

Démarche clinique

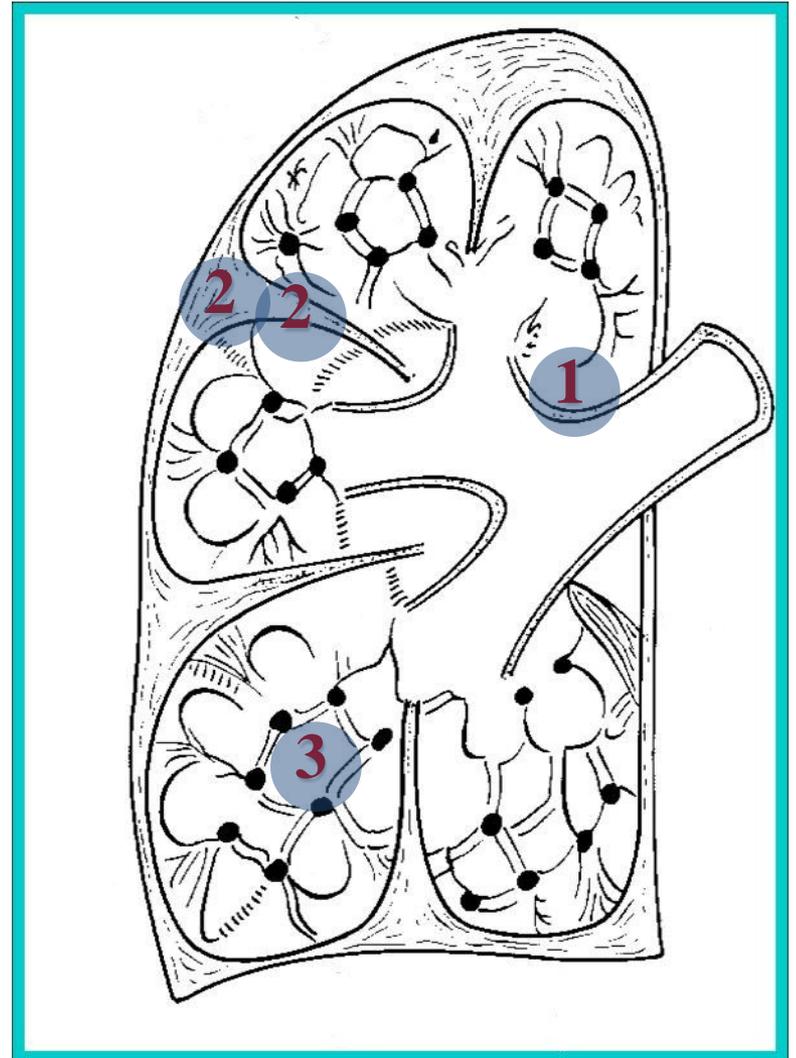
**Le parenchyme pulmonaire:
Le syndrome interstitiel**

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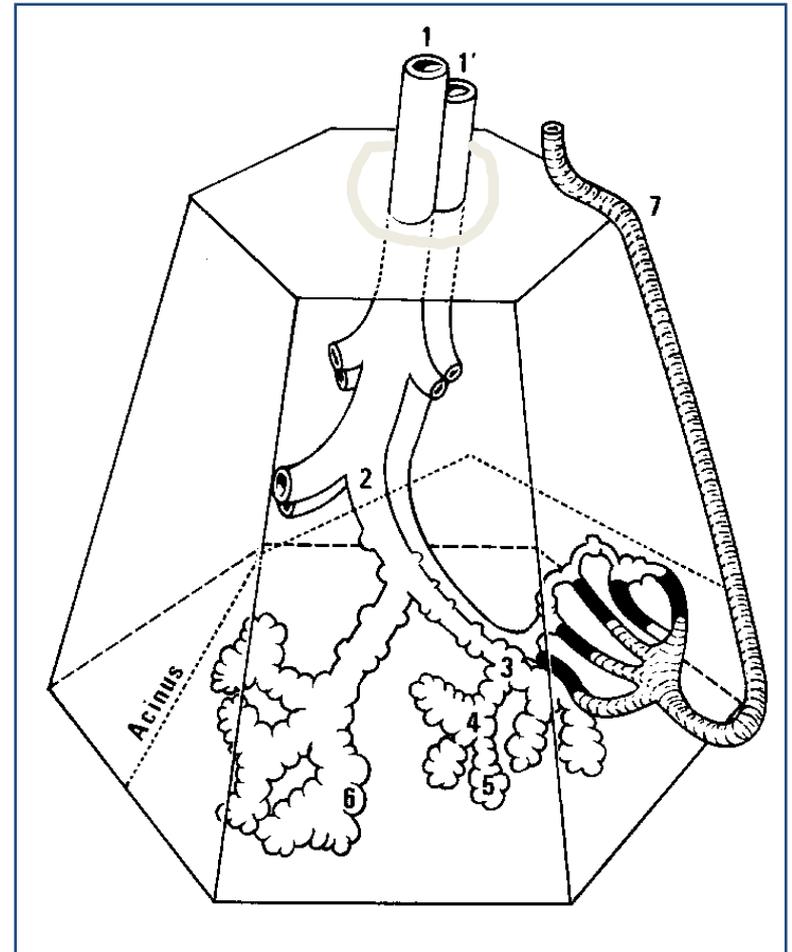
Le Syndrome Interstitiel: définition

- Interstitium = charpente du poumon
 1. Secteur axial = interstitium péribronchovasculaire
 2. Secteur périphérique = septa interlobulaires & tissu sous-pleural
 3. Secteur central = tissu intralobulaire entre les cloisons alvéolaires
- L'interstitium est invisible à l'état normal (RX et TDM)
- S'il est anormal, il s'épaissit et devient visible
 - TDM >>> RX standard



