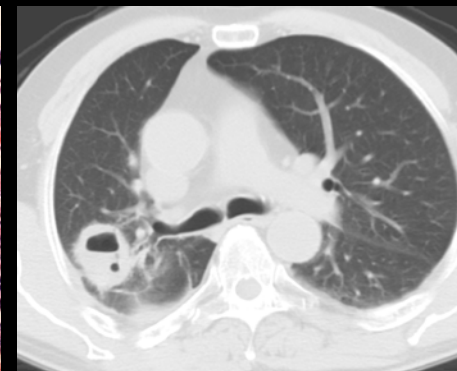
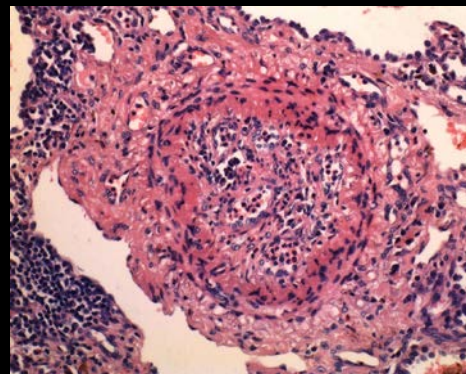
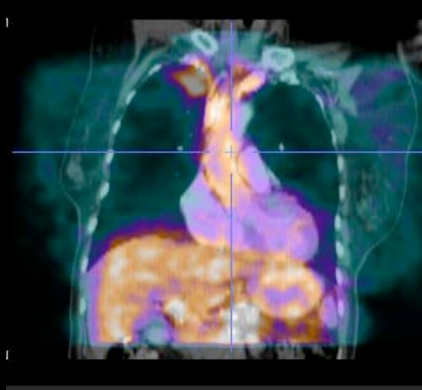




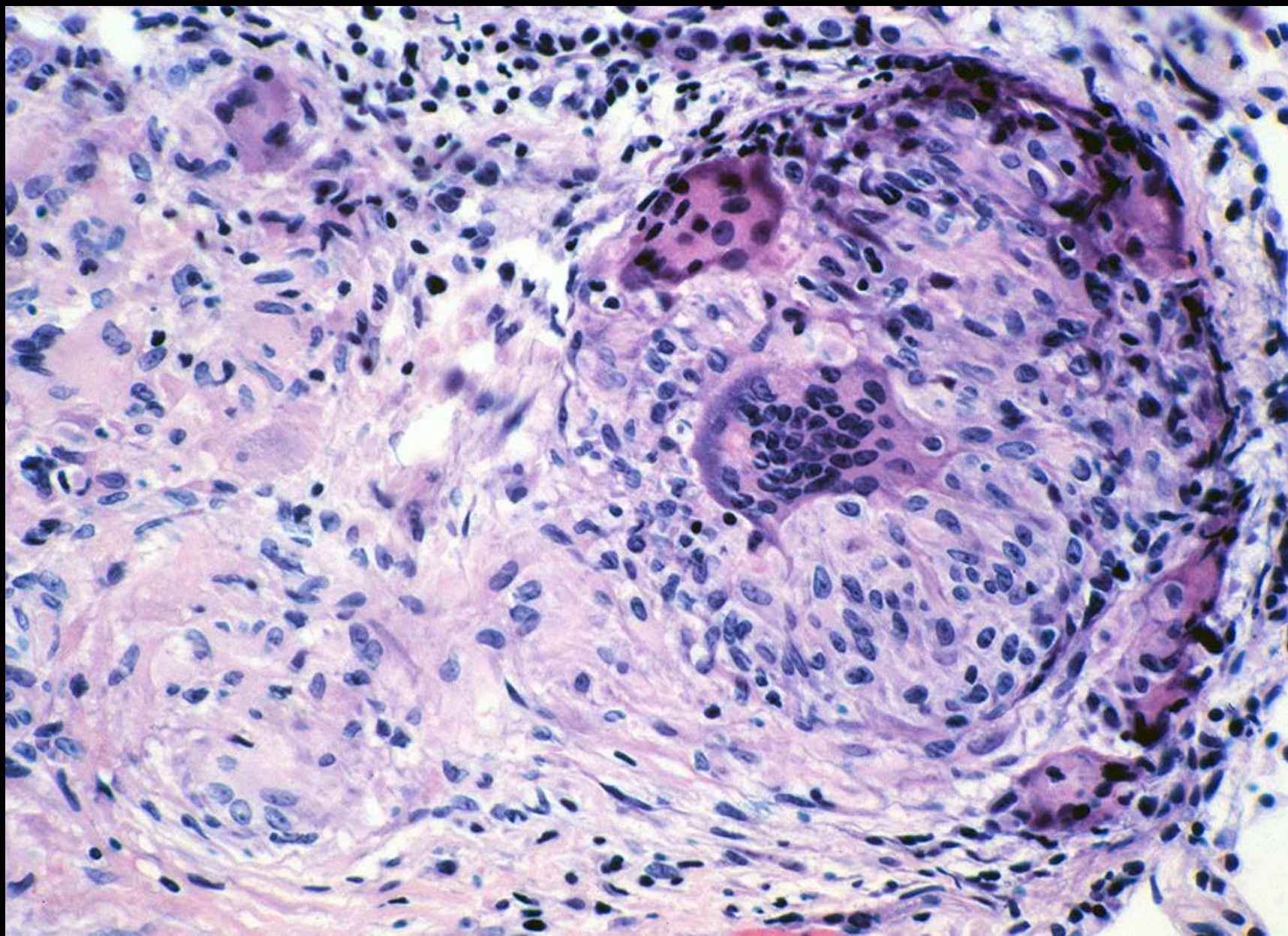
Imagerie des granulomatoses vascularites et connectivites

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

DES Radiologie
2ème cycle
15 décembre 2017

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

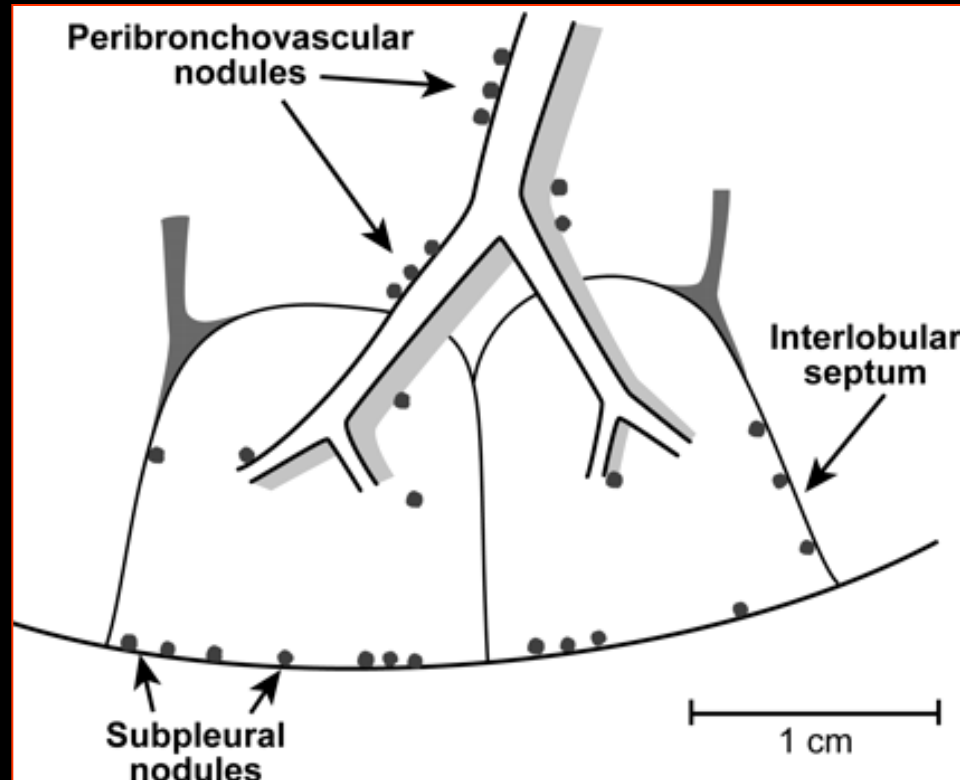
Imagerie

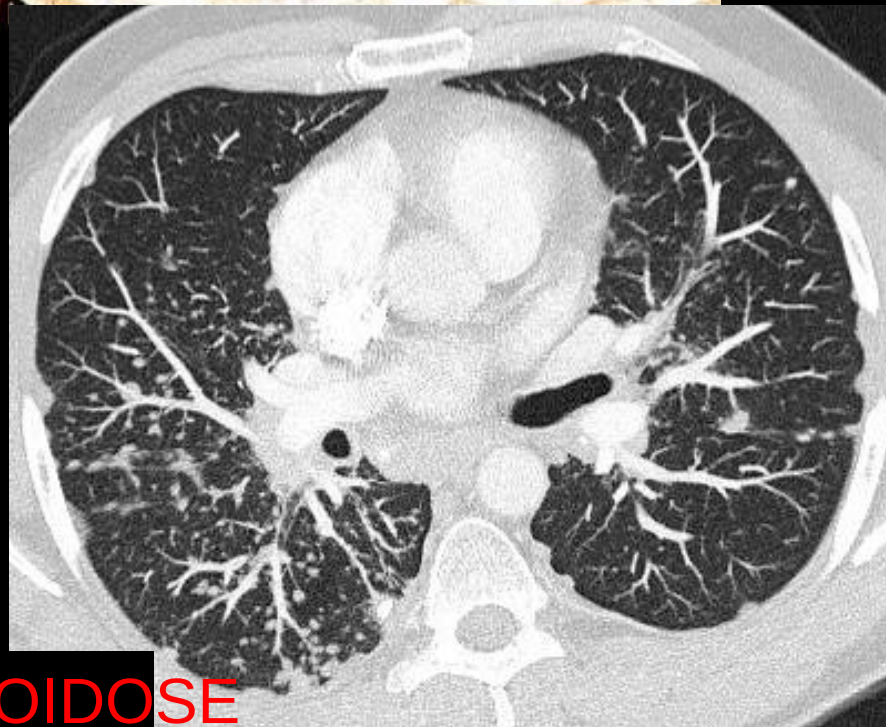
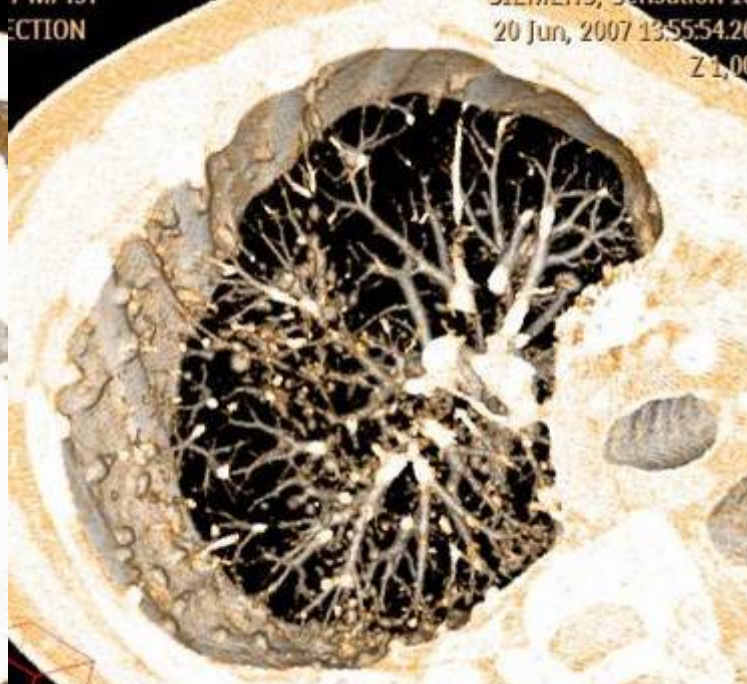
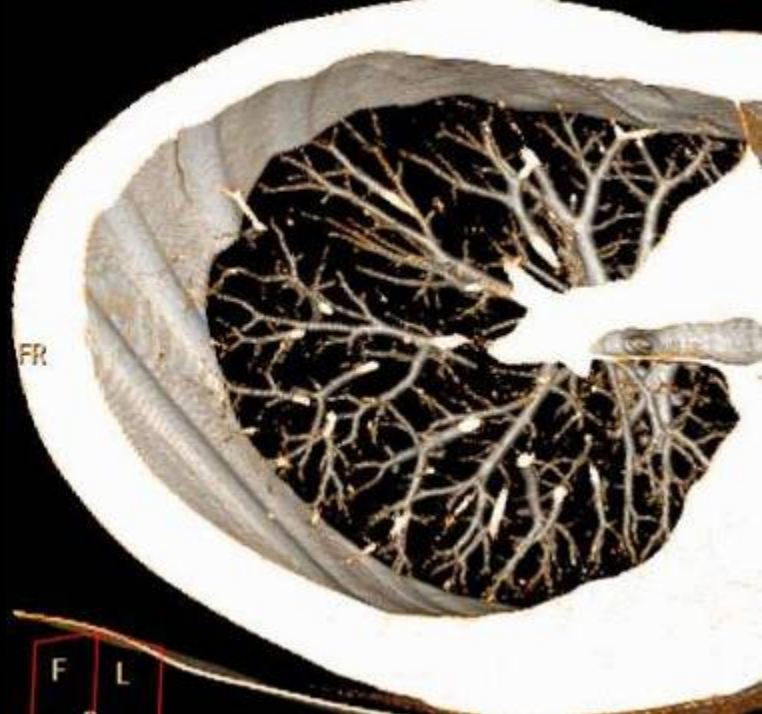
- Adénopathies hilaires et médiastinales symétriques
- Micronodules (1-5 mm)
- Atteinte centri-lobulaire, périvasculaire, lymphatique, septae sous-pleuraux
- Fibrose progressive et massive, kystes, bulles...
- Sarcoïdose alvéolaire: nodules et densifications avec bronchogramme aérique

Modèle micronodulaire péri-lymphatique

Caractéristiques :

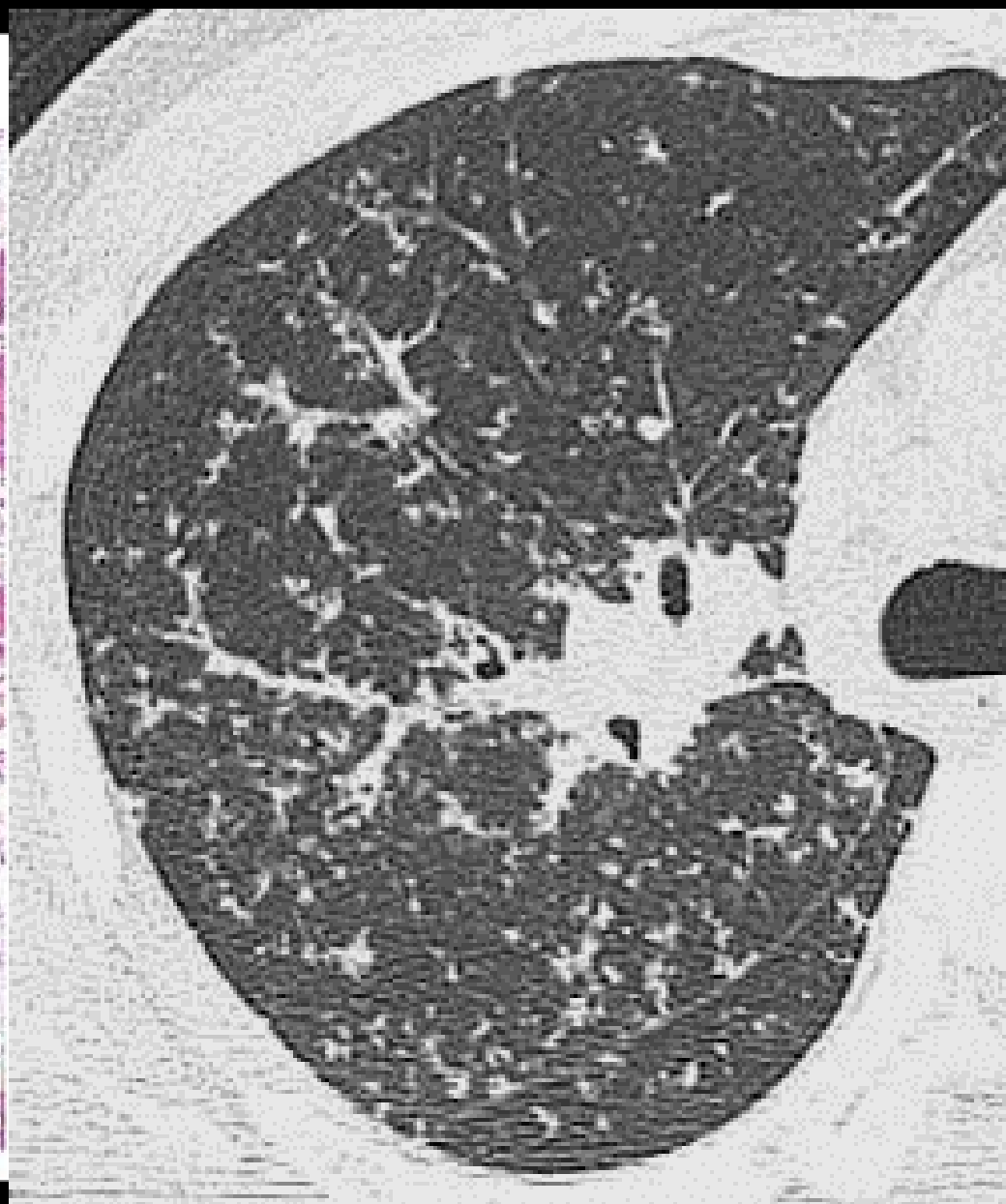
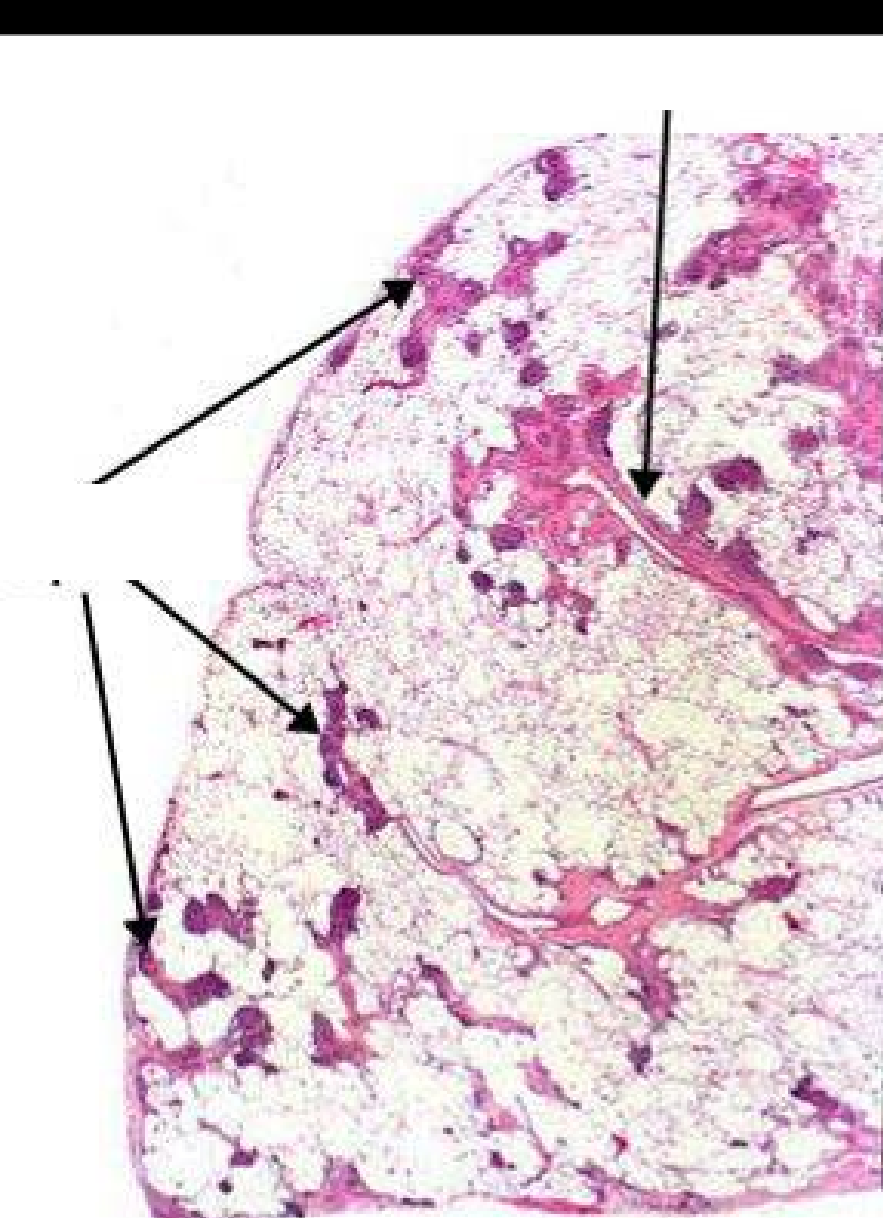
- contours nets
- forte densité
- distribution
 - Le long des scissures et de la plèvre périphérique
 - Le long des septa interlobulaires
 - Le long des trajets vasculaires et bronchiques
 - au cœur du lobule : axes artériolo-bronchiolaires

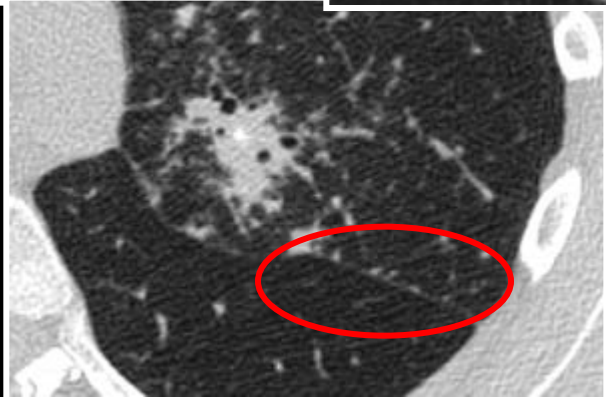
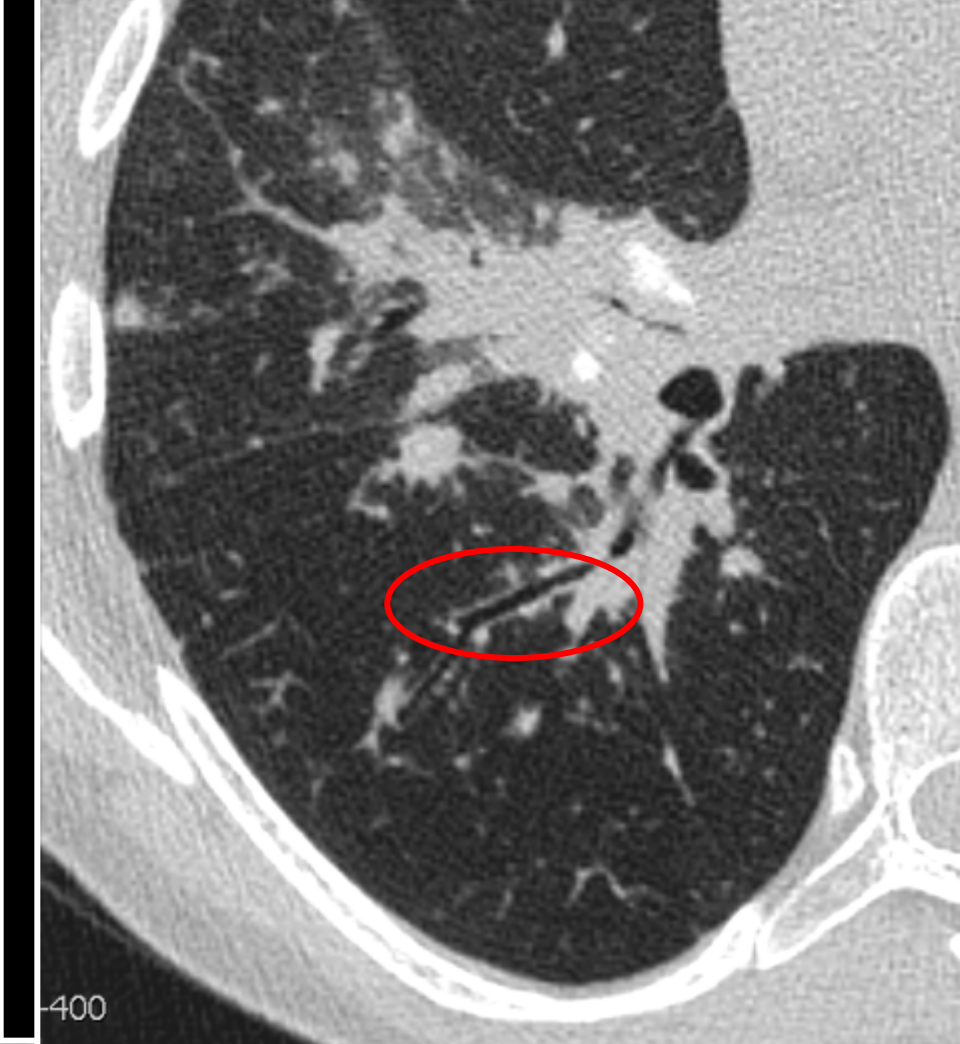
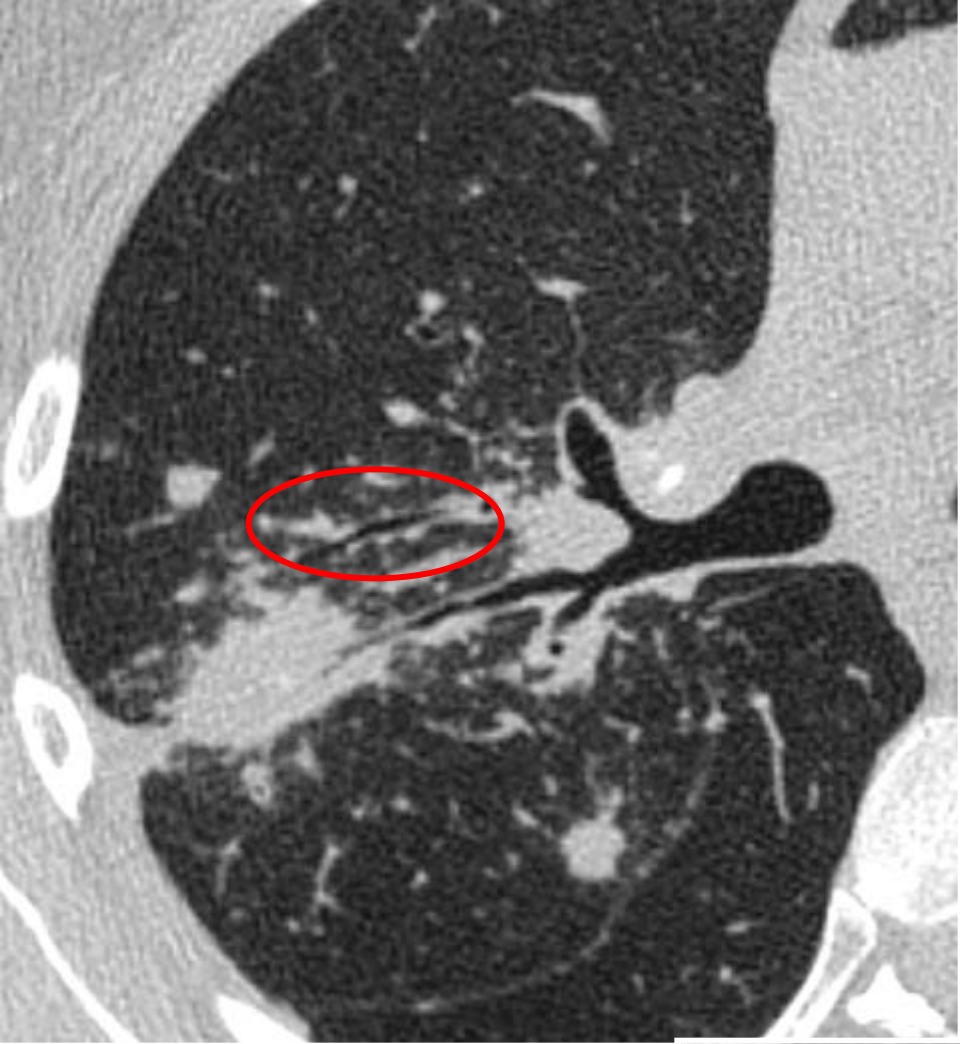


