

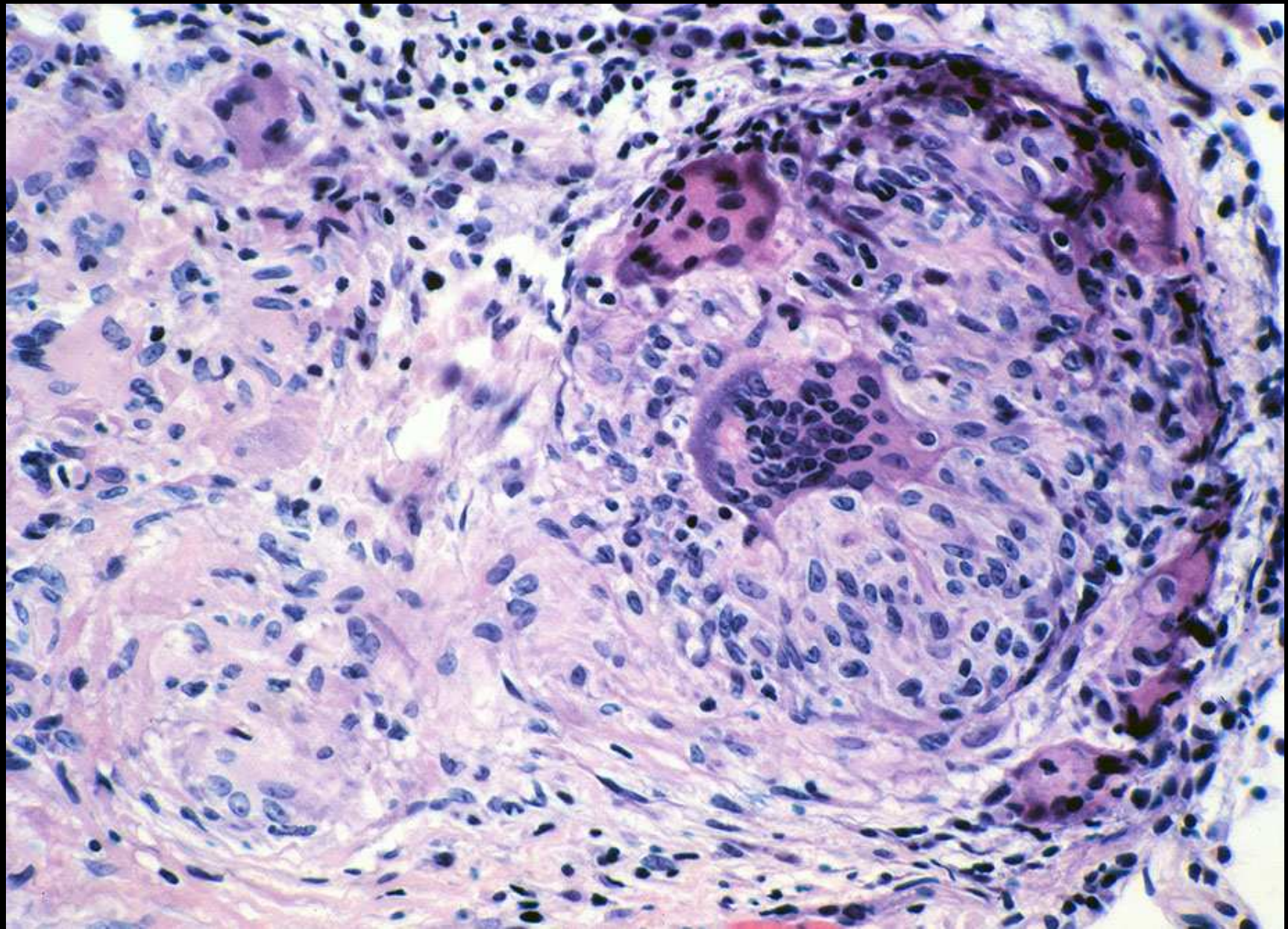
Imagerie des granulomatoses vascularites et connectivites

Emmanuel Coche, MDPHD
Cliniques Universitaires St-Luc
Brussels-Belgium

DES Radiologie
2ème cycle
14 novembre 2015

I/Granulomatoses

- Sarcoidose
- Granulomatose de Wegener
- Granulomatose causées par agents infectieux
 - Mycobactéries typiques /atypiques
 - Aspergillus, coccidioidomycoses, blastomycoses.....
- Granulomatose lymphomatoïde, bronchocentrique
- Granulomatose à cellules de Langerhans...



Sarcoïdose (BBS)

- Maladie granulomateuse d'origine inconnue
- Fatigue, malaise, perte de poids, toux, dyspnée
- Erythème noueux, uvéite, lésions cutanées
- Atteinte systémique: muscles, os, yeux, reins, cœur, tractus gastrointestinal...
- R/ 50% répondent aux stéroïdes, R/immunosuppresseur

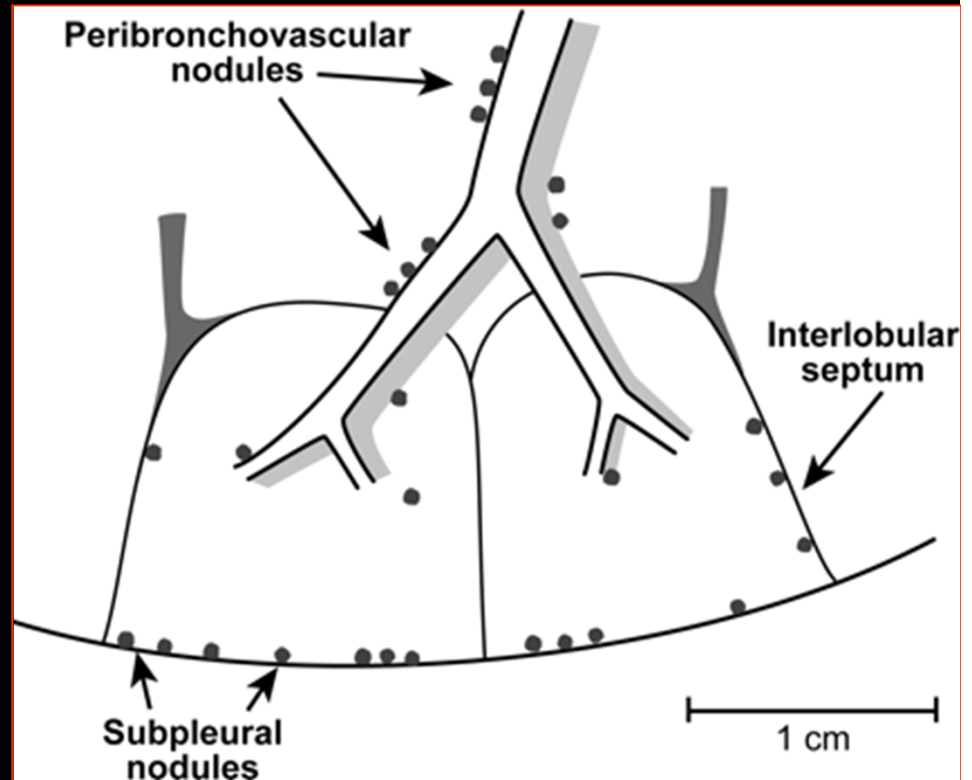
Imagerie

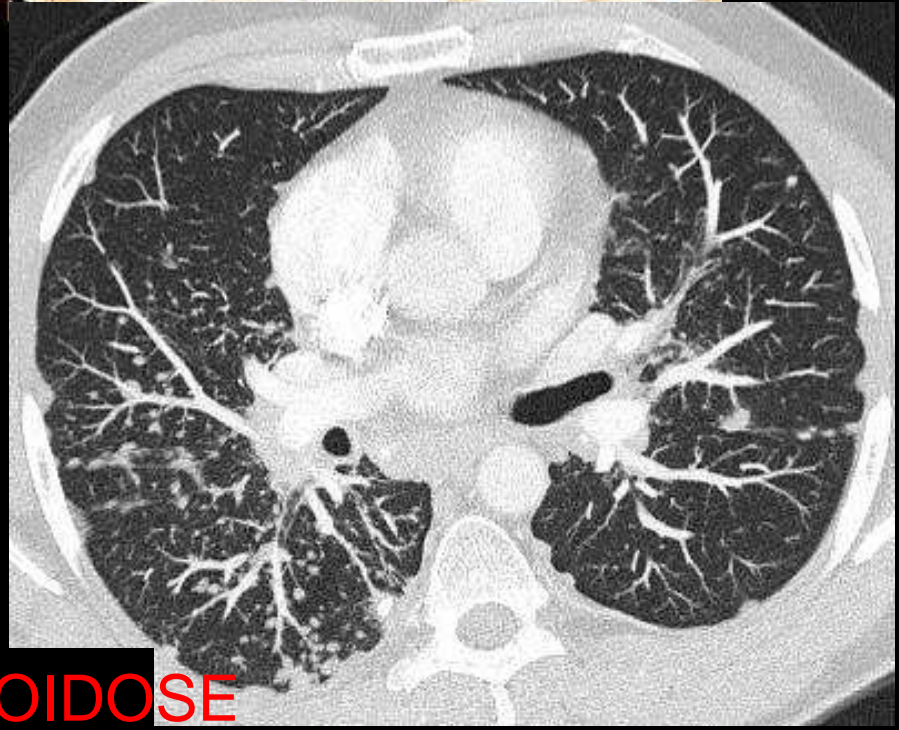
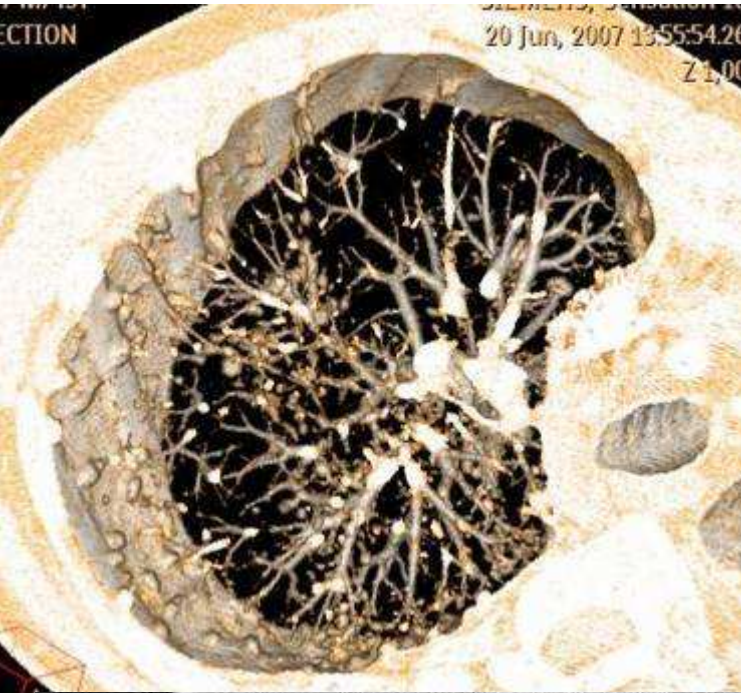
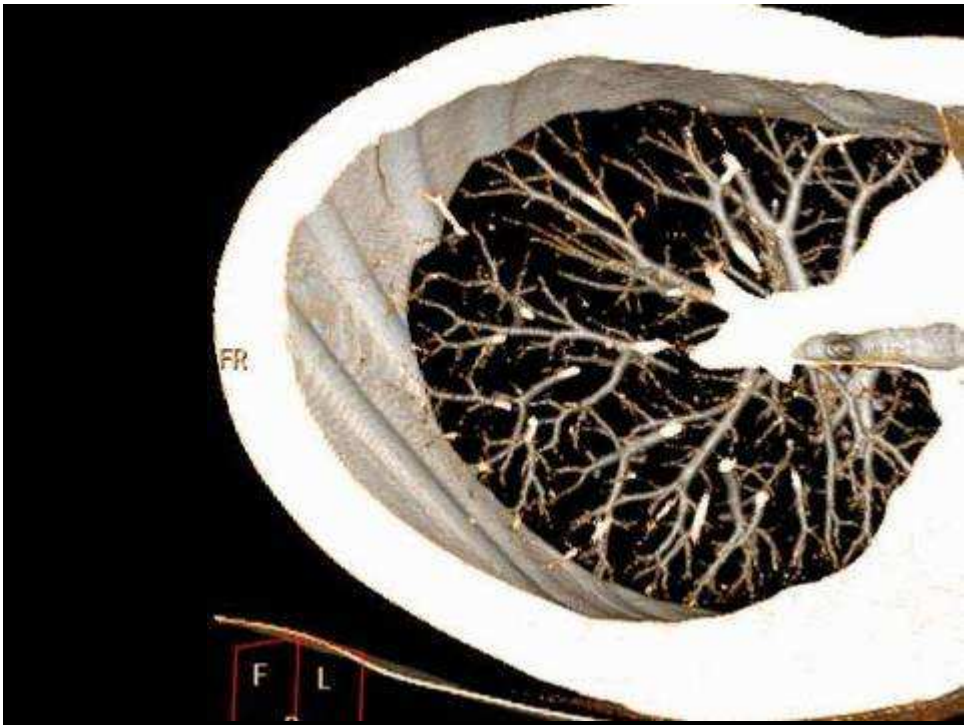
- Adénopathies hilaires et médiastinales symétriques
- Micronodules (1-5 mm)
- Atteinte centri-lobulaire, périvasculaire, lymphatique, septae sous-pleuraux
- Fibrose progressive et massive, kystes, bulles...
- Sarcoidose alvéolaire: nodules et densifications avec bronchogramme aérique

Modèle micronodulaire péri-lymphatique

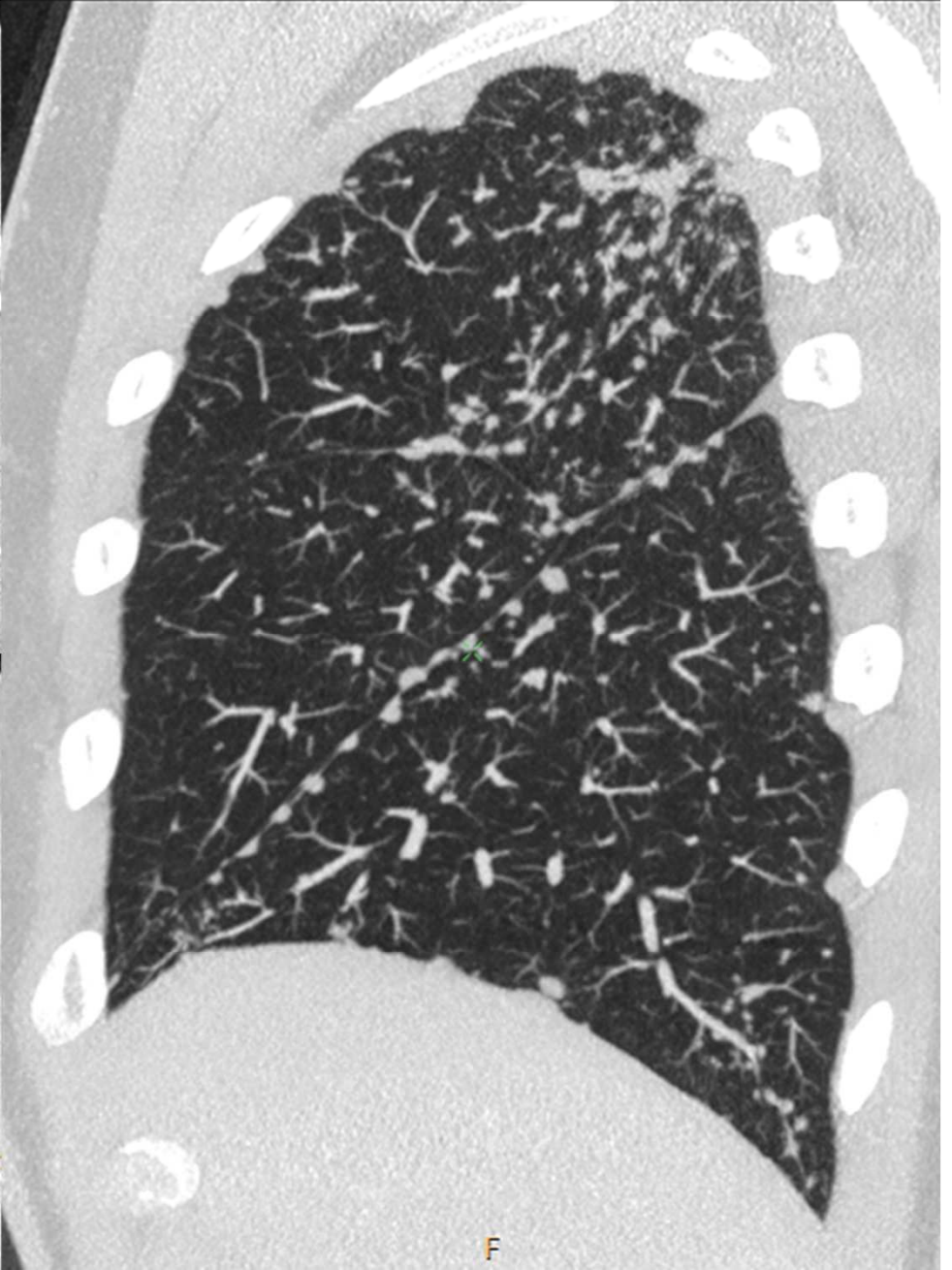
Caractéristiques :

- contours nets
- forte densité
- distribution
 - Le long des scissures et de la plèvre périphérique
 - Le long des septa interlobulaires
 - Le long des trajets vasculaires et bronchiques
 - au cœur du lobule : axes artériolo-bronchiolaires

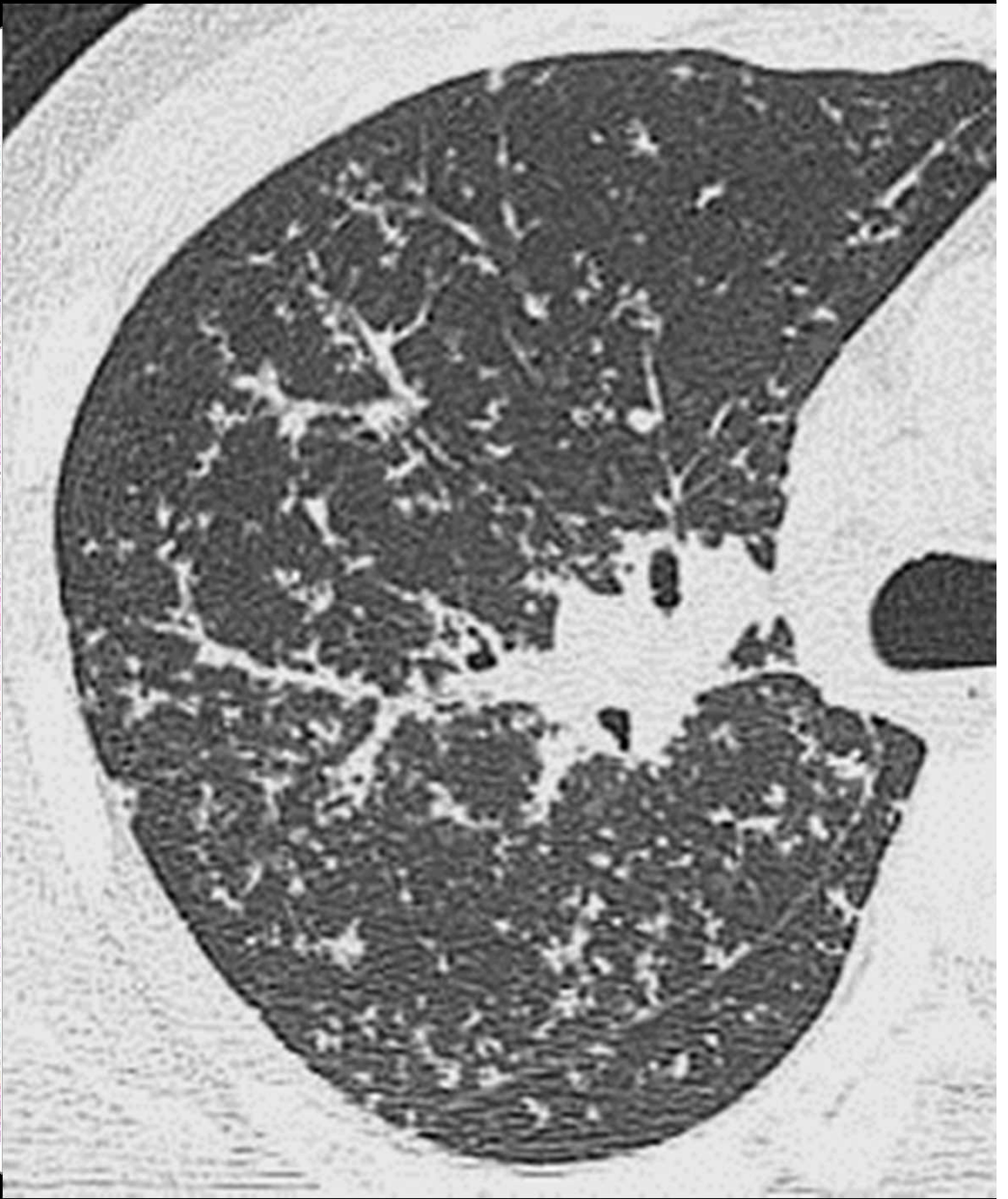
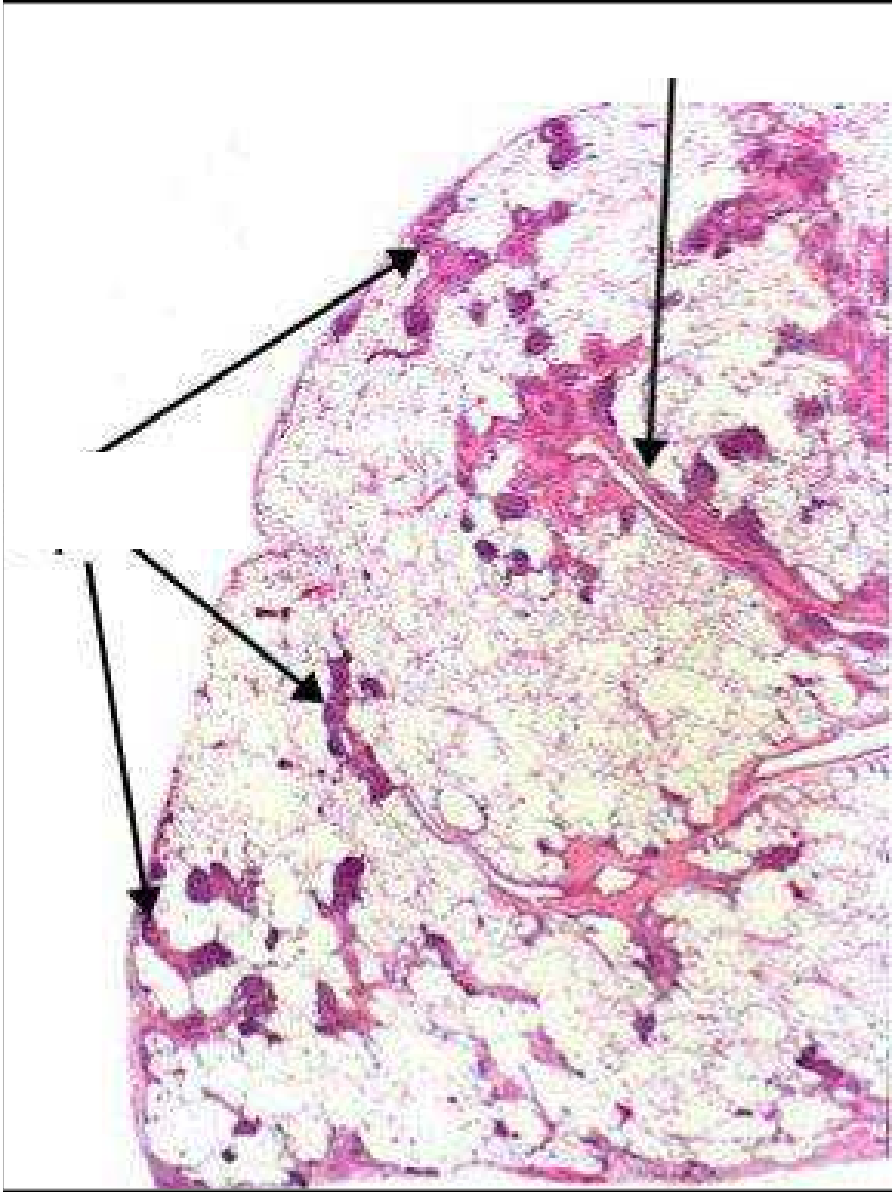