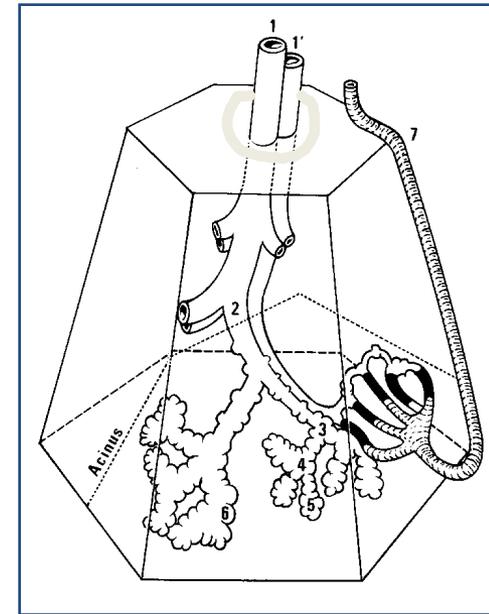
Rappel morphologique: le lobule pulmonaire secondaire

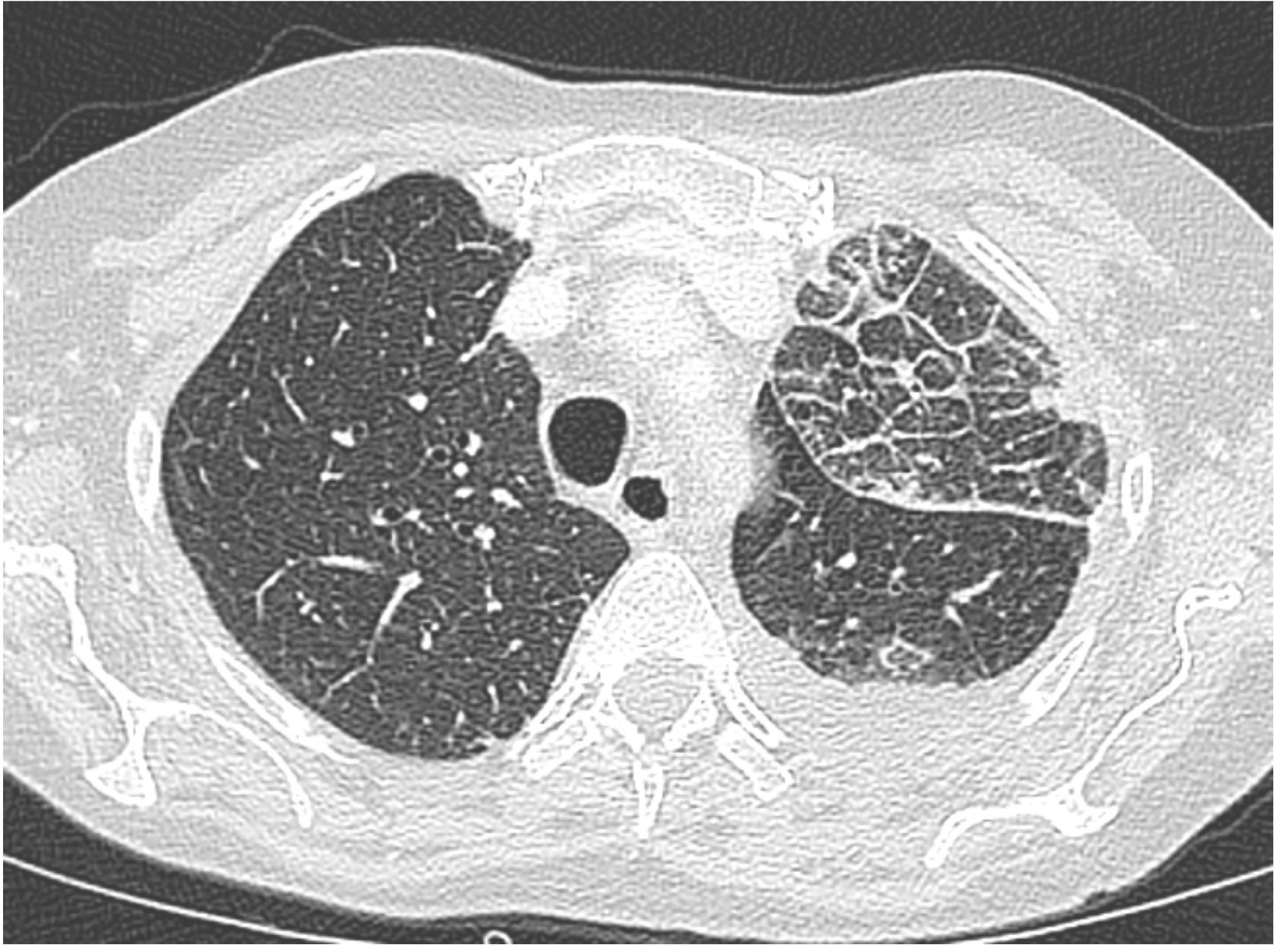
- Unité morphologique et fonctionnelle du poumon
- Polyèdre de 1 à 2 cms de diamètre
- Contient 2 à 4 acini ventilés par des bronchioles terminales
- Centré sur une bronche et une artère centrolobulaires: (secteur interstitiel central)
- La veine est localisée dans le septa interlobulaire: (secteur interstitiel périphérique)