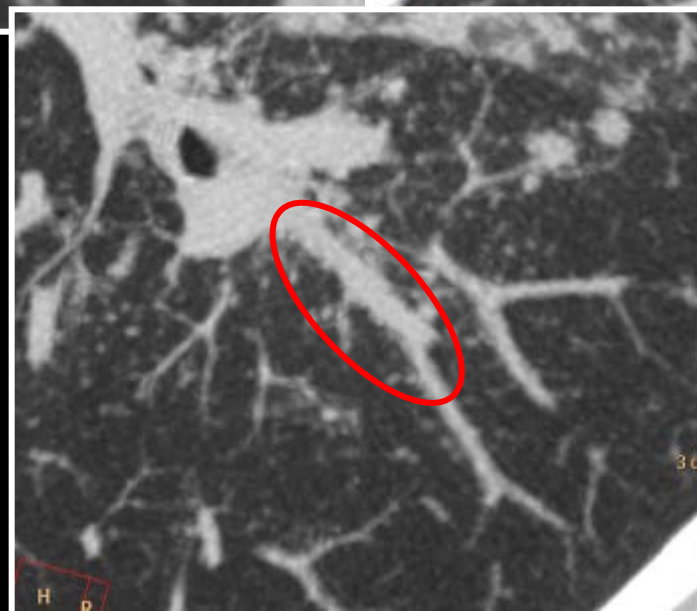
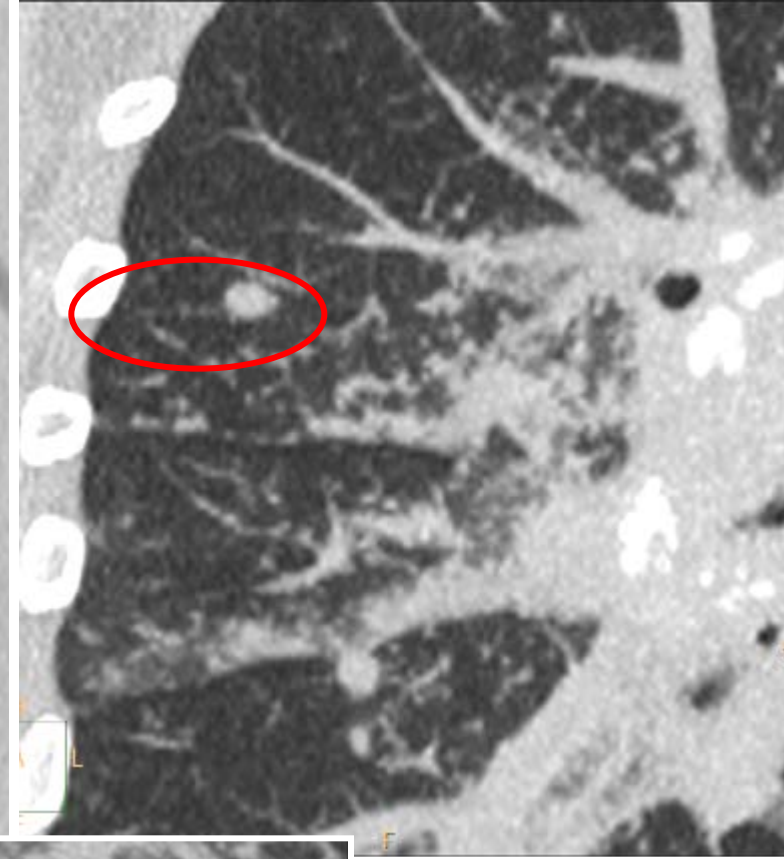


SARCOIDOSE



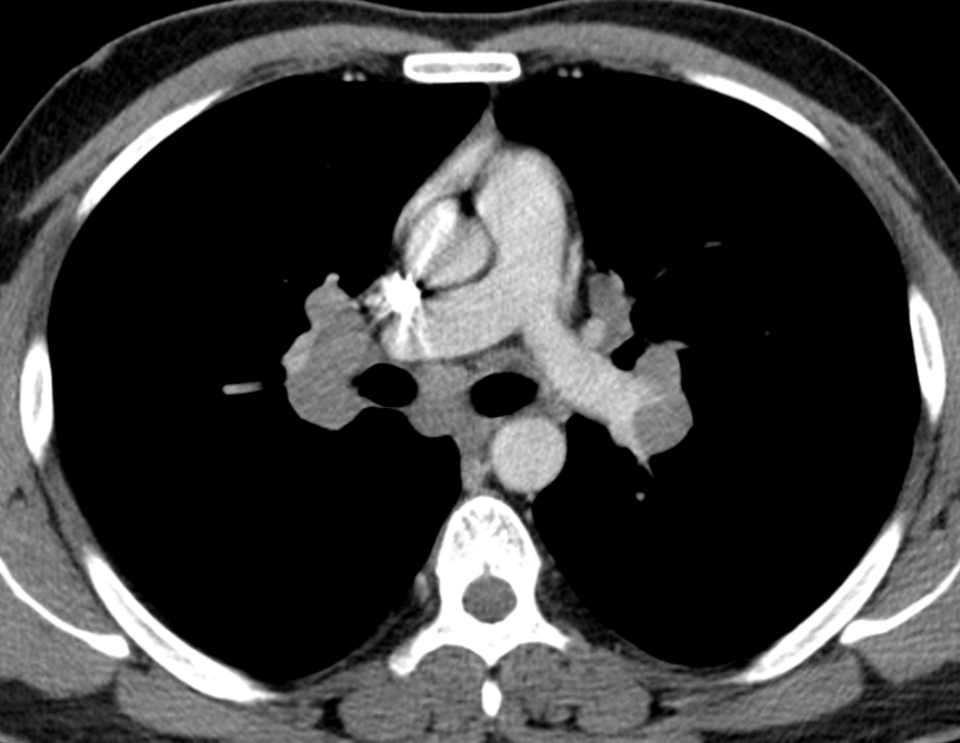






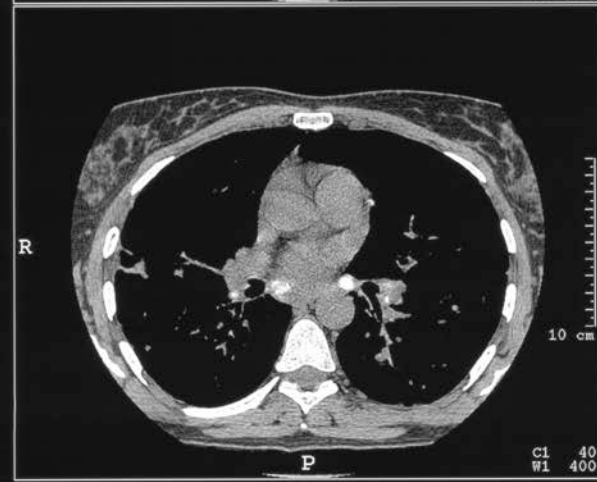
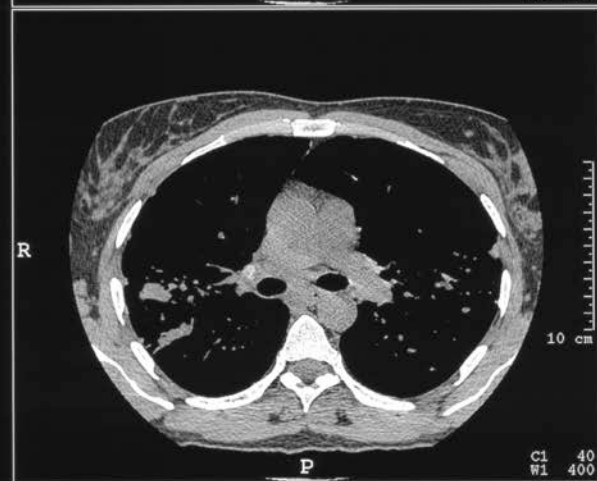
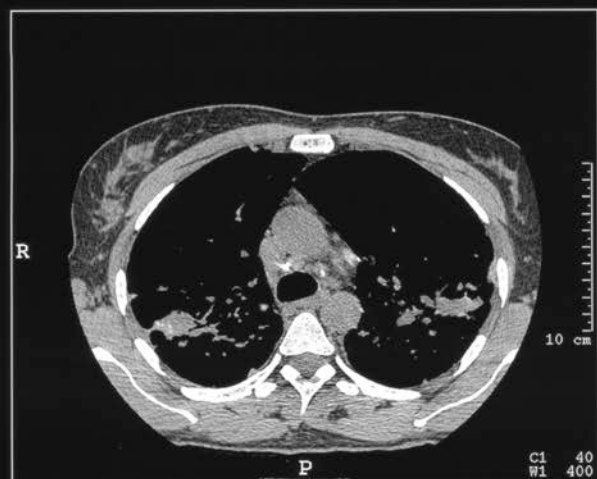
Stadification

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

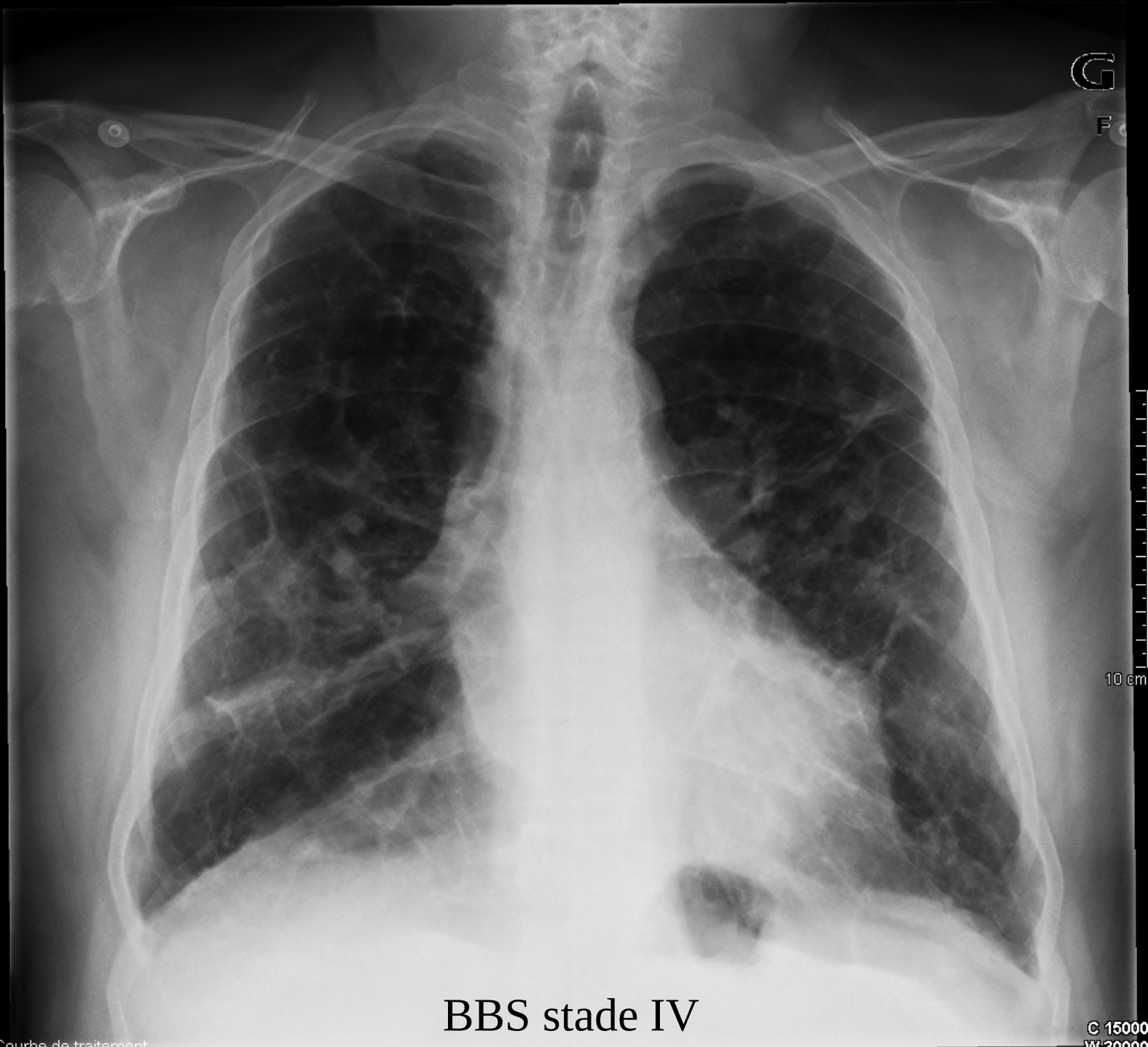




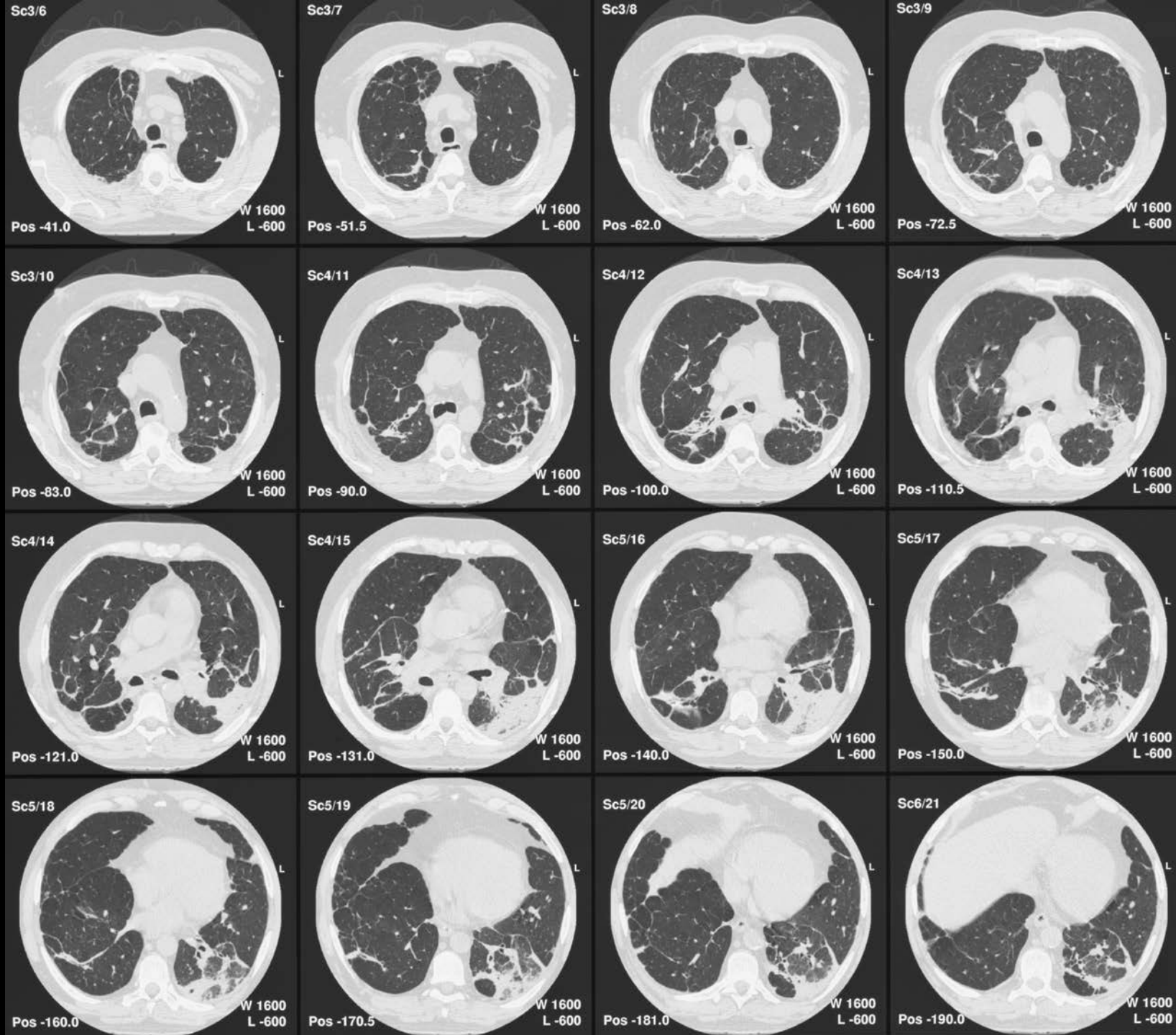
Evolution BBS

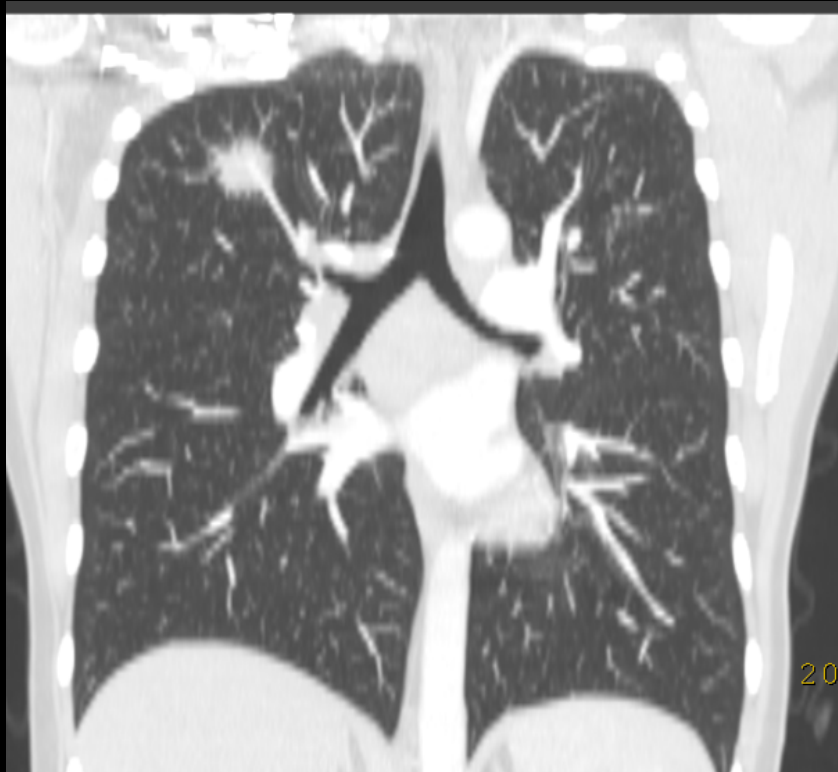
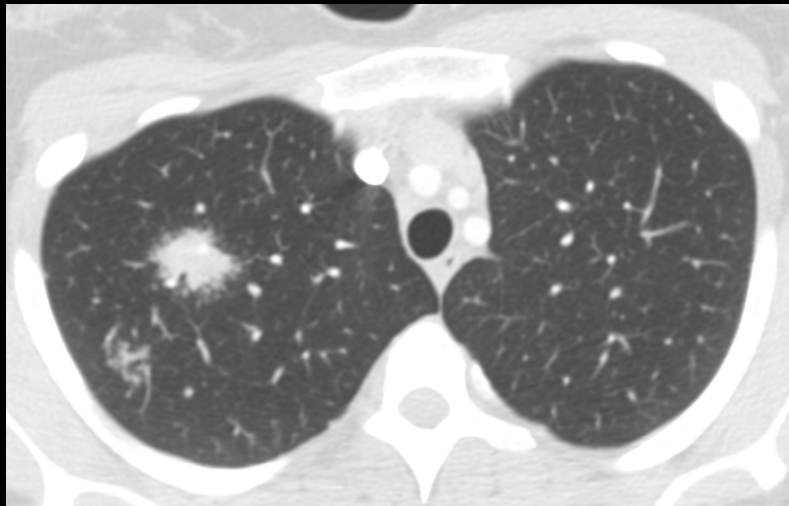


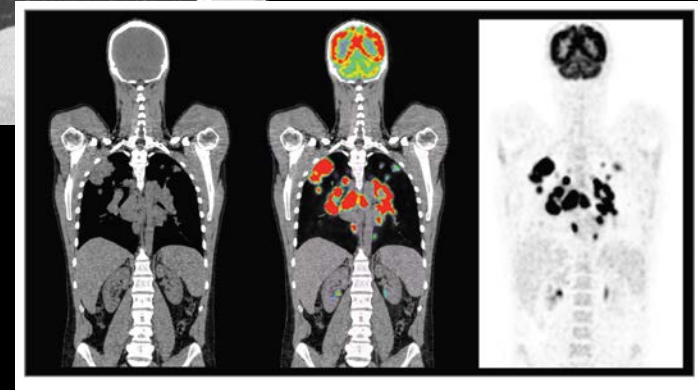
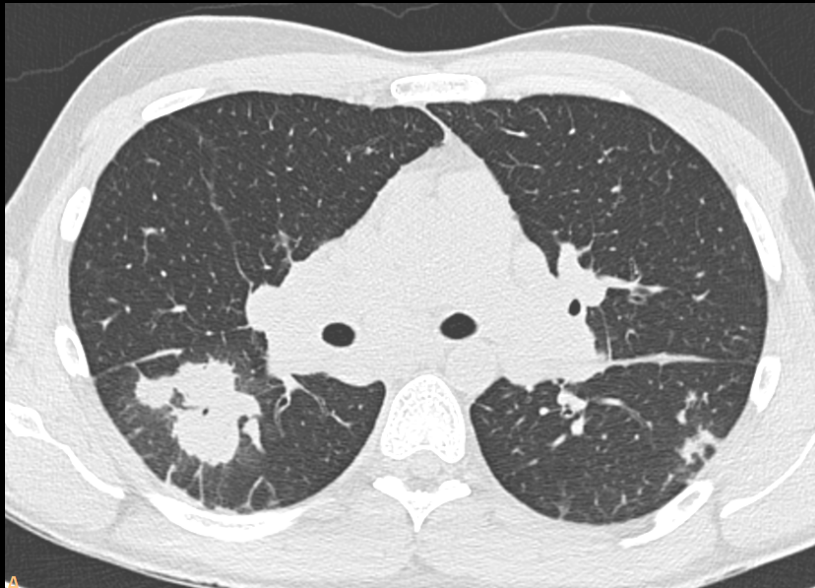
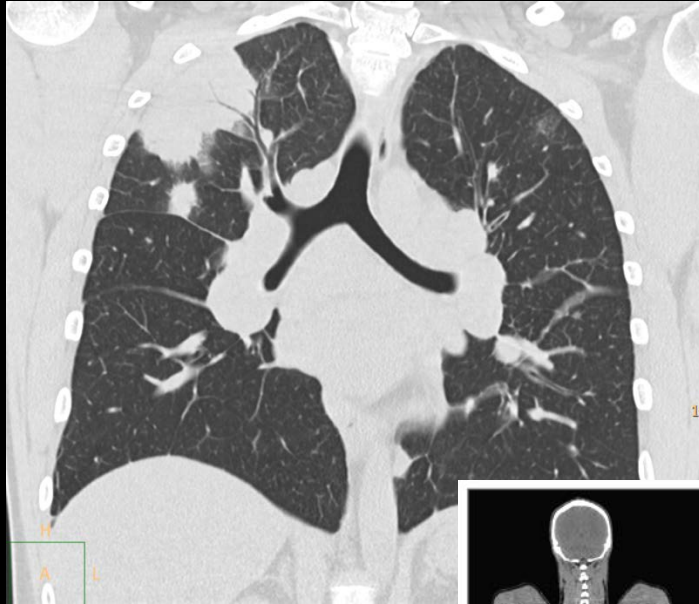
Evolution BBS



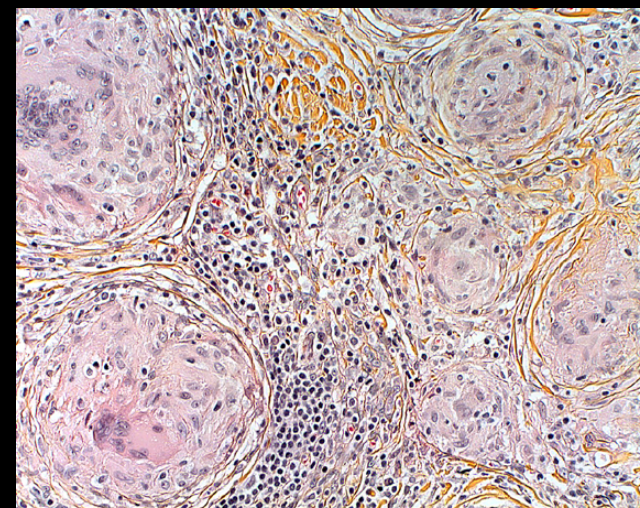
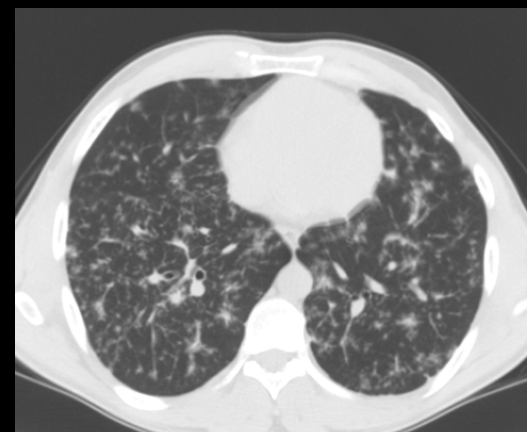
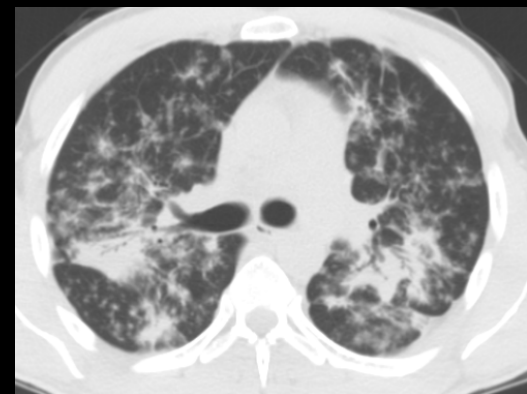
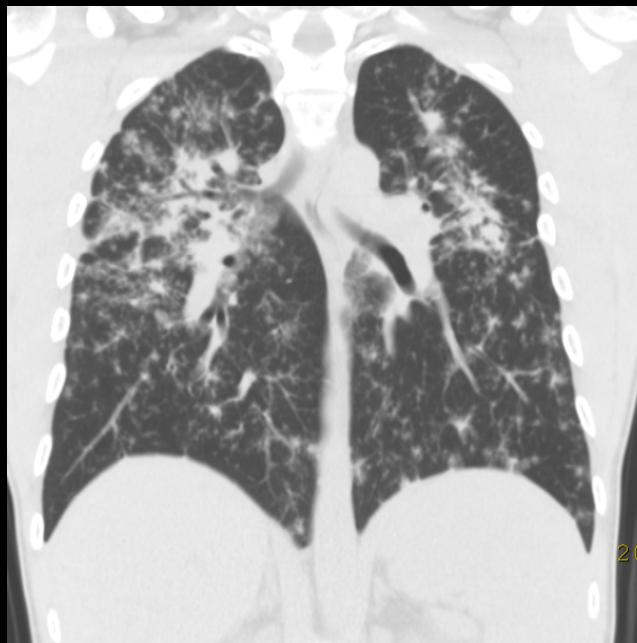
BBS stade IV







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

Radiologic Manifestations of Sarcoidosis in Various Organs¹

CME FEATURE

See accompanying test at http://www.rsna.org/education/irg_cme.html

LEARNING OBJECTIVES FOR TEST 3

After reading this article and taking the test, the reader will be able to:

- Recognize both typical and atypical imaging features of sarcoidosis in various anatomic locations.
- Describe clinical manifestations that play an important role in diagnosing sarcoidosis.
- Discuss various clinical settings and syndromes that are related to sarcoidosis.