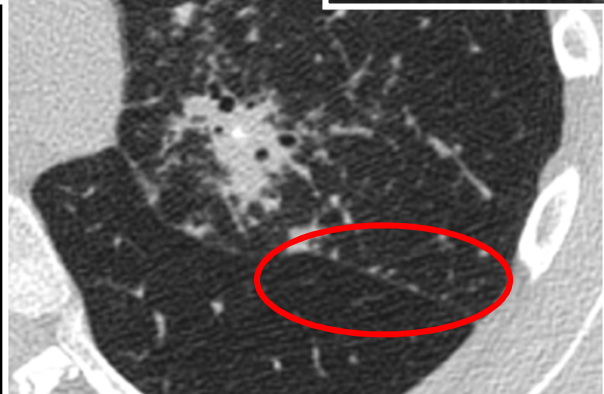
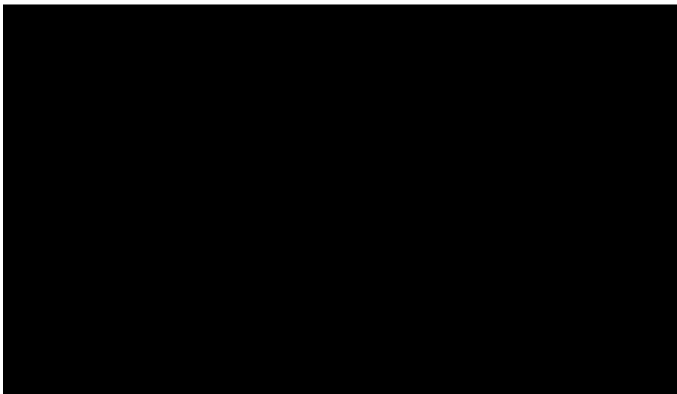
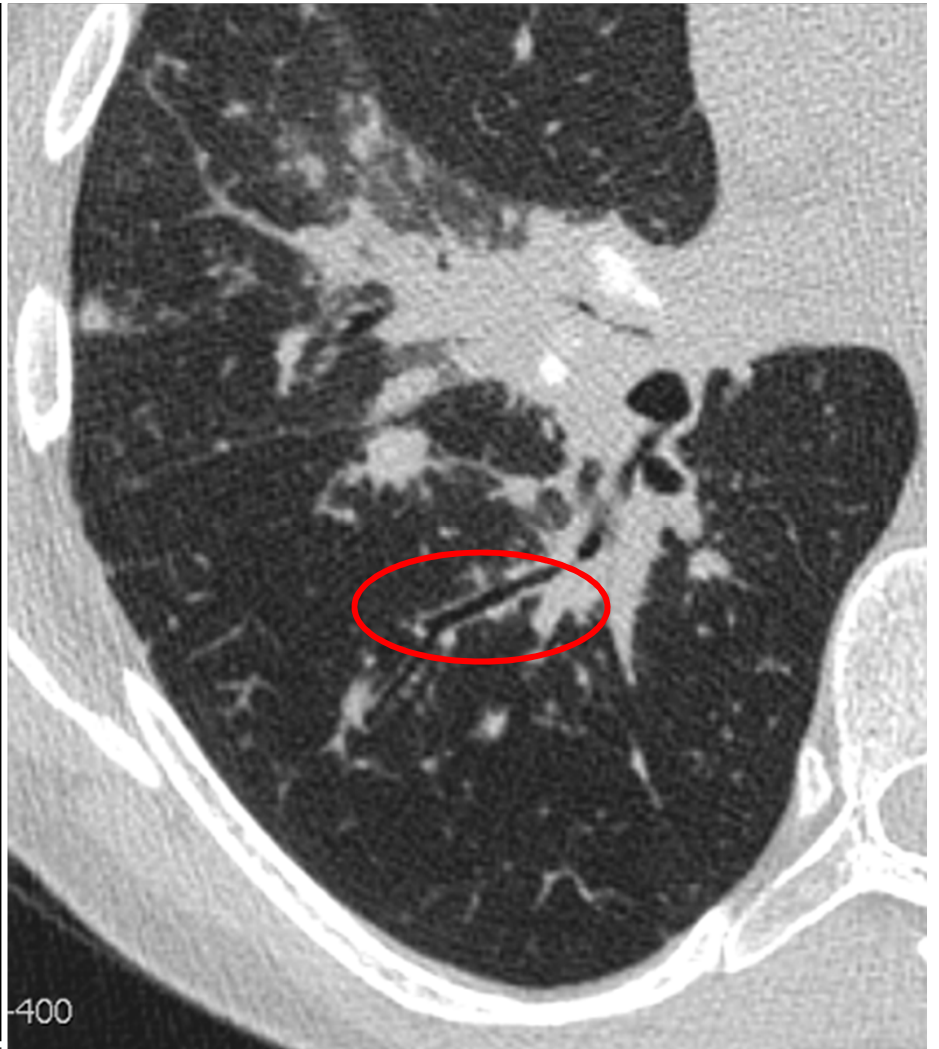
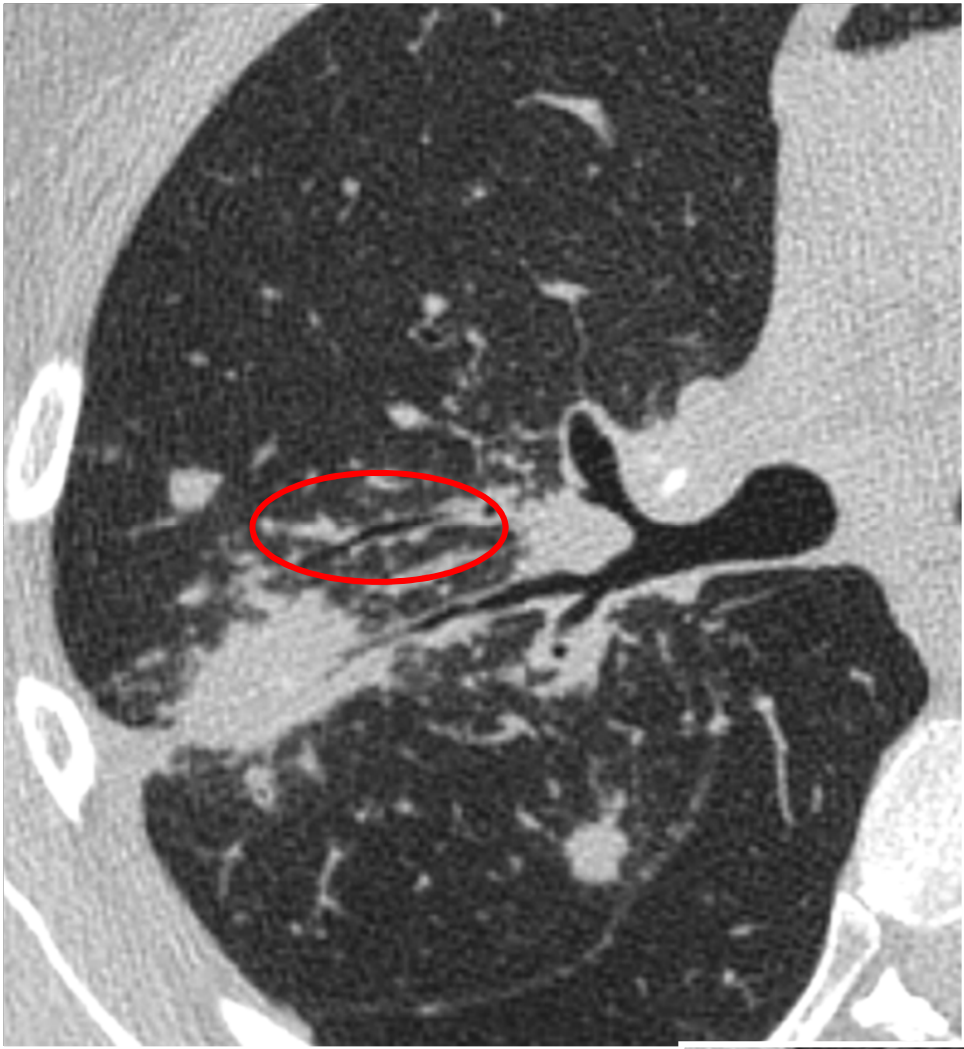


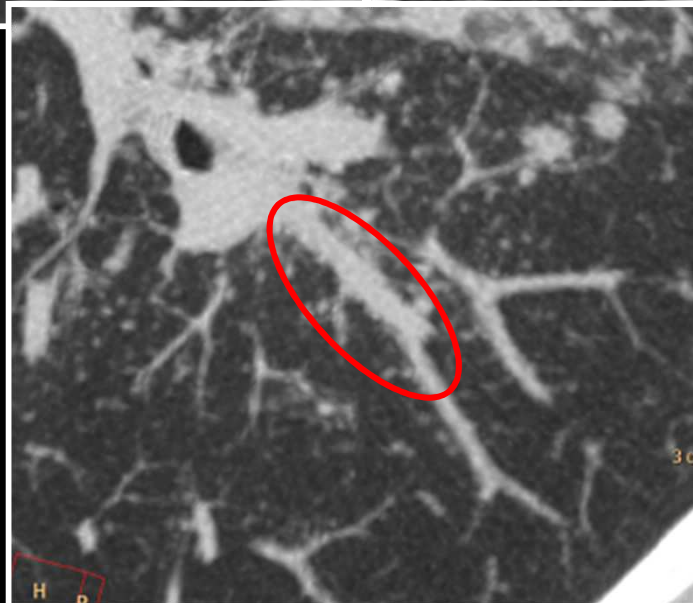
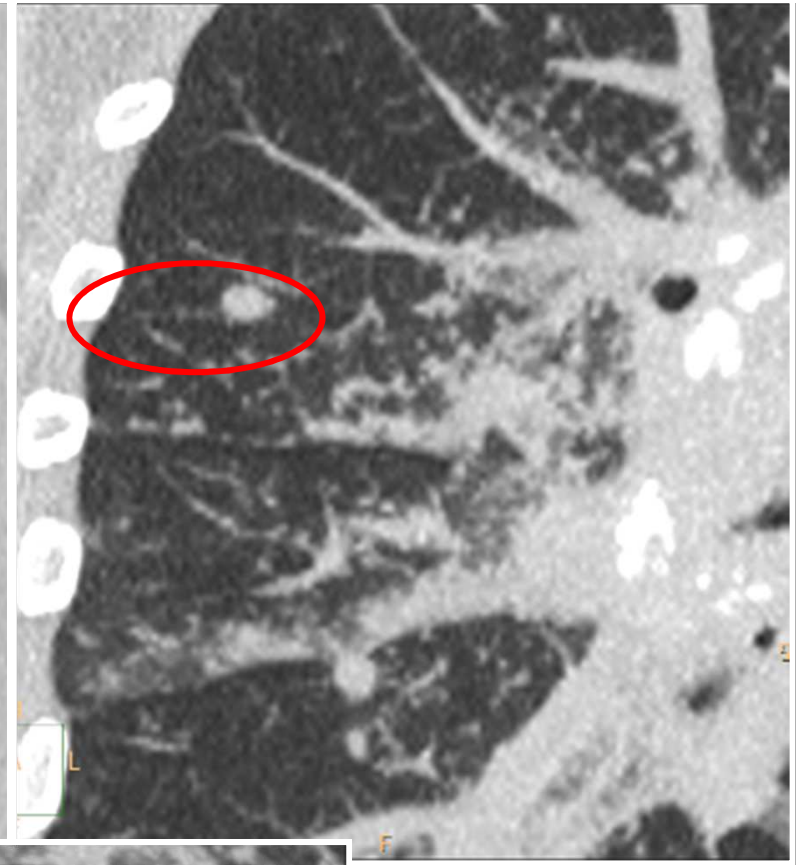
SARCOIDOSE



F

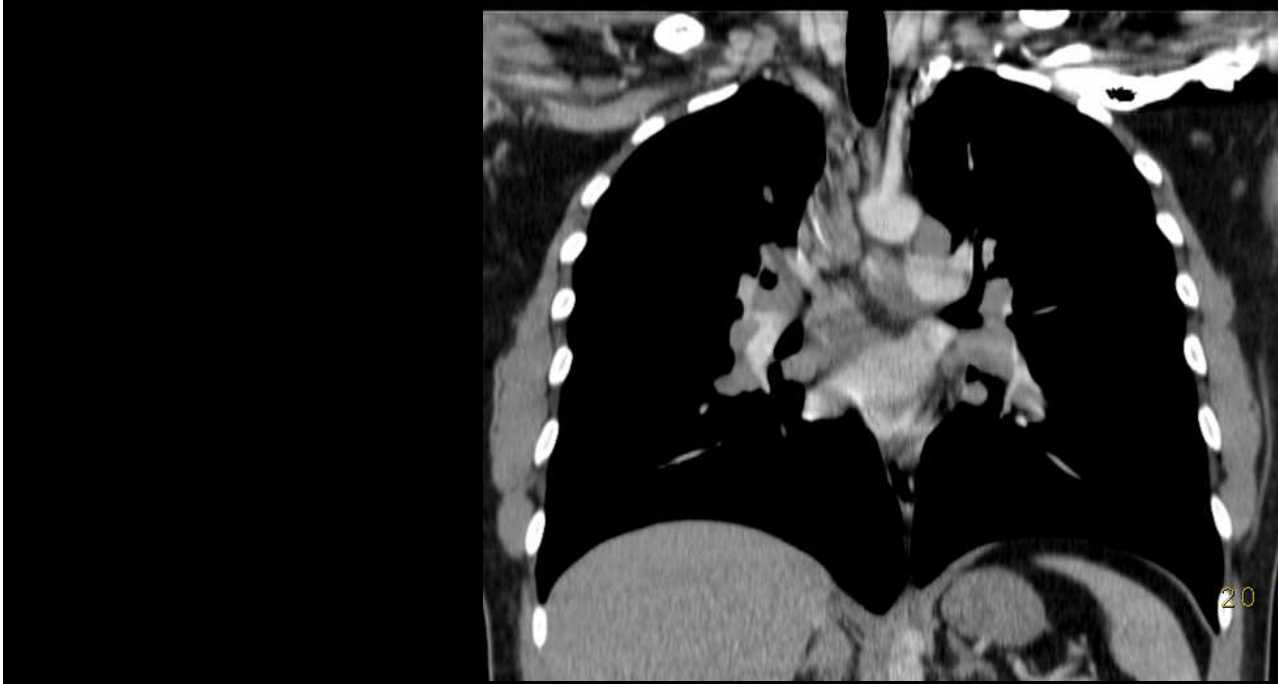
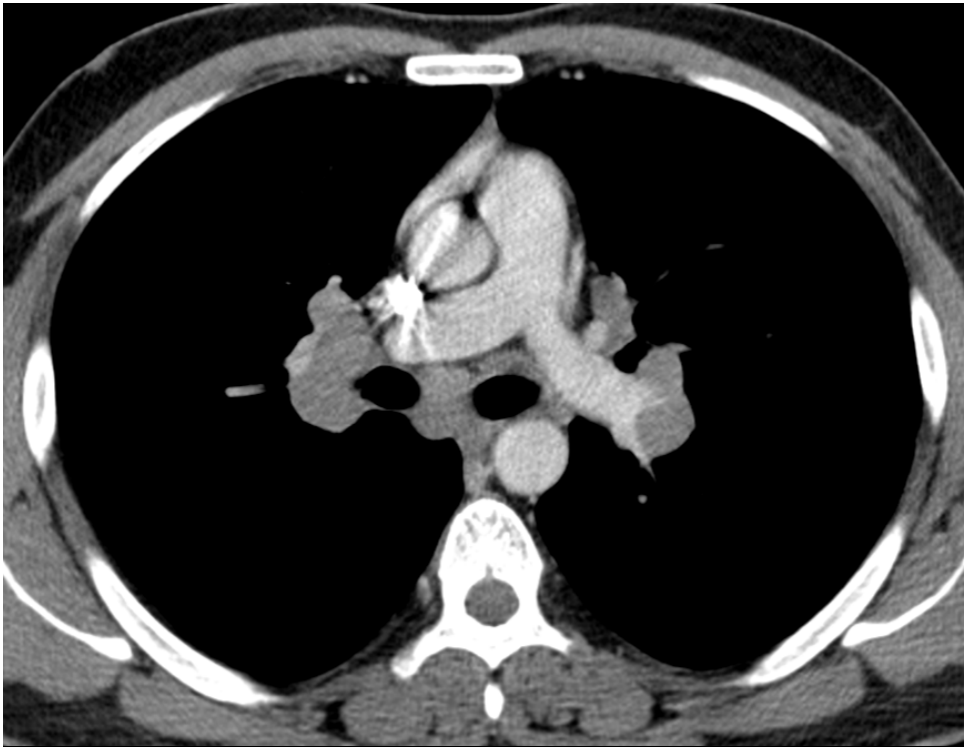


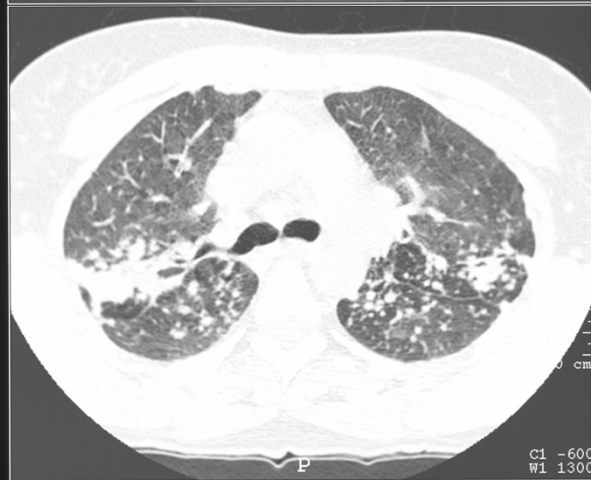




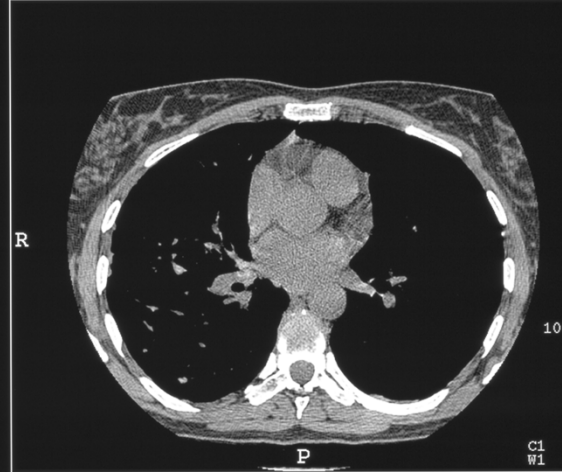
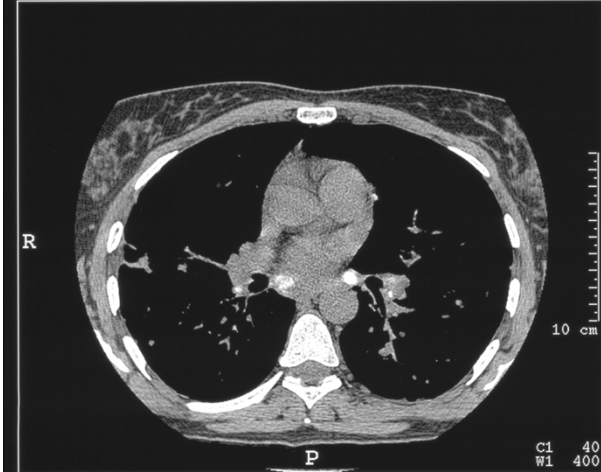
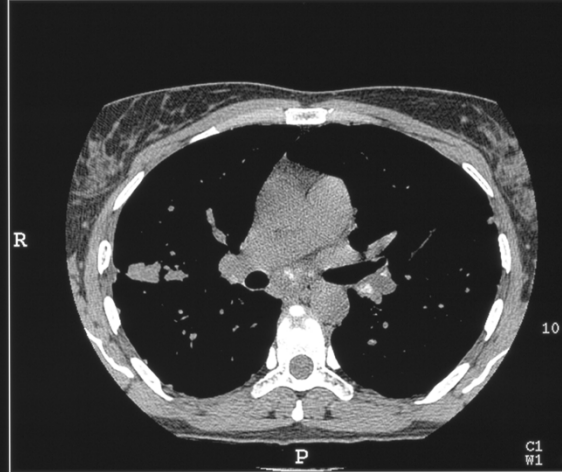
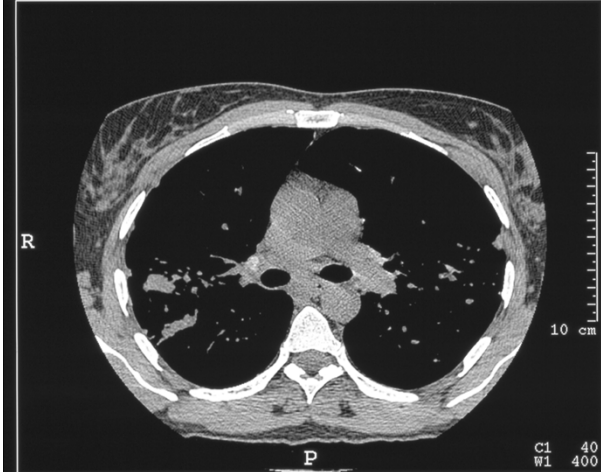
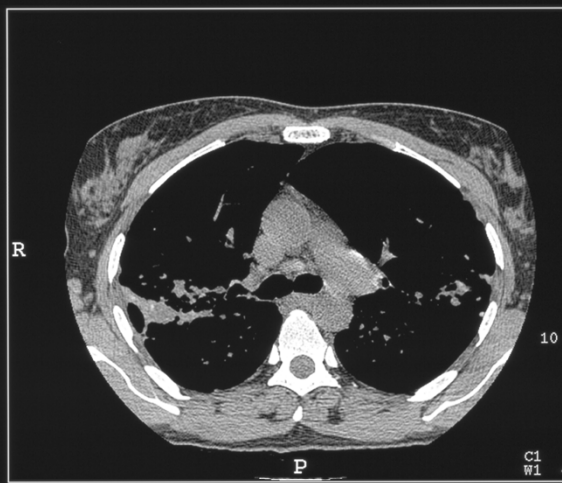
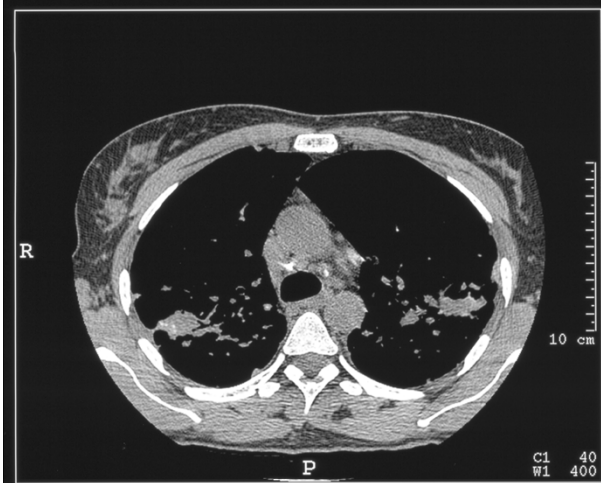
Stadification

- Stade 0: Rx normale
- Stade I: Adénopathies
- Stade II: Adénopathies/ opacités parenchymateuses
- Stade III: Opacités parenchymateuses
- Stade IV: Fibrose pulmonaire