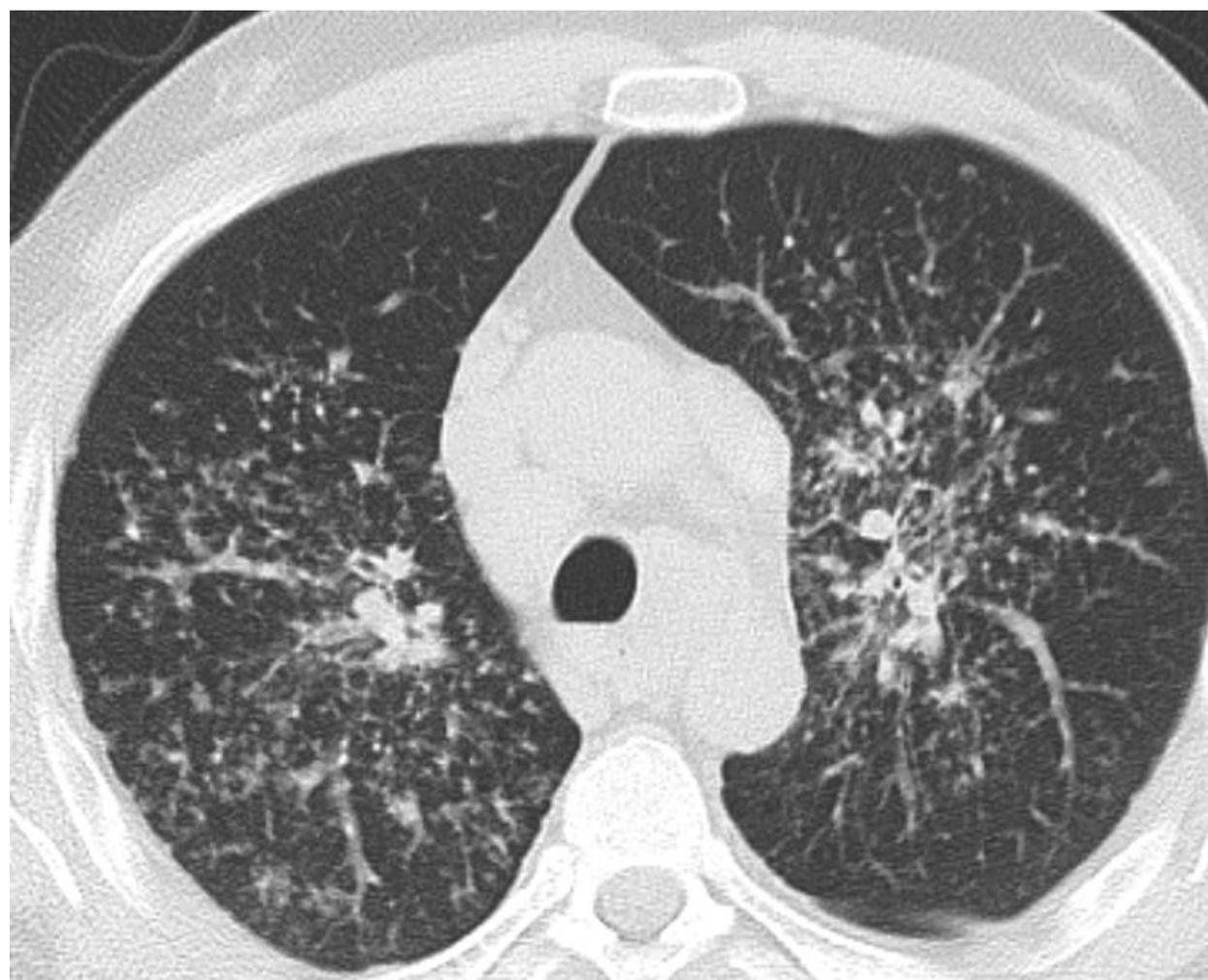


Septa interlobulaires

- Les septa interlobulaires sont donc partie intégrante de l'interstitium pulmonaire
- Ils sont invisibles à l'état normal sur une radiographie standard et sur une TDM
- Ils ne deviennent visibles que si il y a une pathologie interstitielle







Le Syndrome Interstitiel: Etiologies

- **Granulomatoses**

- Tbc
- Coniose
- Alvéolite
extrinsèque
allergique
- BBS
- Histiocytose X

- **Affections interstitielles malignes**

- Lymphangite carcinomateuse

- **Œdème pulmonaire interstitiel**

- Décompensation cardiaque
(avant œdème alvéolaire)