Takashi Koyama, MD • Hiroyuki Ueda, MD • Kaori Togashi, MD
Shigeaki Umeoka, MD • Masako Kataoka, MD • Sonoko Nagai, MD

Sarcoidosis is a systemic disorder of unknown cause with a wide variety of clinical and radiologic manifestations. The diagnosis is usually made on the basis of these manifestations supported by histologic findings. Systemic manifestations (eg, Löfgren syndrome, Heerfordt syndrome) are commonly seen at clinical examination. Bilateral hilar lymphadenopathy is the most common radiologic finding—frequently with associated pulmonary infiltrates—and typically has a characteristic perivascular distribution at high-resolution chest computed tomography. Radiologic findings in the short tubular bones of the hands and feet and magnetic resonance imaging findings of nodular involvement of muscle are often sufficient to raise suspicion for sarcoidosis. In the liver, spleen, kidneys, and scrotum, coalescing granulomas form nodules whose imaging features may occasionally be nonspecific, although familiarity with the relevant clinical settings will be helpful in recognizing the presence of sarcoidosis. Radiologic recognition of cardiac and central nervous system involvement is also important because patients may be only mildly symptomatic. The clinical course and prognosis of sarcoidosis are highly variable, often correlating with the mode of onset. Familiarity with the clinical and radiologic features of sarcoidosis in various anatomic locations plays a crucial role in diagnosis and management.

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Abbreviations: ACE = angiotensin-converting enzyme, CNS = central nervous system, FLAIR = fluid-attenuation inversion recovery

Index terms: Abdomen, diseases, 70.22, 76.22, 77.22, 81.22, 847.22 • Head and neck, 20.22 • Nervous system, diseases, 10.22, 30.22 • Sarcoidosis, 22.22 • Thorax, diseases, 51.22, 60.22, 679.22

RadioGraphics 2004; 24:87–104 • Published online 10.1148/rg.241035076

¹From the Department of Radiology, Kyoto University Hospital, 54 Kawahara-cho, Shogoin, Sakyo-ku, Kyoto 606-8507, Japan (T.K., H.U.); and the Departments of Diagnostic and Interventional Imageology (K.T.), Nuclear Medicine and Diagnostic Imaging (S.U., M.K.), and Respiratory Medicine (S.N.), Graduate School of Medicine, Kyoto University of Medicine, Kyoto, Japan. Recipient of a Certificate of Merit award for an education exhibit at the 2002 RSNA scientific assembly. Received March 21, 2003; revision requested April 23 and received August 28; accepted August 28. All authors have no financial relationships to disclose. Address correspondence to T.K. (e-mail: montpeti@kuhp.kyoto-u.ac.jp).

2** multiple body systems

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RG ■ Volume 24 • Number 1

Koyama et al 93

RadioGraphics

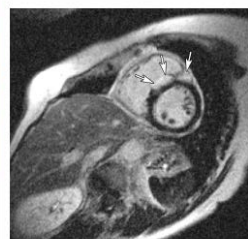


Figure 10. Cardiac sarcoidosis in a 59-year-old woman with abnormal electrocardiographic findings. Contrast-enhanced turbo fast low-angle shot inversion-recovery image (short-axis view) shows enhancement in the interventricular septum (arrows). The long-axis view was deleted.

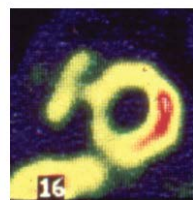


Figure 11. Cardiac sarcoidosis in a 60-year-old man who presented with complete atrioventricular blockage. Image obtained with Ga-67 citrate single photon emission computed tomography (short-axis view) shows diffuse abnormal radiotracer accumulation in the myocardium. Areas of strong uptake represent foci of increased disease activity.

January-February 2004

RG ■ Volume 24 • Number 1

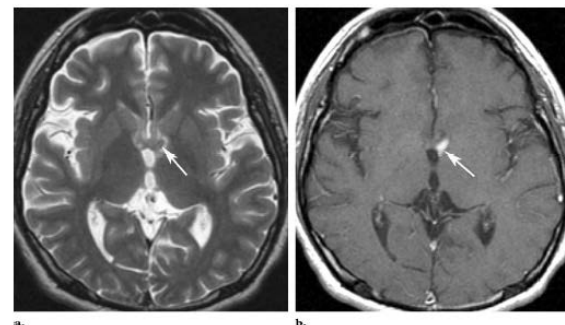


Figure 12. Neurosarcoidosis in a 24-year-old man who presented with diabetes insipidus. (a) Axial T2-weighted MR image demonstrates an iso-intense periventricular lesion (arrow) surrounded by minimal high-signal-intensity edema. (b) On a contrast-enhanced T1-weighted MR image, the lesion demonstrates enhancement (arrow).



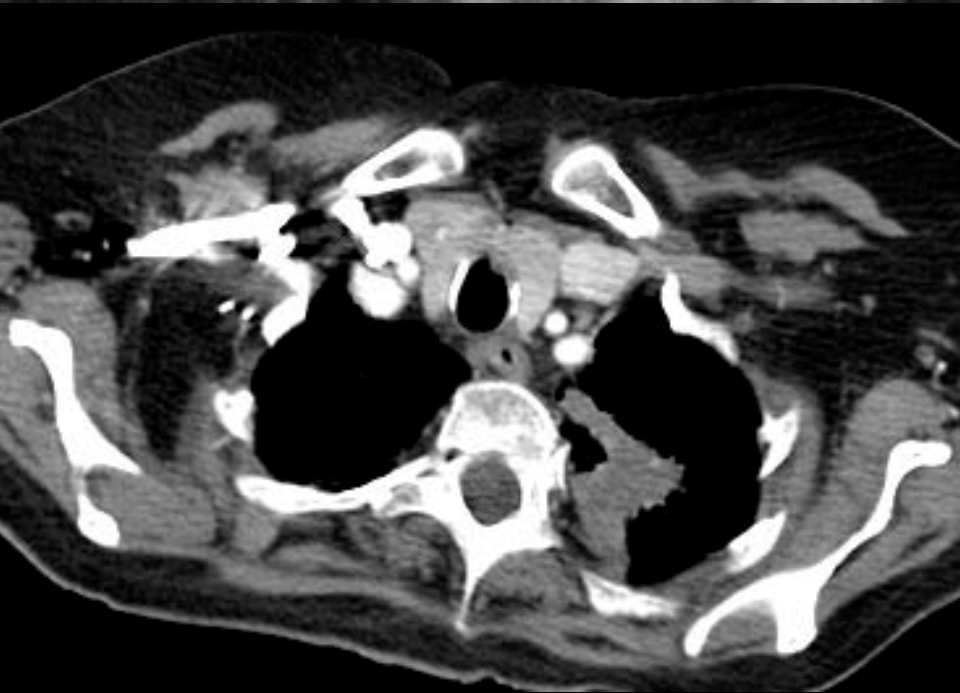
Figure 27. Nodular type muscle involvement in the same patient as in Figure 25. T2-weighted (a) and contrast-enhanced (b) MR images demonstrate a nodular type muscle lesion (arrows). The lesion has a central area of decreased signal intensity (as it did with all sequences). The periphery of the lesion demonstrates increased signal intensity on the T2-weighted image and prominent enhancement on the contrast-enhanced image.

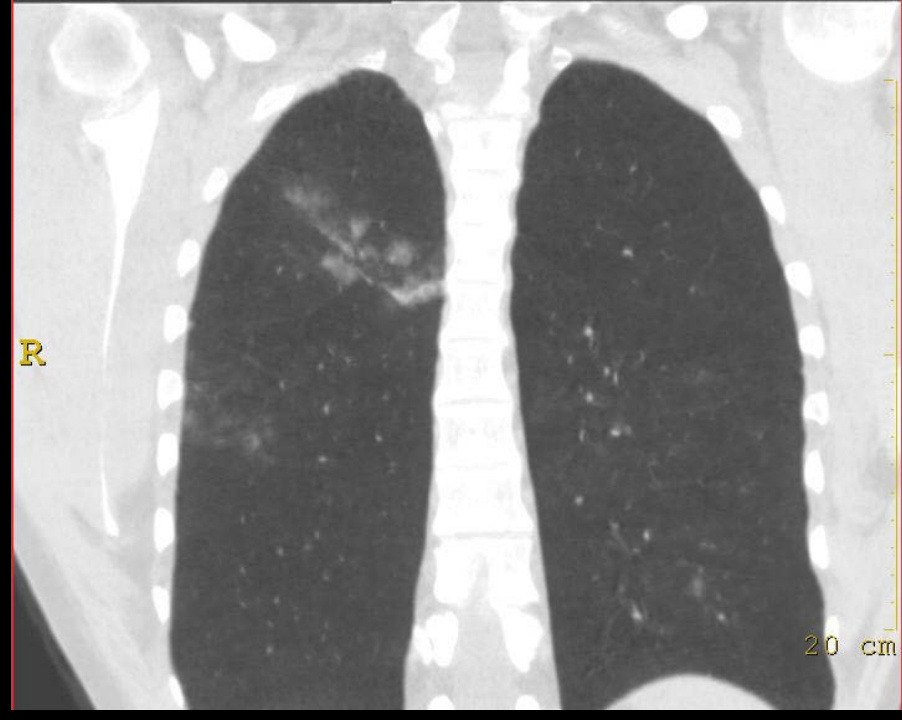
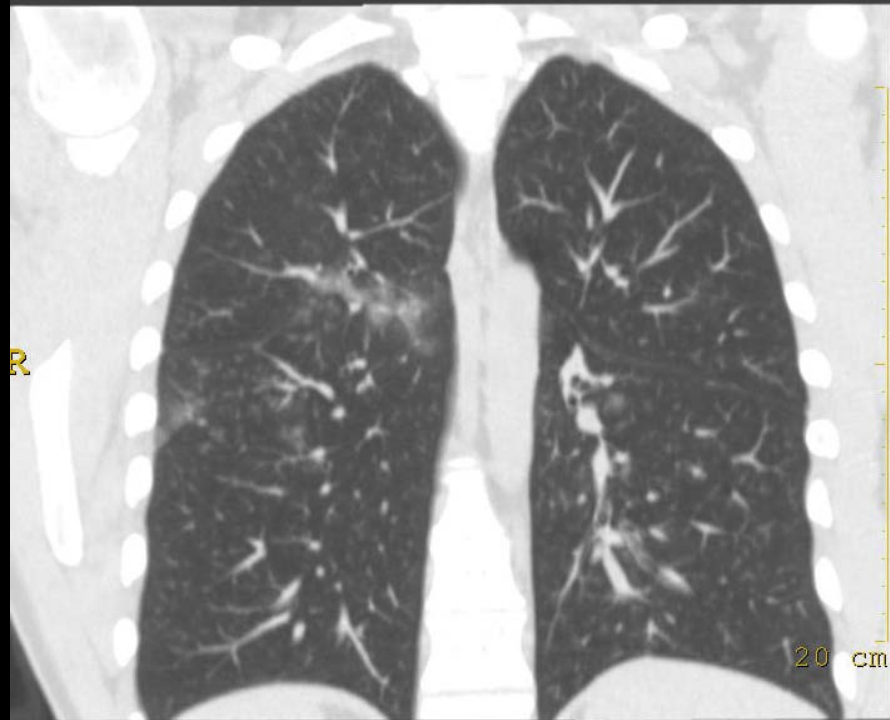
Granulomatose de Wegener

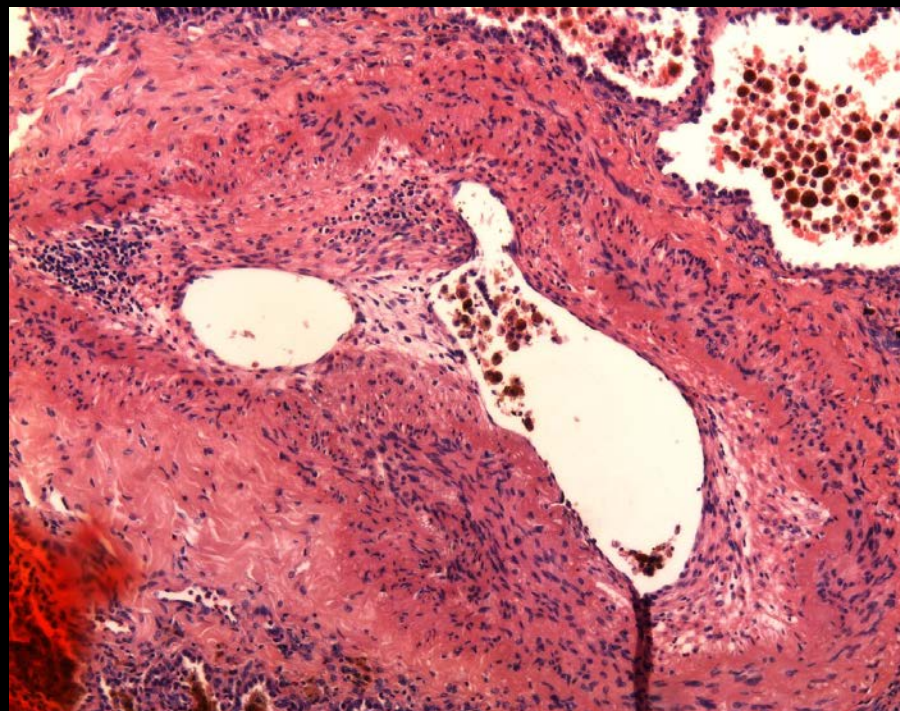
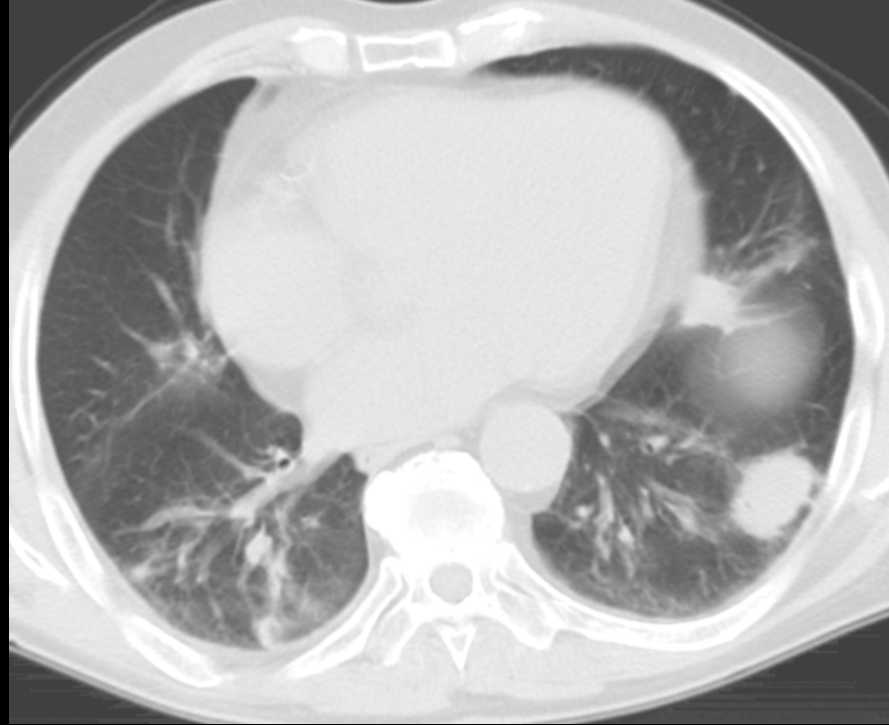
- Maladie multisystémique à expression clinique variable
- Inflammation granulomateuse nécrosante
 - Voies aériennes supérieures et inférieures
 - Reins
 - Organes systémiques et tissus
- 3/100 000 cas/an aux USA
- Pic: 30-50 ans
- C-ANCA, Anticorps anti-cellule endothéliale

Anomalies CT chez 57 patients avec granulomatose de

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

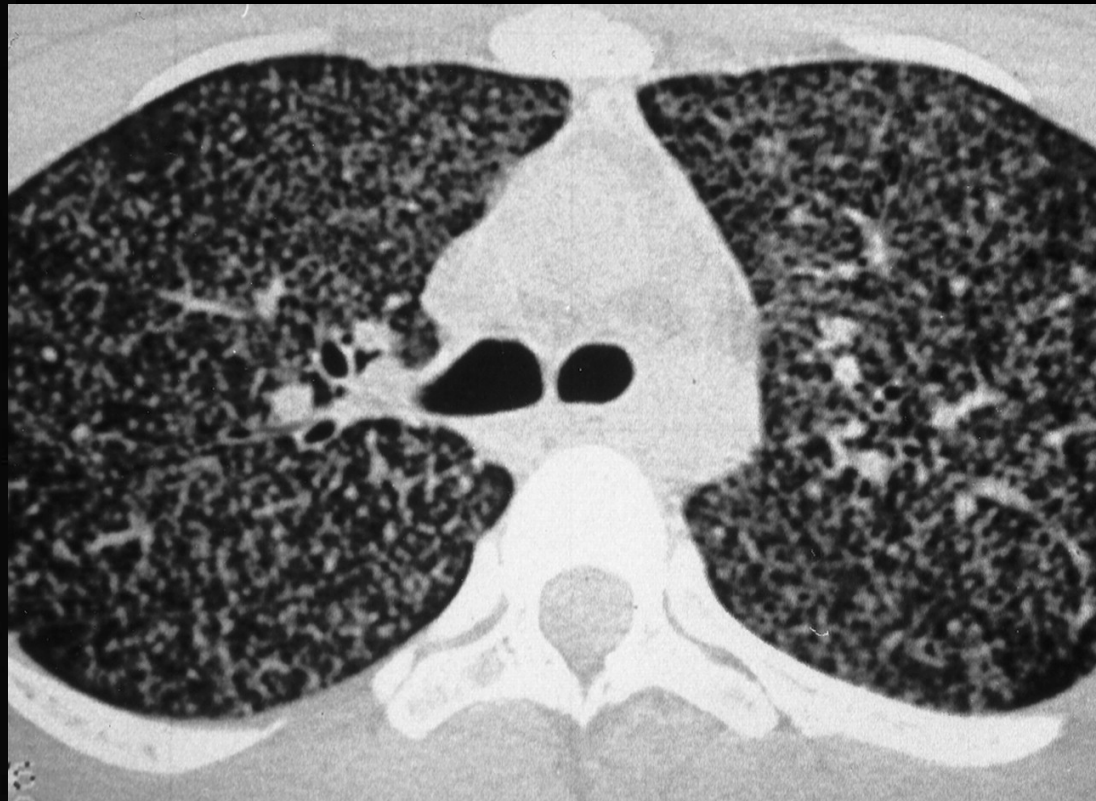
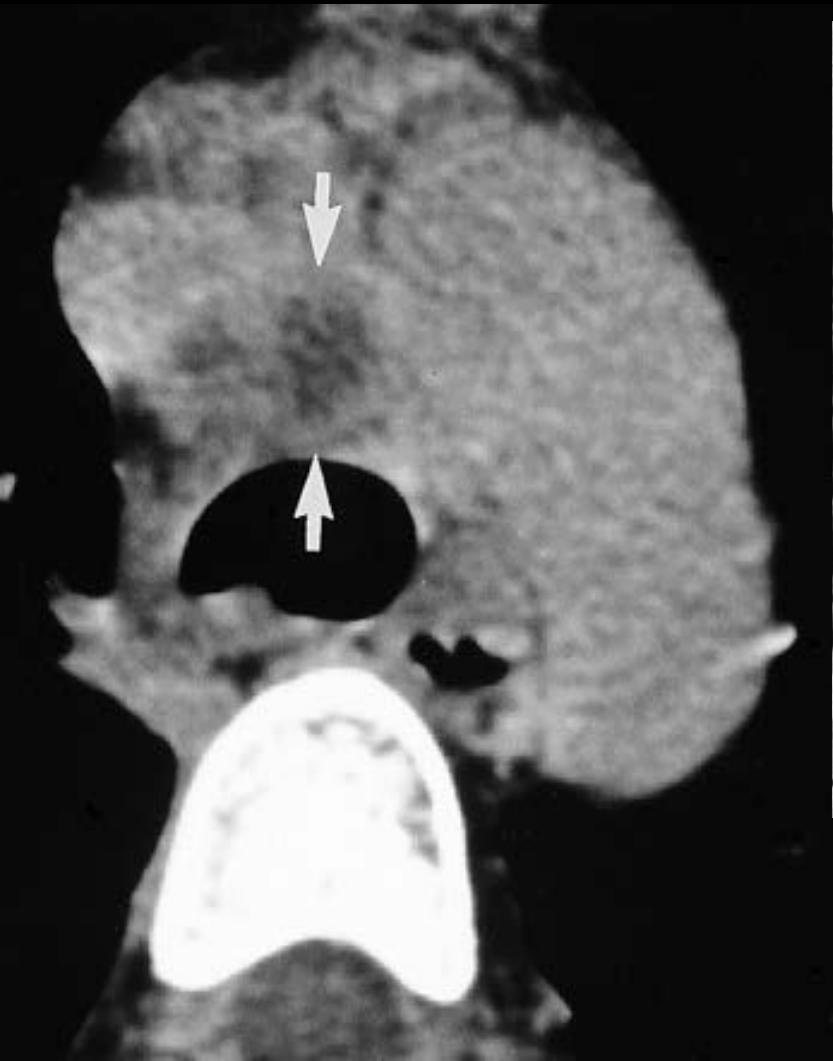






Tuberculose

- 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

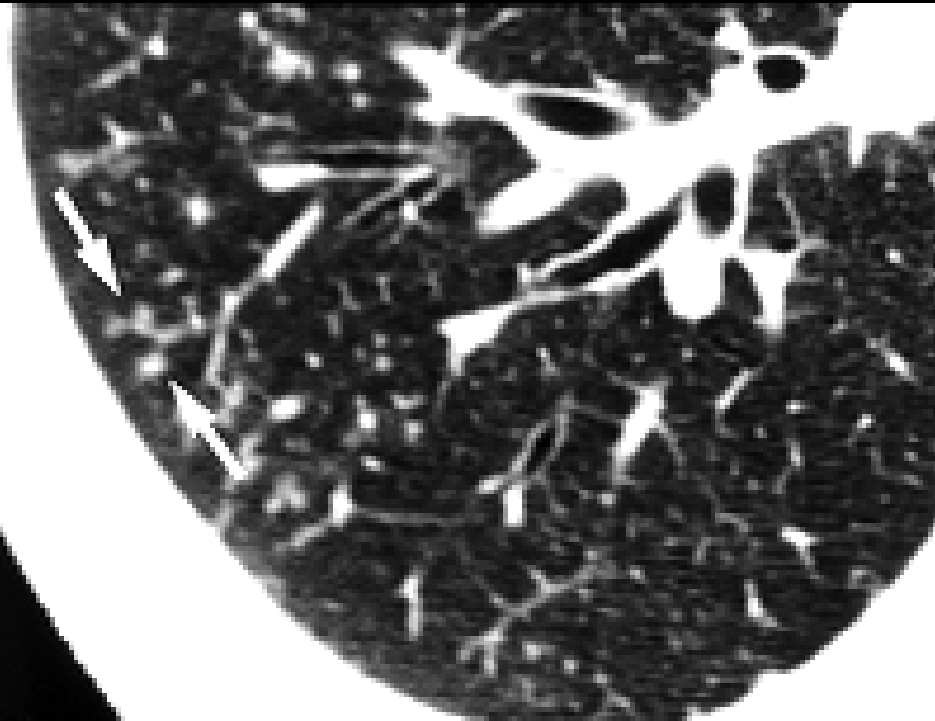
Tuberculose post-primaire

- 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

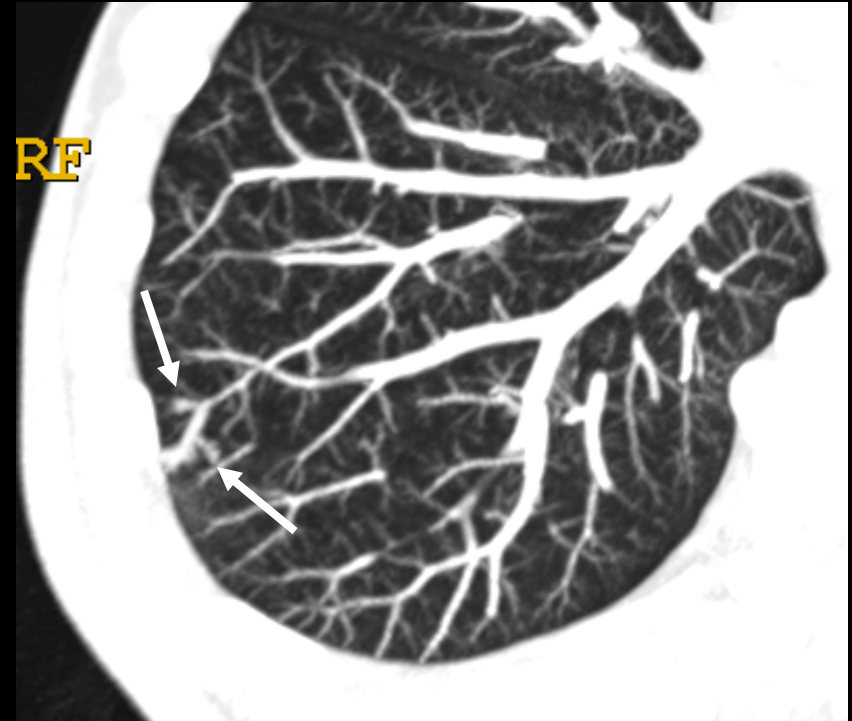




« Tree-in-bud » sign = image non
spécifique:



1. Atteinte
aériennes



2 Atteinte des petites artères

Peripheral airway disease

Infection

Bacterial

Mycobacterium tuberculosis

M avium-intracellulare complex

Staphylococcus aureus

Haemophilus influenzae

Fungal

Aspergillus

Viral

Cytomegalovirus

Respiratory syncytial virus

Congenital disorders

Cystic fibrosis

Kartagener syndrome

Idiopathic disorders

Obliterative bronchiolitis

Diffuse panbronchiolitis

Aspiration

Inhalation

Toxic fumes and gases

Immunologic disorders

Allergic bronchopulmonary aspergillosis

Connective tissue disorders

Rheumatoid arthritis

Sjögren syndrome

Peripheral pulmonary vascular disease

Neoplasms

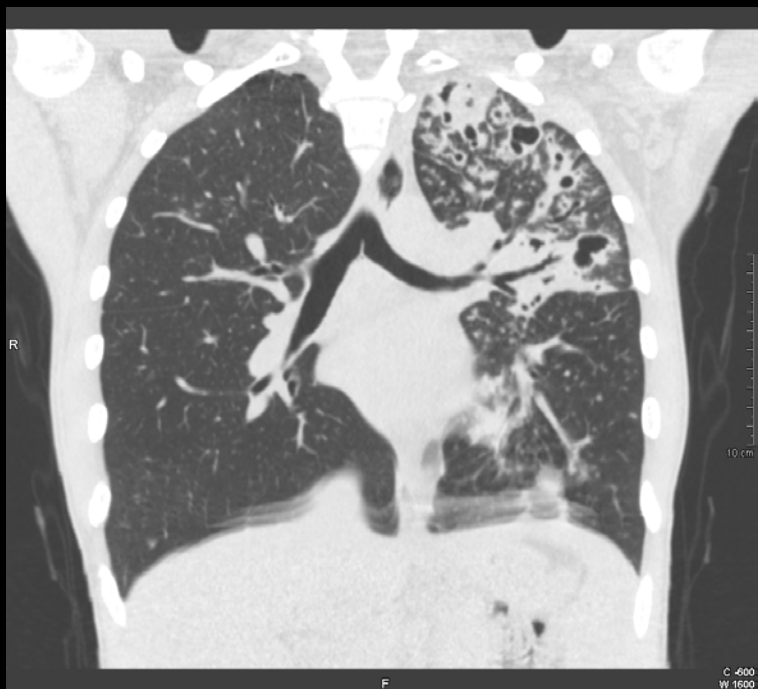
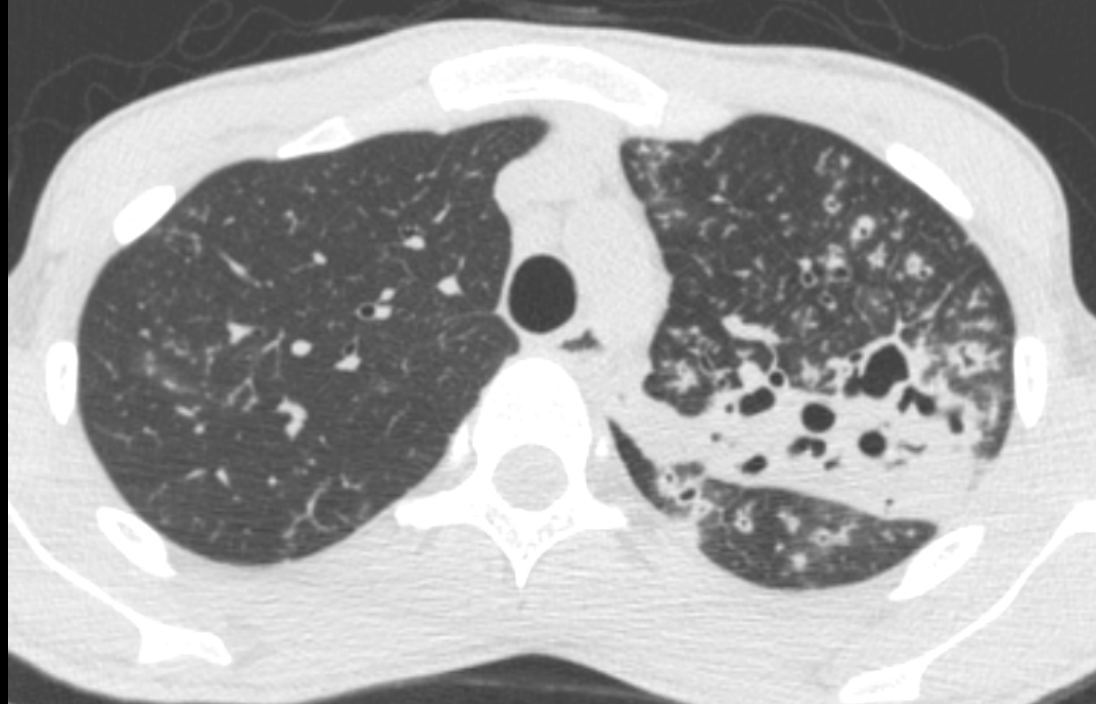
Gastric cancer

Breast cancer

Ewing sarcoma

Renal cancer

**Tree-in-Bud Pattern at Thin-
Section CT of the Lungs:
Radiologic-Pathologic Overview**
Santiago Enrique Rossi, MD,
Radiographics 2005; 25: 789-801







R

10 cm

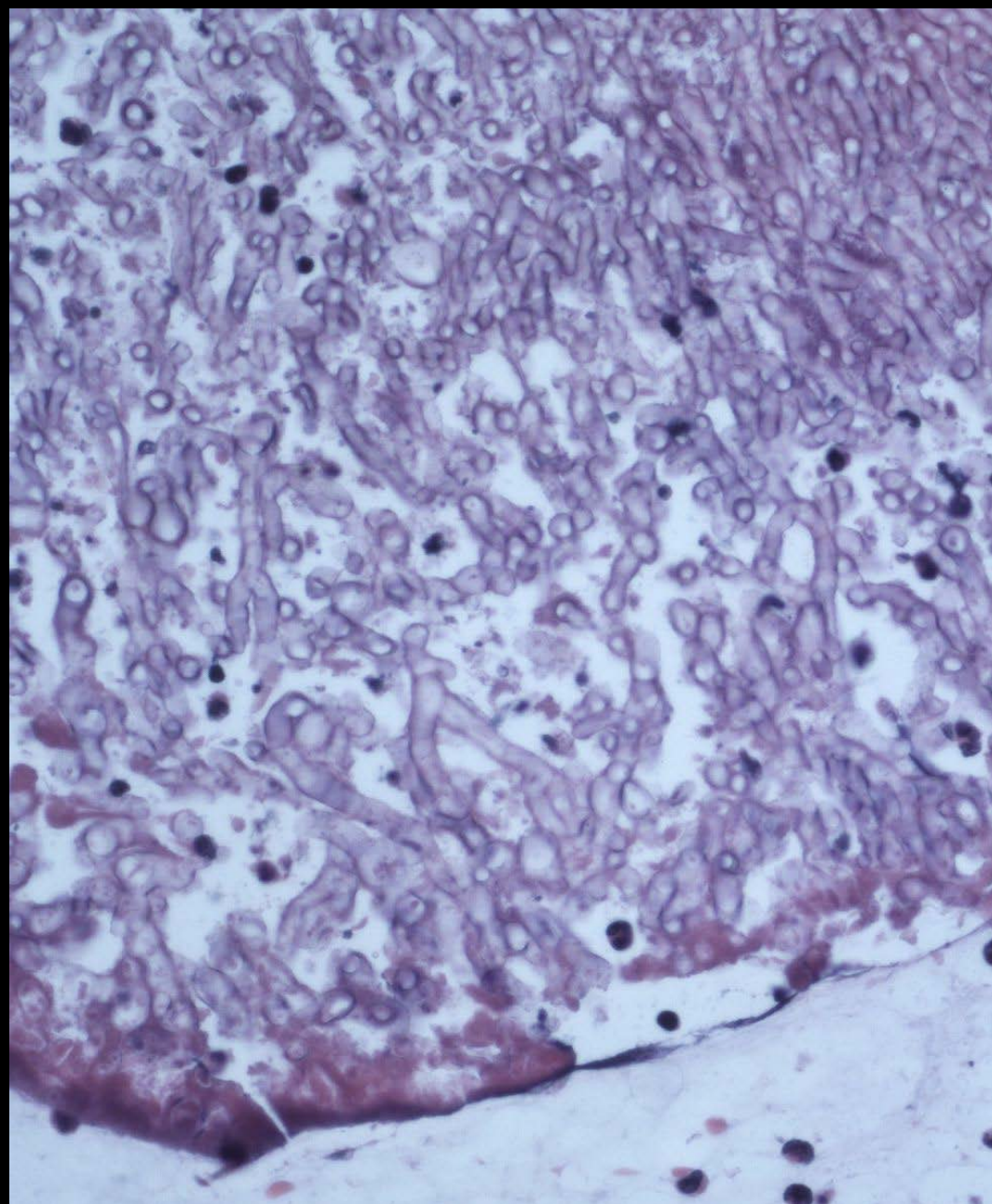
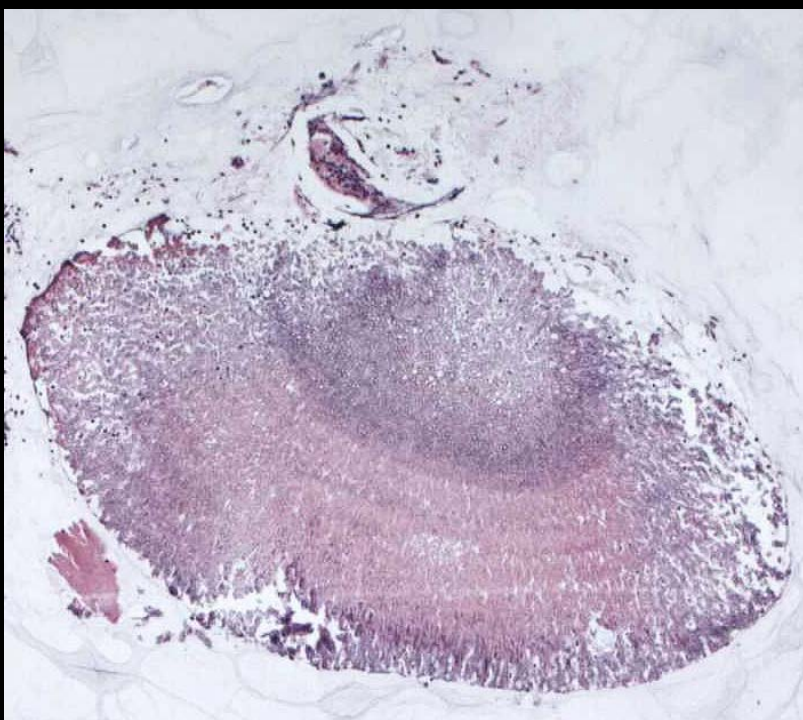
C -600
W 1600

F



Aspergillose

- 300 espèces
- *A. fumigatus*+++; <5% *A. terreus*; rare *A. niger* et *flavus*
- Ubiquitaire dans l'environnement
 - terre, H₂O, matériel organique
 - Colonisation des muqueuses nasales
- Pathogénèse complexe et incomprise

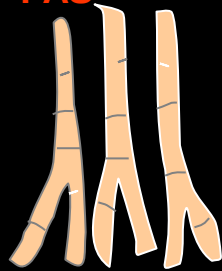


5-9 μm

HE +

G +++

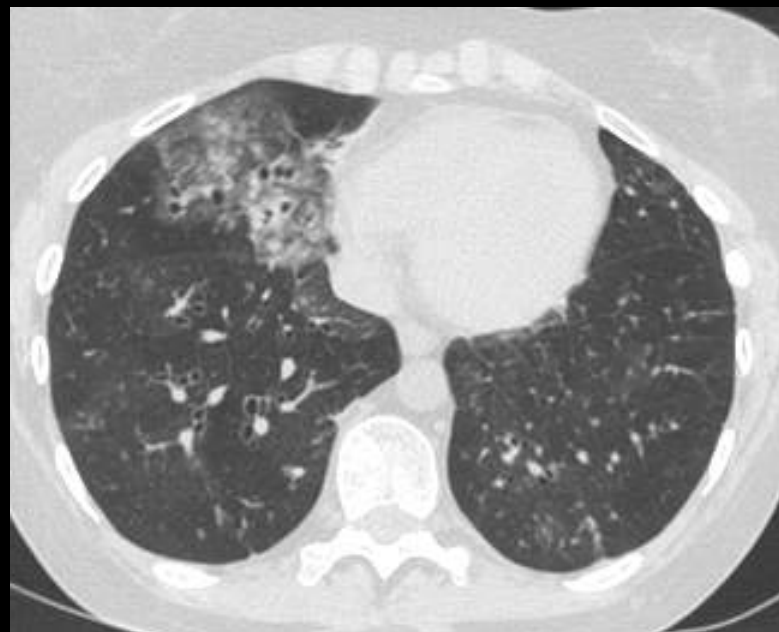
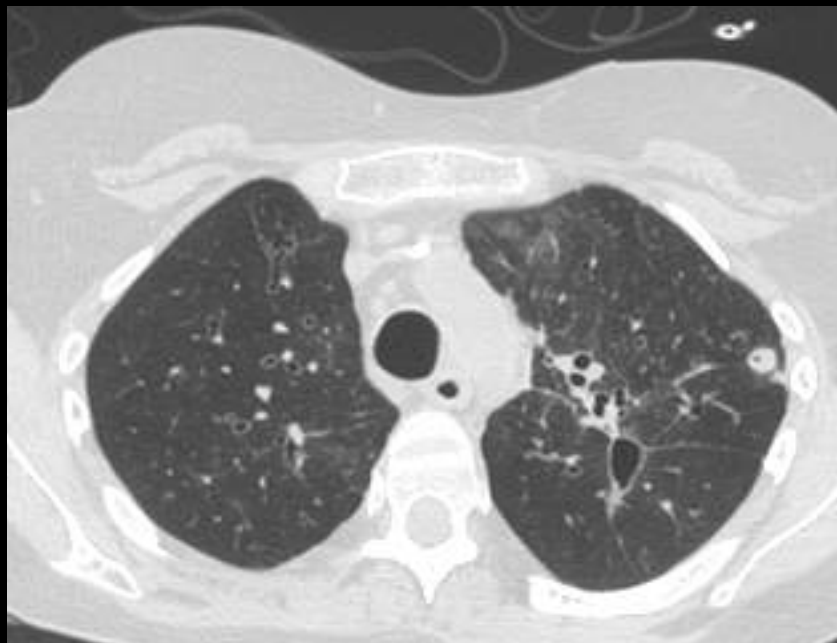
PAS +



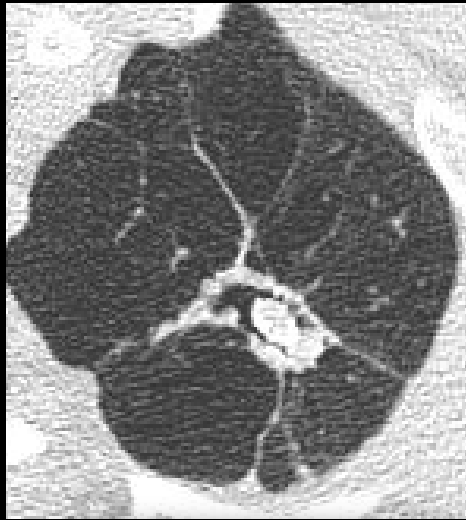
Aspergillus spp.

- **Presentations aiguës**, invasives : neutropénie
leucémie, chimiothérapie, transplantation, SIDA (rare)
→ angioinvasive, moins de réactions
inflammatoires

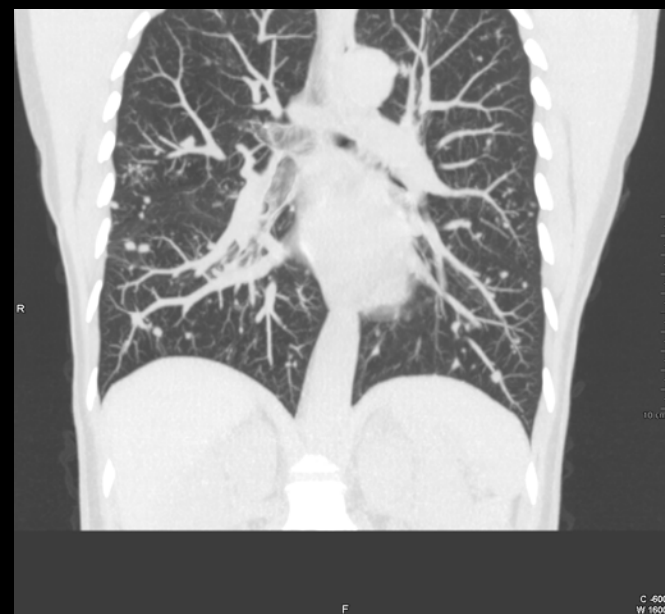
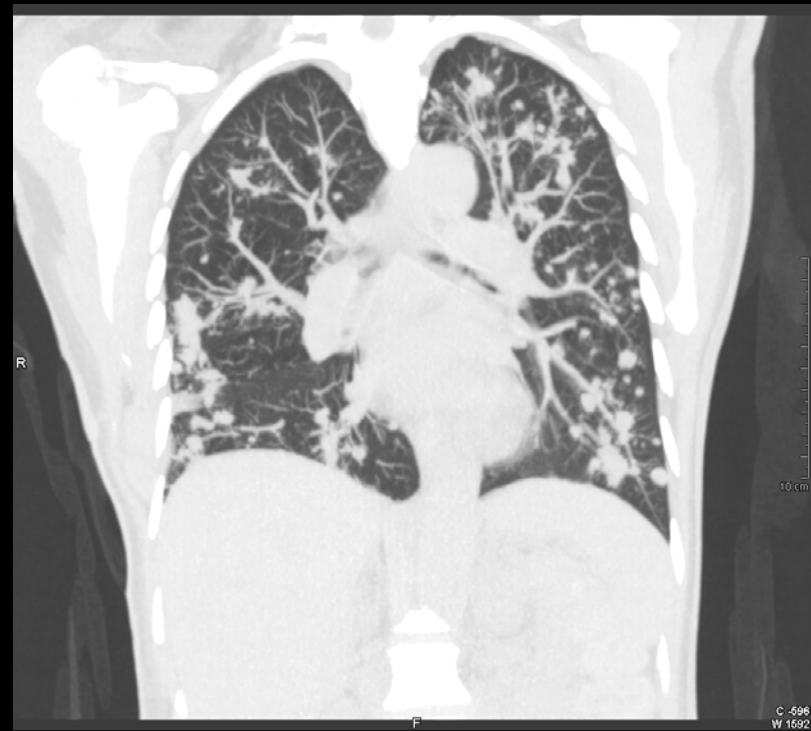
- Chronic presentations** : immunosuppression,
granulomatose, alcool, diabète, cortico-stéroïdes
- Inflammation granulomateuse (« fungus ball »)
 - Aspergillose invasive subaiguë (cavité à parois fines)
 - Aspergillose chronique cavitare
 - Aspergillose chronique fibrosante

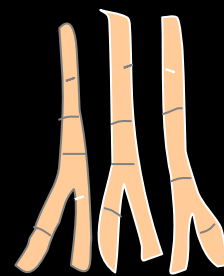
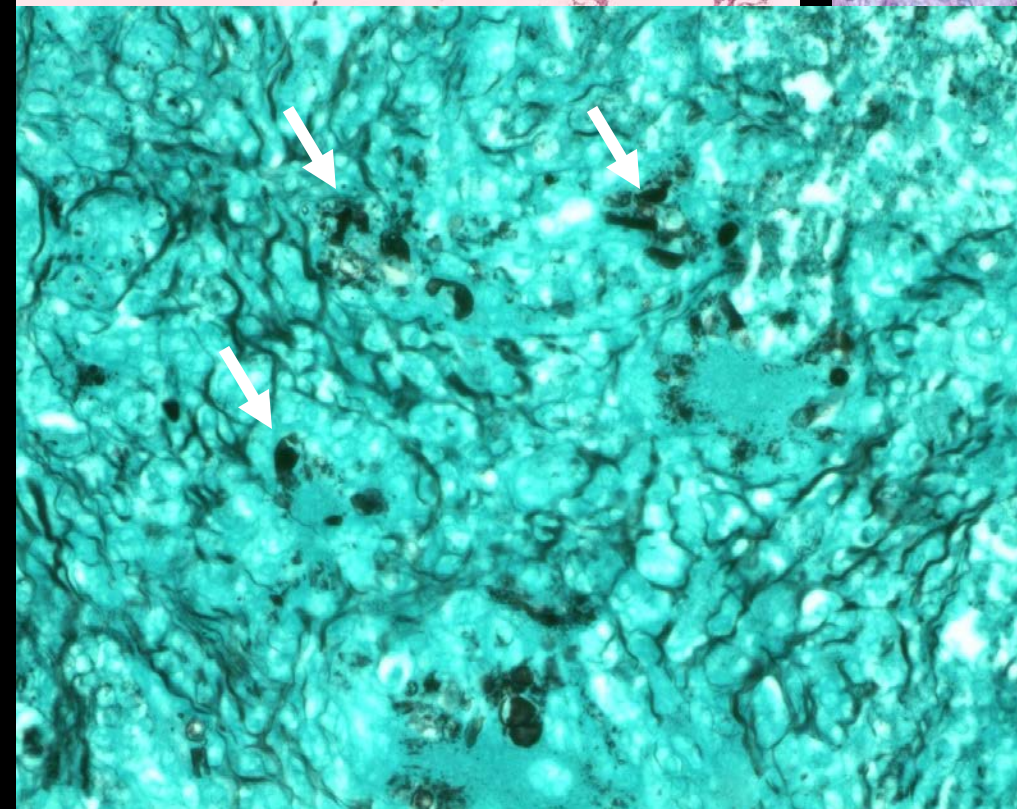
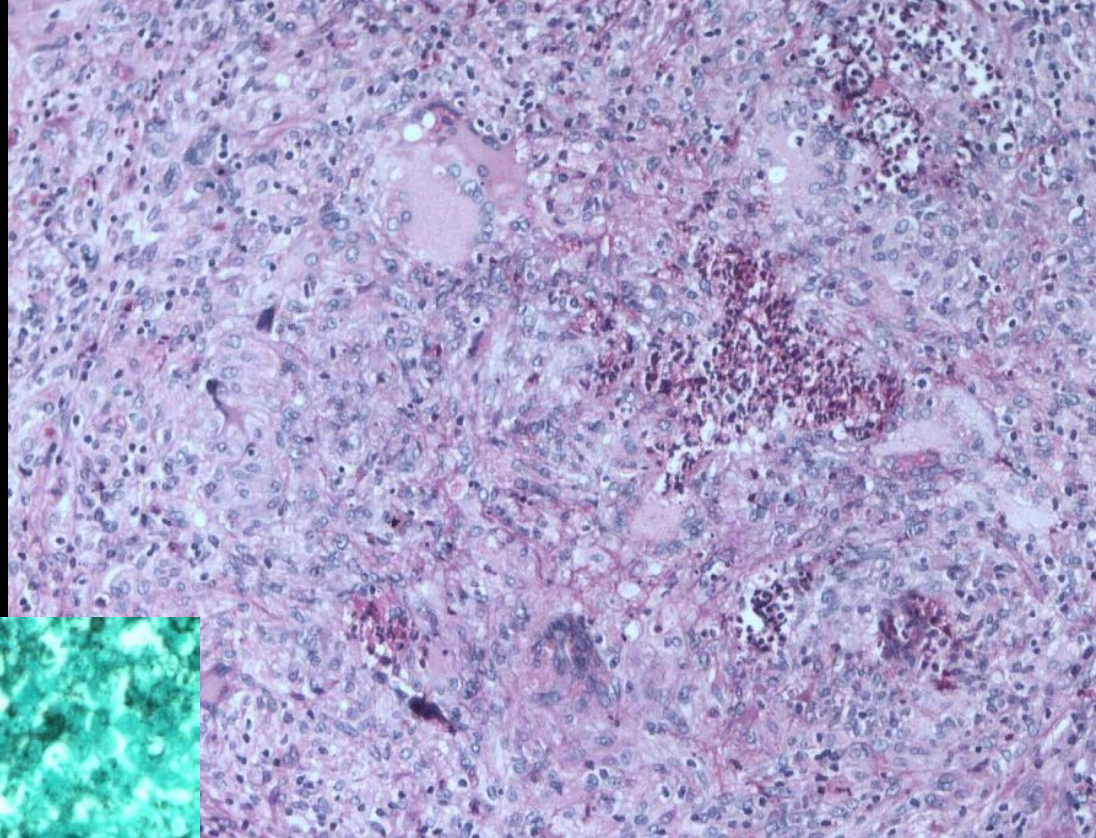
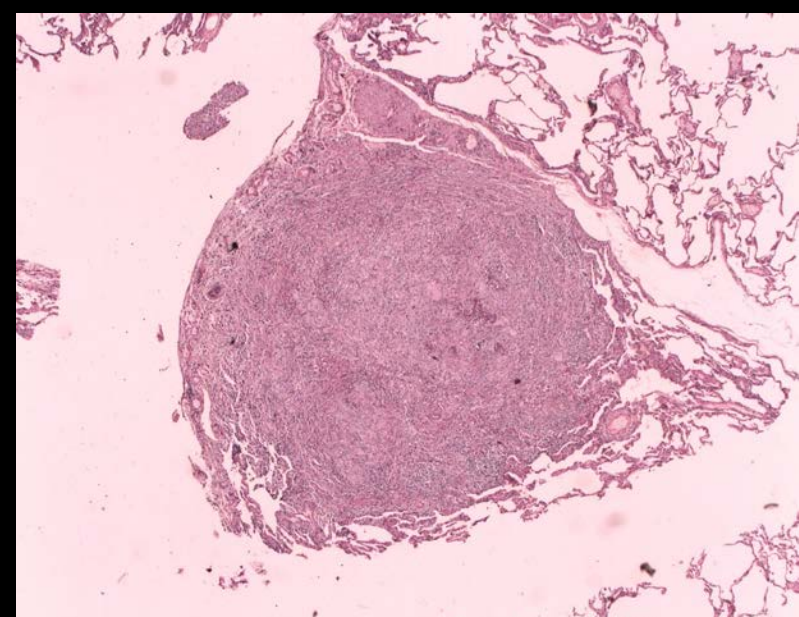


Conditions pré-existantes



- Colonisation d'une cavité ou poumons fragiles
 - Tuberculose
 - Sarcoidose
 - Bronchiectasies
 - Kyste Bronchogénique
 - Carcinome cavitare
 - Fibrose post RT (PR, SA)





Aspergillus spp.