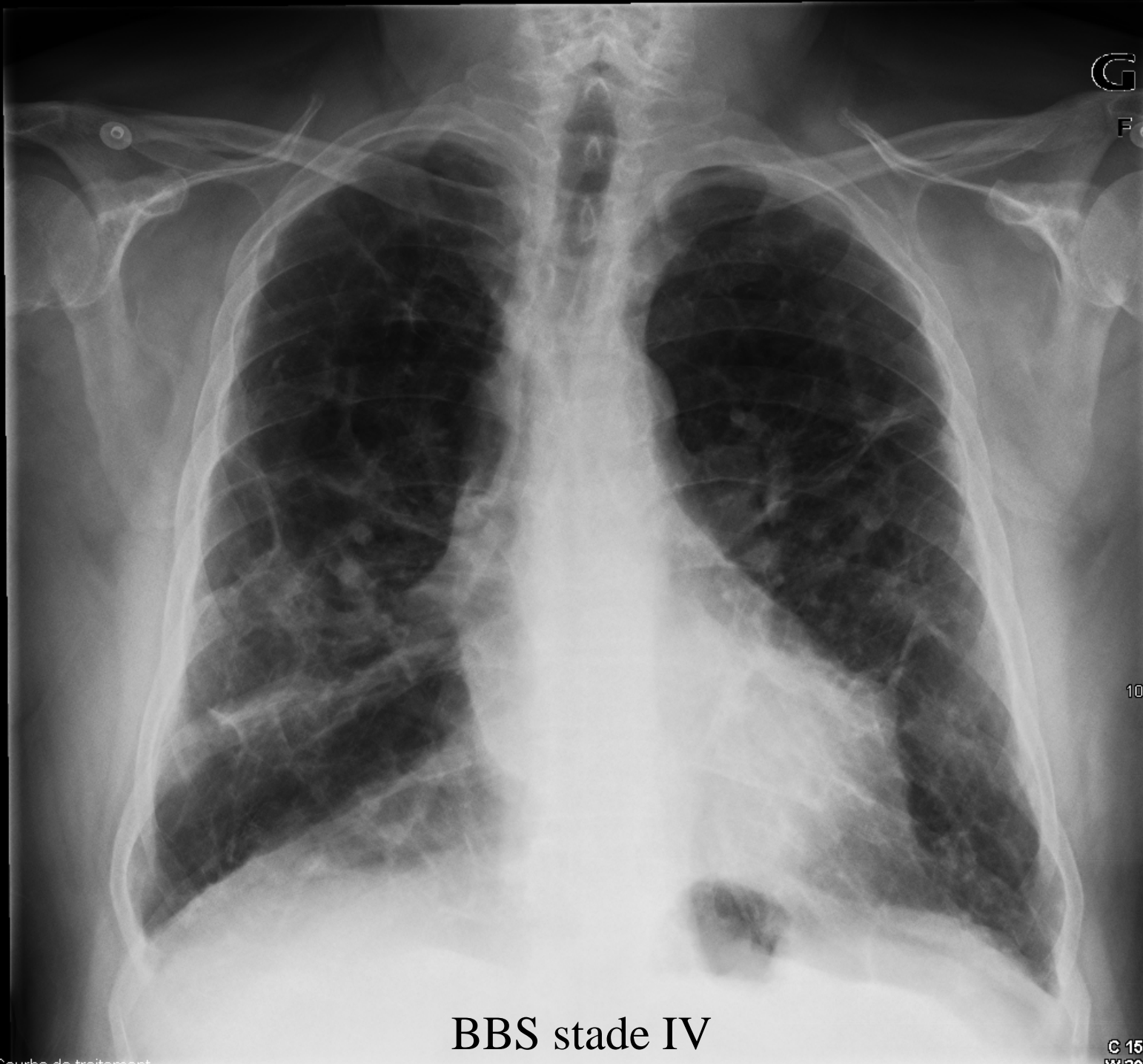




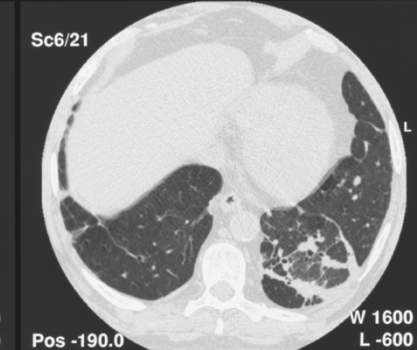
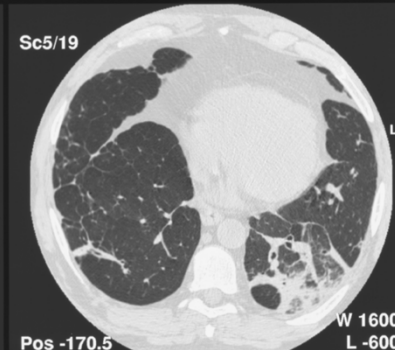
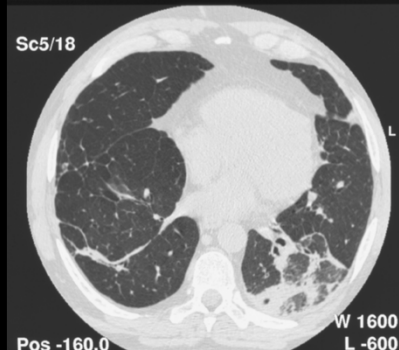
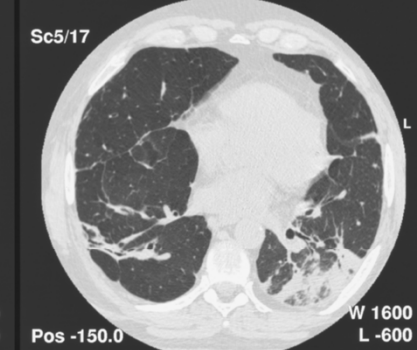
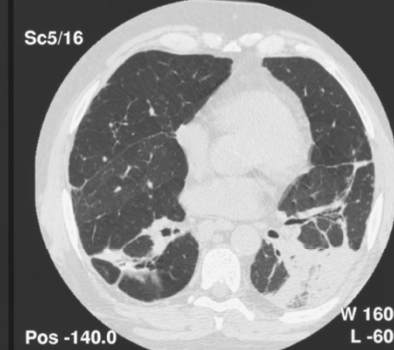
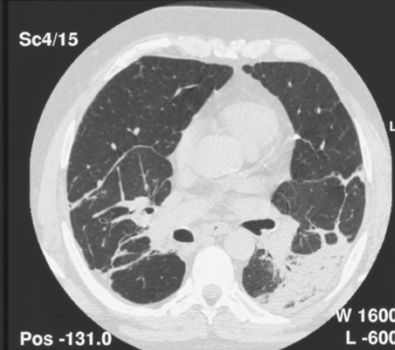
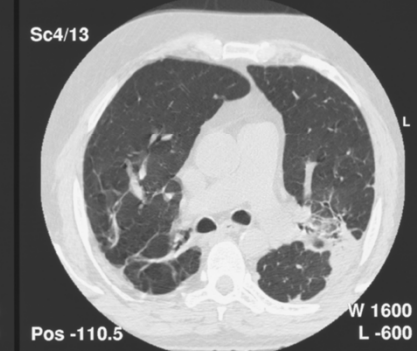
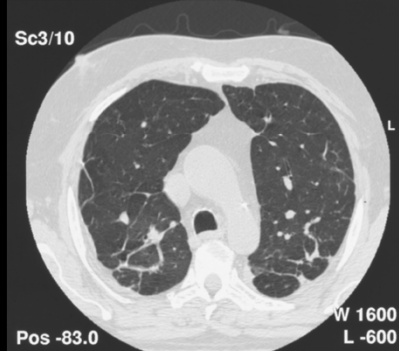
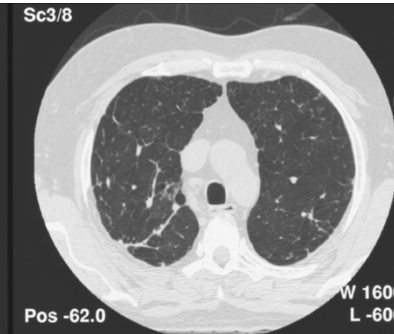
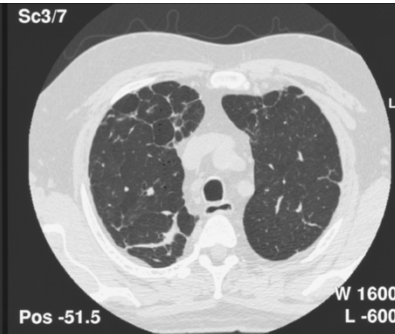
Evolution BBS

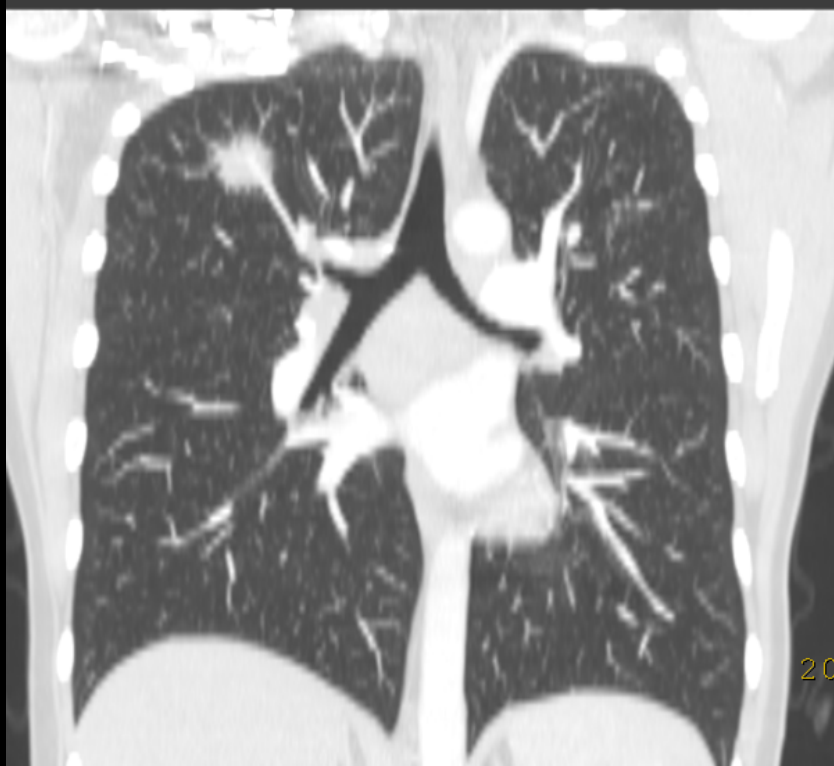
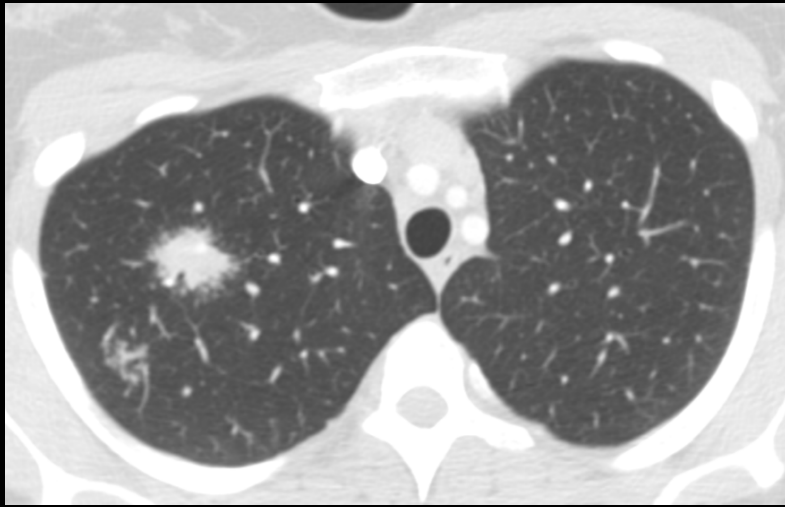


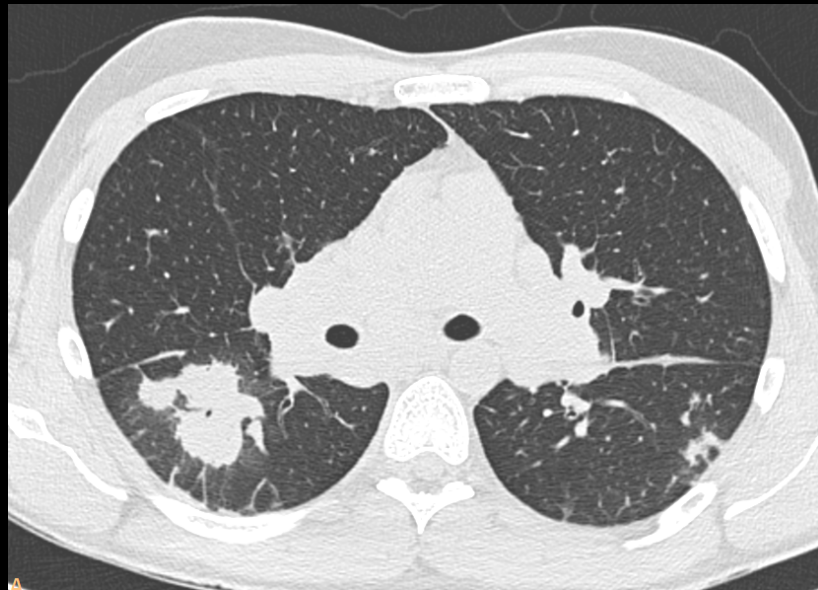
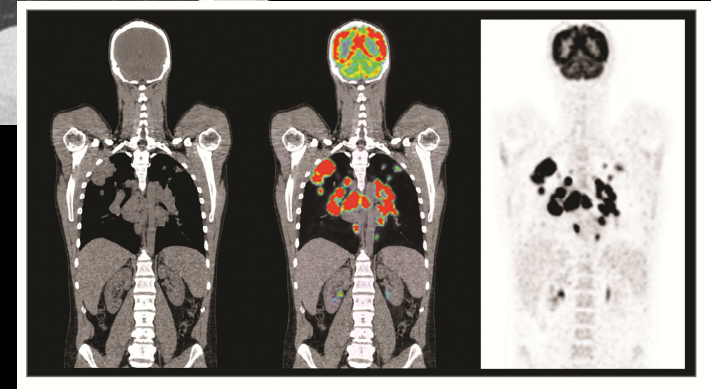
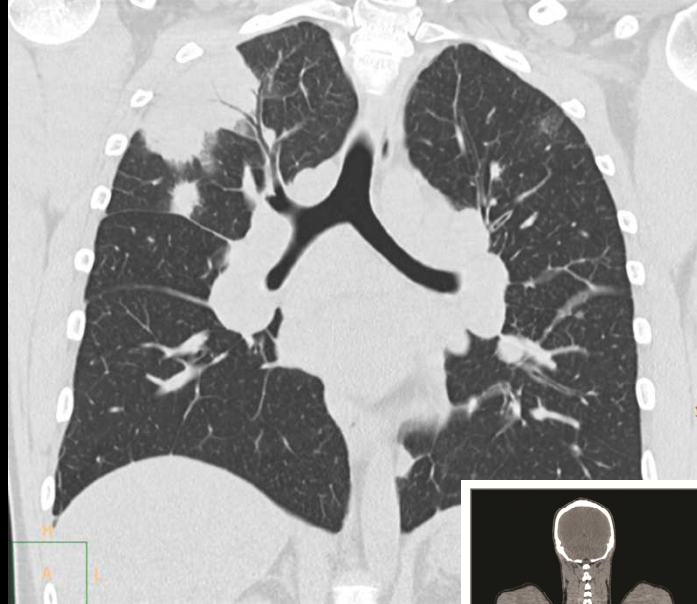
Evolution BBS



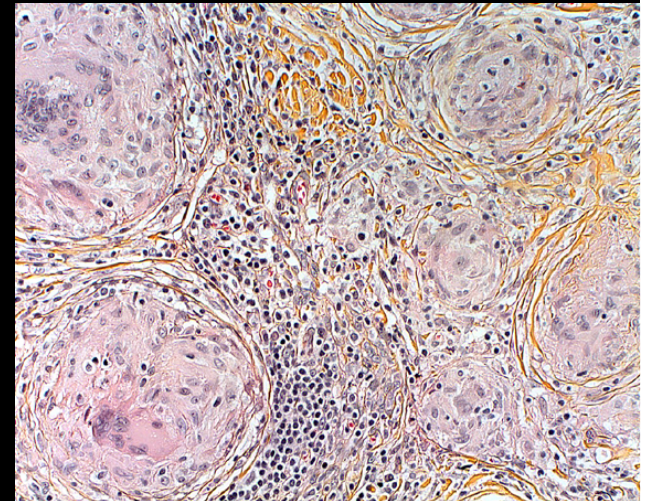
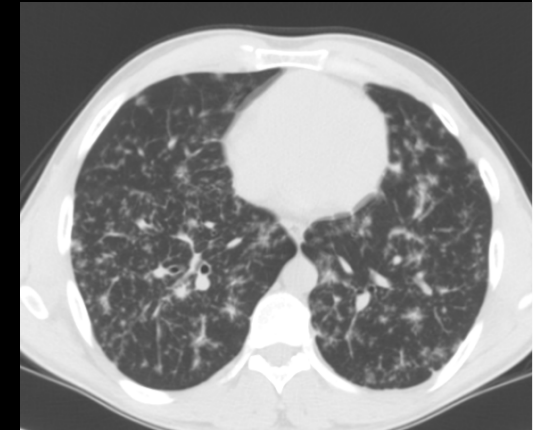
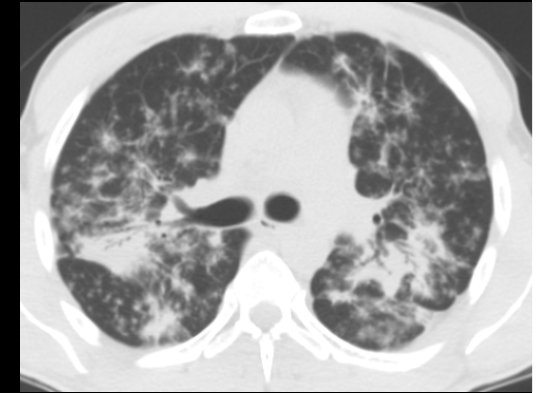
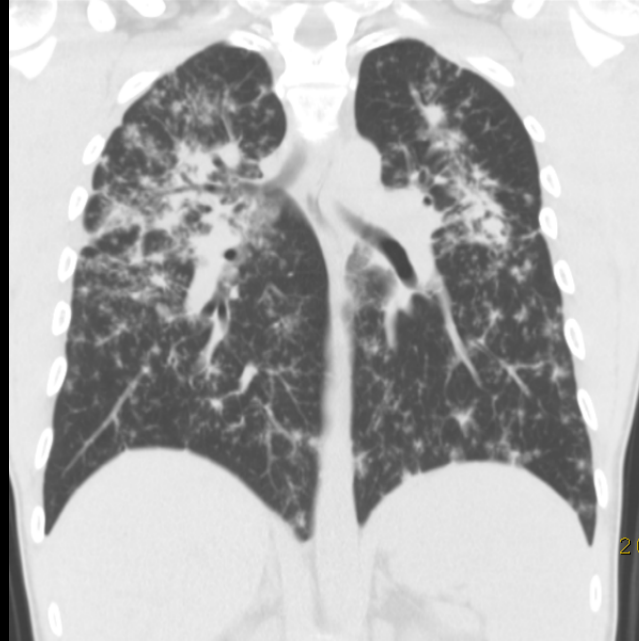
BBS stade IV







A 27-year-old Caucasian smoking male presented with a three-week history of arthralgia and fever. He had some indurated lesions on his lower limbs. Blood tests showed a raised CRP (6,4mg/dl-normal value<1) and serum lysozyme (1880 ng/ml- normal value : 379-819). Pulmonary function tests revealed a restrictive pattern.



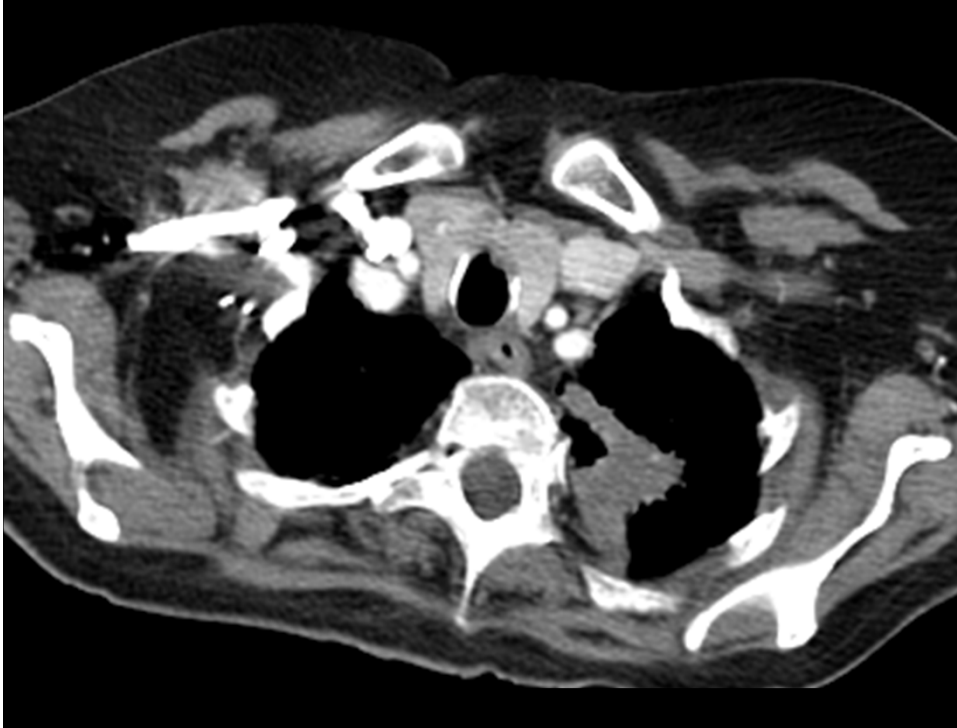
BBS multisystémique

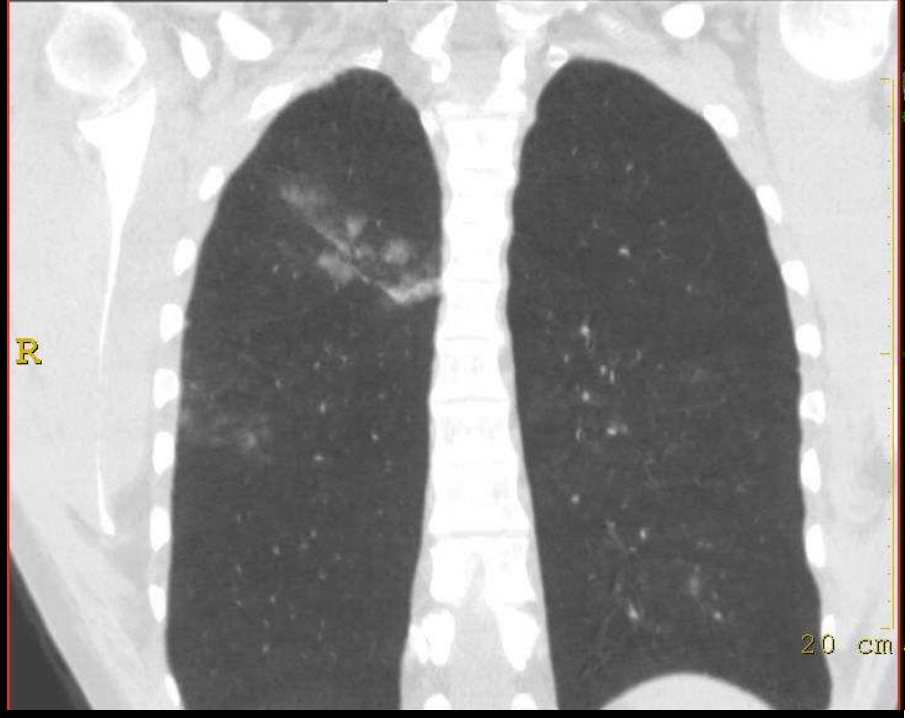
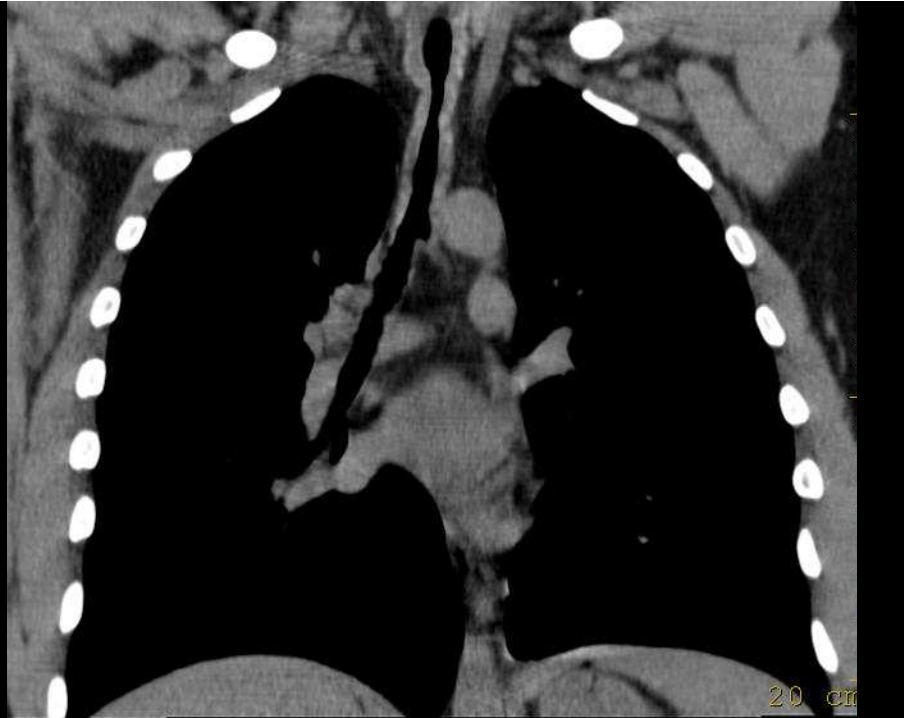
Granulomatose de Wegener