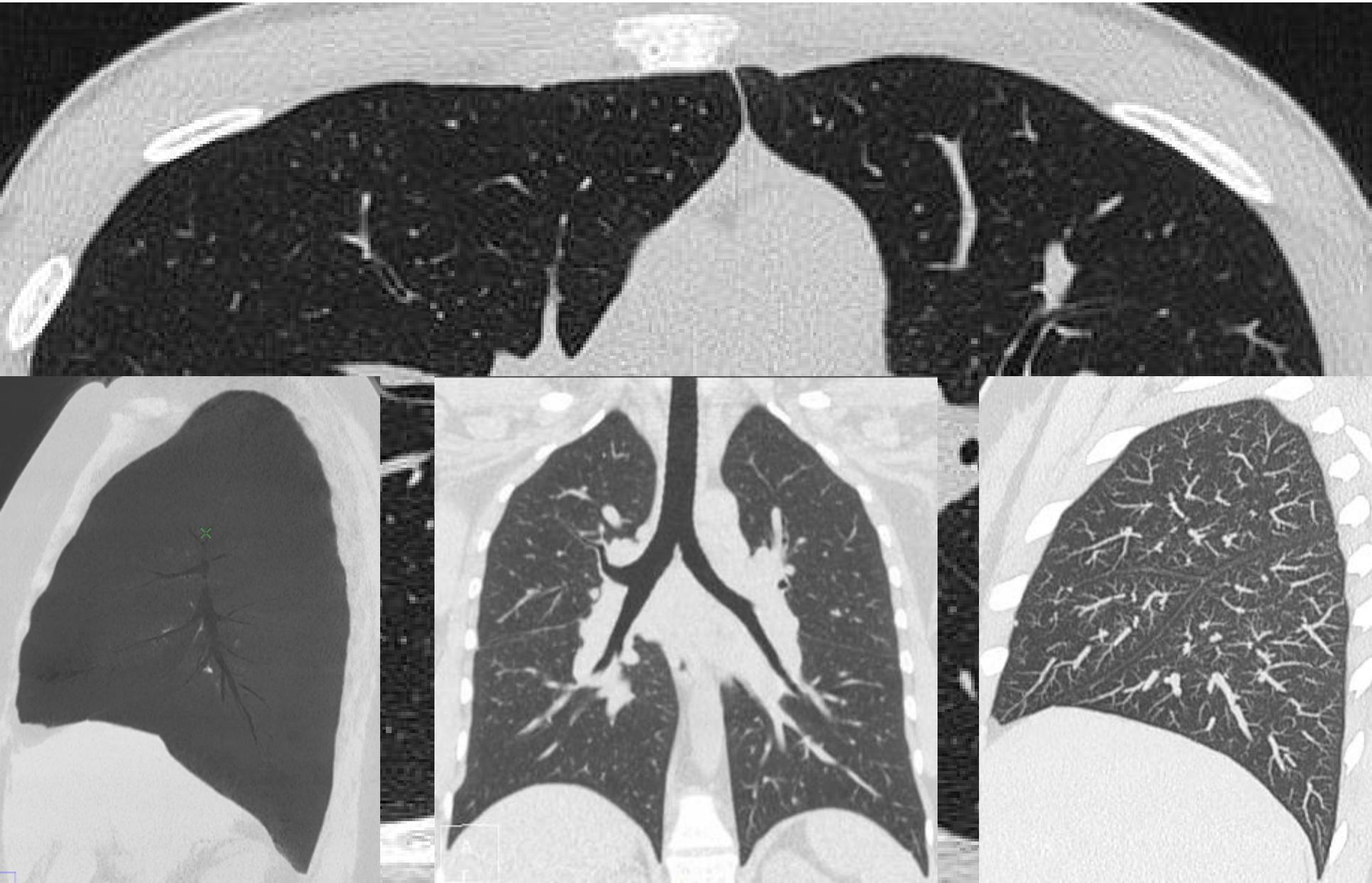
p cap > 25 mm Hg & < 35 mmHg

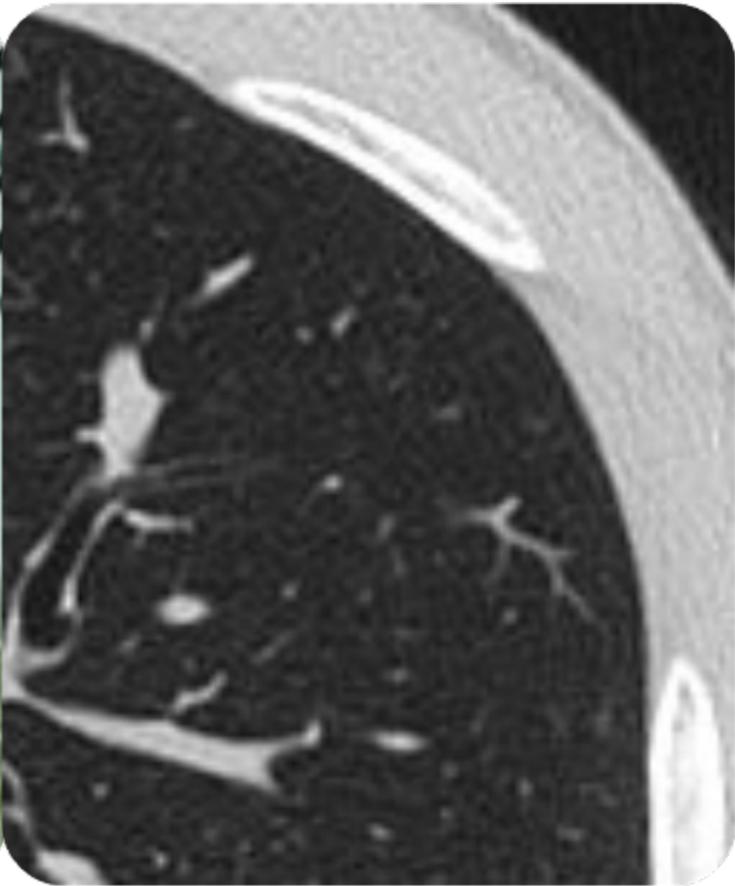
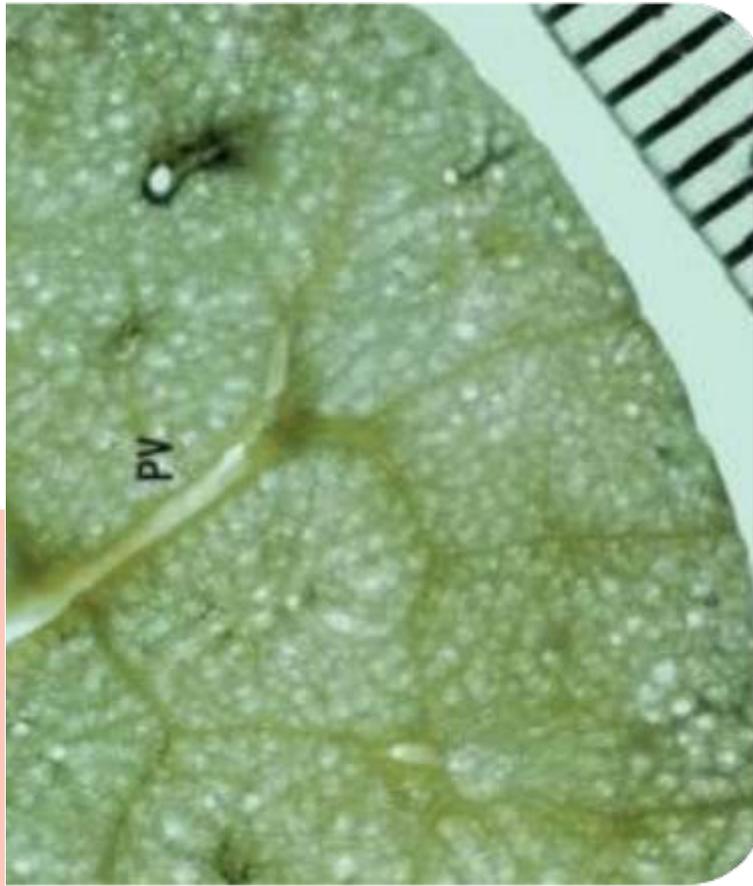
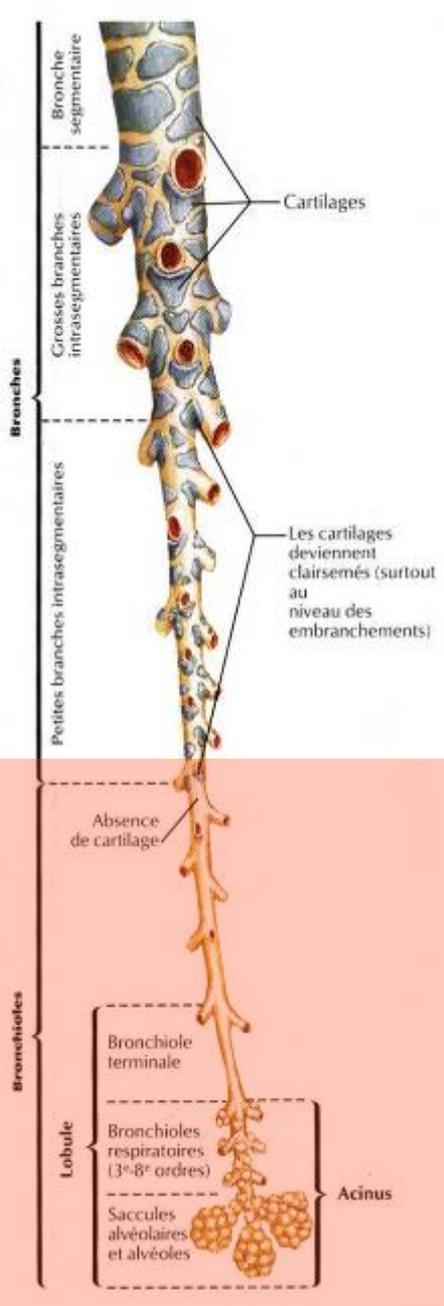
- **Fibrose interstitielle diffuse**