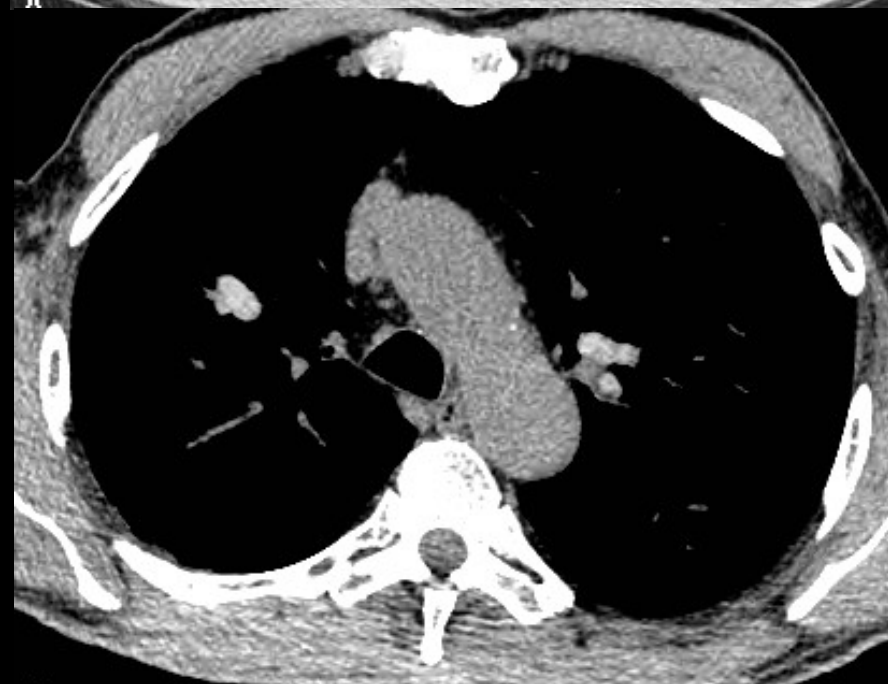
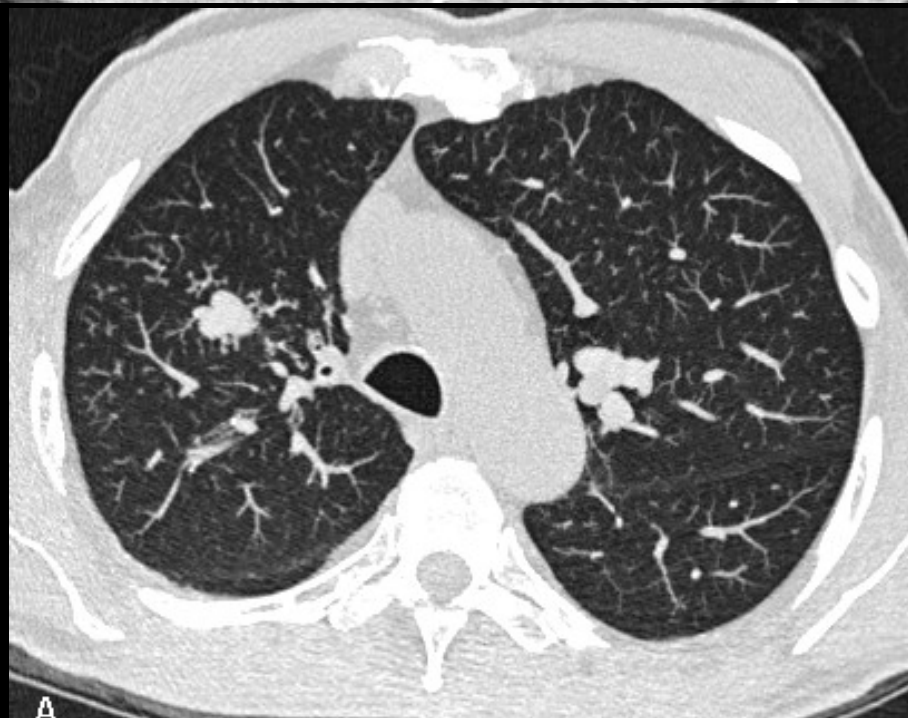


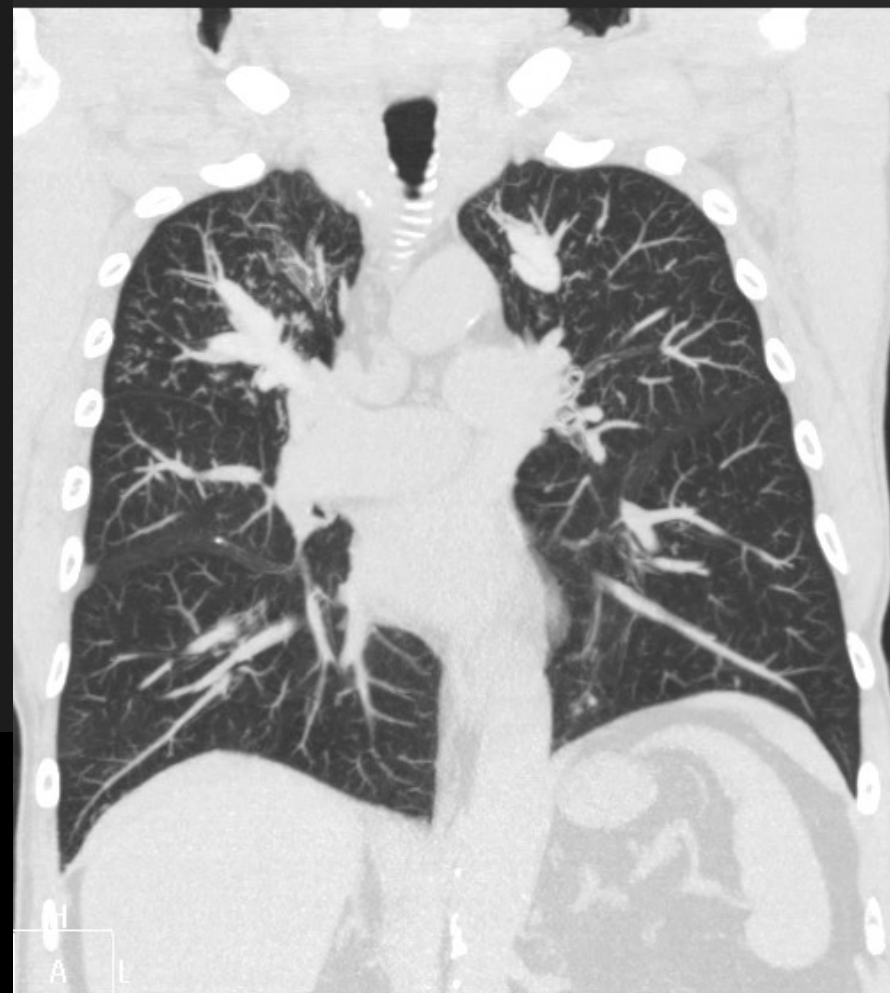
Aspergillose bronchopulmonaire Allergique (ABPA)

- Impactions mucoïdes
 - Image en « doigt de gant » homogène
 - Hyperdense
 - Lobes supérieurs de préférence
 - Bronches centrales et segmentaires > atélectasies périphériques

Présentation clinique

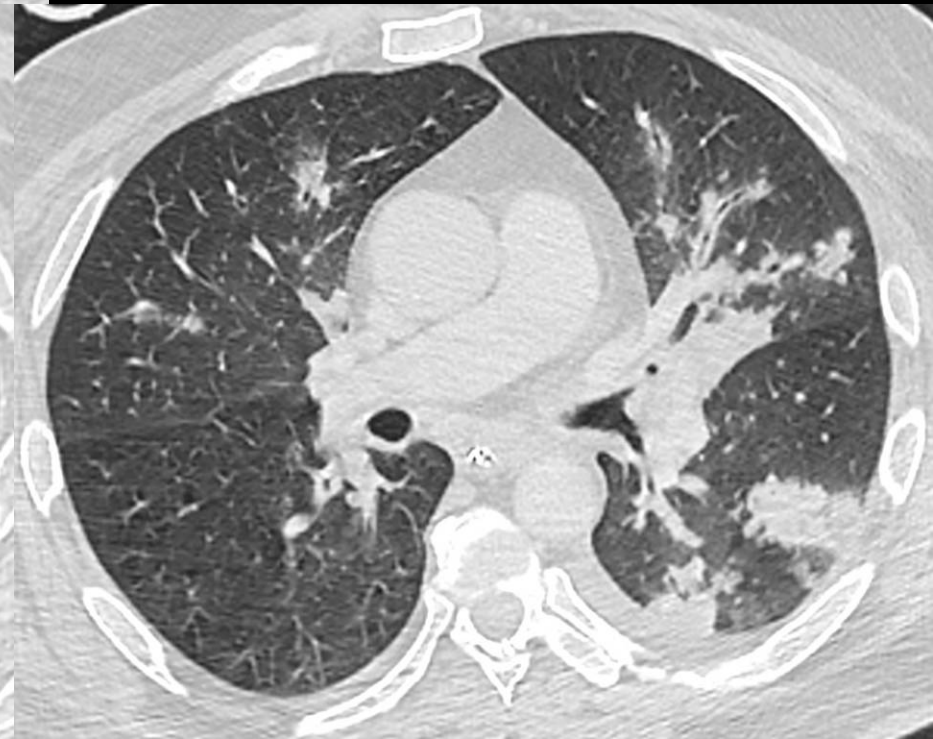
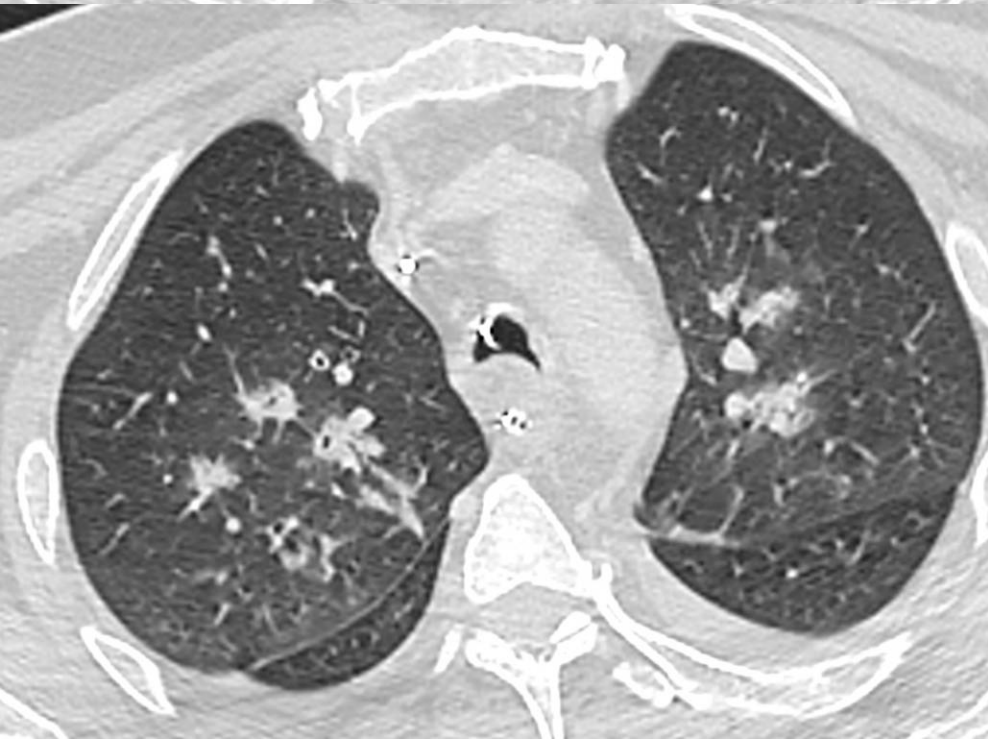
- Manifestations allergiques
 - Rhinite
 - Conjonctivite
 - Asthme , sinusite chronique
- Exacerbations aiguës
 - toux, hémoptysie, dyspnée, wheezing





Trachéobronchite aiguë

- Relativement rare
- Prédilection pour les patients avec transplantation cardiaque ou pulmonaire
- Histologie: ulcérations focales ou diffuses, pseudomembranes, impactions obstructives
- Rx/CT: atélectasie, épaissement des voies aériennes, nodules centrilobulaires, « ground glass » péribronchique, fistules



DR
RIGAUD

19/03/2013

12:11:00

CVP:A1/4

G:N E:A5

DR
RIGAUD

19/03/2013

12:12:25

CVP:A2/4

G:N E:A5

Invasive Aspergillosis

DR
RIGAUD

19/03/2013

12:12:56

CVP:A3/4

G:N E:A5

DR
RIGAUD

19/03/2013

12:20:50

CVP:A4/4

G:N E:A5

Histiocytose Langerhansienne

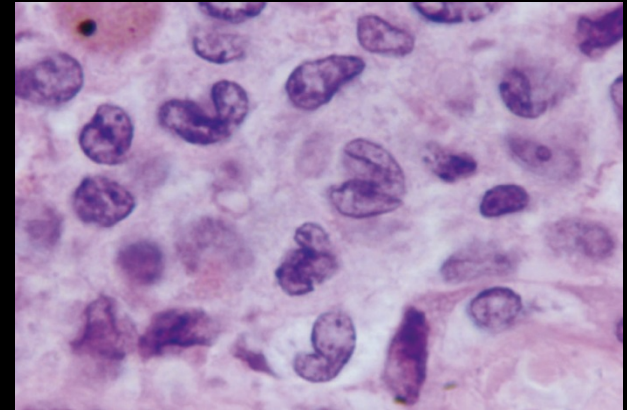
- Prolifération d'histiocytes CD1+ (cellules de Langerhans) formant des granulomes.



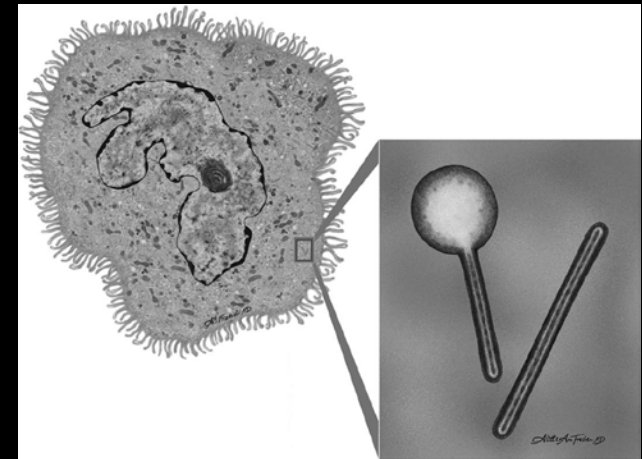
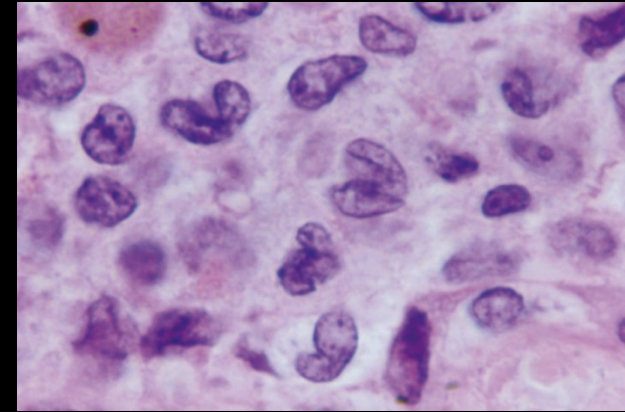
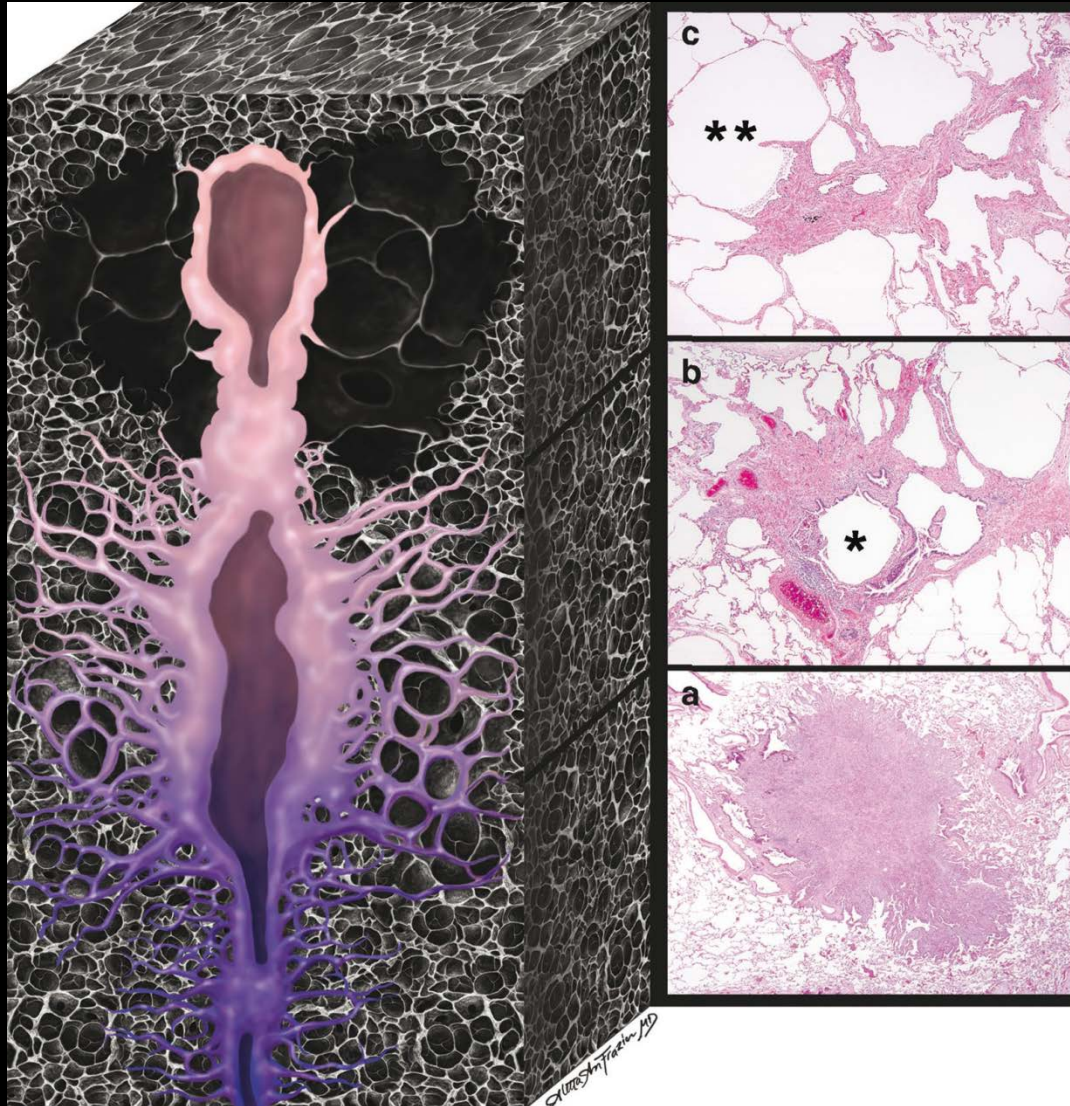
Bronchiolectasies/cavitation



Kystes

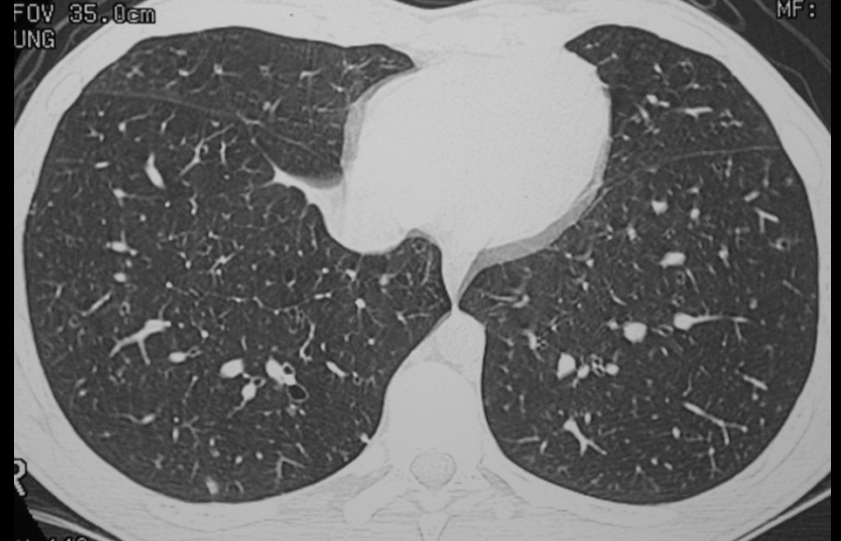
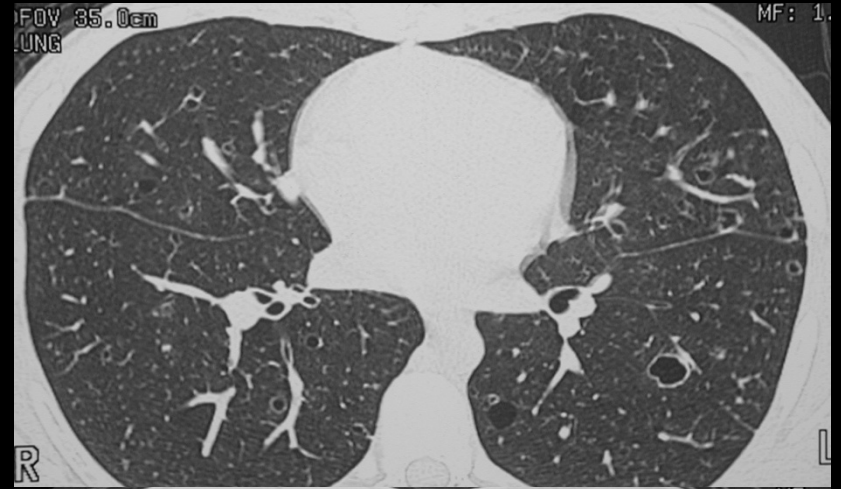


Histiocytose Langerhansienne

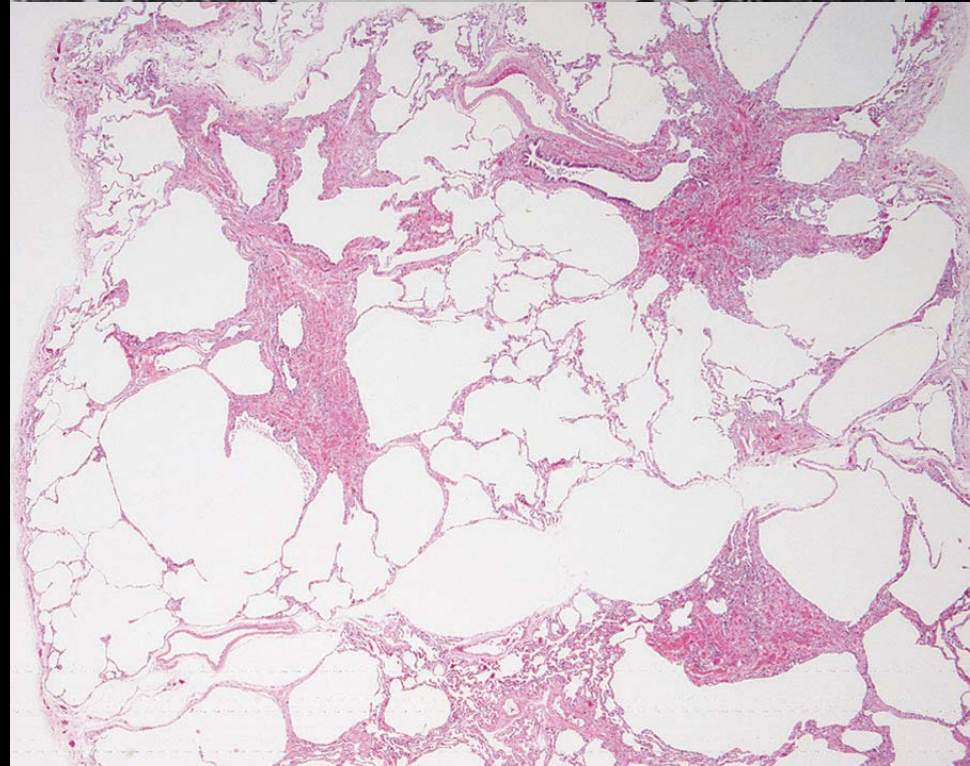
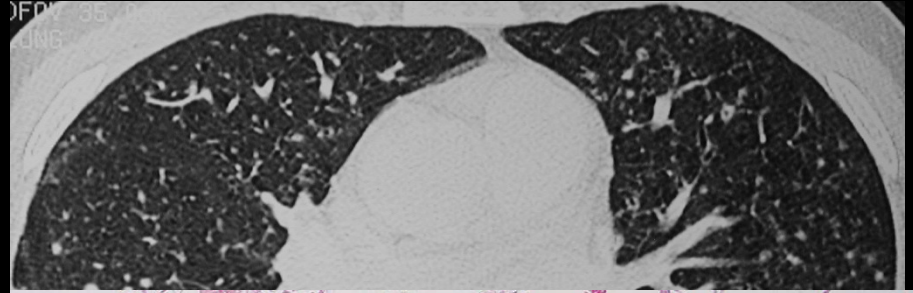