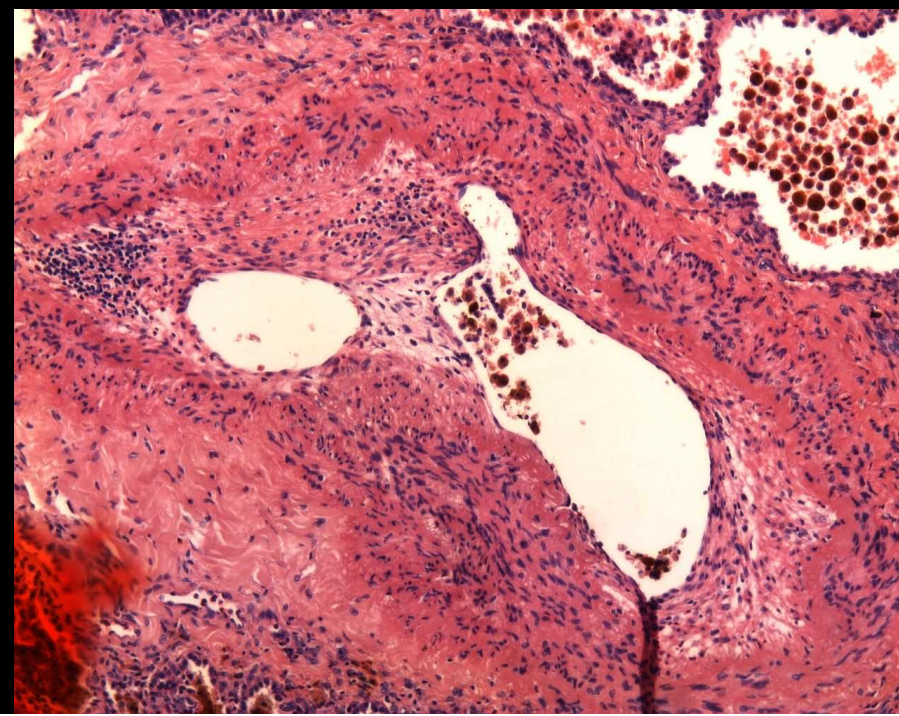
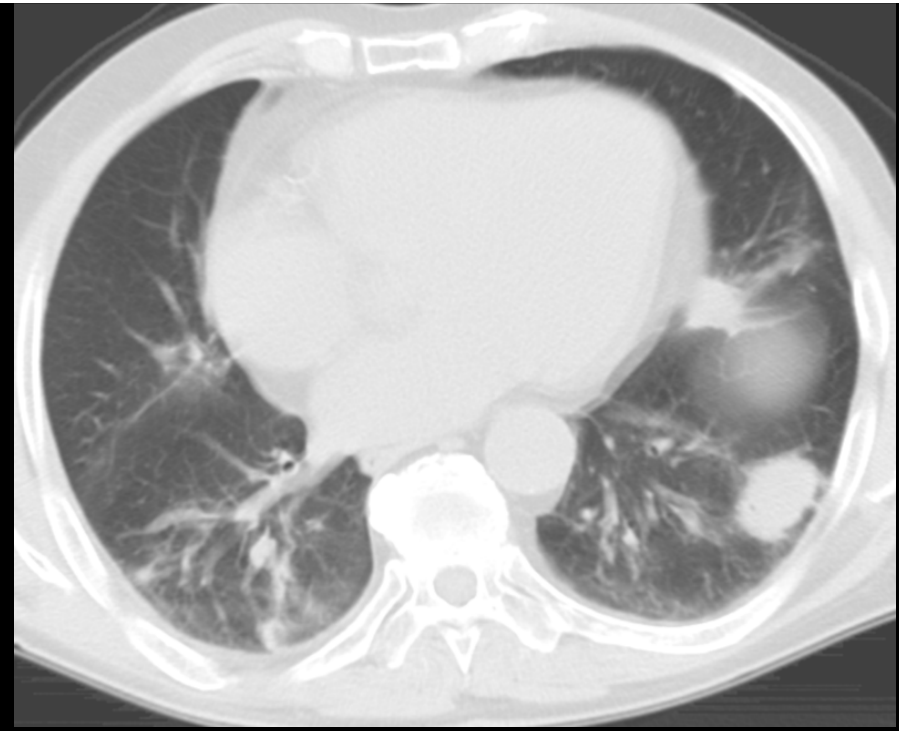
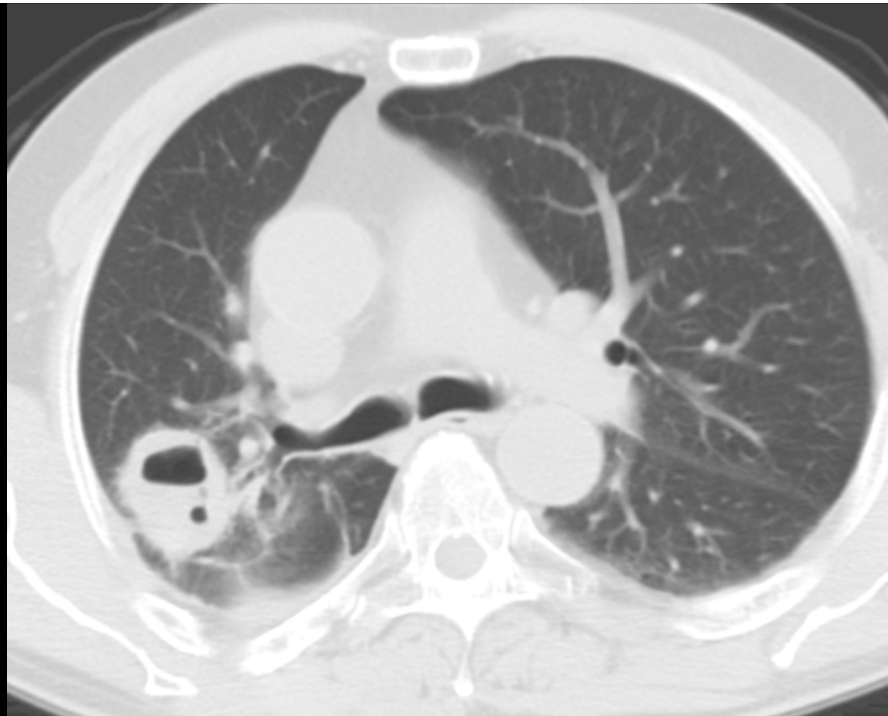
- Maladie multisystémique à expression clinique variable
- Inflammation granulomateuse nécrosante
 - Voies aériennes supérieures et inférieures
 - Reins
 - Organes systémiques et tissus
- 3/100 000 cas/an aux USA
- Pic: 30-50 ans
- C-ANCA, Anticorps anti-cellule endothéliale

Anomalies CT chez 57 patients avec granulomatose de

• Nodules-masses (1-10 cm)	165	
• Nodules excavés et masses	36	
• Densifications		50
• Verre dépoli		32
• Epaissement paroi bronchique		
– Niveau Segmentaire/ ss segm		64
– Niveau trachéal		9
– Bronchiectasies	21	
• Epcht pleural		9
• Ggl hilaire/médiastinal		

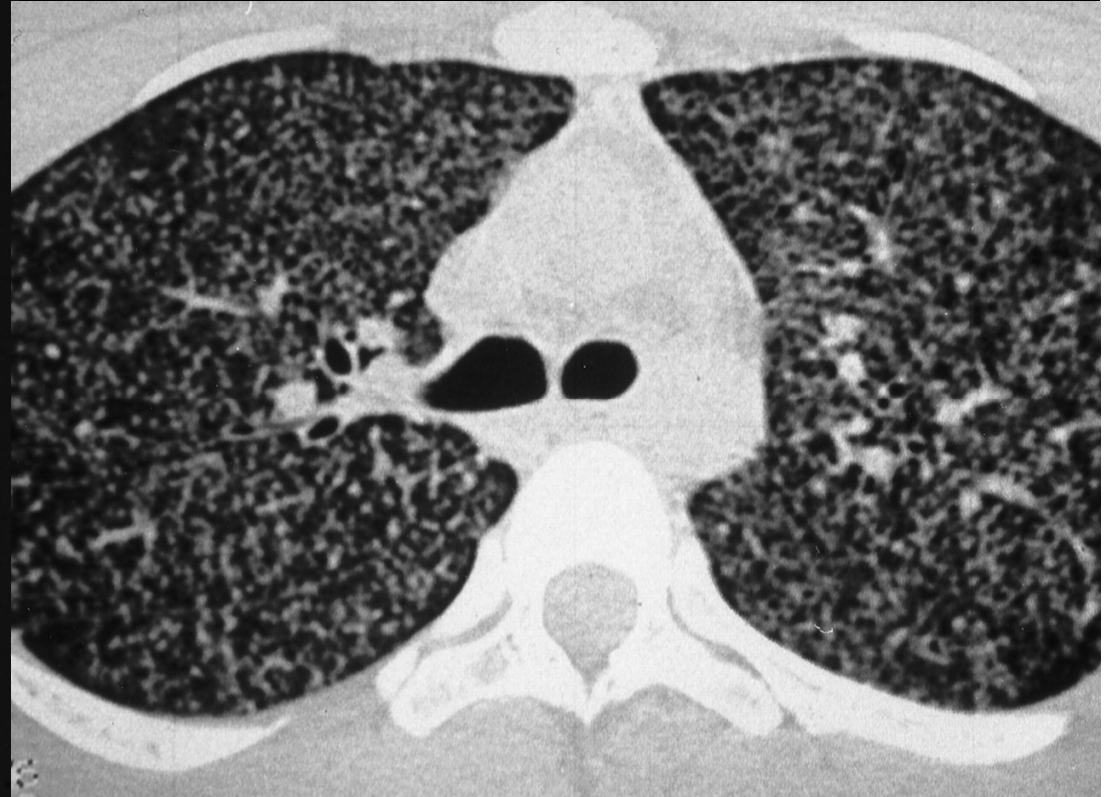






Tuberculose

- Recrudescence (HIV)-résistance
- Mycobacterium tuberculosis, non tuberculosis
- Tuberculose systémique
 - Squelette, SNC, tractus génito-urinaire
- Tbc primaire
 - Lobe moyen, lobes inférieurs, segm ant. lobe sup
 - Ganglions (nécrose centrale, périphérie rehaussante)
 - Parenchyme: densifications, masses, atélectasie
 - Epanchement pleural



Harisinghani MG et al. Tuberculosis from head to toe. Radiographics 2000;20:449-470

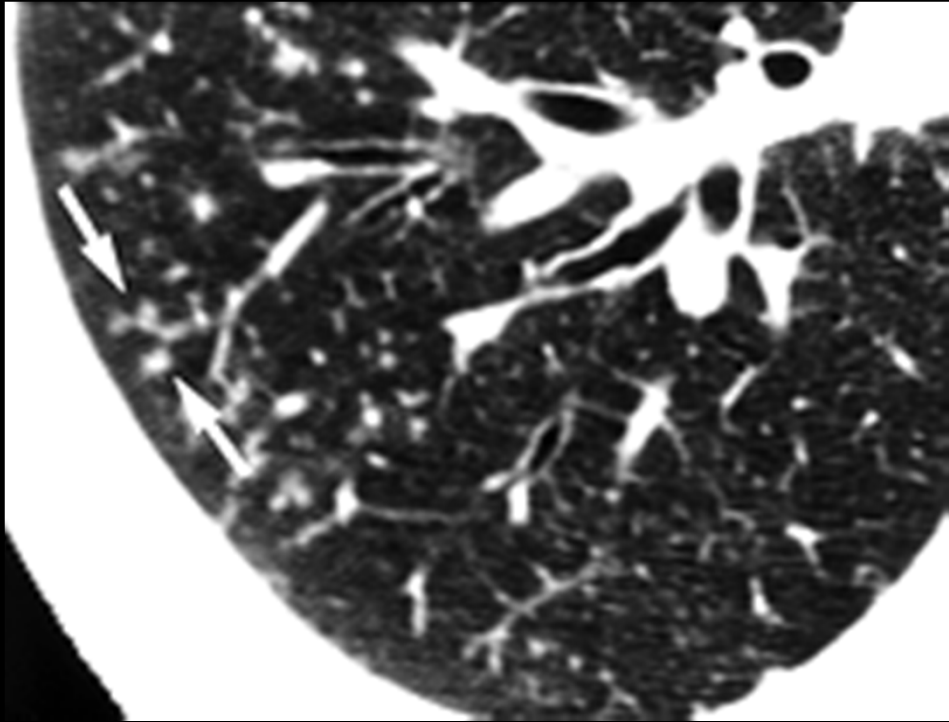
Tuberculose post-primaire

- Réactivation Tbc ¹aire dans 90% des cas
- Atteinte parenchymateuse
 - Nodules à limites floues (segments apicaux/dorsaux L sup, segments apical LI)
 - Cavitation
- Atteinte aérienne: sténose bronchique, atélectasie, pneumonie obstructive, DDB...
- Extension pleurale: collection encapsulée, fistule broncho-pleurale
- Atteinte paroi thoracique

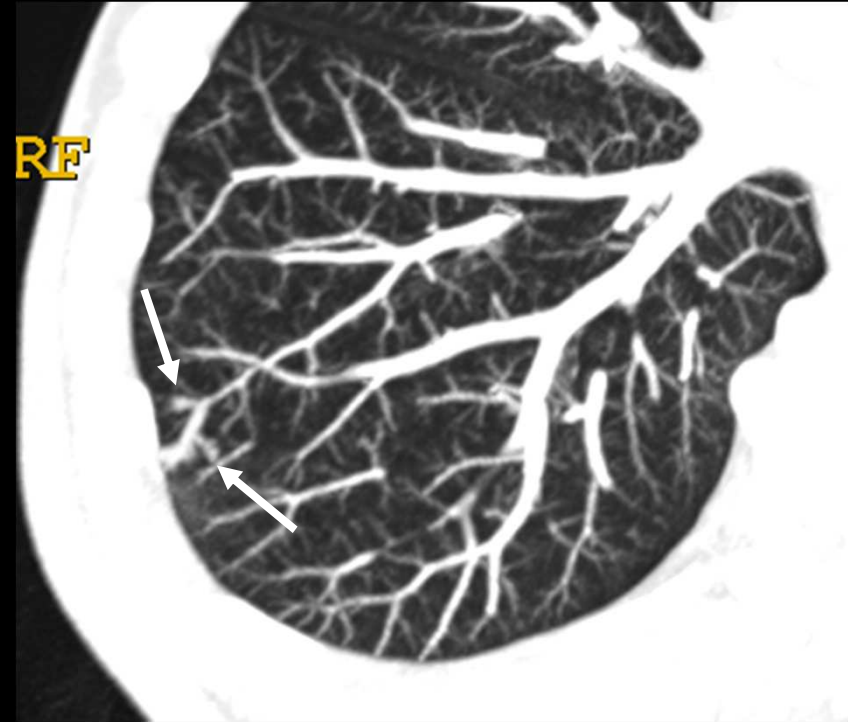




« Tree-in-bud » sign = image non spécifique:



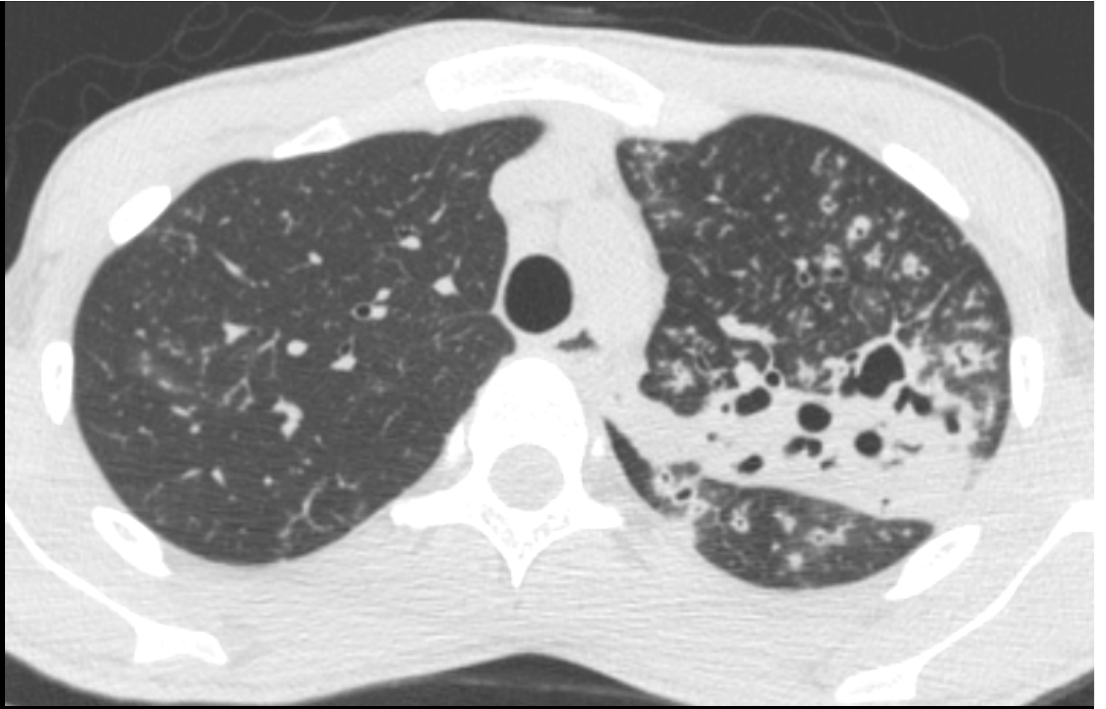
1. Atteinte aériennes

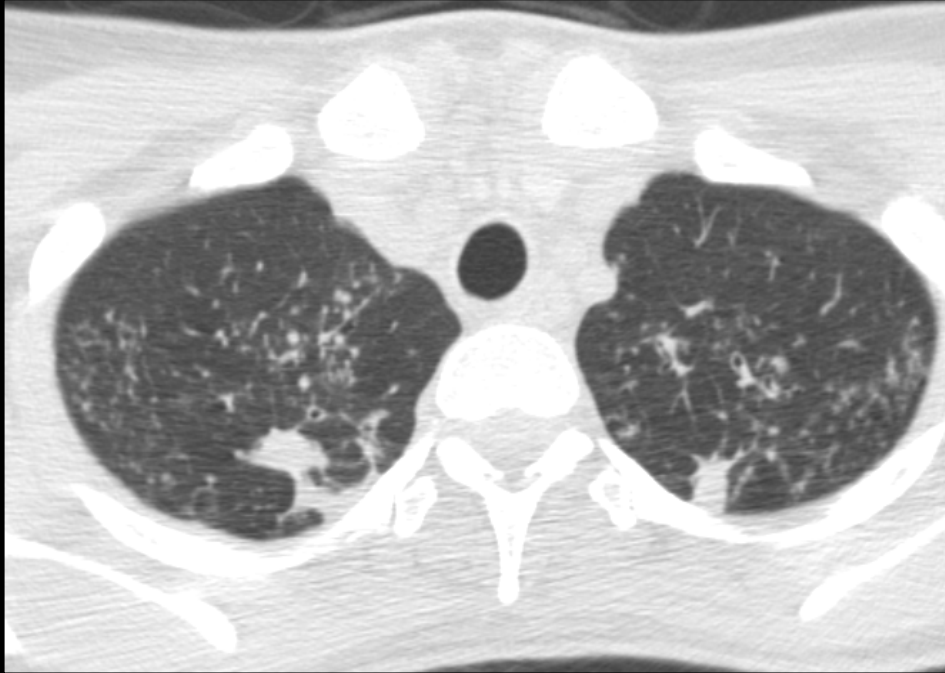


2 Atteinte des petites artères

Arbre en bourgeon « tree in bud »

- Tuberculose par voie aérienne
- Métastases angiocentrique hémotogène
- Infection hémotogène
 - Tbc
 - Champignons
- Pneumonie d'hypersensibilité
- Bronchiolite respiratoire
- Tumeur lymphangitique







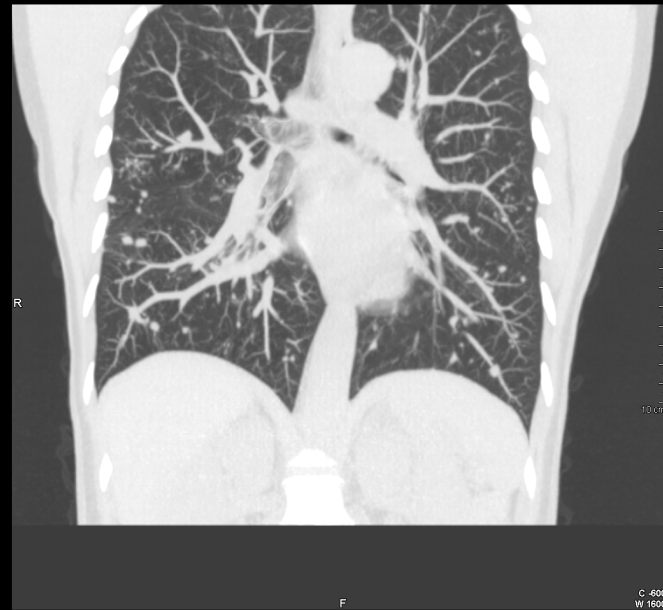
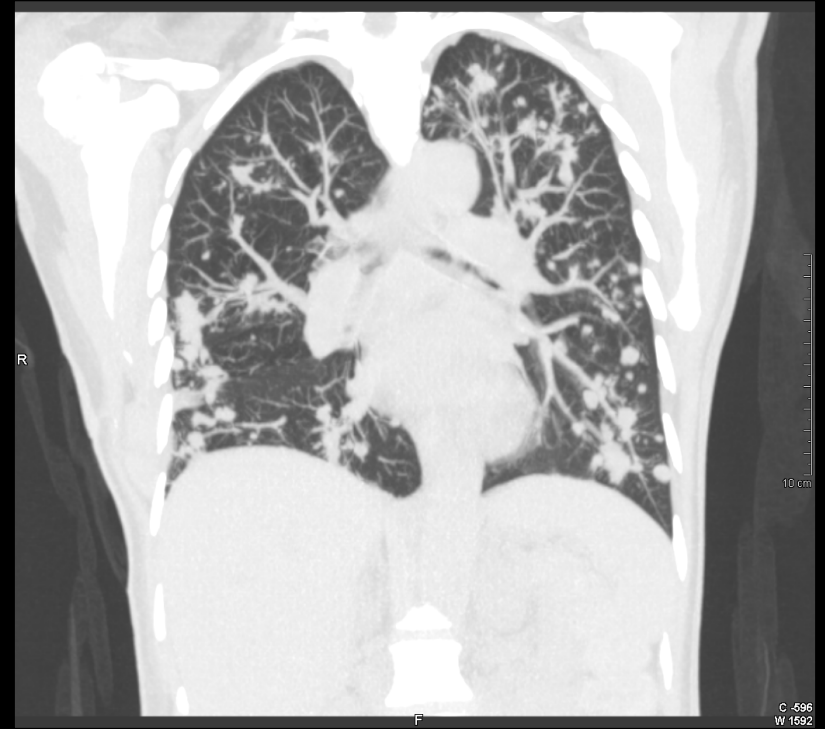
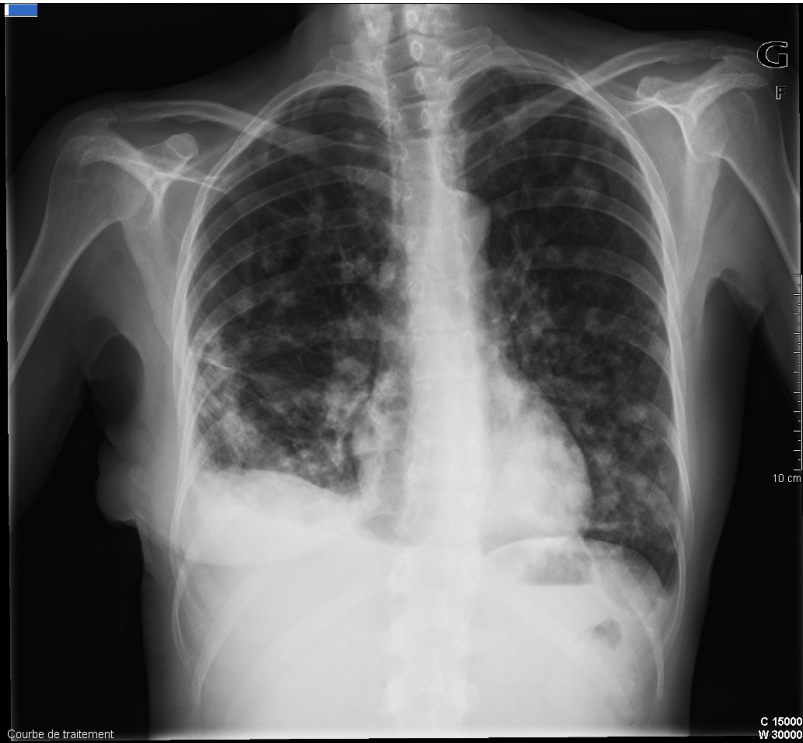
R