- Idiopathique: UIP et variantes
- Spécifiques: sclérodermie, PR
- Médicamenteuses

Diagnostic d' une micronodulation

technique





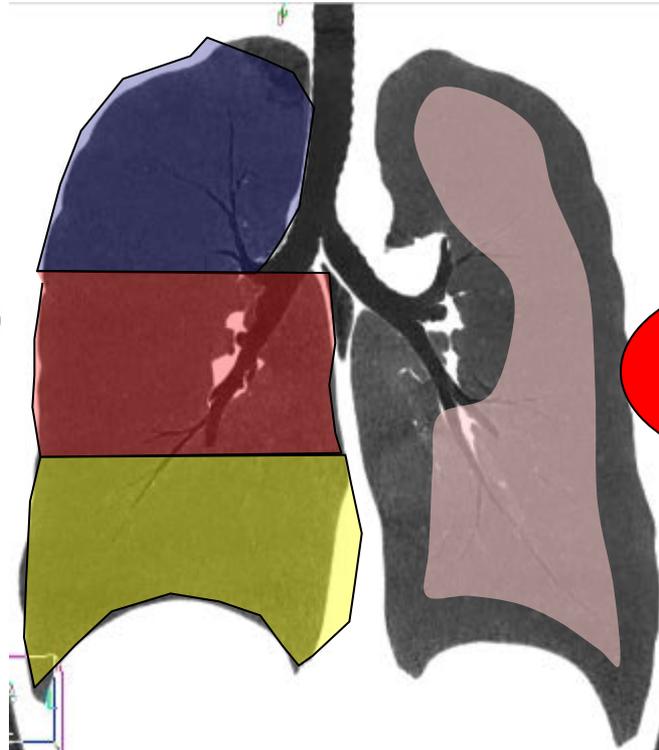
Avantages de la TDM-HRV

- Exhaustivité de l'exploration
 - Accès aux techniques de rendu volumique
 - ✓ reconstructions MPR
 - ✓ reconstruction minIP
 - ✓ reconstruction MIP
 - ✓ VR et 3D surface
- Détection**
Caractérisation
Localisation
Quantification