Abbot Radiographics 2004

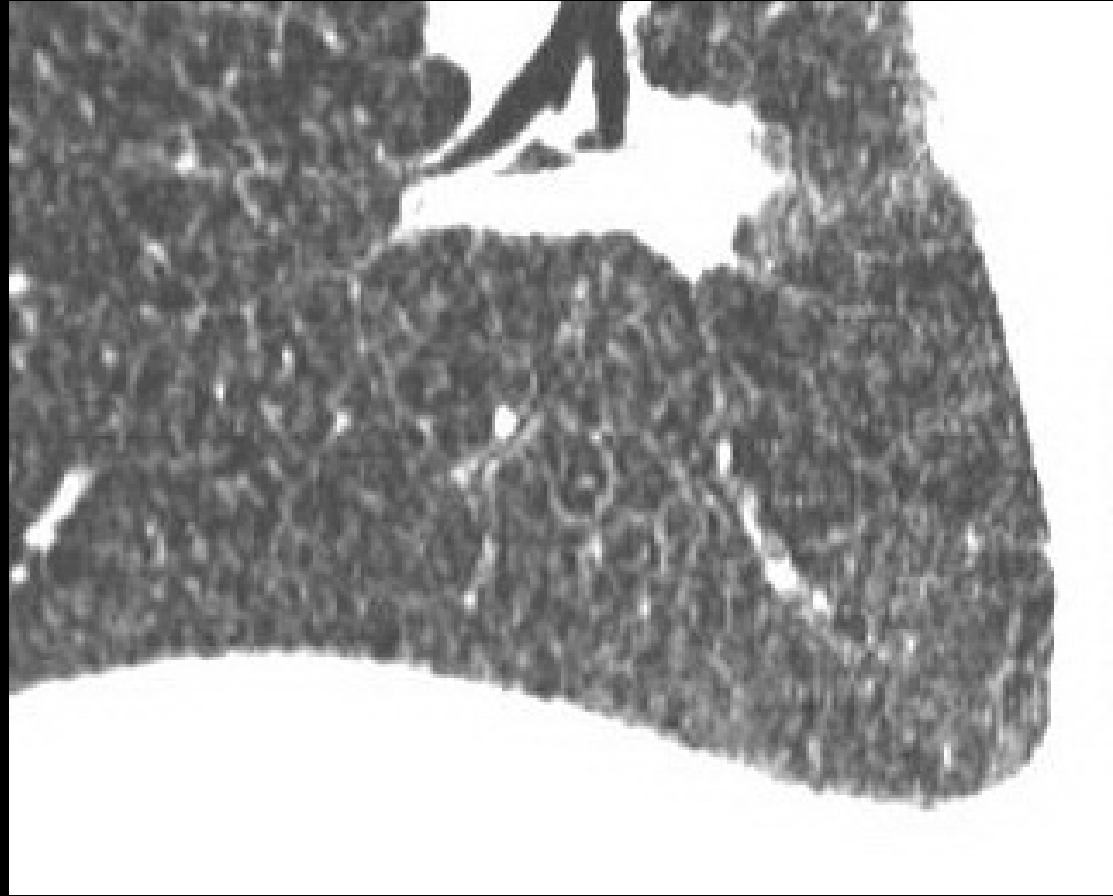
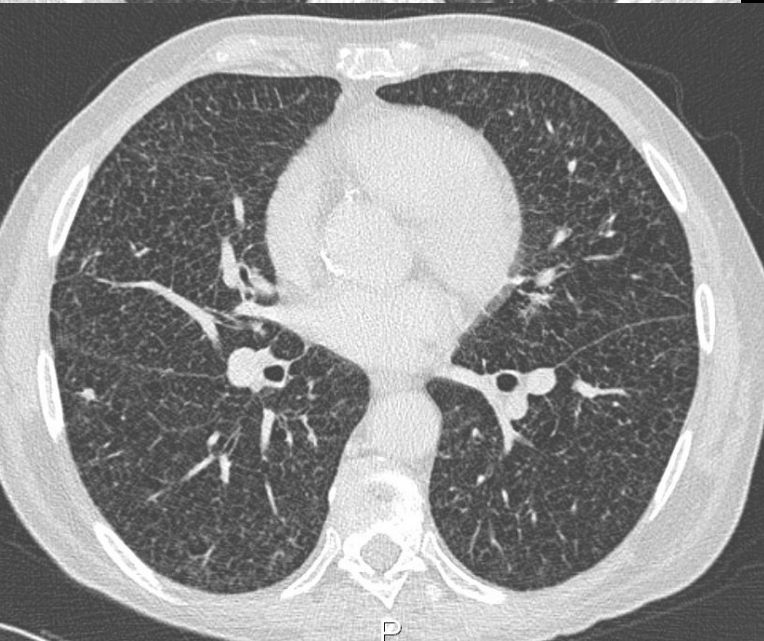
Histiocytose Langerhansienne



Histiocytose Langerhansienne



Histiocytose Langerhansienne



Histiocytose Langerhansienne



T0

ARRET
DU
TABAC



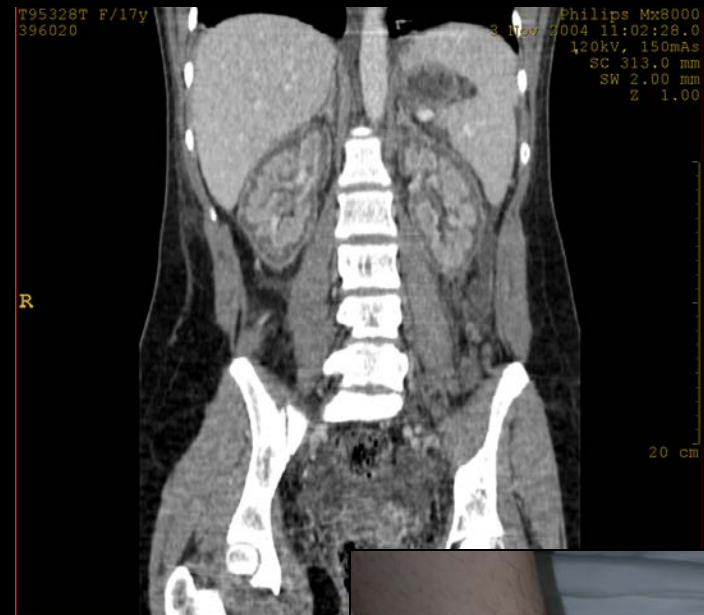
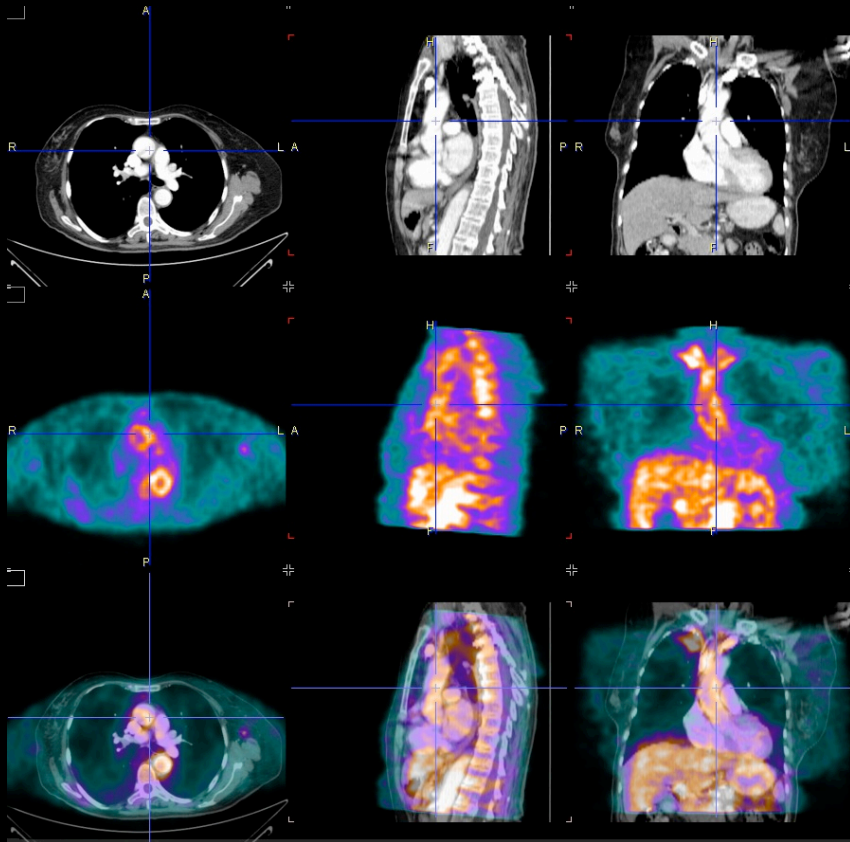
T1

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

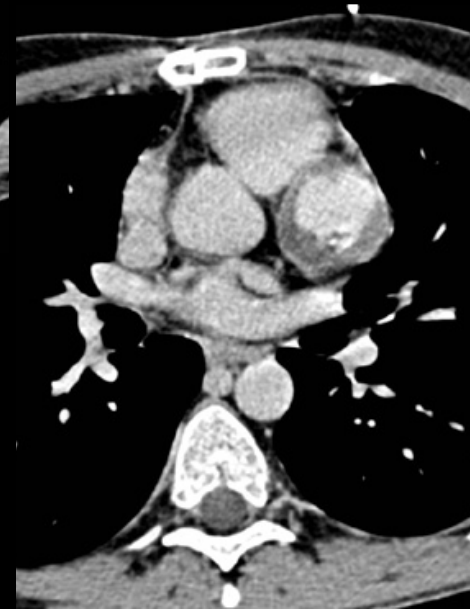
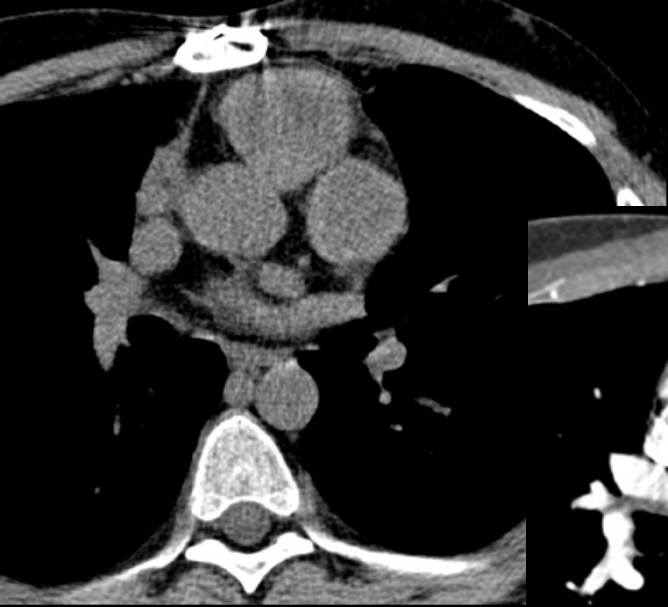
- Granulomatose de Wegener
- Maladie de Behçet
- Artérite de Takayasu
- Syndrome de Churg-Strauss
- Polyartérite noueuse
- Artérite à cellules géantes
- Divers

Maladie de Behçet

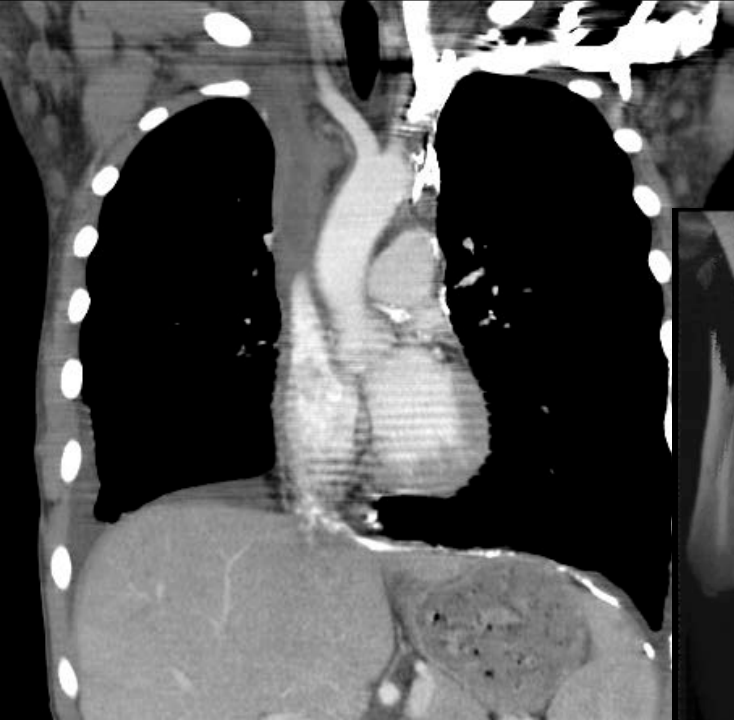
- Maladie systémique rare: 80-370/100 10³ en Turquie
 - stomatite aptheuse récurrente
 - Ulcères génitaux
 - Lésions dermatologiques et uvéite
- Atteinte pulmonaire rare
 - Anérysme artère pulmonaire : simple ou multiples
 - Thrombose ou occlusion de la VCS, A. pulmonaires
 - Infarctus pulmonaire, hémorragie,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

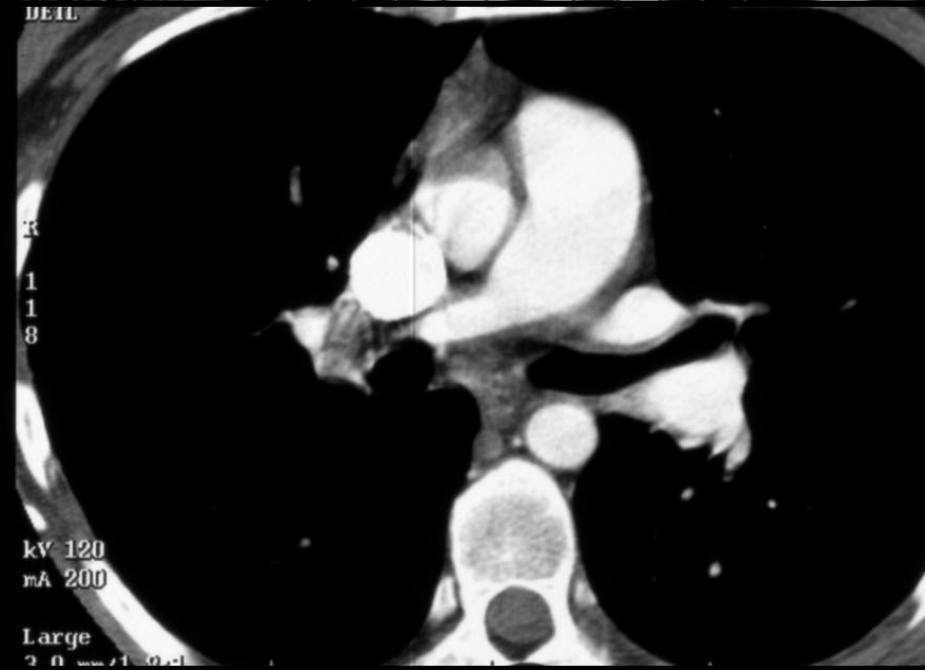
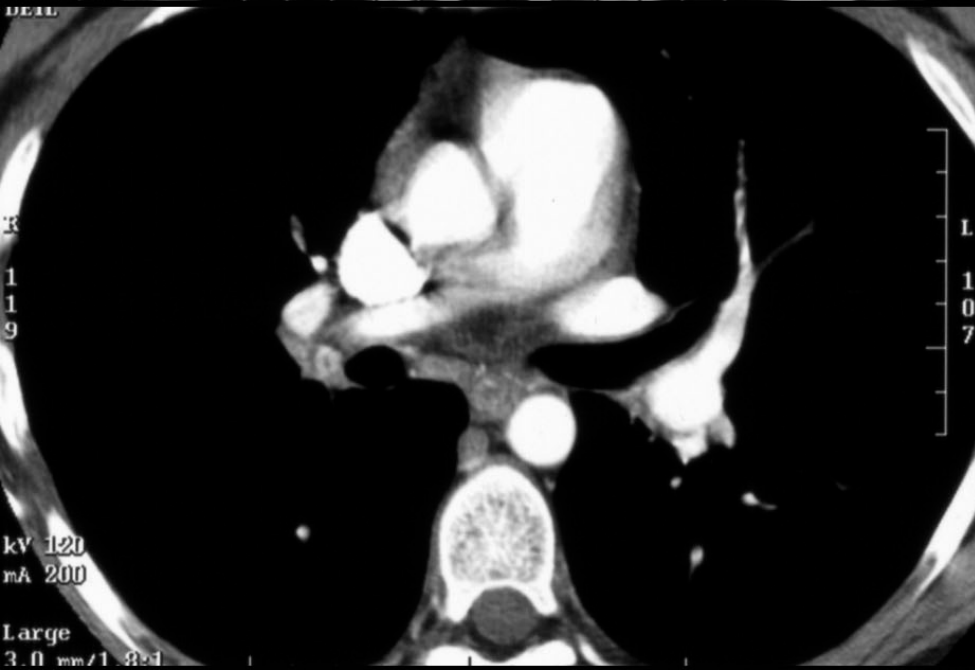
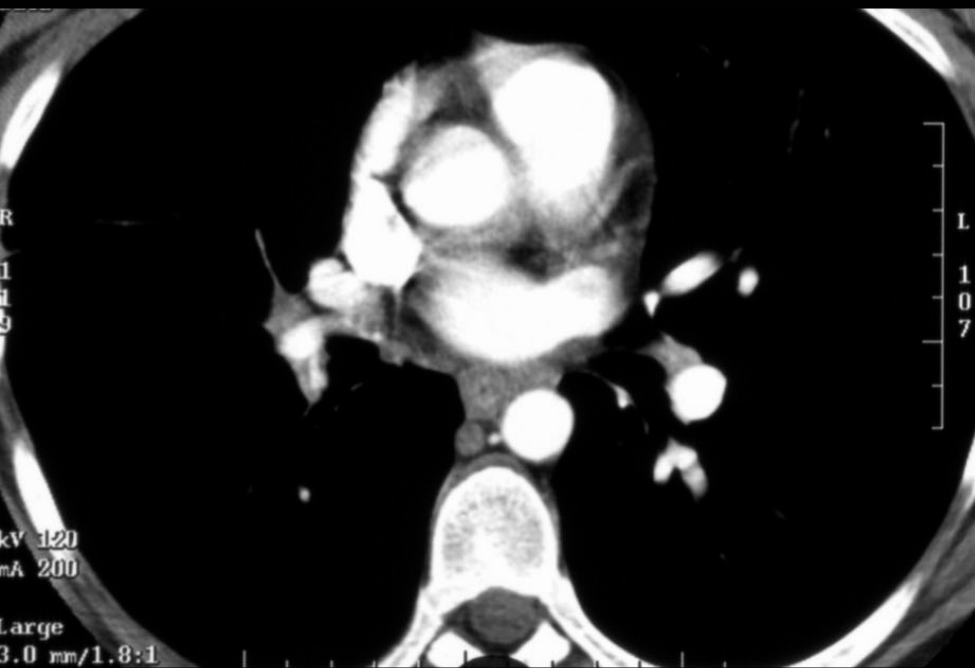
Syndrome veine cave supérieur

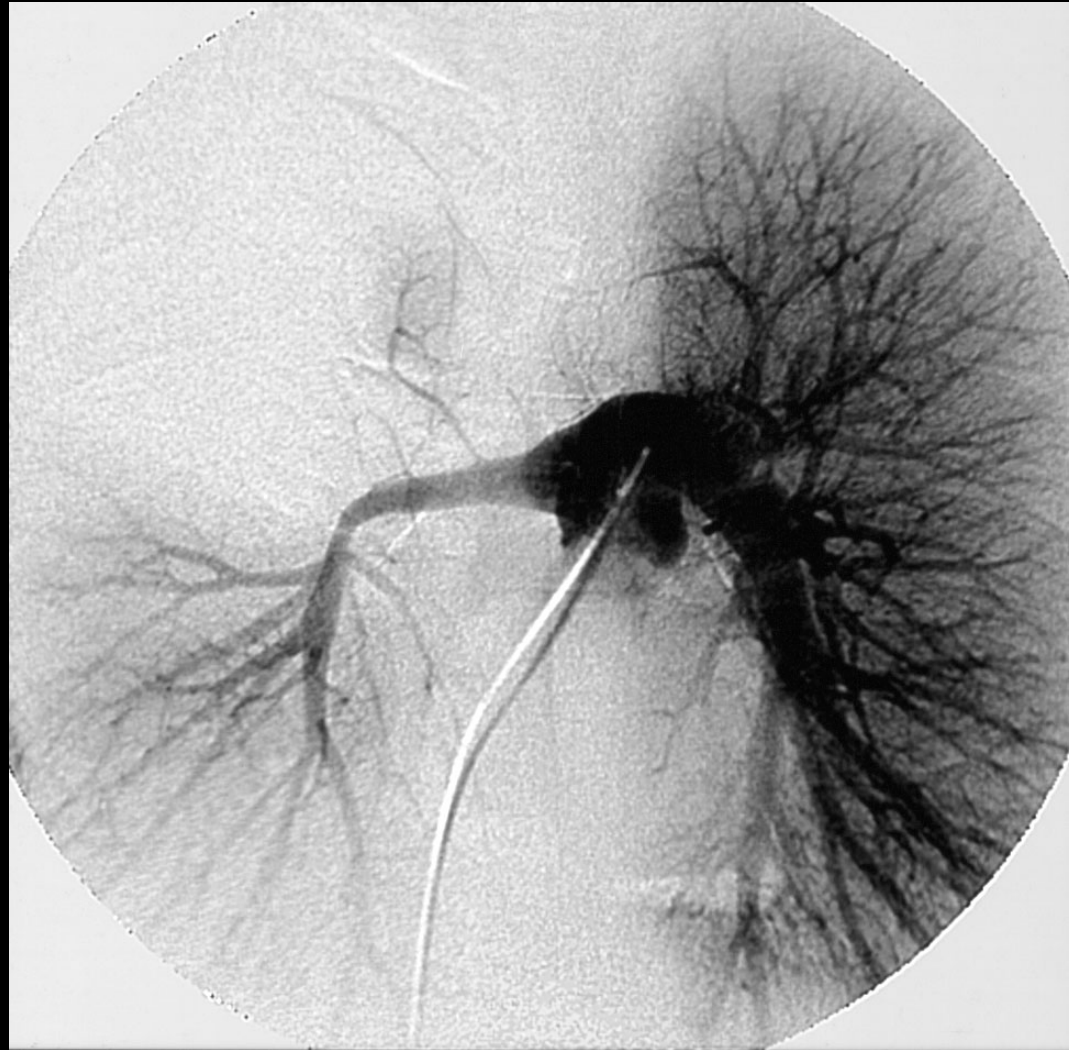
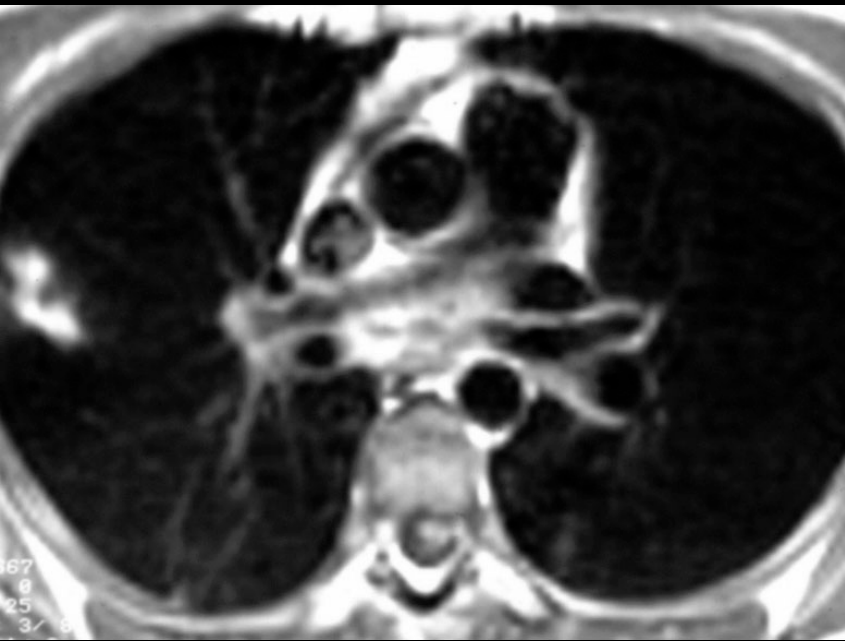
CT multicoupe



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

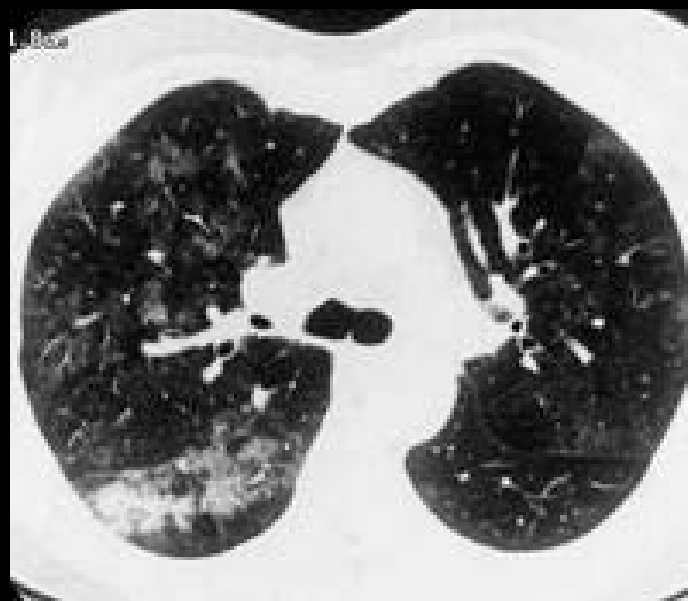




Ferretti G, et al Initial isolated Takayasu's arteritis of the right pulmonary artery: MR appearance. Eur Radiol. 1996;6:429-32