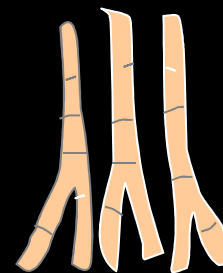
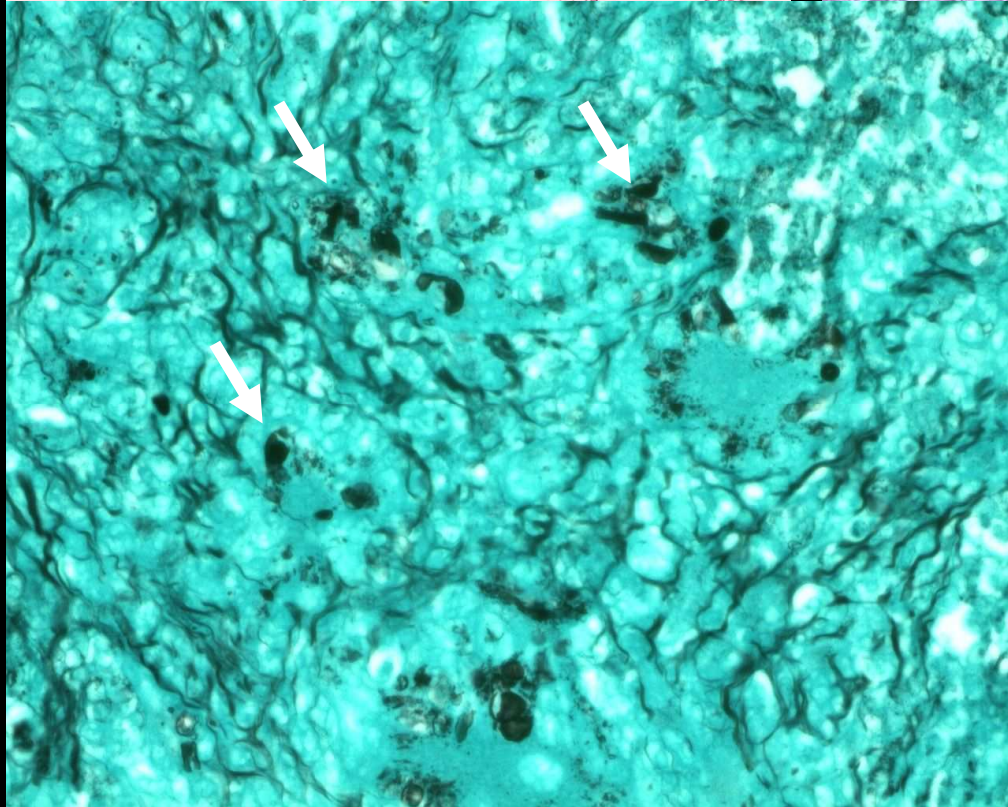
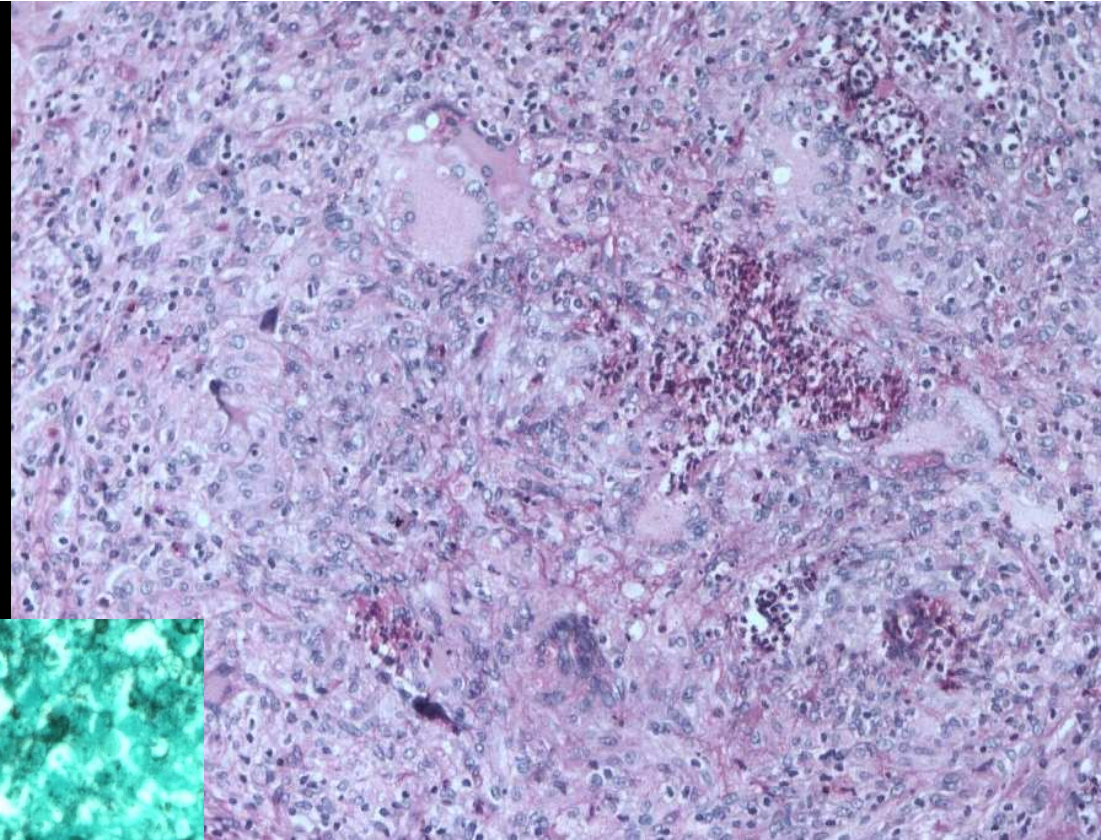
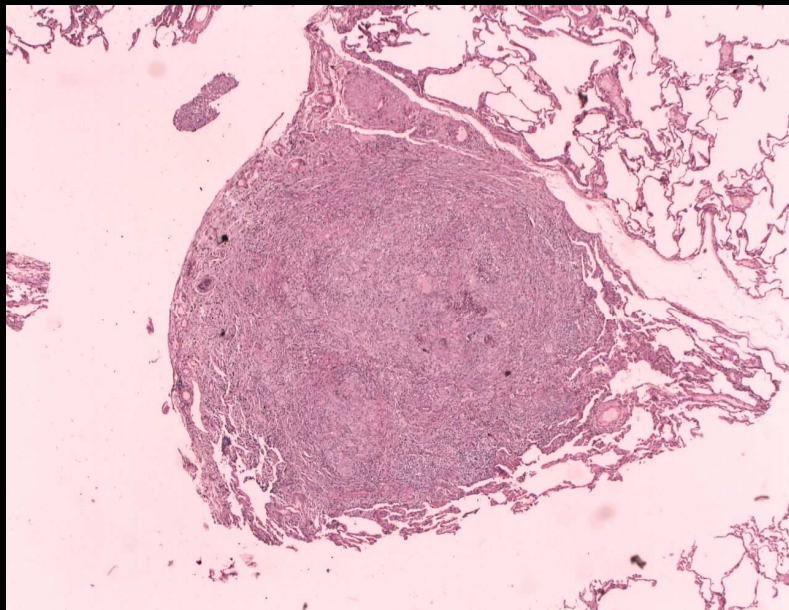
10 cm

F

C -600
W 1600







Aspergillus spp.



Histiocytose Langerhansienne

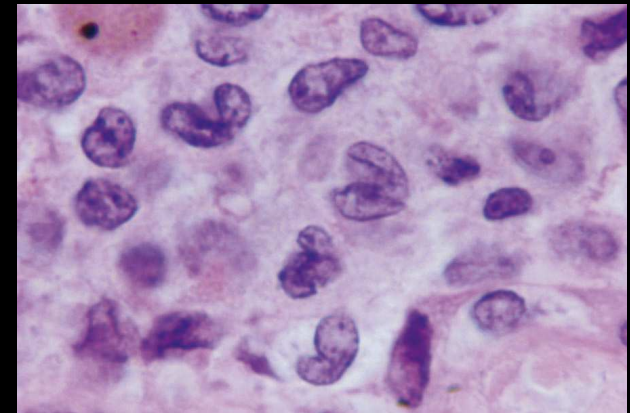
- Prolifération d'histiocytes CD1+ (cellules de Langerhans) formant des granulomes.
- Développement au niveau des parois des bronchioles



Bronchioectasies/cavitation

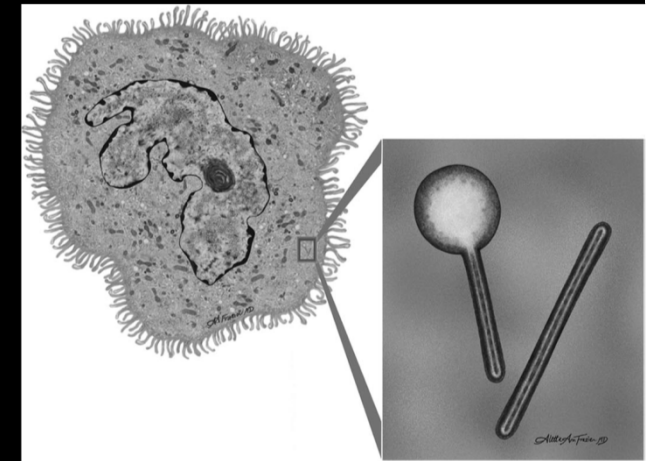
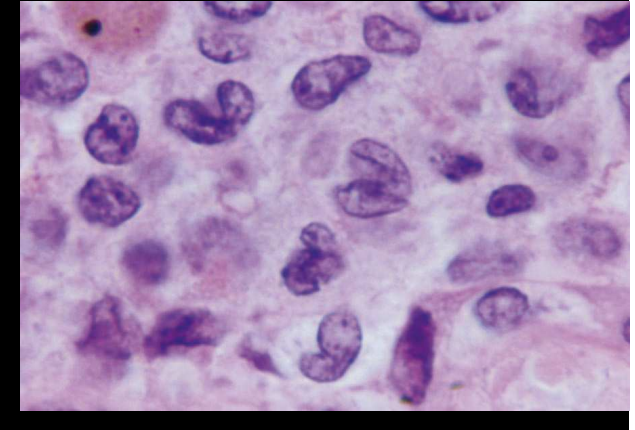
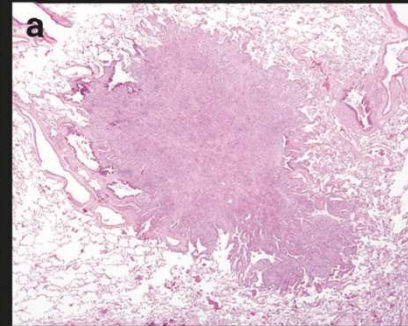
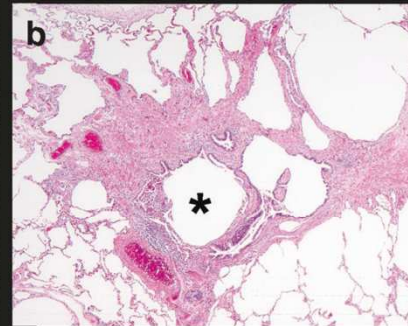
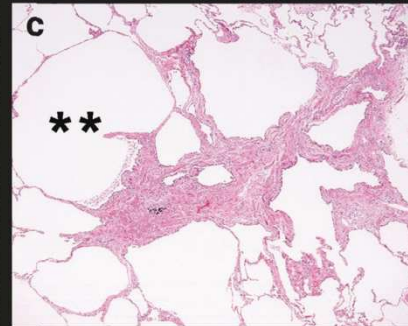


Kystes



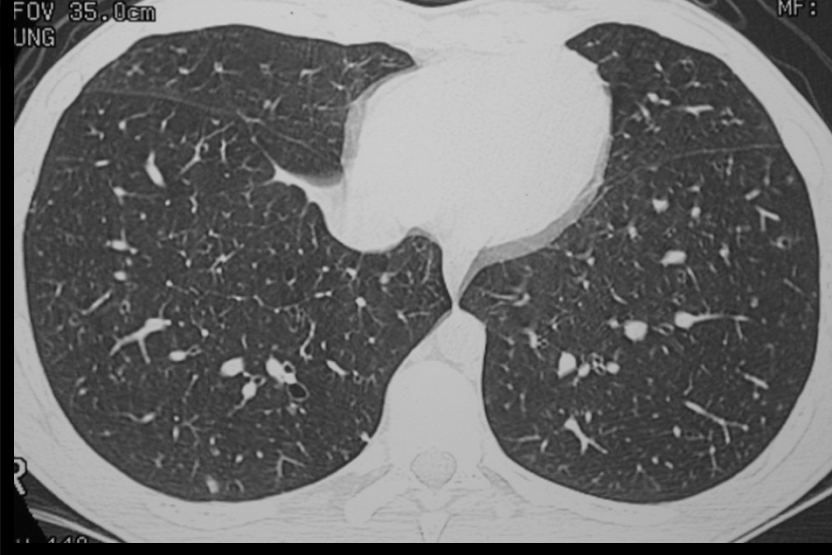
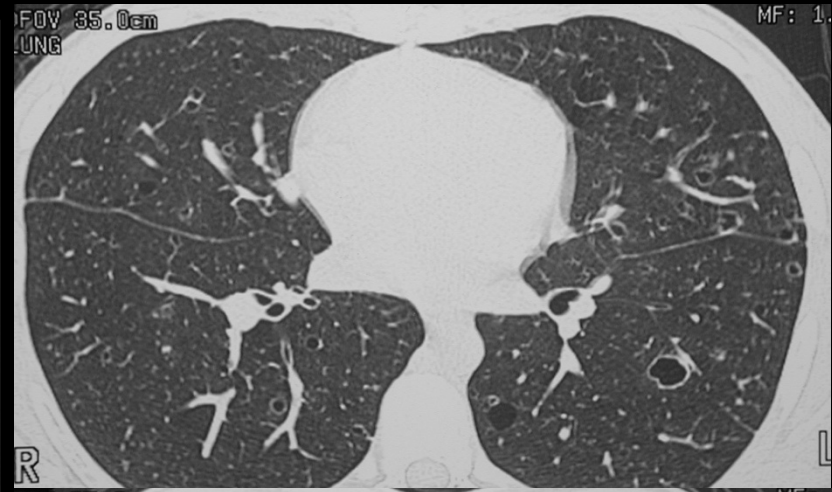
90 % des patients atteints sont fumeurs. Association fréquente avec l'emphysème ou des lésions de fibrose

Histiocytose Langerhansienne

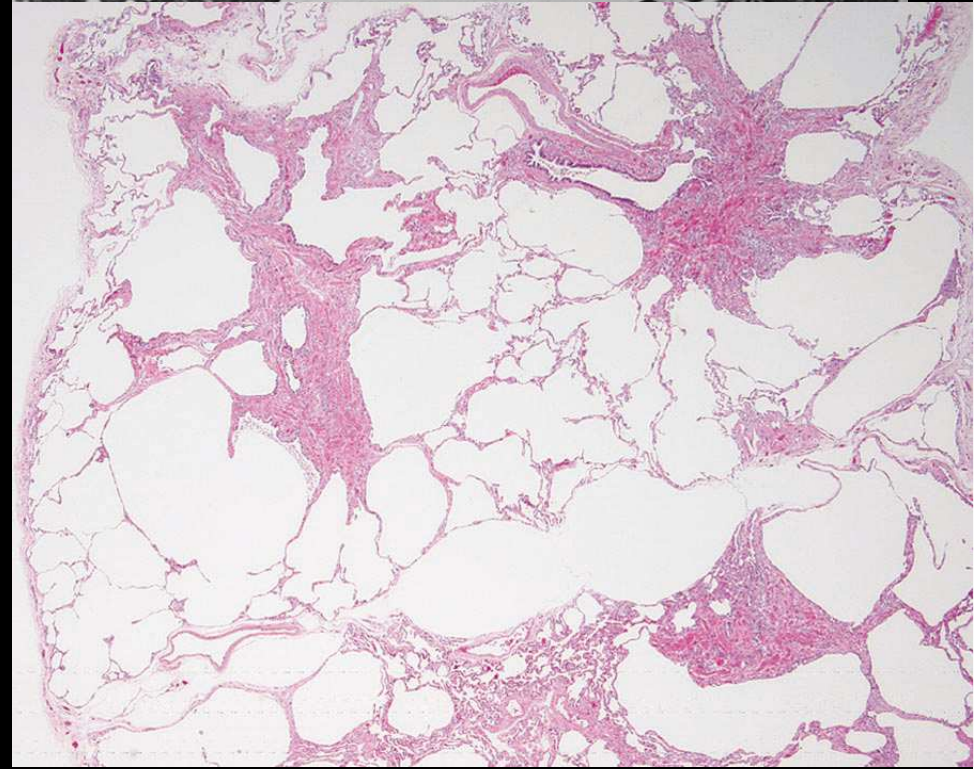
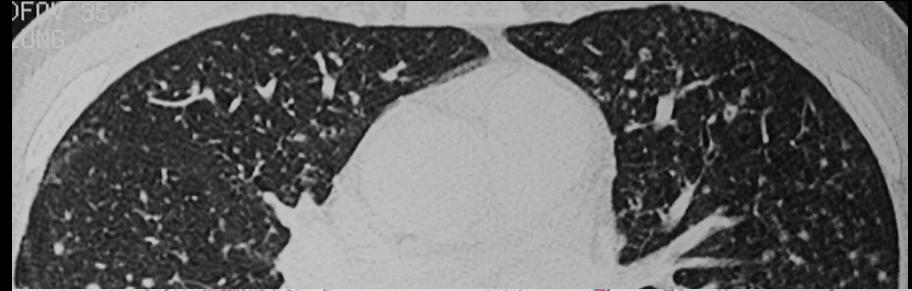


Abbot Radiographics 2004

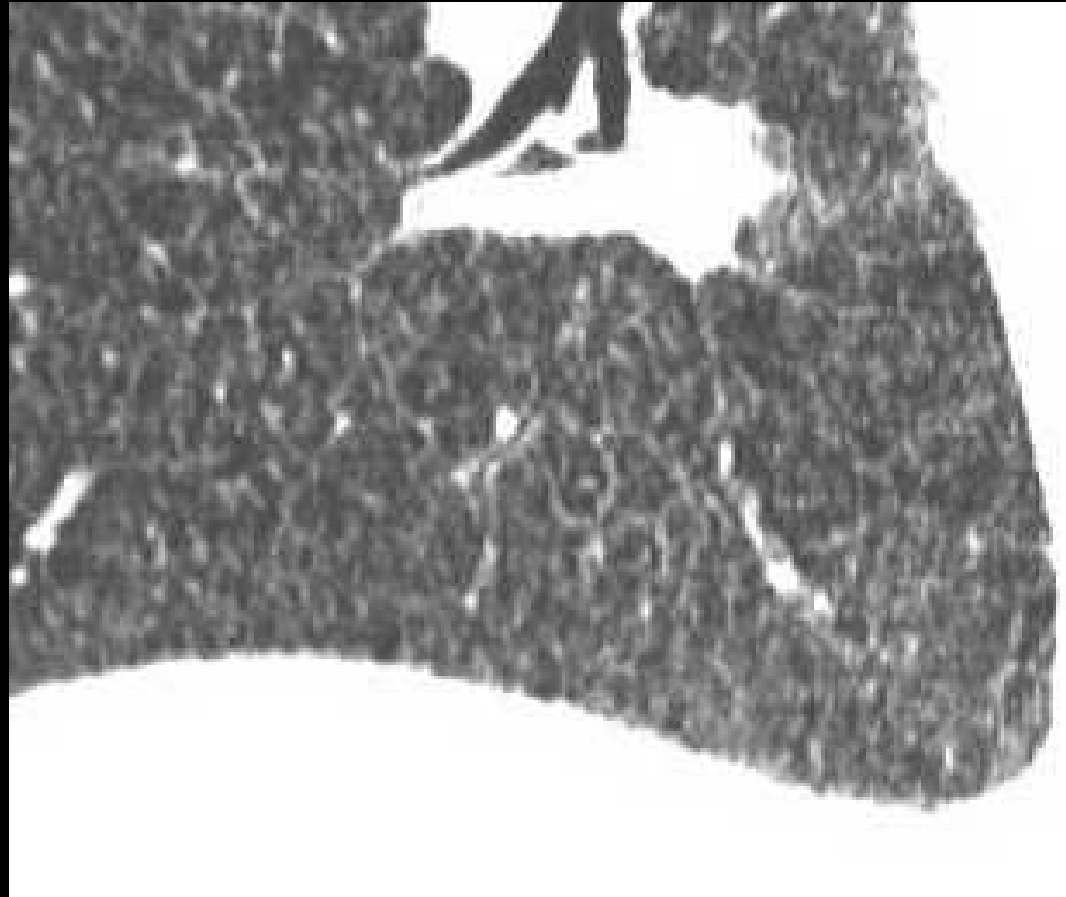
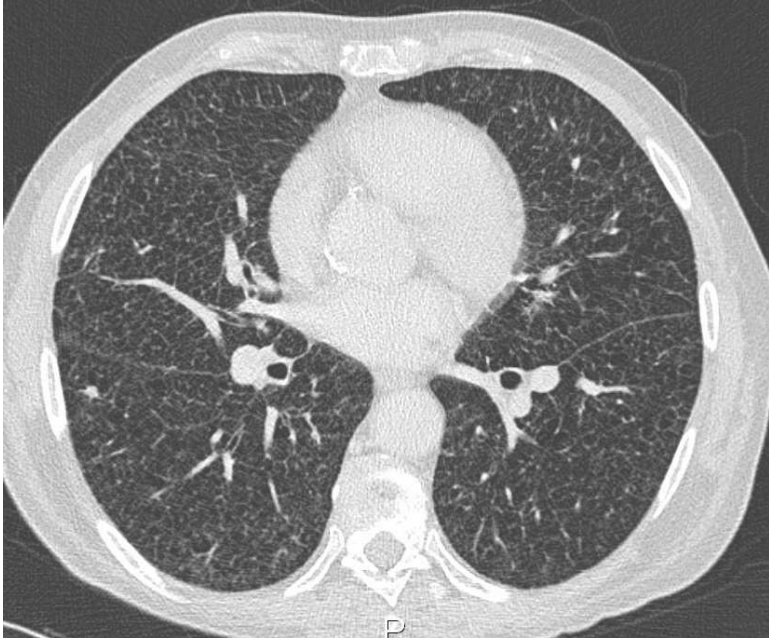
Histiocytose Langerhansienne



