulmonary fibrosis: functional correlations and comparison with changes in visual CT scores. Eur Radiol. 2017 Sep 29.

CALIPER: İPF ve MORTALİTE

- **PVV artışı**

- Zaman içinde periferik retikülasyon artışı → vasküler volüm artışı
- Bu artış **korunmuş alanda özellikle orta zonda (küçük damarlar)**

- Negatif intratorasik basınç → nonkompliyant akciğer → damarlarda genişleme

veya

- Plevraparankimal veya bronşiyal arter-PA anastomozlar

Mortality prediction in IPF: evaluation of automated computer tomographic analysis with conventional severity measures. Jacob et al. Eur Respir J 2016

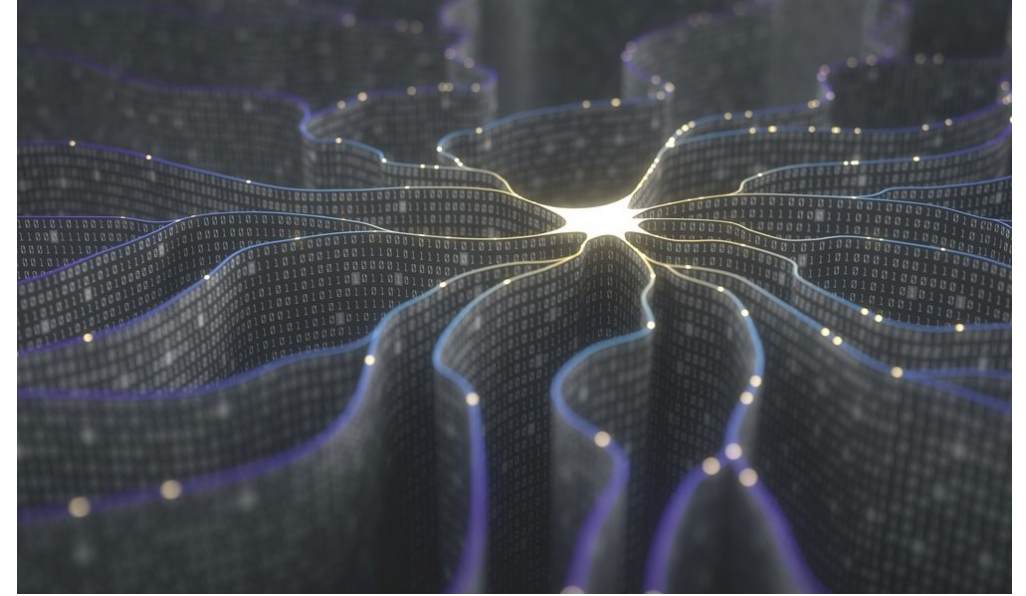
CALIPER : İPF VE MORTALİTE

- **PVV artışı mortalite için önemli bir belirteç**
- Amfizem klinik olarak anlamlı değil
- IPF patogenezinde angiogenesis? PVV artışı ?

Mortality prediction in IPF: evaluation of automated computer tomographic analysis with conventional severity measures. Jacob et al. Eur Respir J 2016

SANAL ZEKA VE İPF RADIOMICS

- Hastaların tüm verileri güçlü bilgisayarlarca depolanır.
(Radyolojik, patolojik, laboratuvar ve diğer tüm veriler)
- Daha önce verileri mevcut hastalarla kıyaslanarak **KİŞİSEL** yorum yapılır.



SONUÇ:

- İPF ile ilgili bilgilerimiz gün geçtikçe artıyor ve derinleşiyor.
- Radyolojik tipik vakalarda sorun yok ama yaklaşık yarısı atipik özellikte olabilmekte
- Günümüzde bilgisayar destekli tanı giderek artıyor.
- Bu konu İPF içinde umut vaat edici görünümde.