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

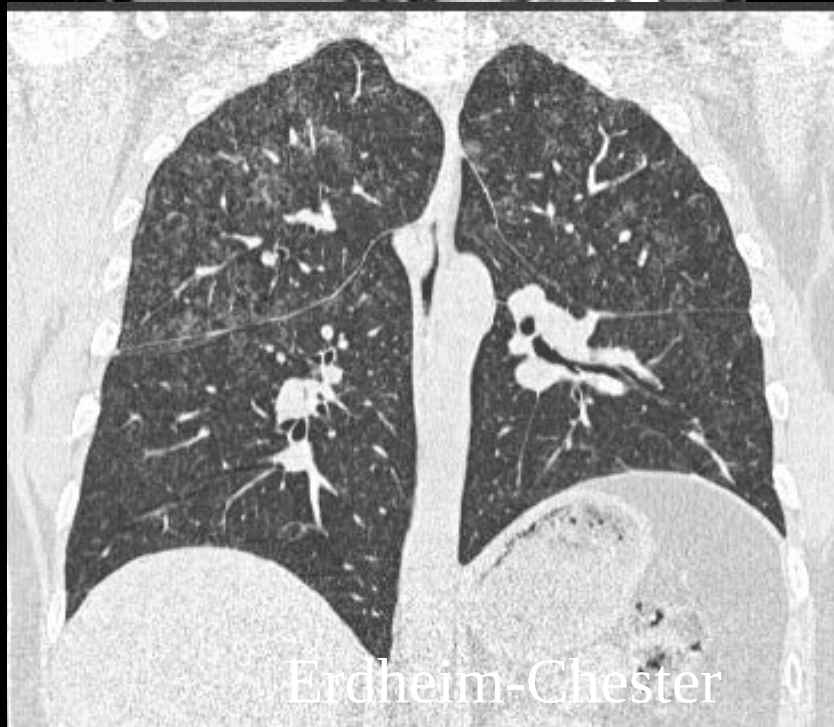
- Periartérite Nodulaire:
Vascularite nécrosante / artères musculaires de petite à moyenne taille
Vascularite de la circulation pulmonaire est rare
- Maladie d'Erdheim-Chester :
Épaississement lisse des septae interlobulaires ,
nodules centri-lobulaires, épaississement
scissuraux, épanchements pleuraux, aortite,
masses extra-thoraciques
- Artérite à cellules géantes:
Aortite (20-40%), atteinte coronaire, péricardite



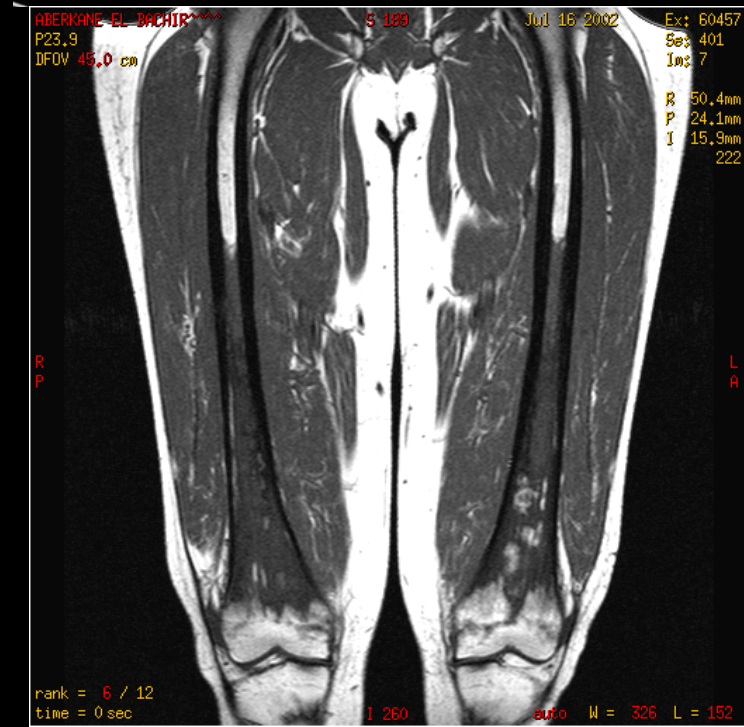
2



Aorte infiltrée



Erdheim-Chester



ABERKANE EL BACHIR S 189 Jul 15 2002 Ex: 60457
P23.9 Seq: 401
DFOV 45.0 cm Im: 7
R 50.4mm
P 24.1mm
I 15.9mm
222
R P L A
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

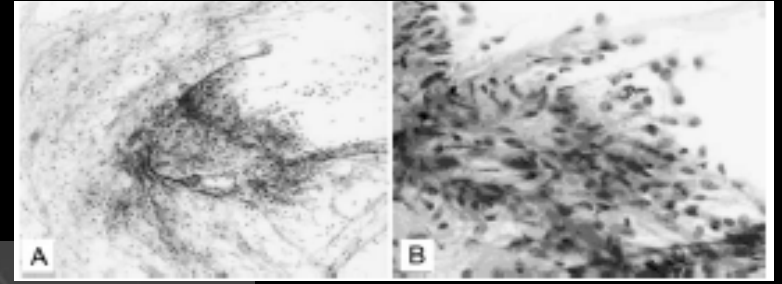
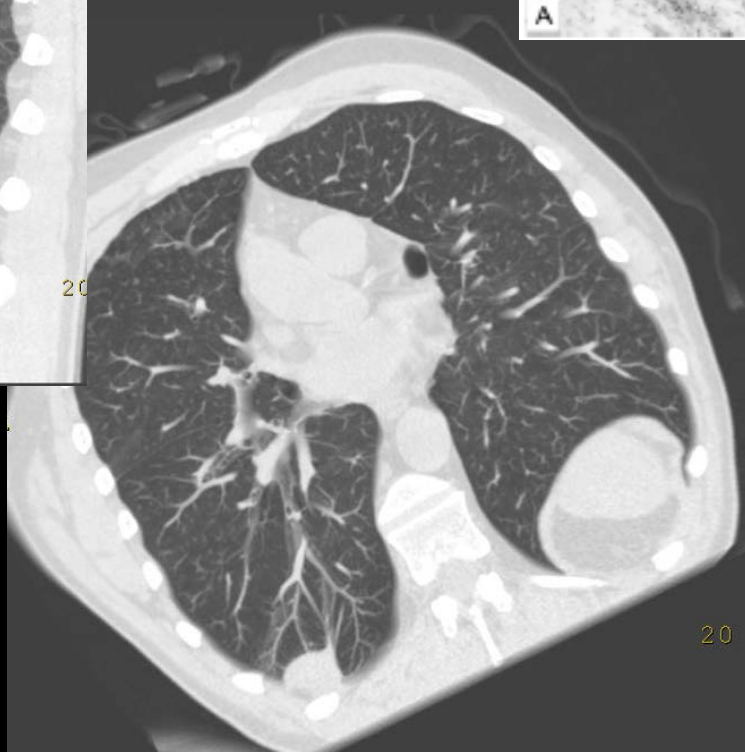
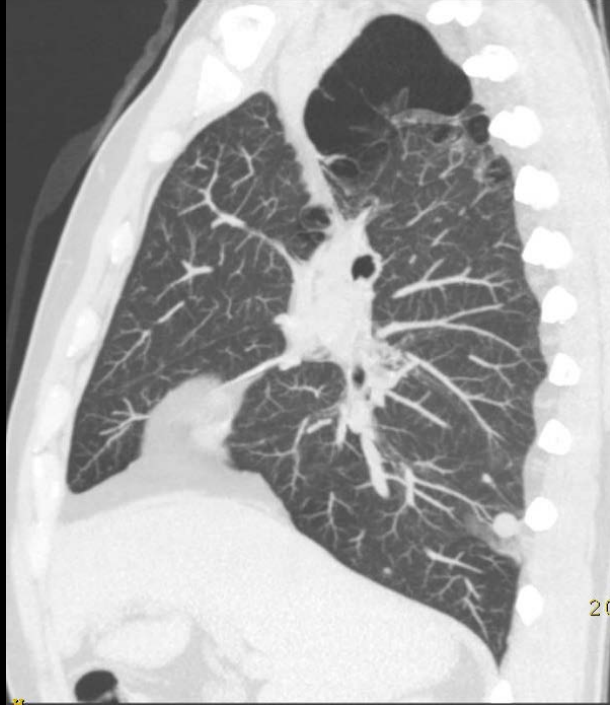
Classification

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

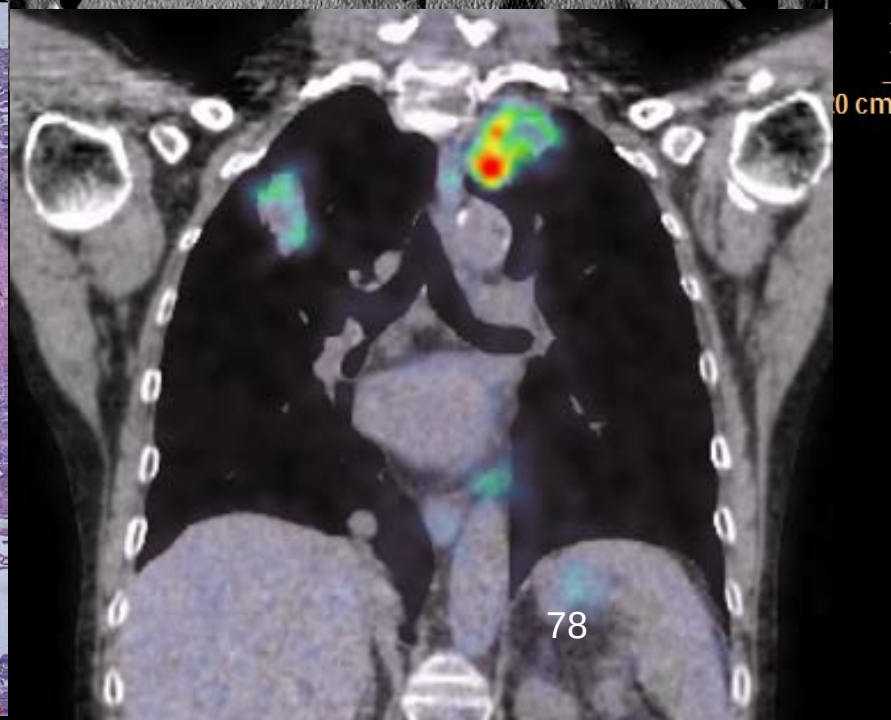
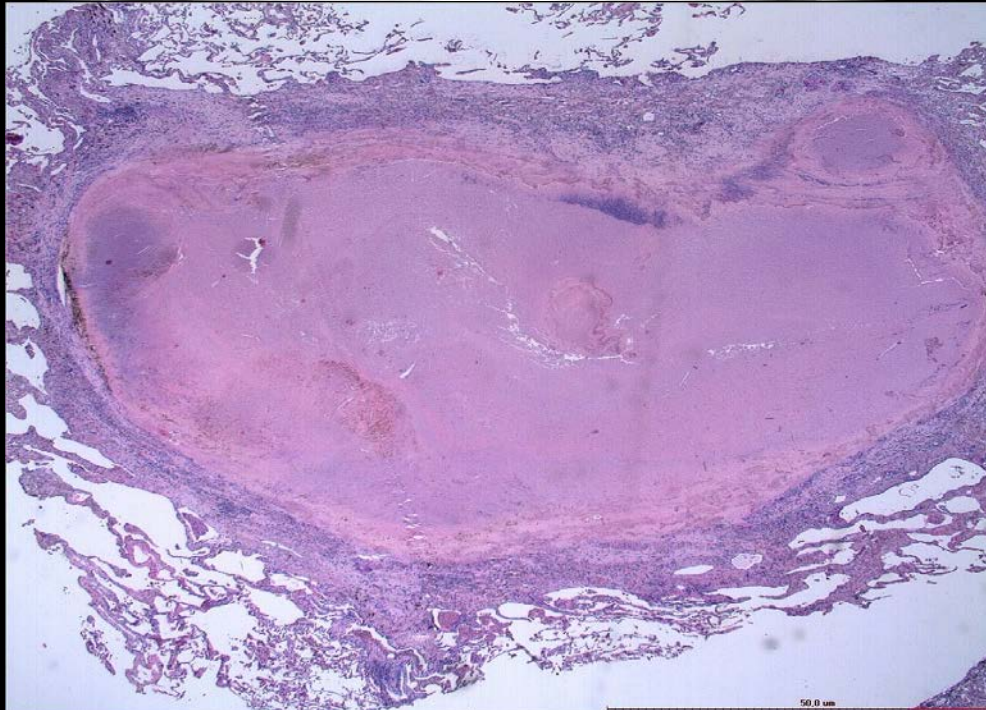
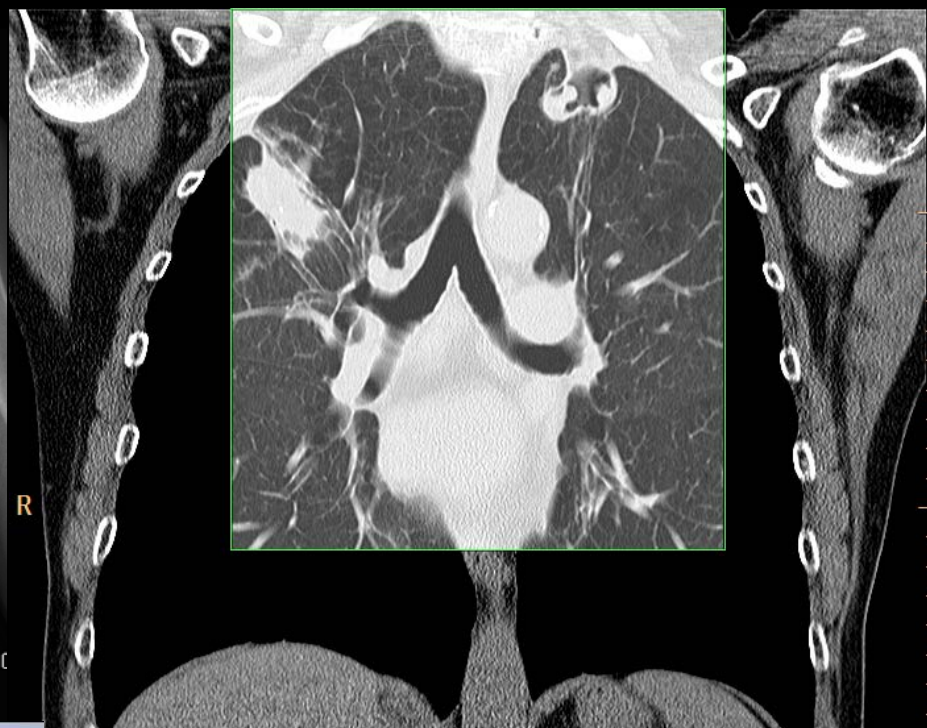
Maladie rhumatoïde

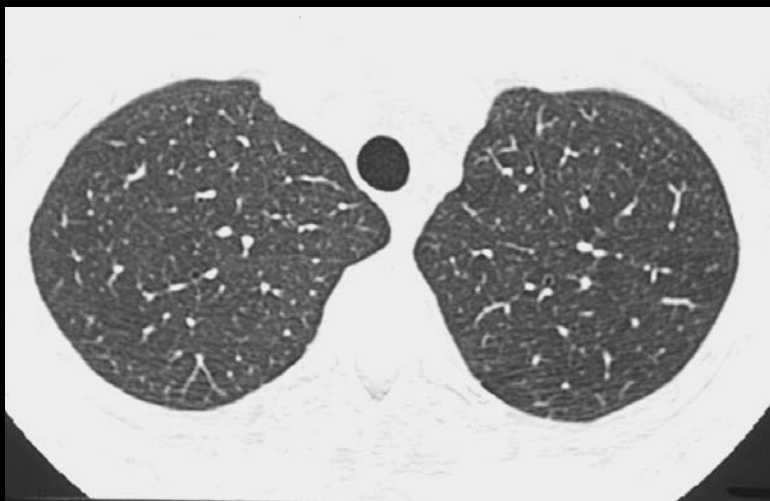
- Maladie de système fréquente (0.8% de la population)
- Manifestations pleuro-pulmonaires fréquentes
 - Épanchements pleuraux
 - Nodules rhumatoïdes
 - Pneumonie interstitielle : UIP, NSIP, BOOP
 - Vascularite pulmonaire
 - Atteinte des voies aériennes: bronchiectasies, Bronchiolite oblitérante, bronchiolite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





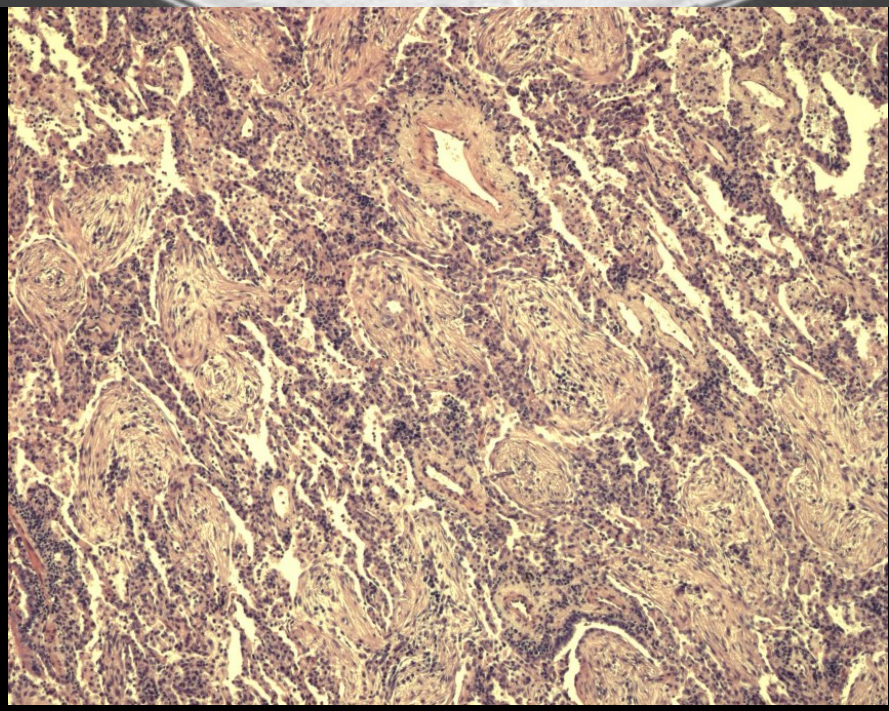
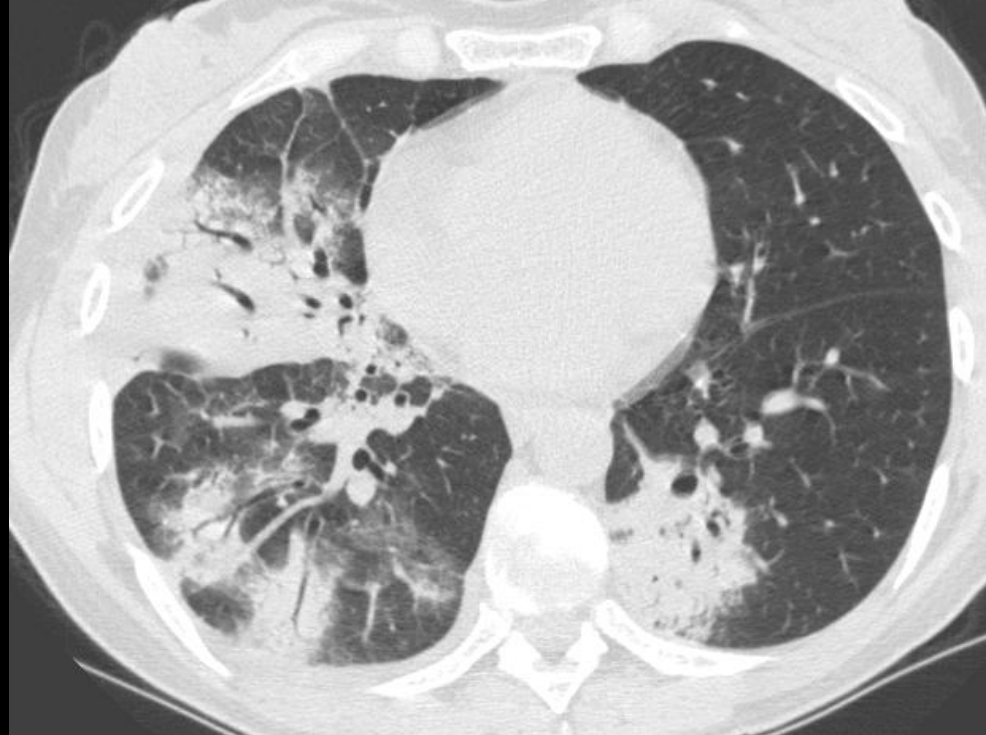
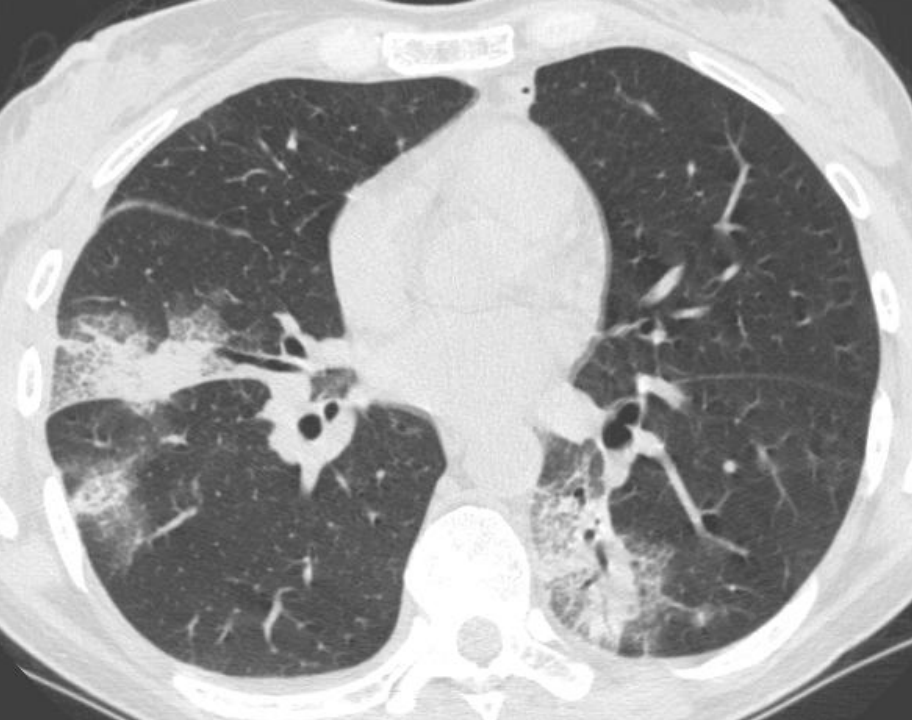
Nodules rhumatoïdes sont identifiés sur 1% des radiographies thoraciques
Qqs mm à qqs cms de diamètre, simple ou multiple, cavitation centrale (50%)



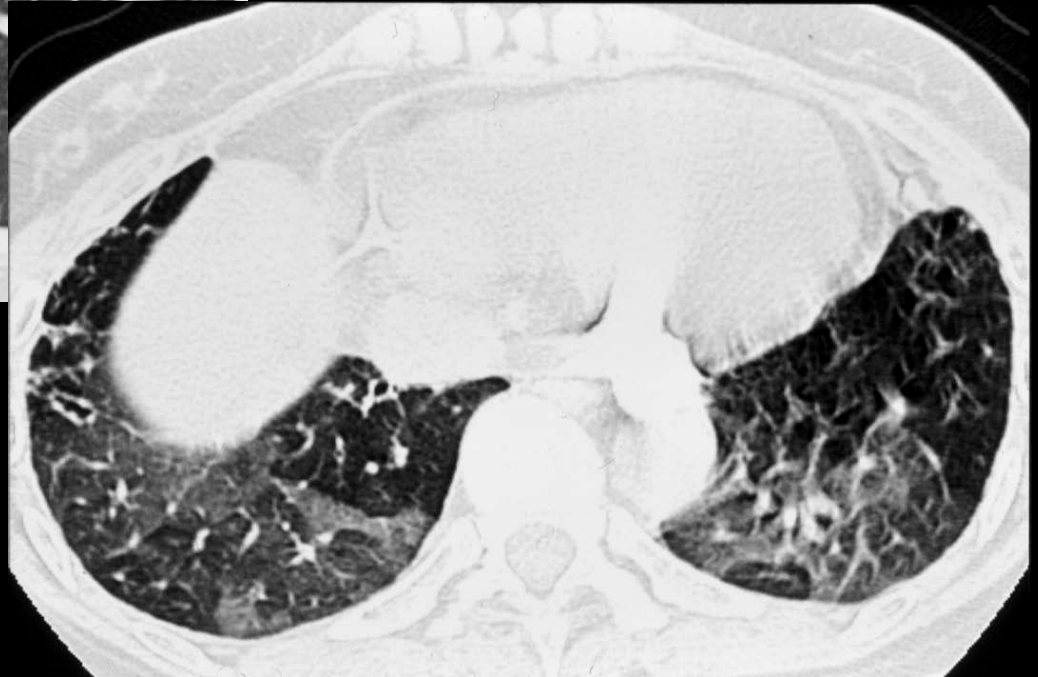


Lupus érythémateux disséminé

- 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, BOOP
 - 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.