Histiocytose Langerhansienne



Histiocytose Langerhansienne

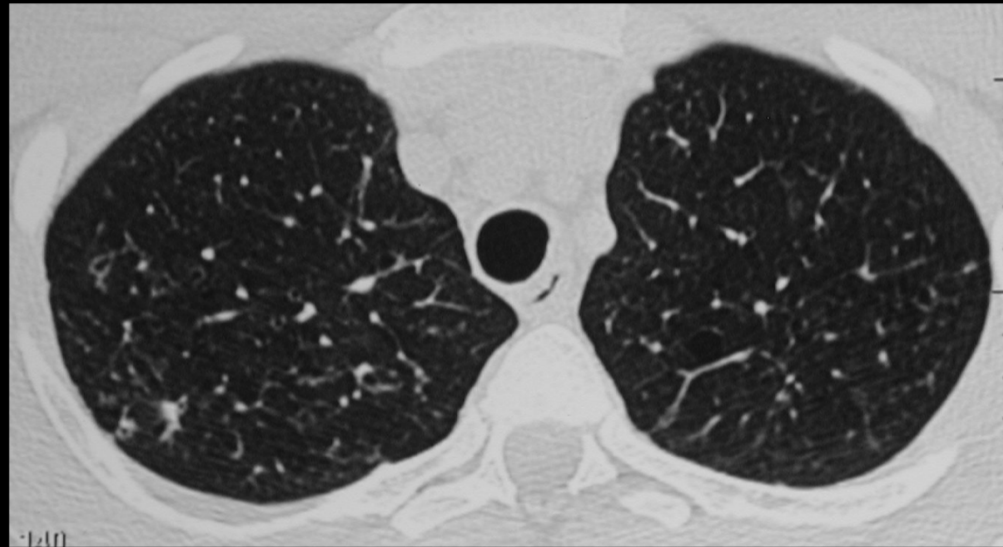


Histiocytose Langerhansienne



T
0

ARRET
DU
TABAC



T
1

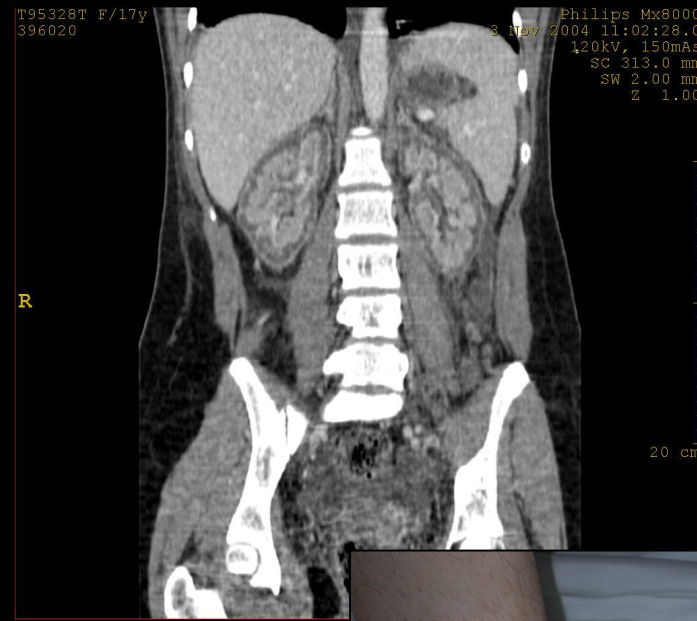
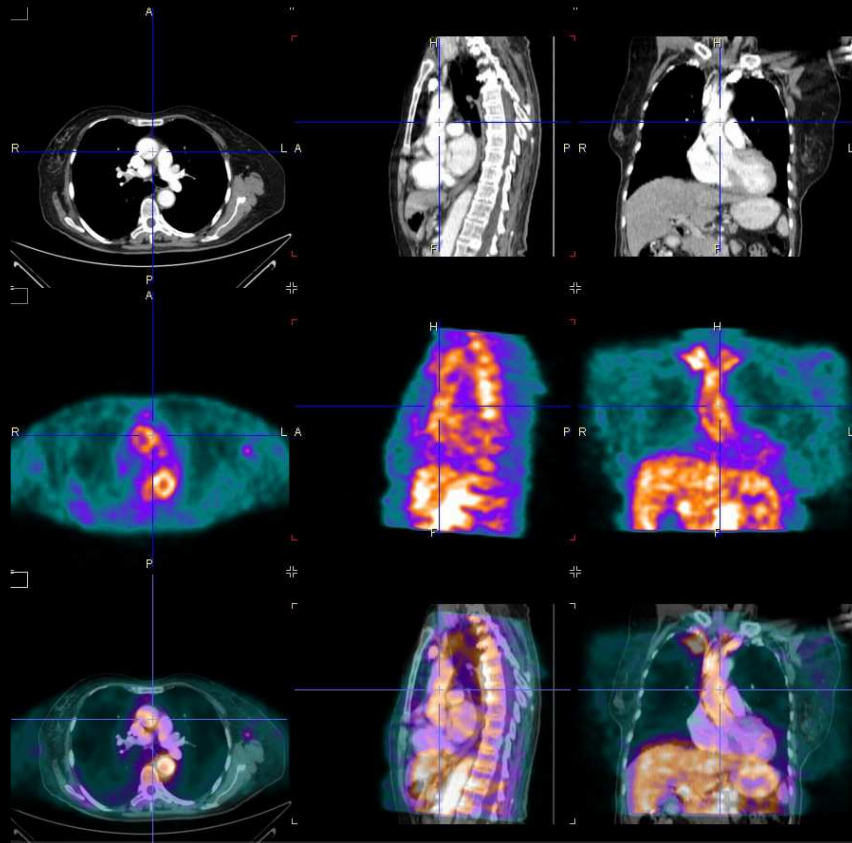
II/Vascularites

- Inflammation des vaisseaux
- Maladie auto-immune
- La lumière vasculaire est souvent compromise
- Ischémie des tissus dans les territoires concernés
- Vasculite ou ses conséquences peuvent être la seule manifestation de la maladie

Vascularite

Maladie rare d'origine immunologique présumée

vaisseau



Classification

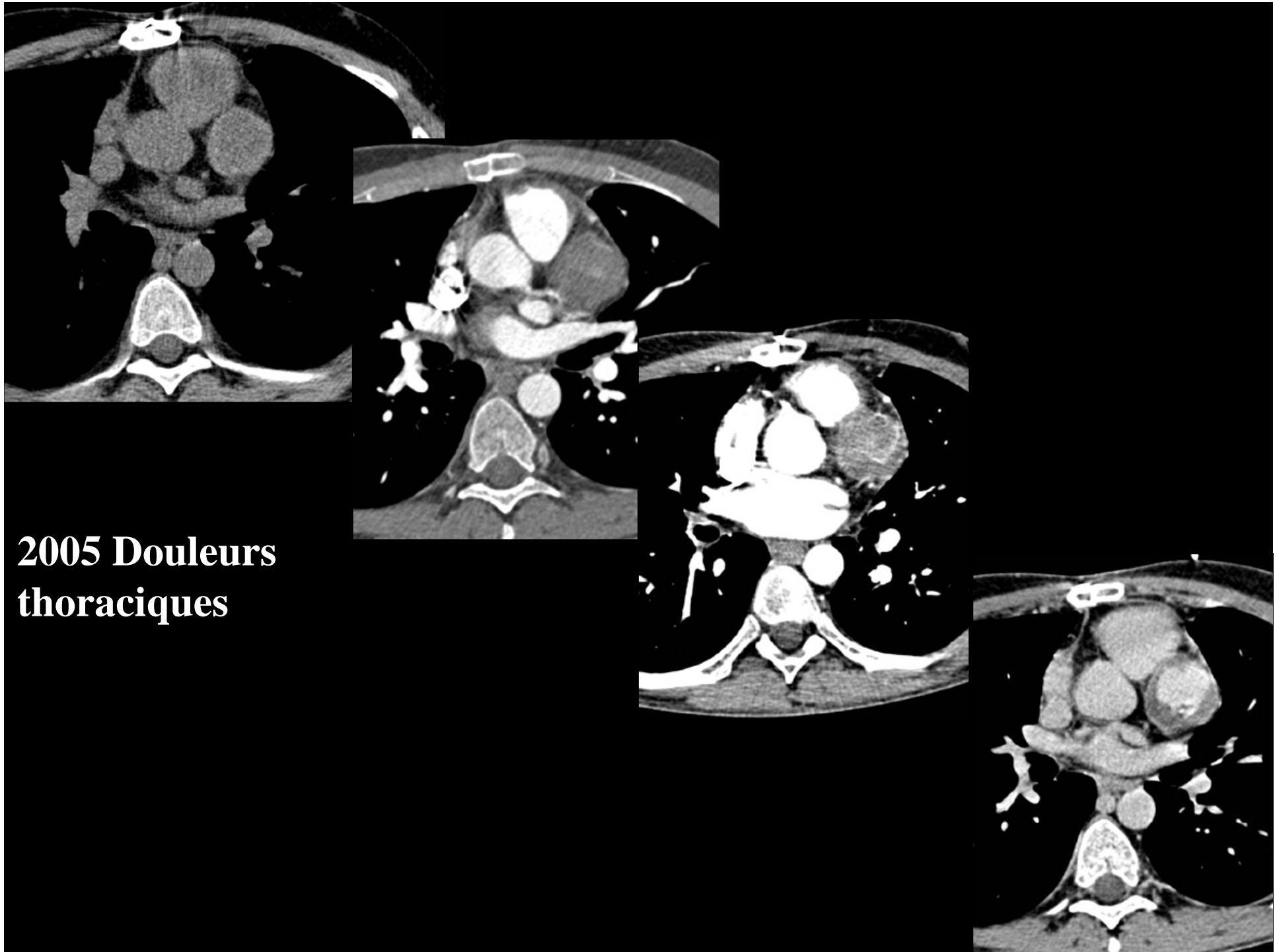
- Granulomatose de Wegener
- Maladie de Behçet
- Artérite de Takayasu
- Syndrome de Churg-Strauss
- Polyartérite noueuse
- Artérite à cellules géantes
- Divers

Maladie de Behçet

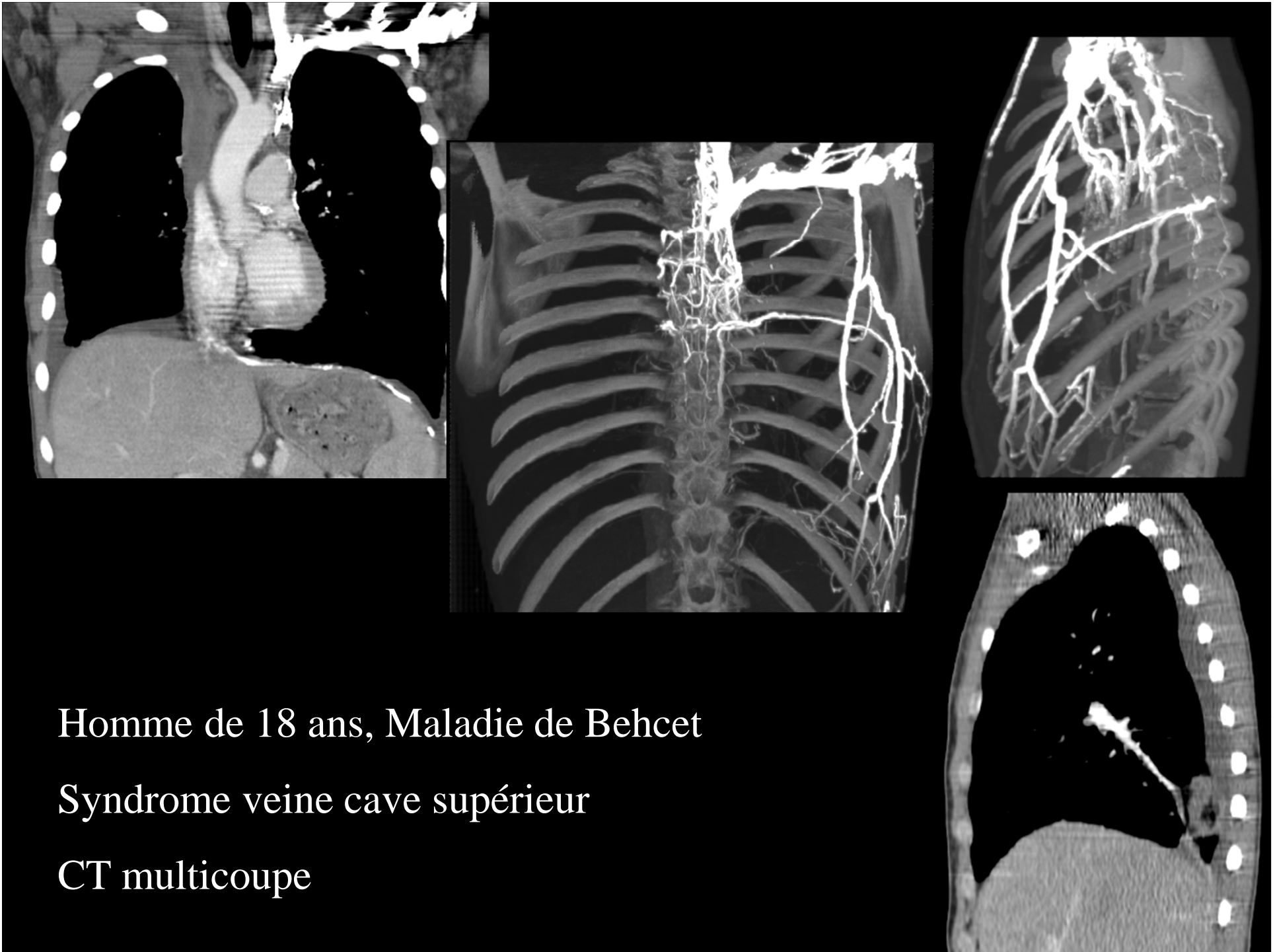
- Maladie systémique rare: 80-370/100 10³ en Turquie
 - stomatite aptheuse récurrente
 - Ulcères génitaux
 - Lésions dermatologiques et uvéite
- Atteinte pulmonaire rare
 - Anérysme artère pulmonaire : simple ou multiples
 - Thrombose ou occlusion de la VCS, A. pulmonaires
 - Infarctus pulmonaire, hémorragie, COP, fibrose, emphysème, adénopathies, épanchement péricardique..



1997: Homme de 18 ans, arthralgies diffuses, augmentation des paramètres inflammatoires



**2005 Douleurs
thoraciques**



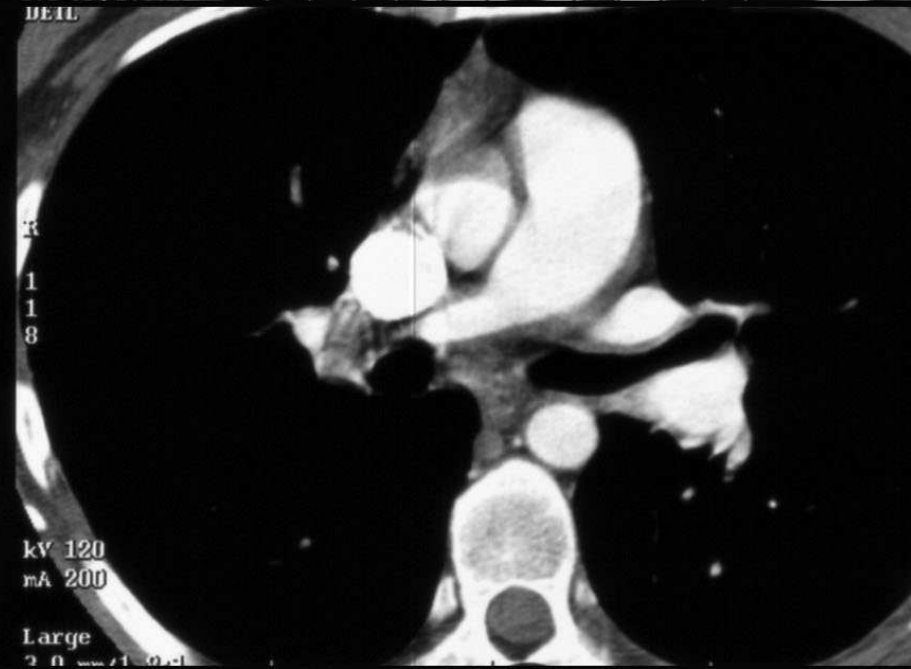
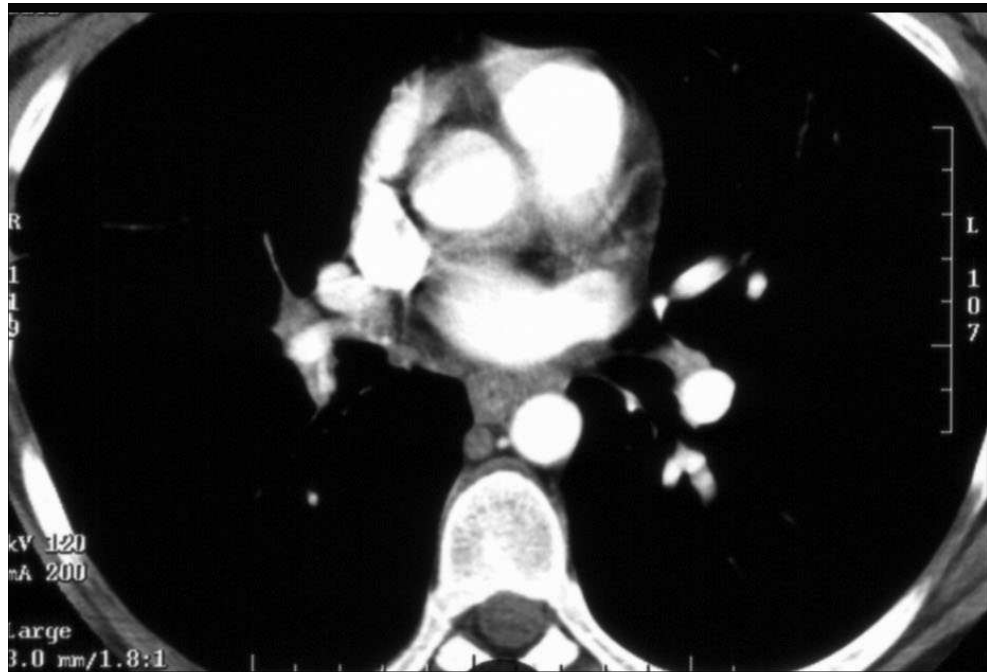
Homme de 18 ans, Maladie de Behcet

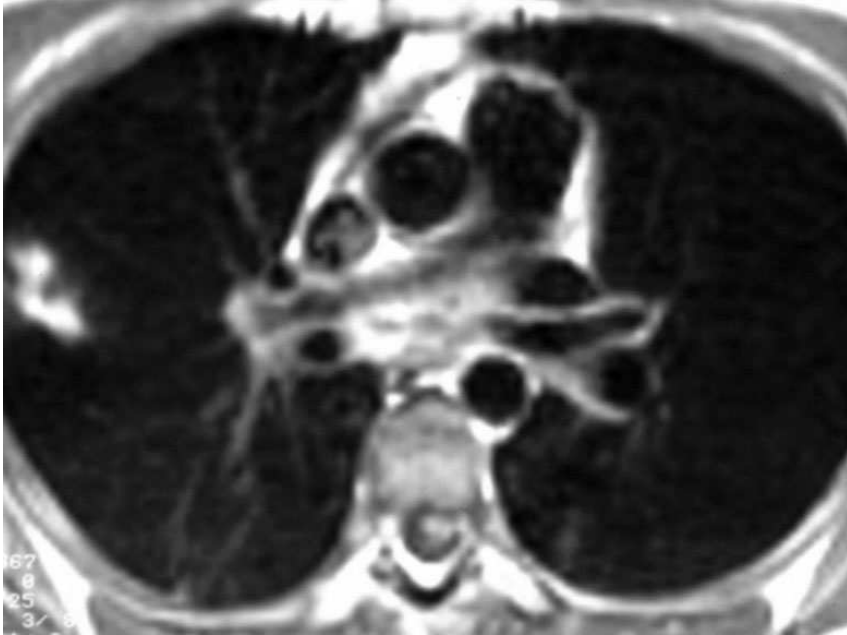
Syndrome veine cave supérieur

CT multicoupe

Artérite de Takayasu

- Artérite rare atteignant principalement l' aorte et ses branches principales
- Atteinte préférentielle des femmes
- Installation entre 10 et 40 ans
- Manifestations radiologiques
 - Aorte (10-75%): contours irréguliers, calcifications, ectasie de la crosse aortique
 - Artères pulmonaires (50-70%): sténose, occlusion de branches segmentaires ou sous-segmentaires





Ferretti G, et al Initial isolated Takayasu's arteritis of the right pulmonary artery: MR appearance. Eur Radiol. 1996;6:429-32

Syndrome de Churg-Strauss

- Maladie rare: 2.4/10⁶ cas
- Asthme, fièvre, éosinophilie sanguine
- Vascularite nécrosante et inflammation extravasculaire granulomateuse
- Manifestations HRCT (communes)
 - Densifications parenchymateuses et « verre dépoli »
 - Distribution hétérogène et plutôt périphérique
 - Epaississement des septae interlobulaires
 - Nodules centrilobulaires
 - Epaississement des parois bronchiques
 - Epanchements pleuraux



Divers

- Periartérite Noyeuse:

Vascularite nécrosante / artères musculaires de petite à moyenne taille

Vascularite de la circulation pulmonaire est rare

- Maladie d'Erdheim-Chester :

Épaississement lisse des septae interlobulaires , nodules centri-lobulaires, épaississement scissuraux, épanchements pleuraux, aortite, masses extra-thoraciques

- Artérite à cellules géantes:

Aortite (20-40%), atteinte coronaire, péricardite



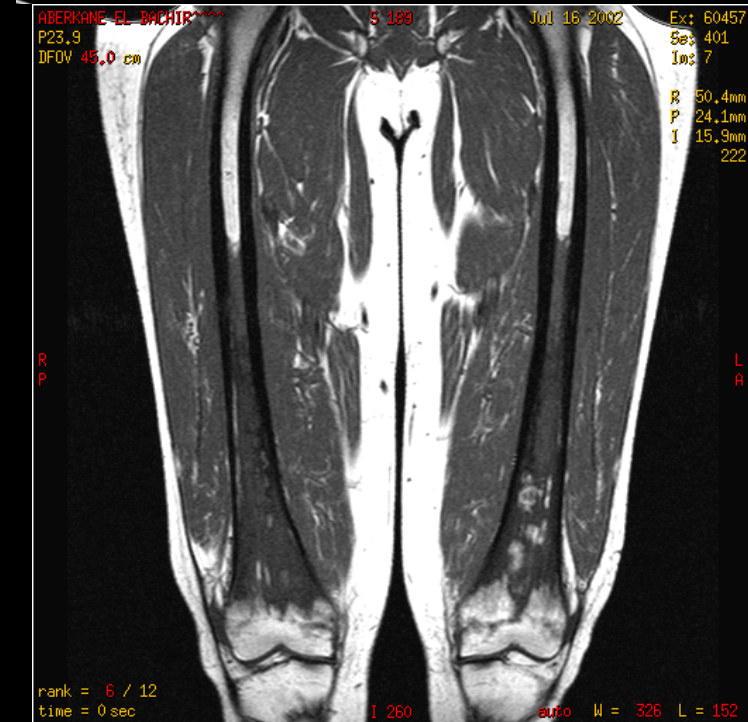
2



Aorte infiltrée



Erdheim-Chester



ABERKANE-EL-BACHIR
P23.9
DFOV 45.0 cm
S 189
Jul 16 2002
Ex: 60457
Se: 401
Im: 7
R 50.4mm
P 24.1mm
I 15.9mm
222

rank = 6 / 12
time = 0 sec
I 260
auto W = 326 L = 152

III/Connectivites

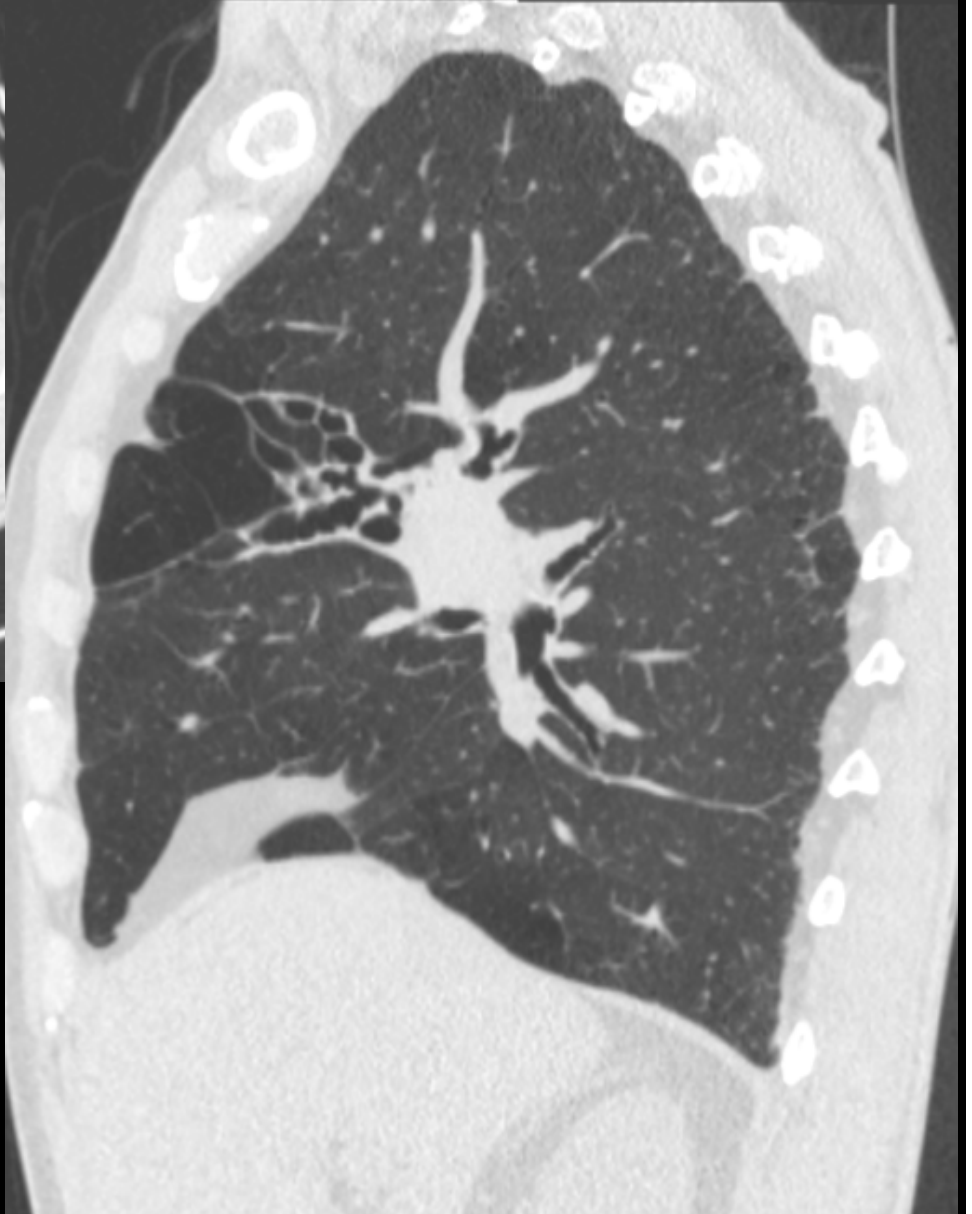
- Groupe hétérogène, affectant plusieurs organes
- Atteinte inflammatoire.
- Manifestations pulmonaires sont relativement fréquentes.
- Toutes les composantes du poumon peuvent être atteintes.
- Les symptômes respiratoires peuvent passer inaperçus en raison
 - Manque d'exercice
 - Atteinte du système musculo-squelettique