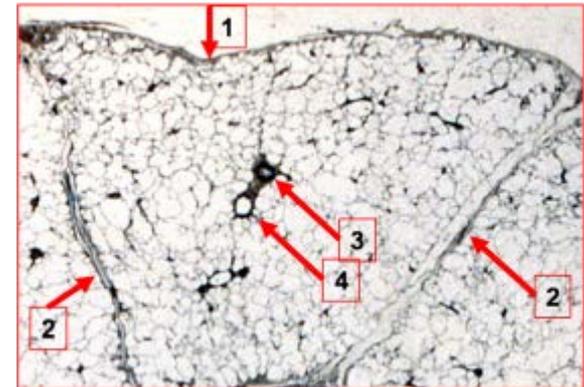
Diagnostic des PID par TDM-HR

Signe prédominant

Signes associés



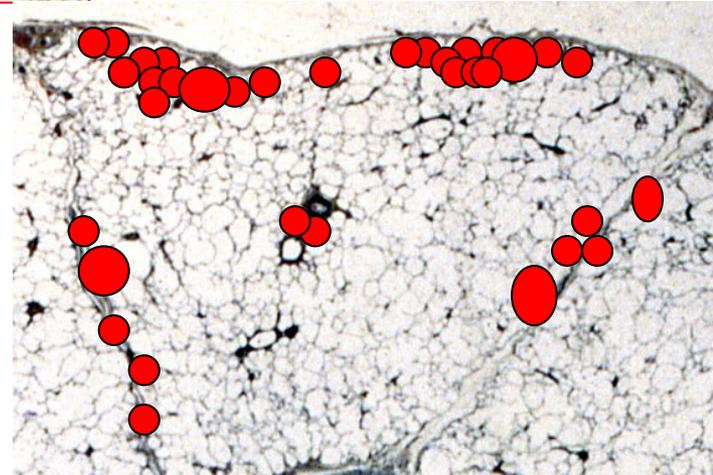
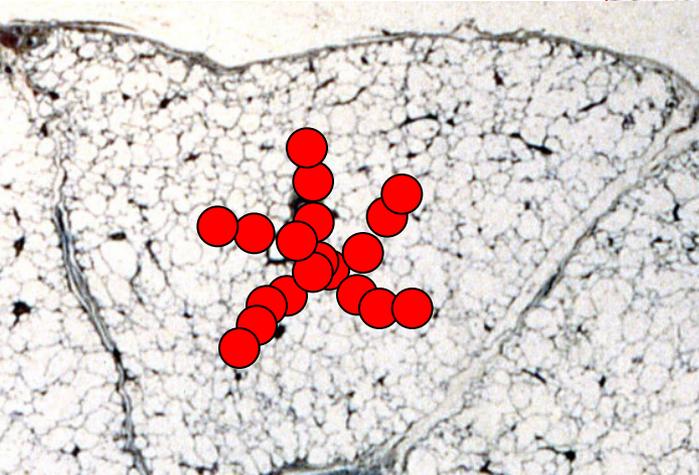
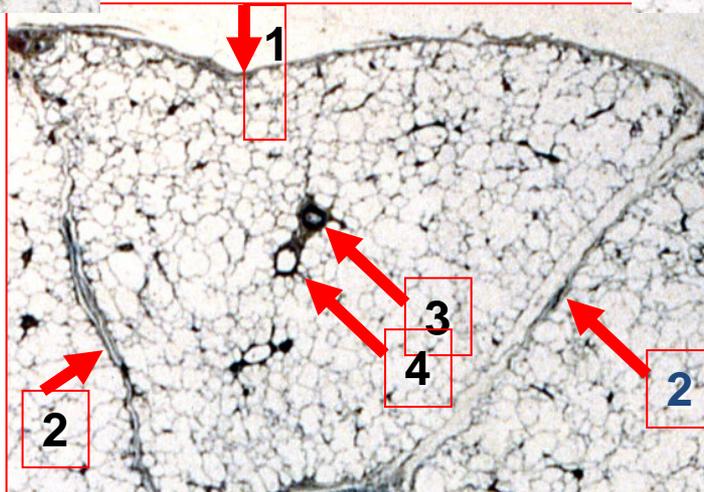
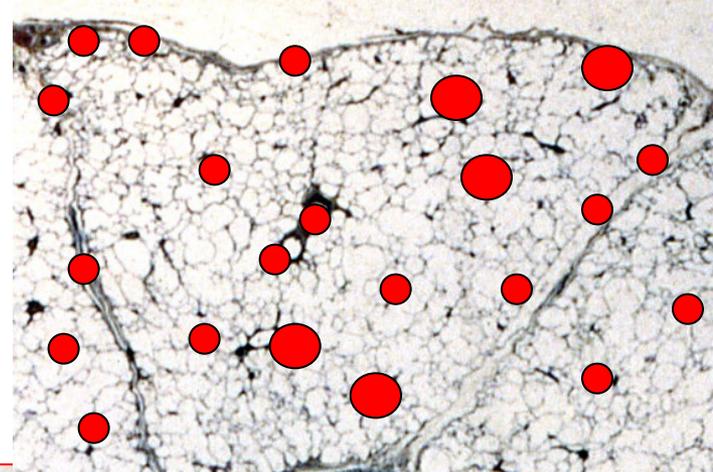
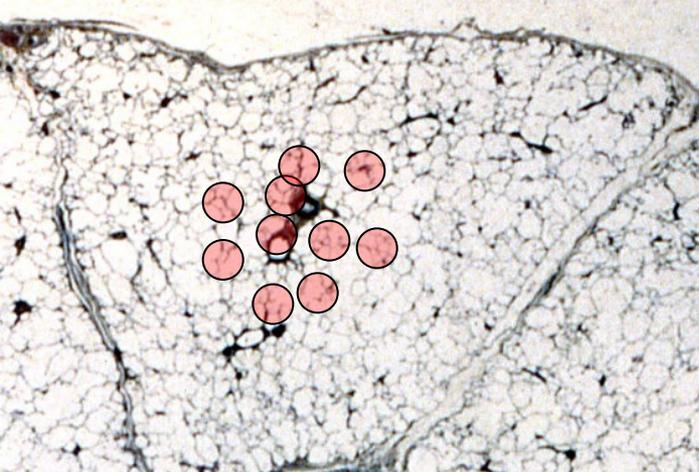
distribution