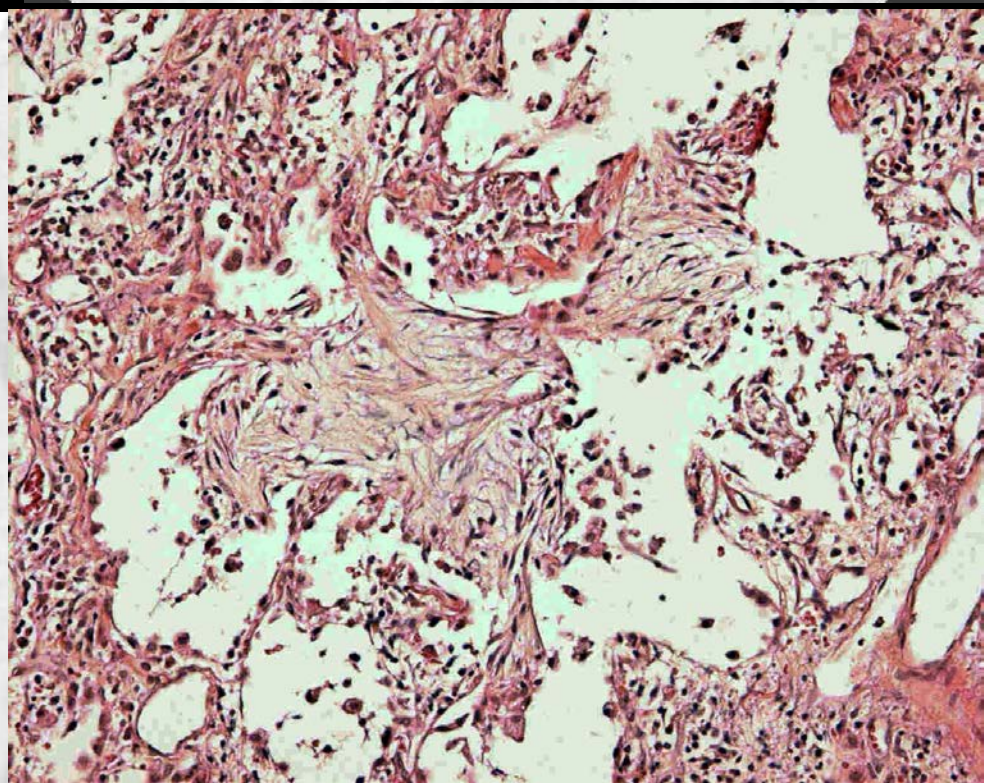
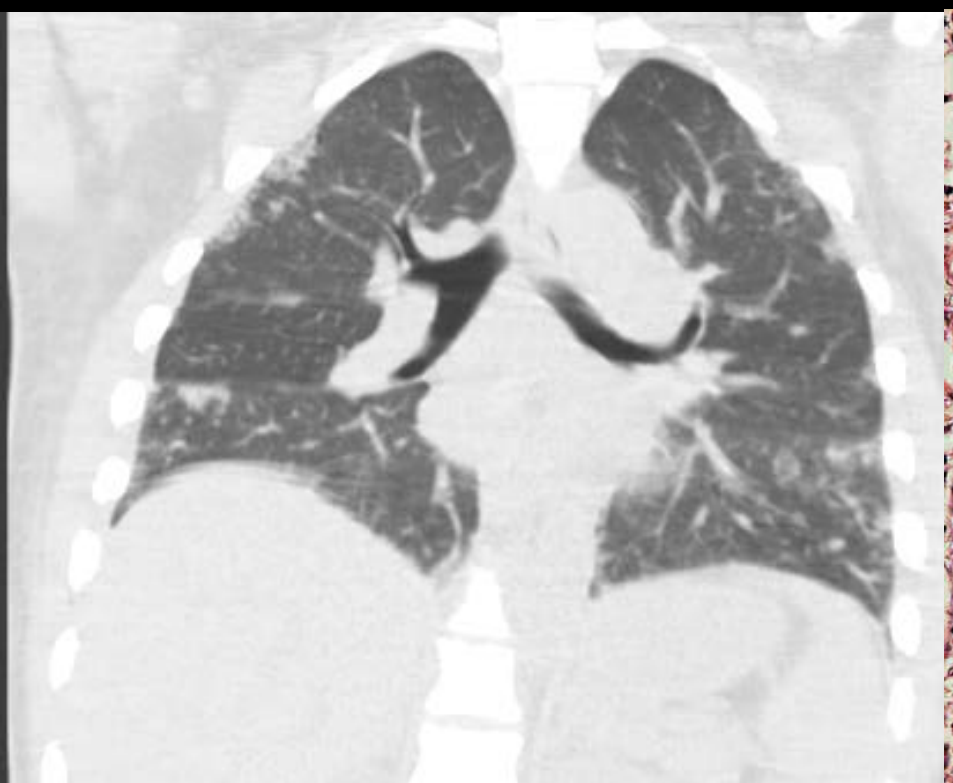
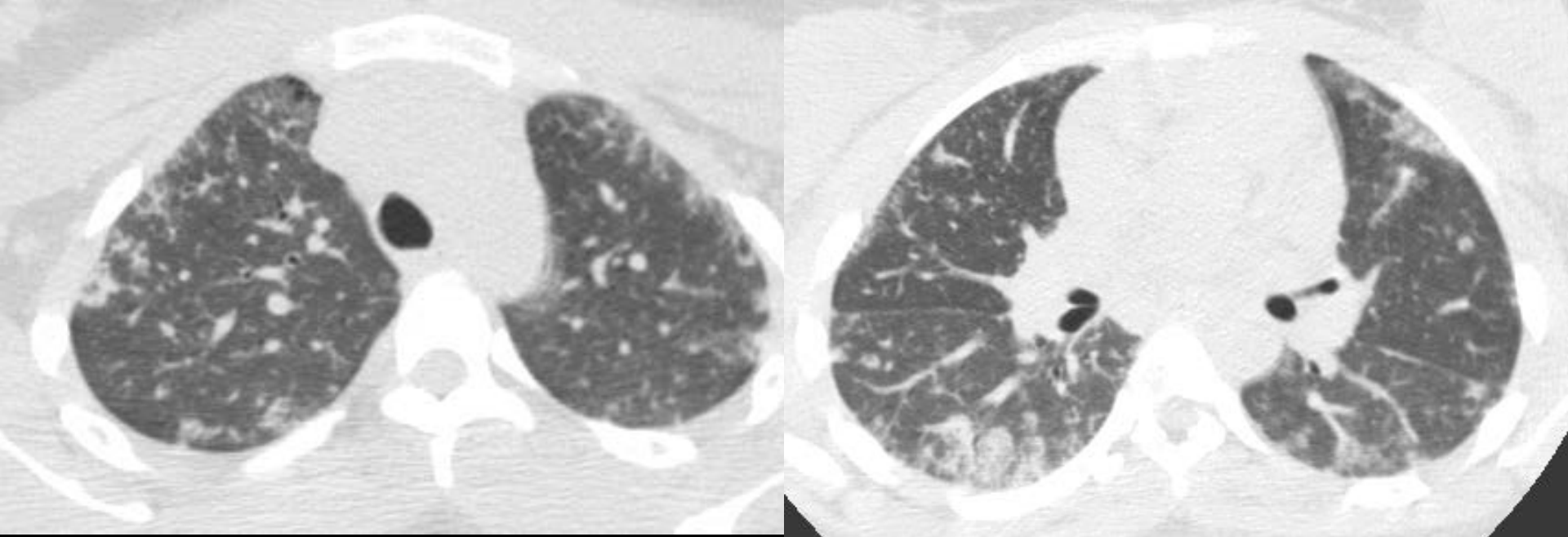
AJR Am J Roentgenol. 1993;161(2):269-72.

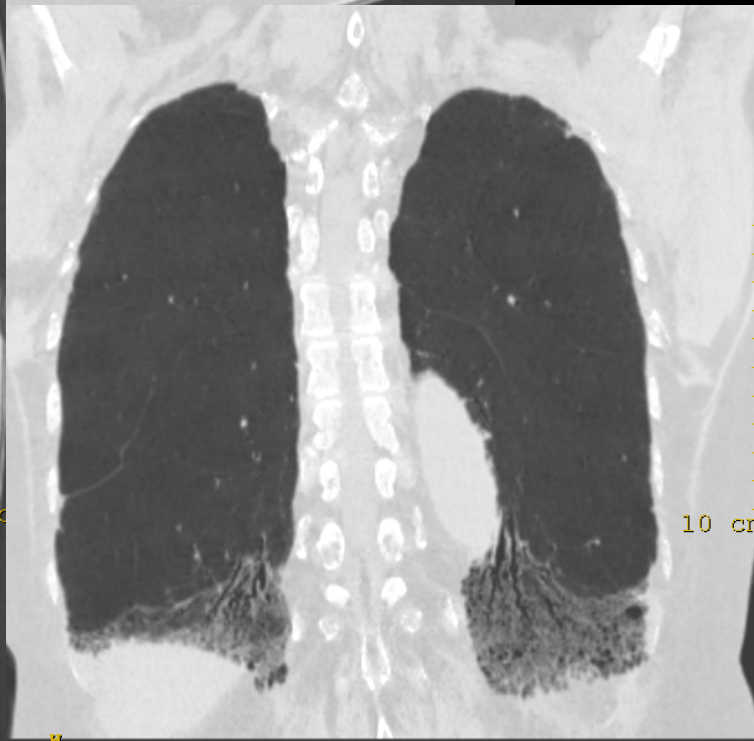
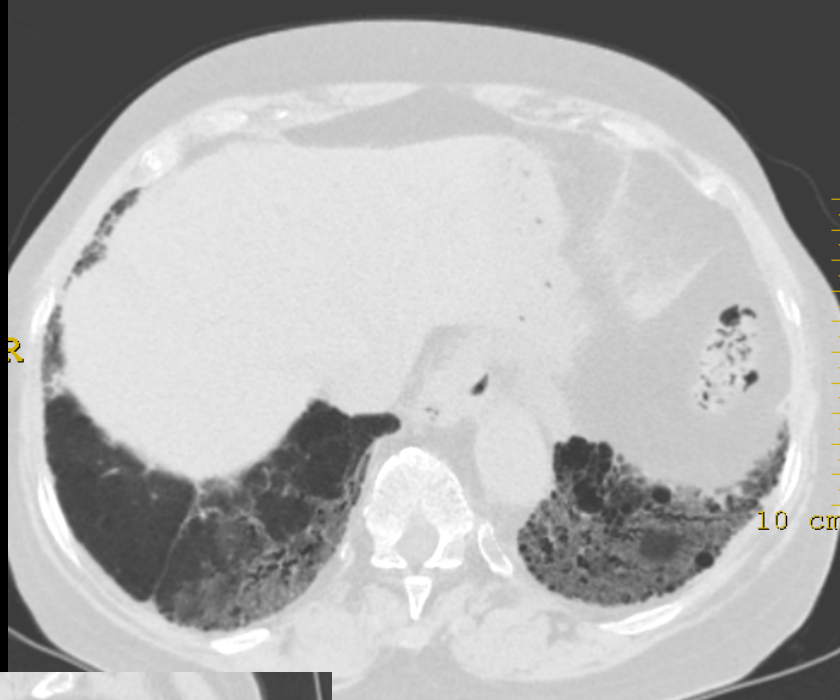


Dermatomyosite et polymyosite

- 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

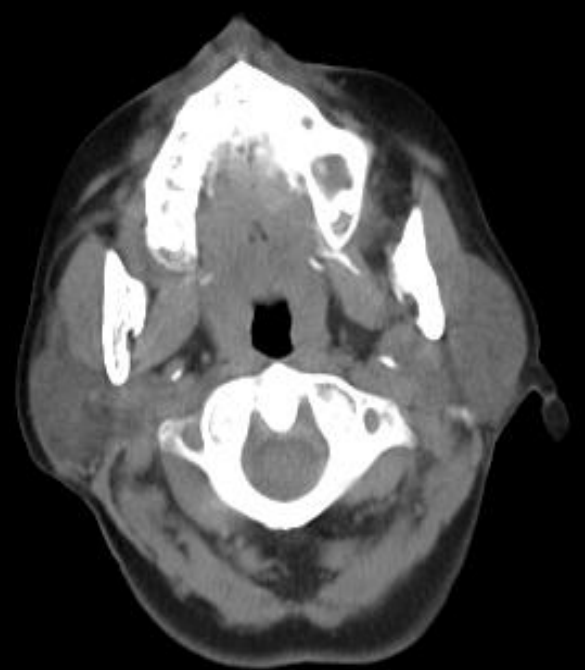
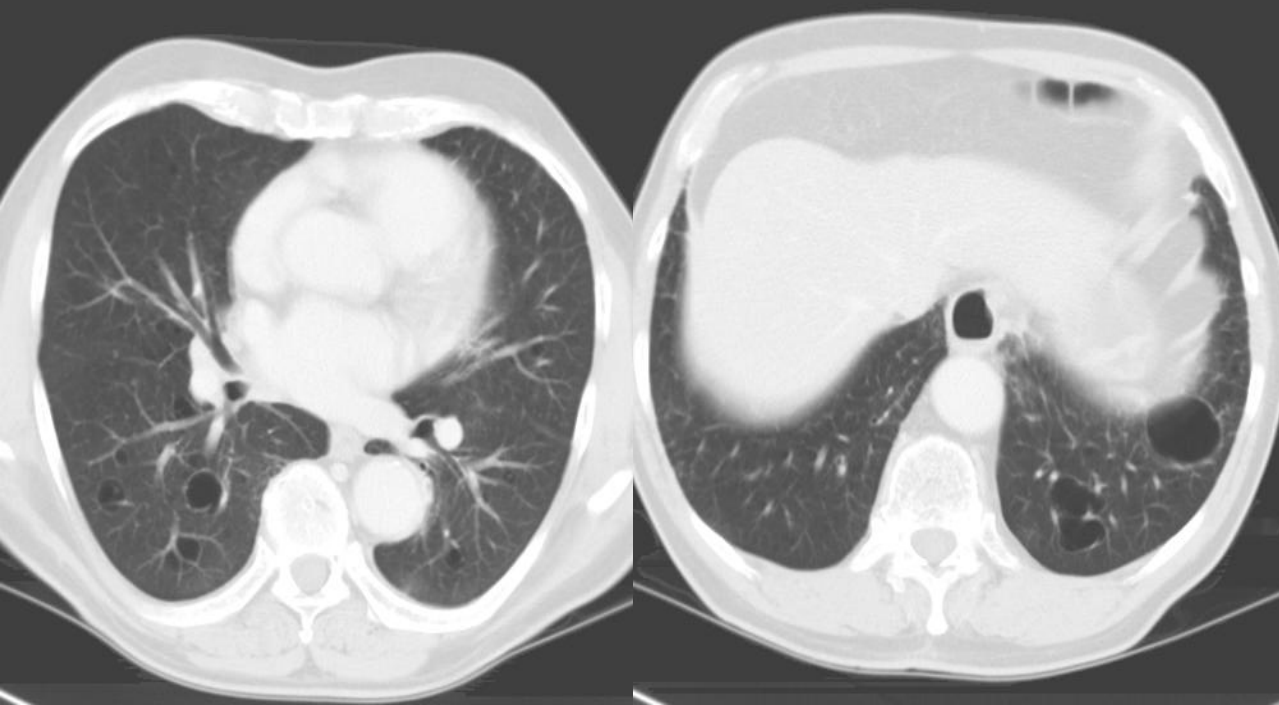
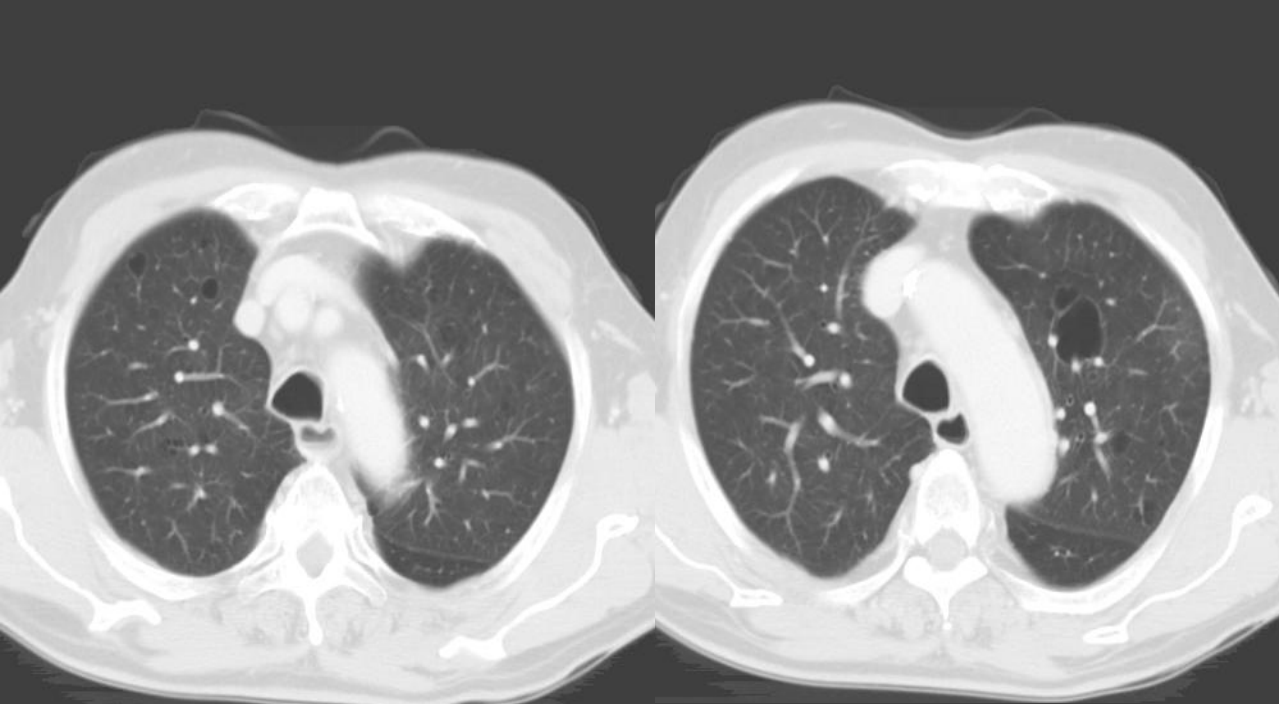


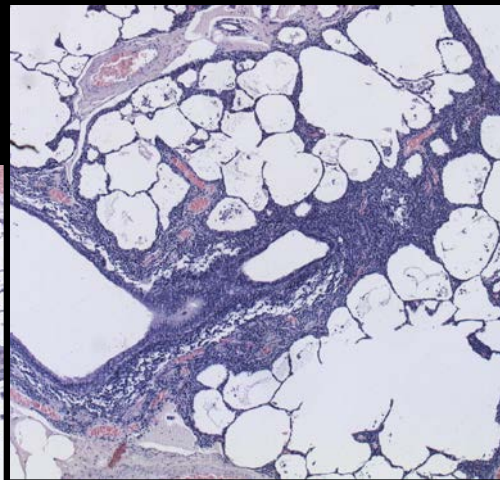
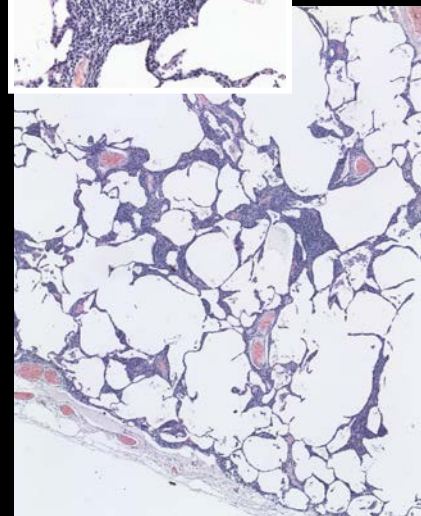
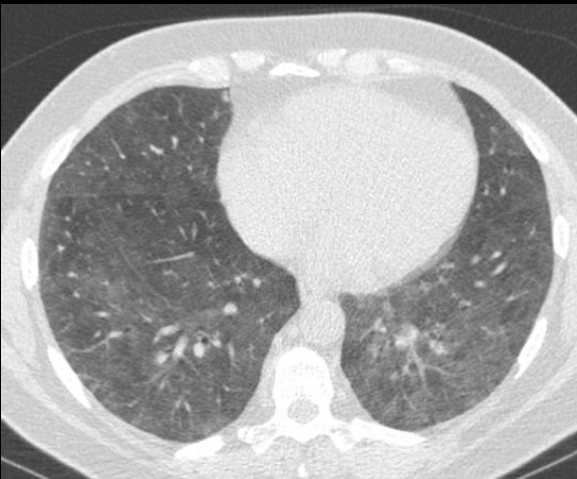
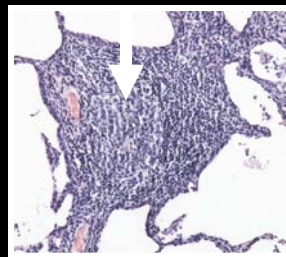
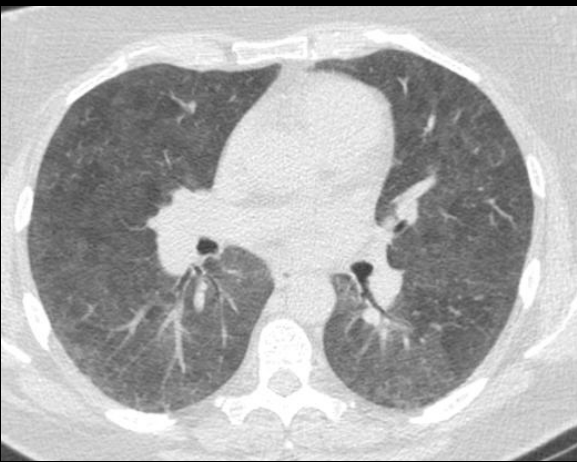
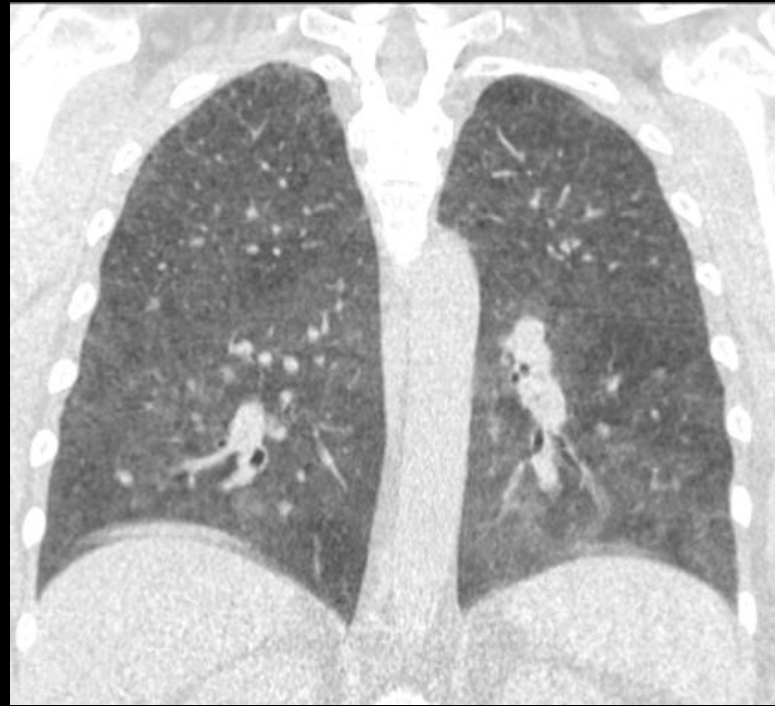


Syndrome de Sjögren

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

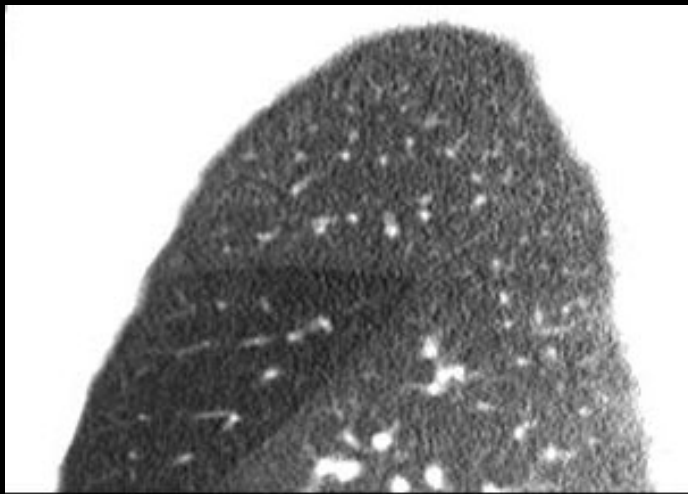
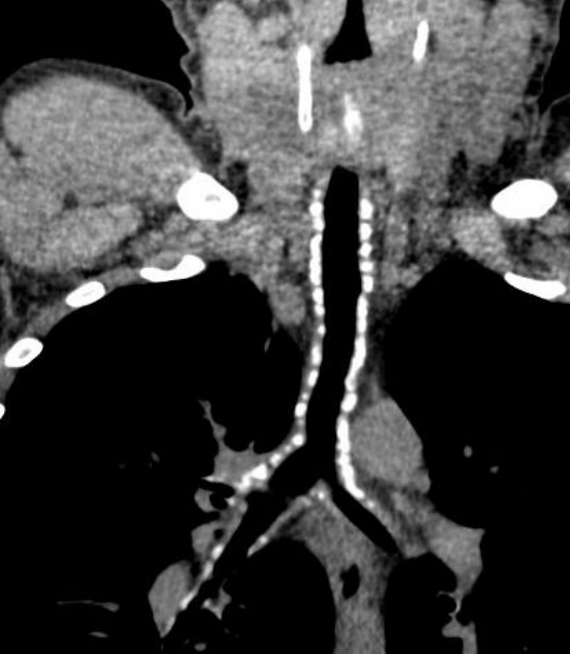
Tannoue LT. Pulmonary involvement in collagen vascular disease: a review of the pulmonary manifestations of the Marfan syndrome, ankylosing spondylitis, Sjögren's syndrome, and relapsing polychondritis. J Thorac imaging 1992;7:62-77





Polychondrite atrophiante

- Maladie rare (qqs 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



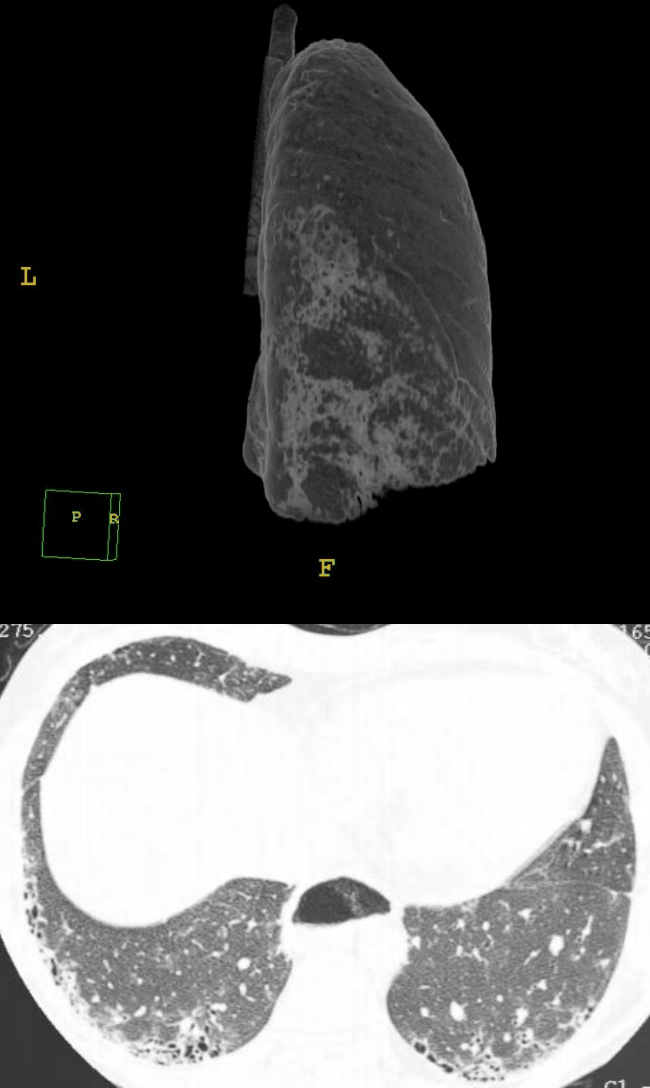
Spondylarthrite ankylosante

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



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