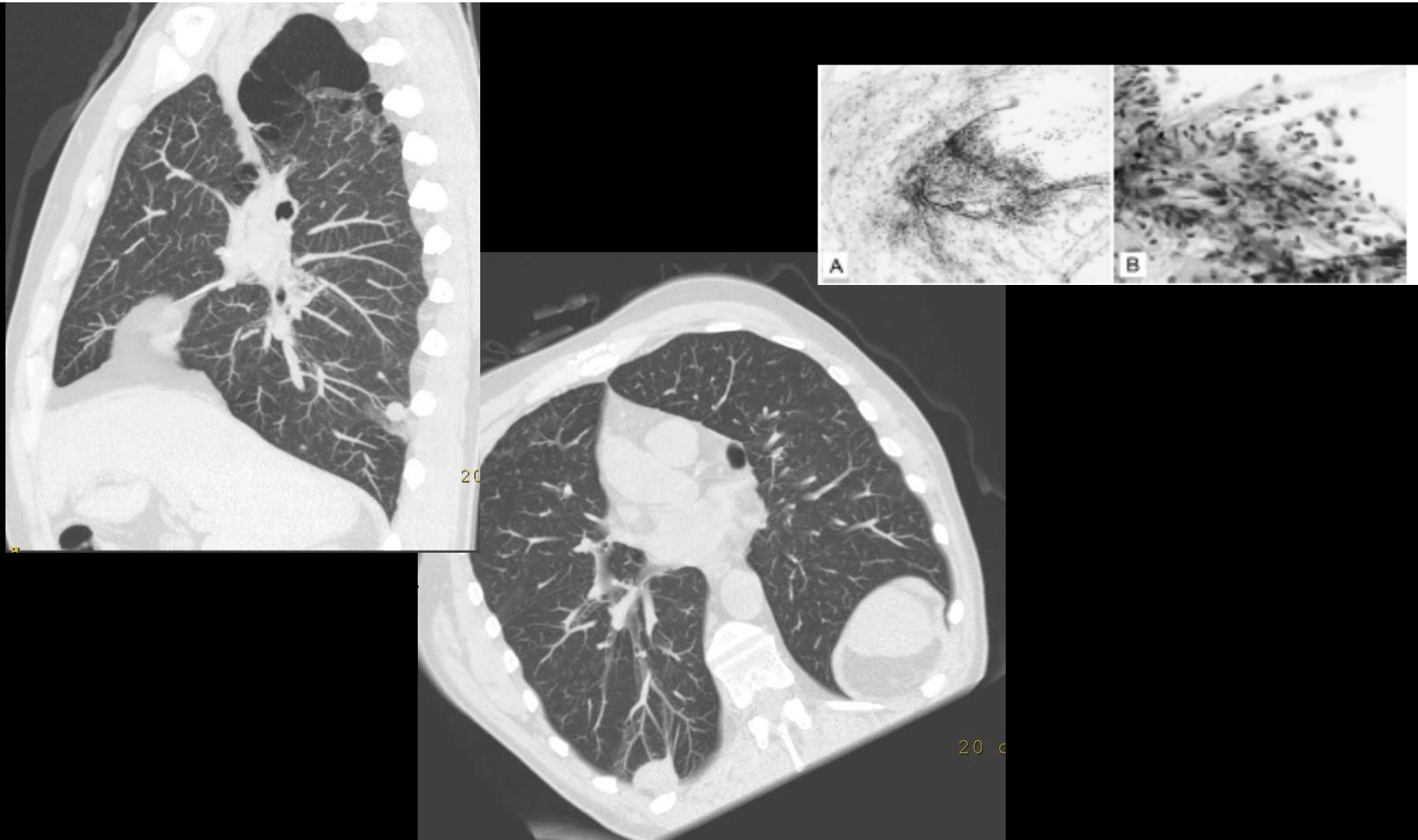
Classification

- Maladie rhumatoïde
- Lupus érythémateux disséminé
- Sclérose systémique progressive
- Dermatomyosite and polymyosite
- Syndrome de Sjögren
- Polychondrite atrophiante
- Spondylarthrite ankylosante

Maladie rhumatoïde

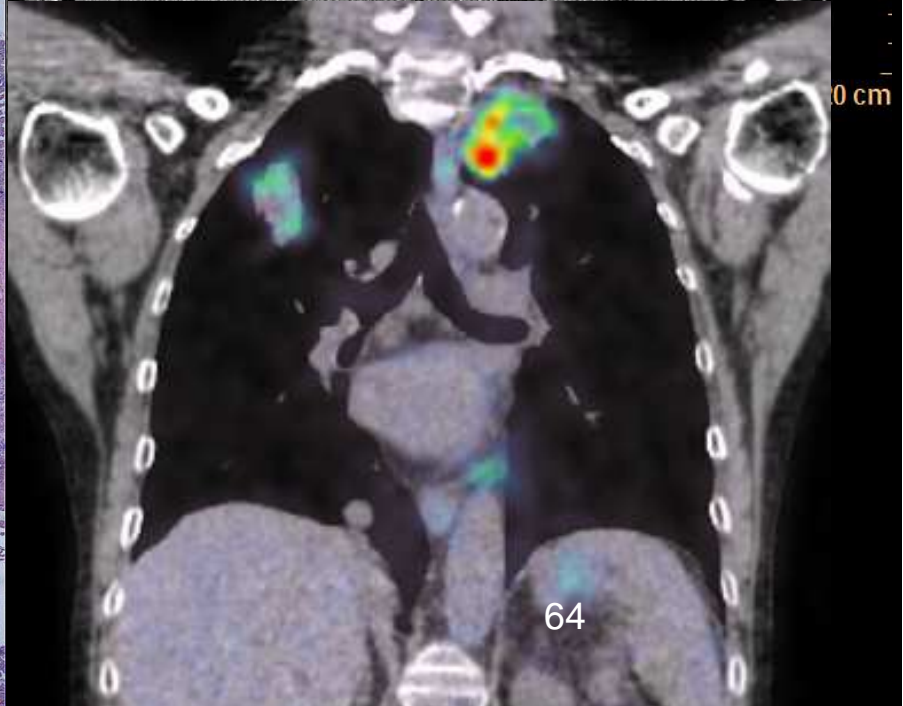
- Maladie de système fréquente (0.8% de la population)
- Manifestations pleuro-pulmonaires fréquentes
 - Épanchements pleuraux
 - Nodules rhumatoïdes
 - Pneumonie interstitielle : UIP, NSIP, COP
 - Vascularite pulmonaire
 - Atteinte des voies aériennes: bronchiectasies, Bronchiolite oblitérante, bronchiolite folliculaire

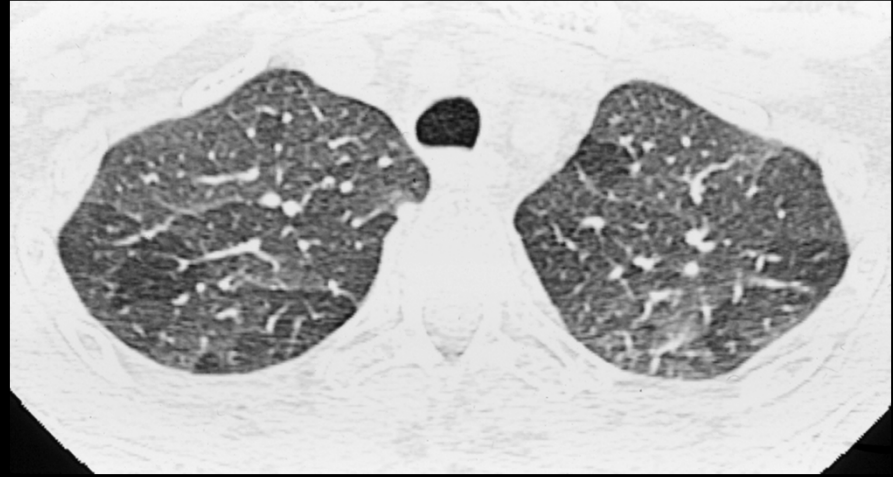
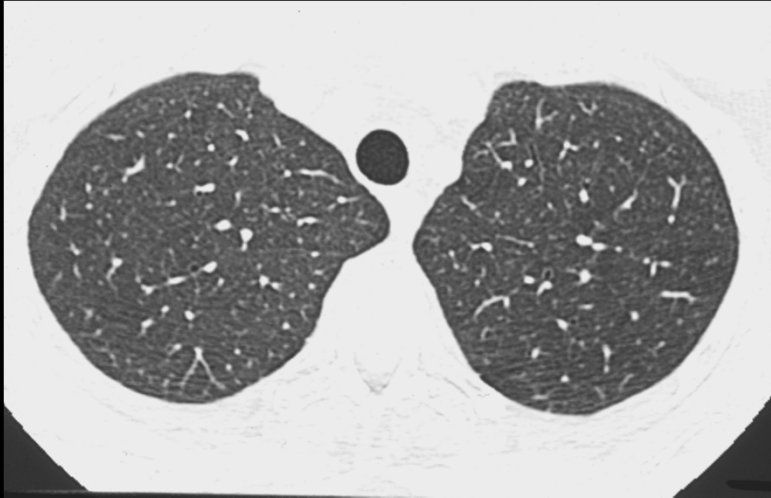




Nodules rhumatoïdes sont identifiés sur 1% des radiographies thoraciques
Qqs mm à qqs cms de diamètre, simple ou multiple, cavitation centrale (50%)

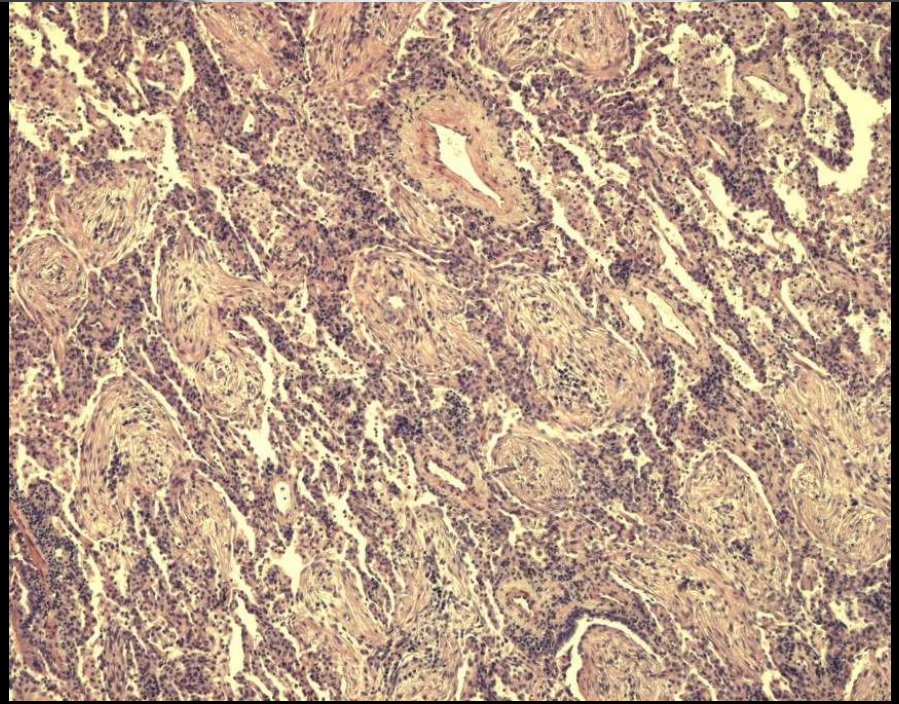
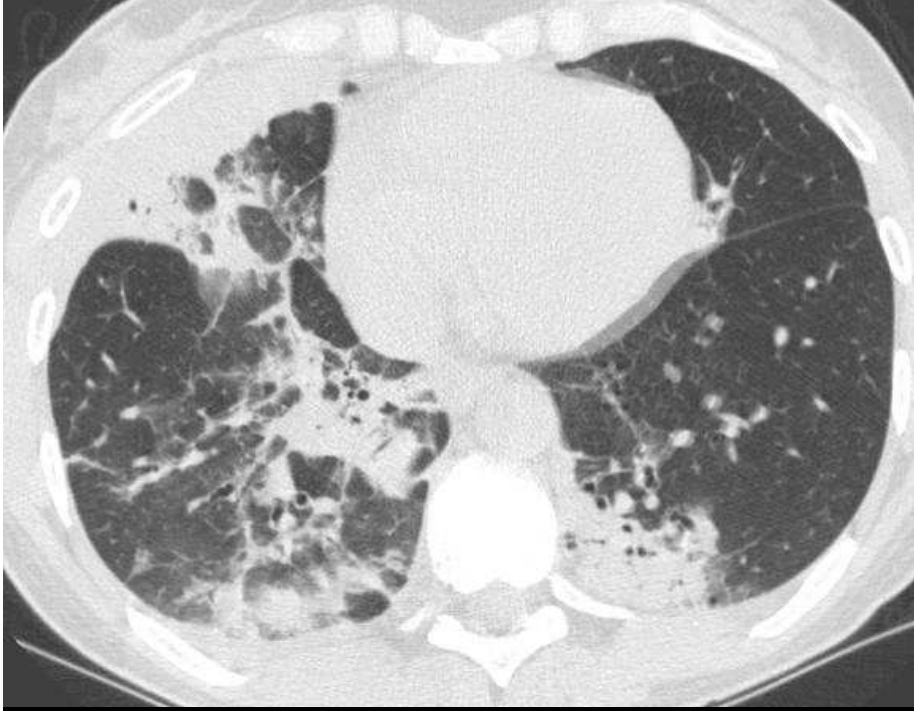
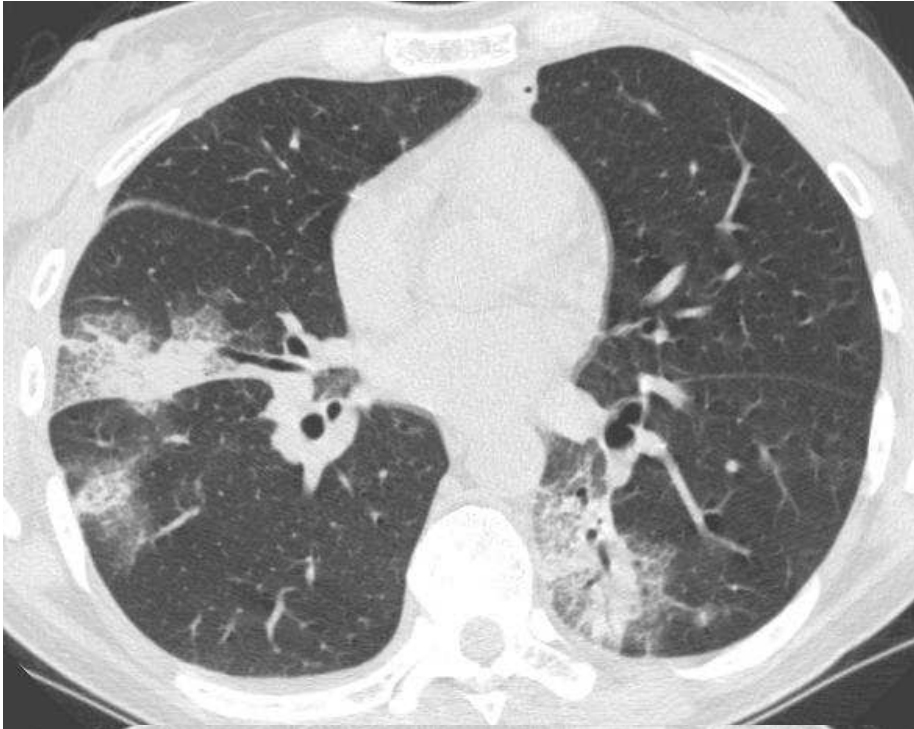
Gupta P, et al. Clin Rheumatol 2005;24:402-405



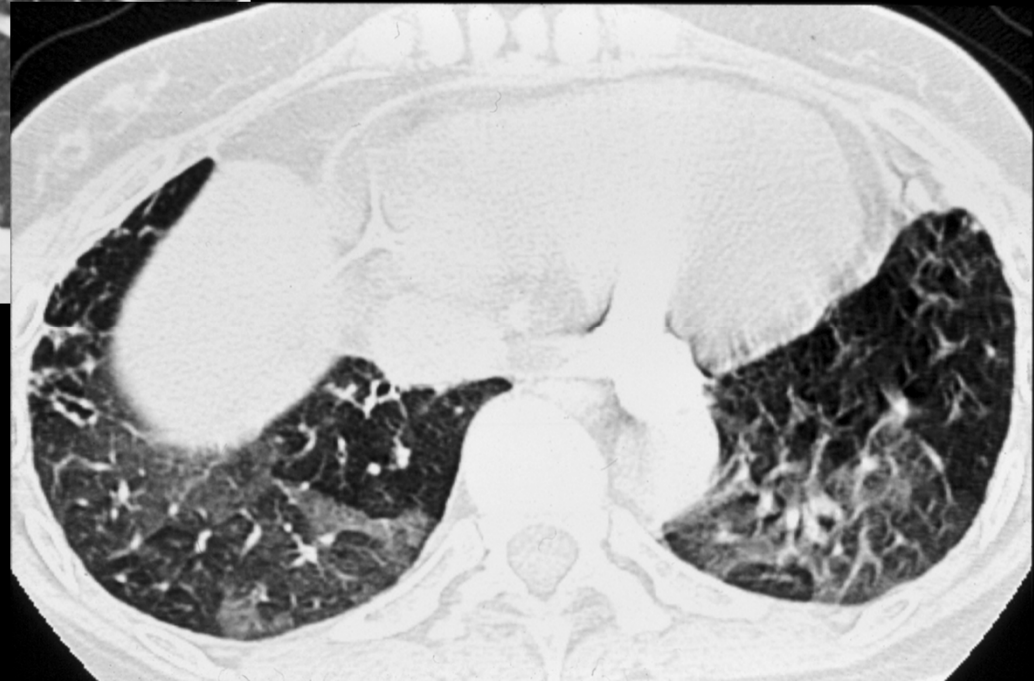


Lupus érythémateux disséminé

- Maladie multisystémique d'origine autoimmune (peau, reins, poumons, coeur)
- 25/100 000 sujets
- Maladie pleuro-pulmonaire (7-100%)
 - Epanchements pleuraux (50-83%)
 - Pneumonie lupique aiguë, pneumonie interstitielle /fibrose, hémorragie pulmonaire
 - Thrombose artère pulmonaire, EP, Hypertension AP
 - Bronchiolite oblitérante, COP
 - Dysfonction diaphragmatique



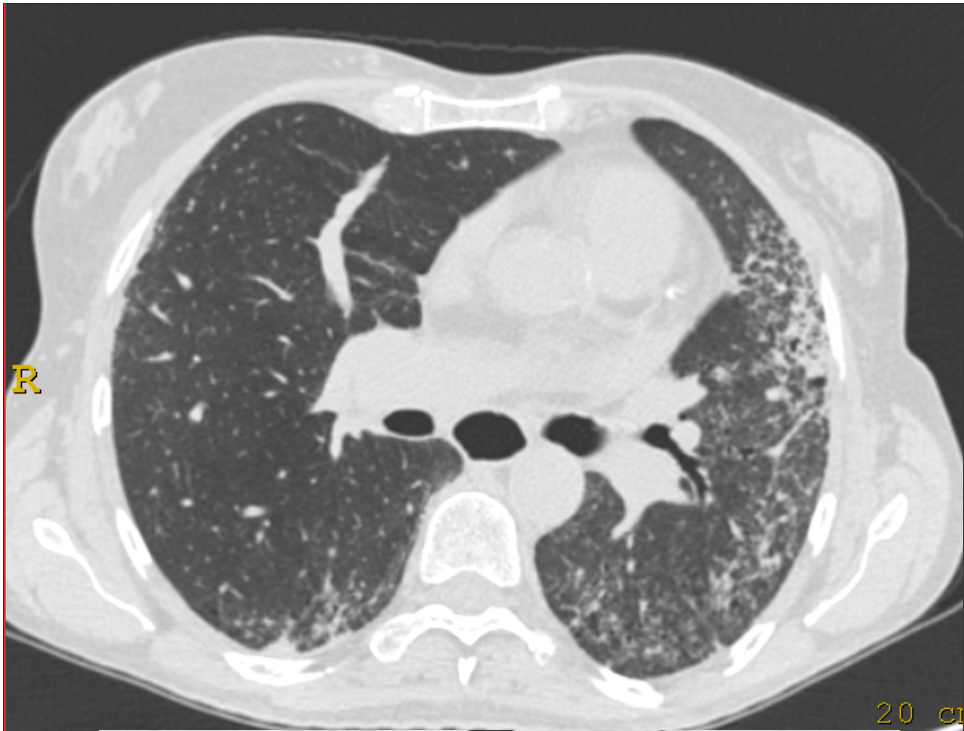
Bronchiectasies et piégeage expiratoire



Sclérose systémique progressive

- Maladie systémique de cause inconnue
- Fibrose cutanée, vsx sanguins, et organes viscéraux
- Incidence 12 /10⁶ cas/an
- Poumons: 2nd organe atteint
- Pneumonie Interstitielle et fibrose (NSIP, UIP)
- Hypertension pulmonaire , adénomégalies
- Atrophie et atonie de l'oesophage (12-40 mm plan coronal)