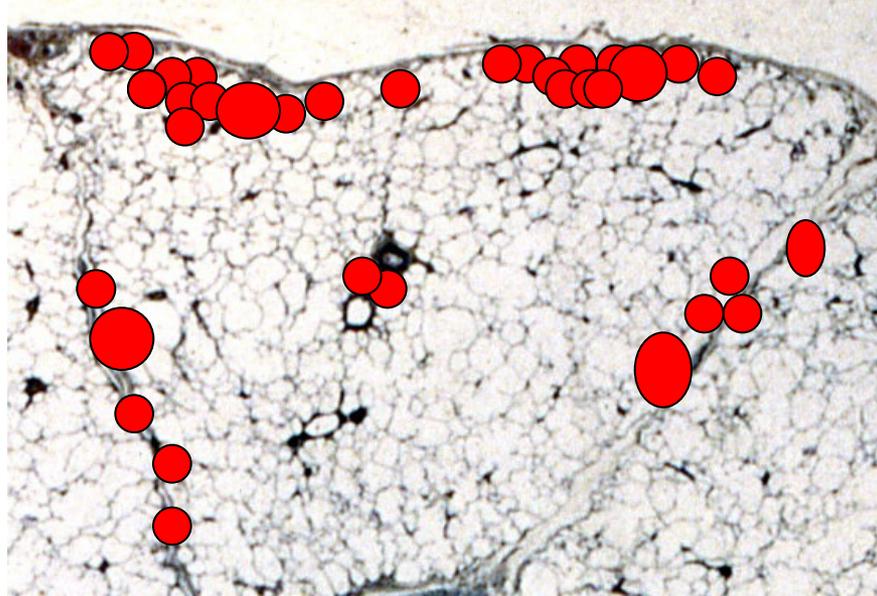


temps

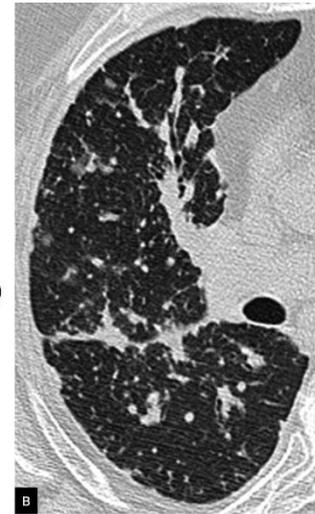
Image nodulaire

- **Infiltration pulmonaire**
 - opacités de forme arrondie 1 à 30 mm
 - micronodule < 5 mm
 - 5 mm < nodules < 30 mm
- **La composition histologique du nodule : cellulaire, fibreuse ou mixte**
 - Infiltration bronchiolaire et péribronchiolaire
 - Confluence de granulomes (*sarcoïdose*)
 - Prolifération cellulaire (*métastases*)
 - Fibrose mutilante nodulaire (*silicose*)
- **Confluence possible avec rétraction : masses pseudo-tumorales**
- **Le nodule est homogène ou présente une clarté centrale (*nodule excavé*)**