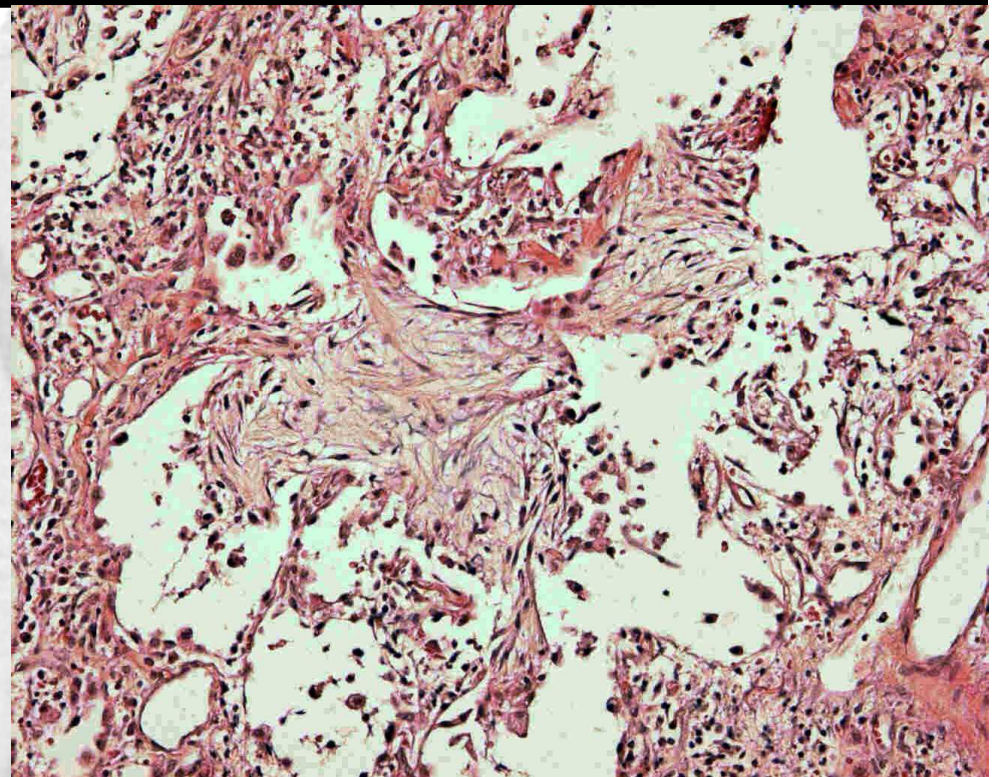
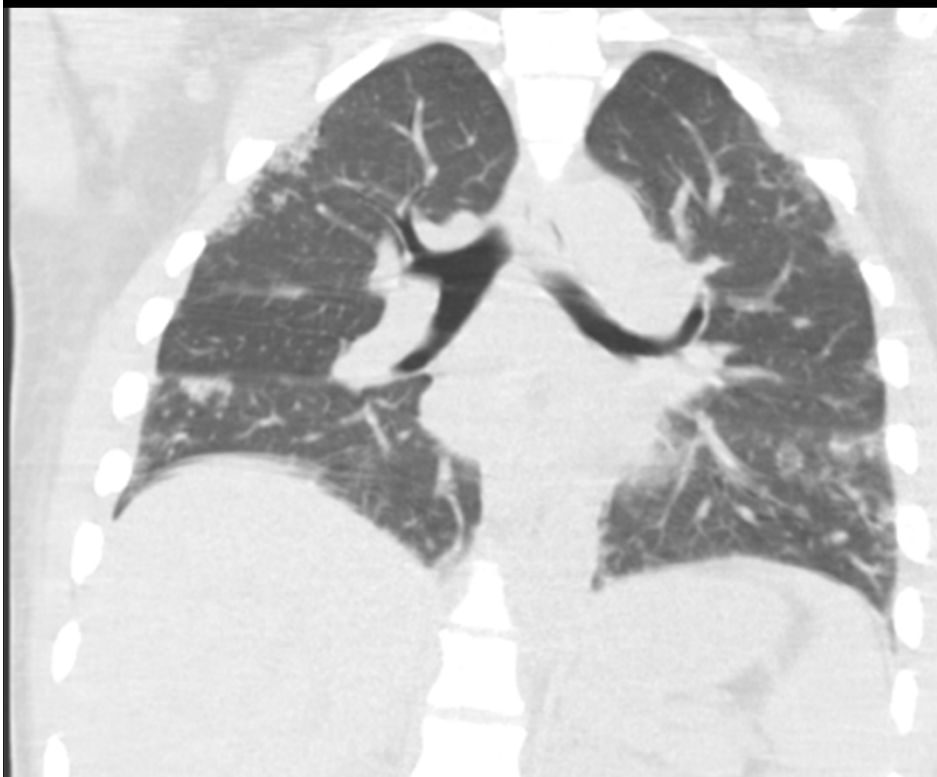
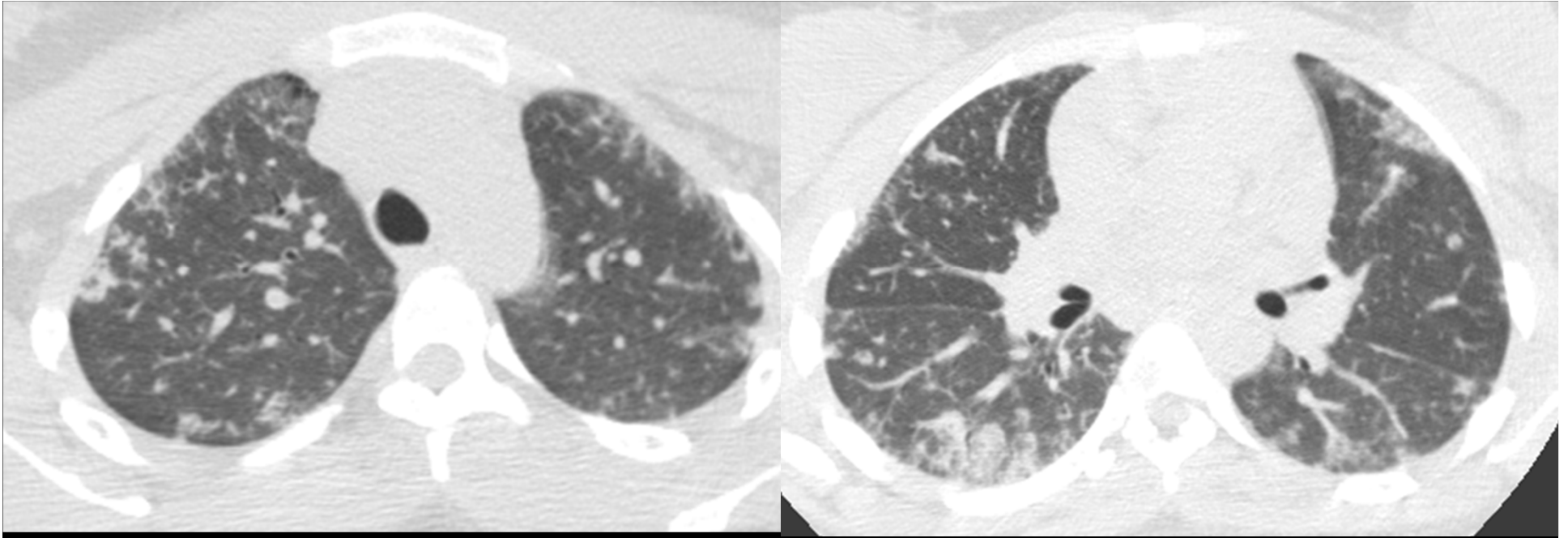
Bhalla M, Silver RM, Shepard JA, McLoud TC. Chest CT in patients with scleroderma: prevalence of asymptomatic esophageal dilatation and mediastinal lymphadenopathy. *AJR Am J Roentgenol.* 1993;161(2):269-72.

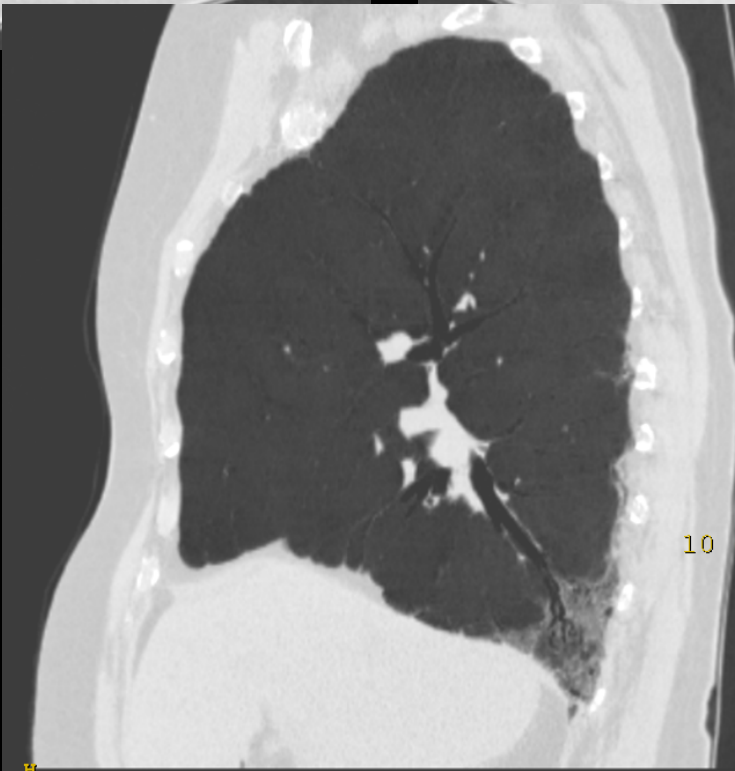
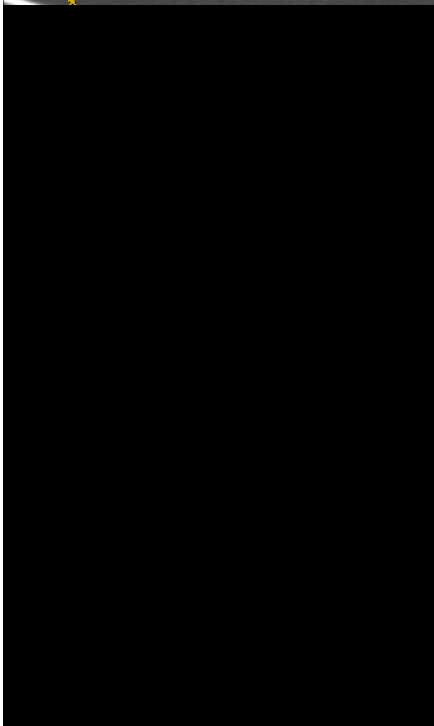
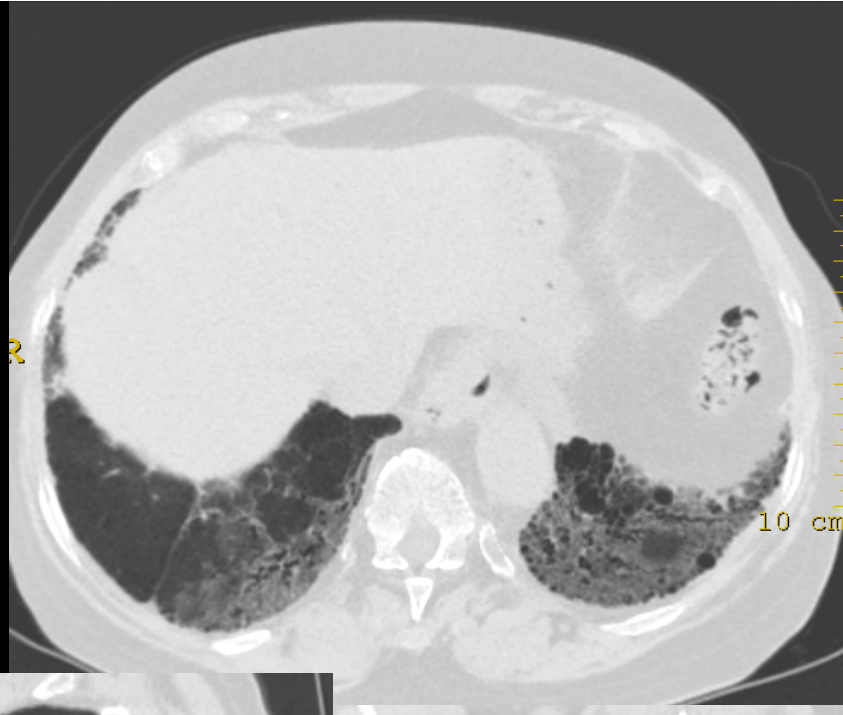


Dermatomyosite et polymyosite

- Faiblesse et pfs douleurs dans les muscles proximaux
- Hypoventilation et détresse respiratoire
- Pneumonie interstitielle
 - Fibrose pulmonaire idiopathique
 - COP, NSIP
- Pneumonie d'aspiration

Douglas WW, et al. Polymyositis-Dermatomyositis-associated interstitial lung disease. Am J Respir Crit Care Med 2001;164: 1182-1185

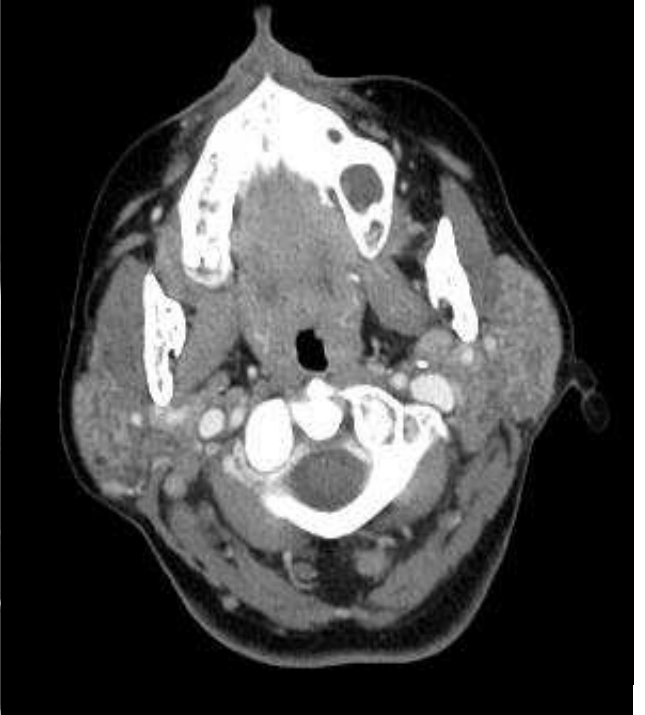
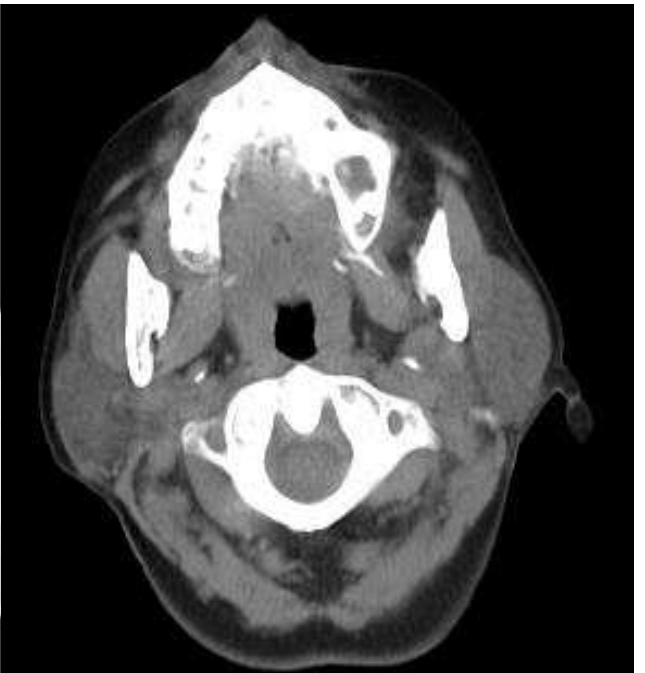
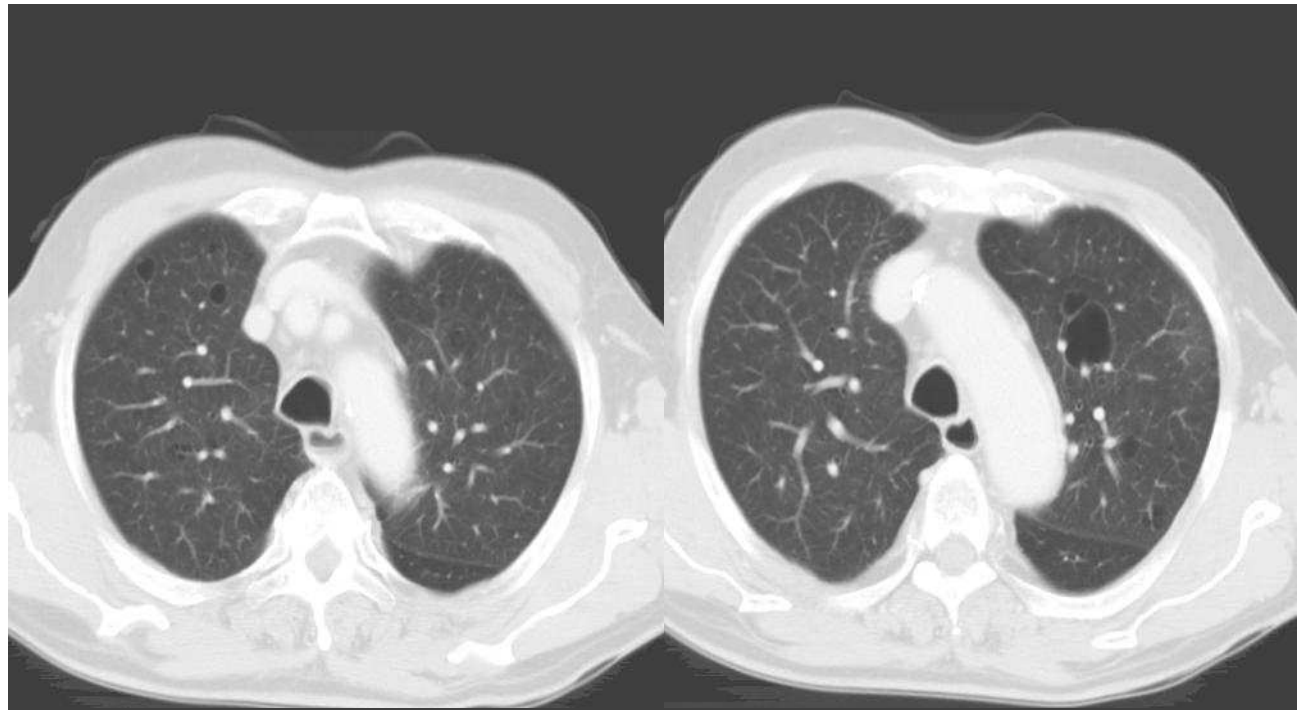




Syndrome de Sjögren

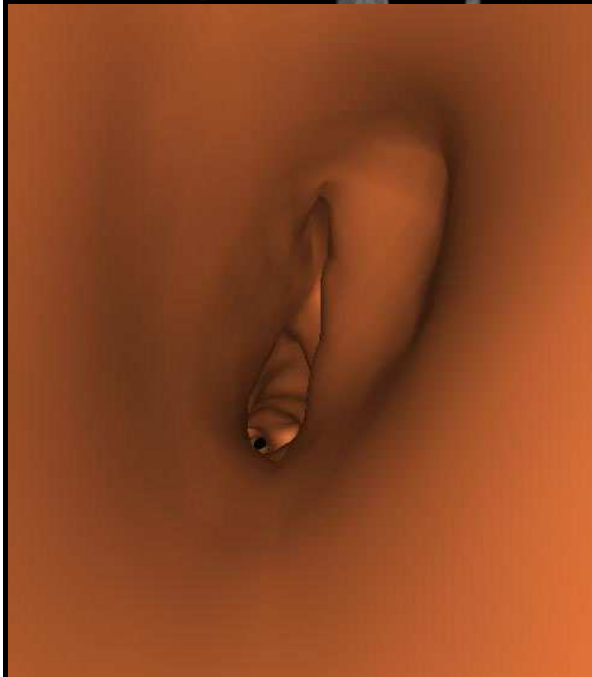
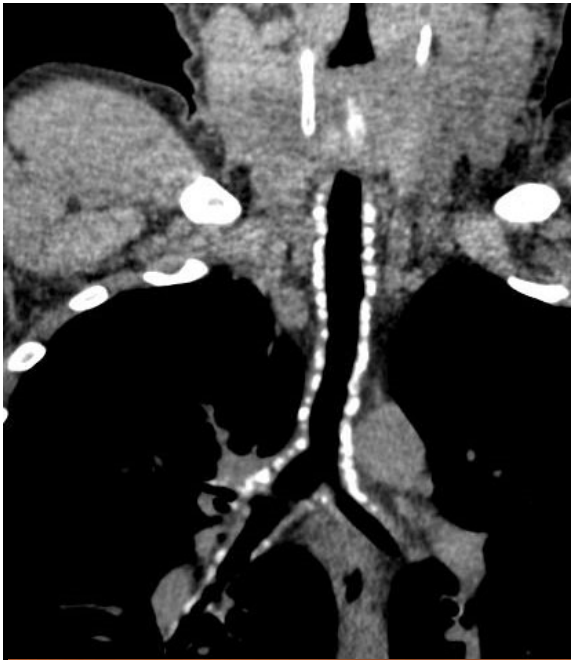
- Keratoconjunctivite, xerostomie, gonflement récurrent des parotides
- Processus autoimmun
- Infiltrats lymphoplasmocytaire au niveau de la trachée et des bronches
- Pneumonie lymphoïde interstitielle, fibrose interstitielle, lymphome, amyloïdose, bronchiolite oblitérante, épanchements pleuraux

Tannoue LT. Pulmonary involvement in collagen vascular disease: a review of the pulmonary manifestations of the Marfan syndrome, ankylosing spondylitis, Sjögren's syndrome, and relapsing polychondritis. J Thorac imaging 1992;7:62-77



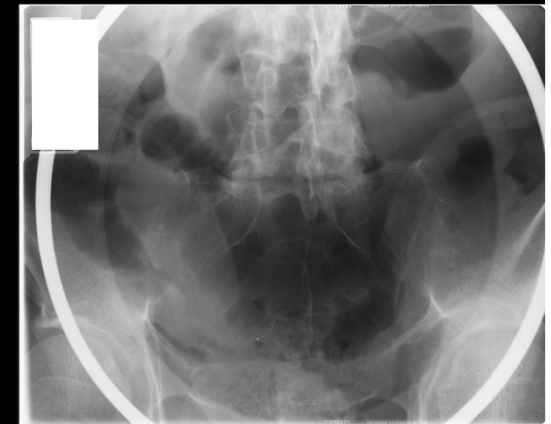
Polychondrite atrophiante

- Maladie rare (qqqs 100 cas)
- Inflammation épisodique des cartilages
 - Chondrite auriculaire, chondrite nasale, inflammation oculaire, dommage cochléaire et vestibulaire
 - Chondrite laryngée, trachéale, bronchique
- Manifestations radiologiques
 - Sténose trachéale-bronchique
 - Pneumonie-atelectasie
 - CT expiratoire



Spondylarthrite ankylosante

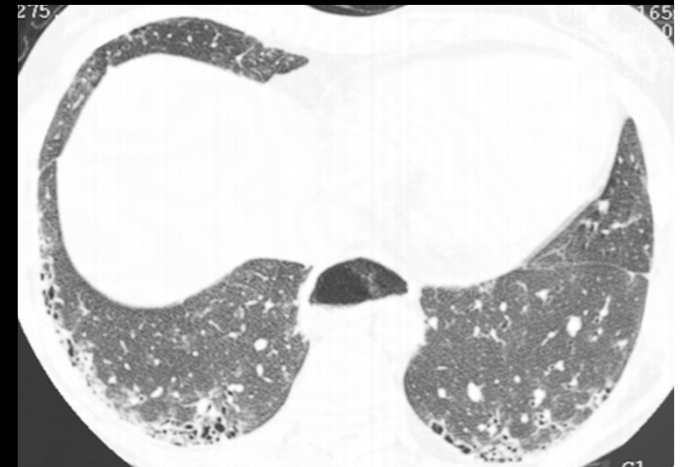
- Spondylarthrite séronégative
- Atteinte préférentielle du squelette axial
- Rigidité progressive de la colonne
- Anomalies pulmonaires (1%)
 - Fibrose des LS, atteinte interstitielles, DDB, Emphysème
 - Piégeage expiratoire



Souza AS Jr, Muller NL, Marchiori E, Soares-Souza LV, de Souza Rocha M. Pulmonary abnormalities in ankylosing spondylitis: inspiratory and expiratory high-resolution CT findings in 17 patients. *J Thorac Imaging.* 2004;19(4):259-63

Rôle du CT

- Détecter les anomalies parenchymateuses de façon précoce
 - Caractérisation des anomalies
 - Localisation pour biopsie
- Détecter les atteintes vasculaires/des voies aériennes
 - Modifications transmurales, maladies thrombo-emboliques
 - Trachée, bronche, piégeage expiratoire
- Illustrer les anomalies associées
 - coeur, plèvre, péricarde, tissus mous
- Suivre les effets du traitement



Schurawitzki H, Stiglbauer R, Graninger W. Interstitial lung disease in progressive systemic sclerosis: high-resolution CT versus radiography. Radiology. 1990 Sep;176(3):755-9