NODULES PÉRILYMPHATIQUES



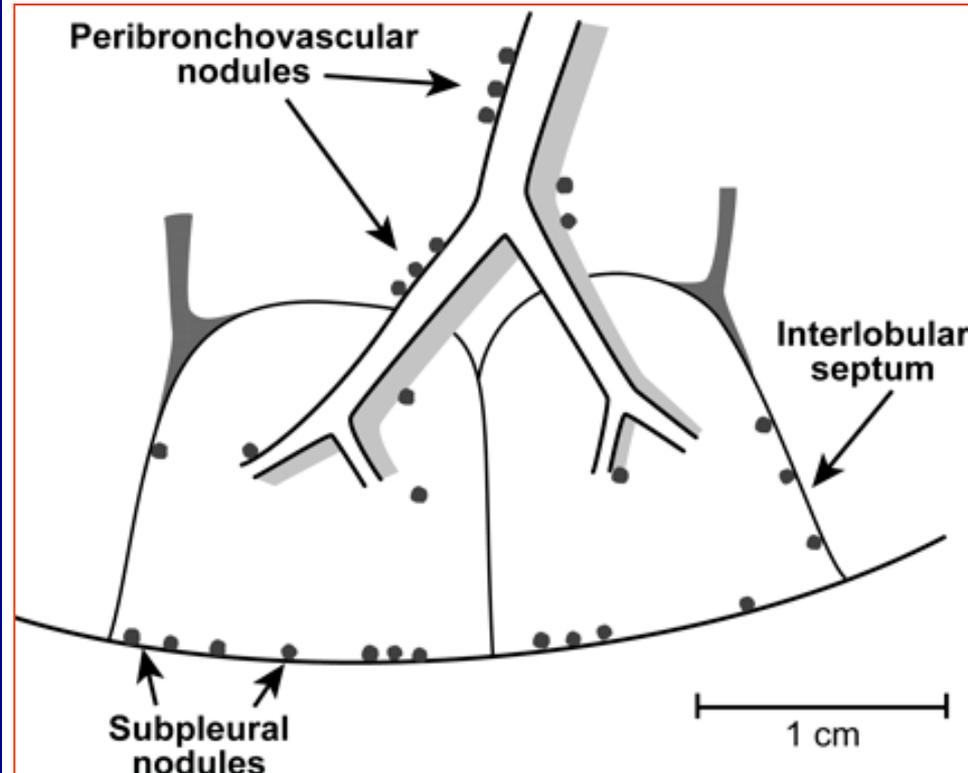
Modèle micronodulaire péri-lymphatique

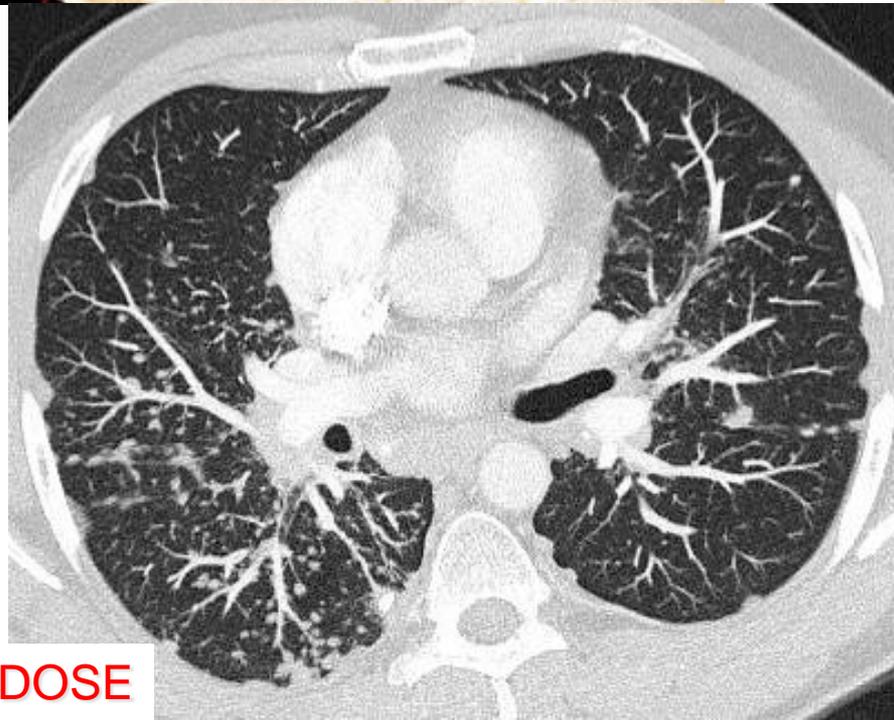
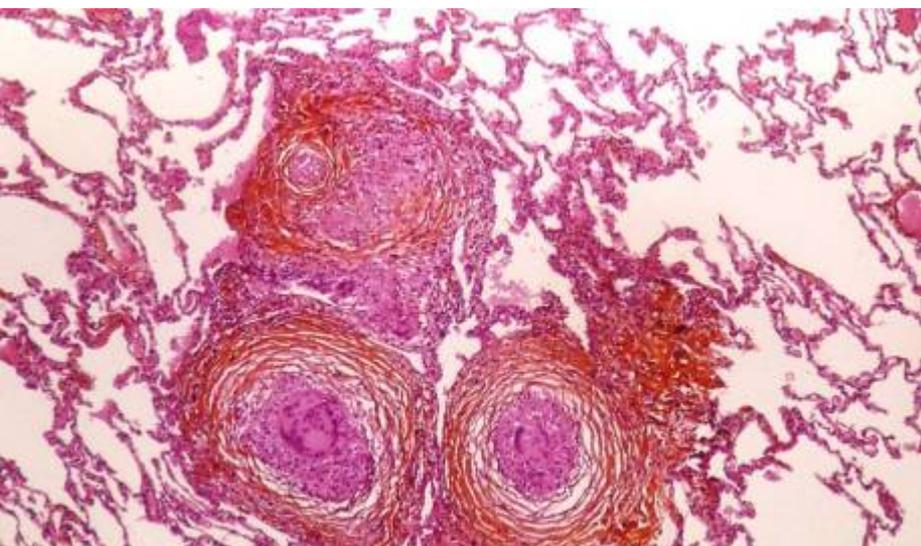
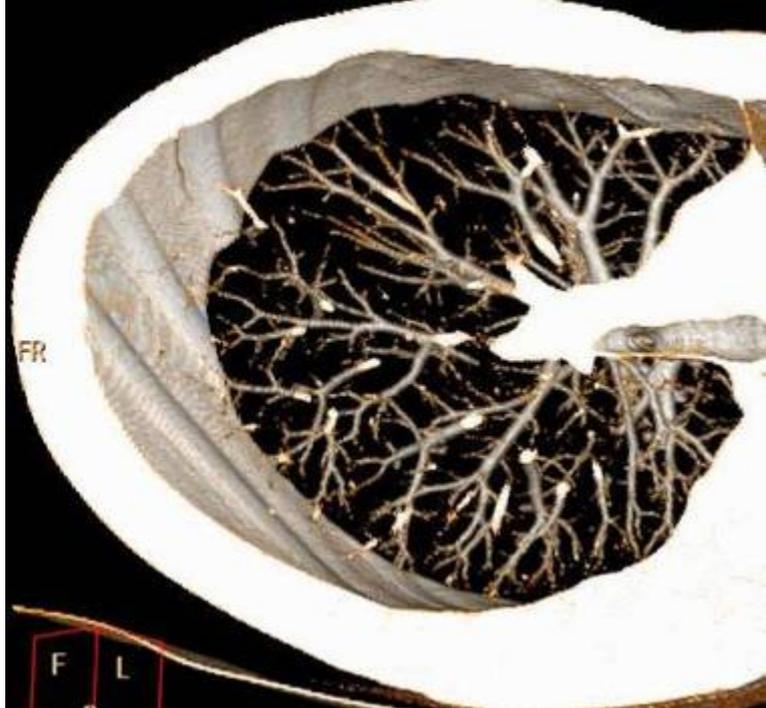
Caractéristiques :

- 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

D'autres images peuvent se trouver associées en fonction de l'étiologie :

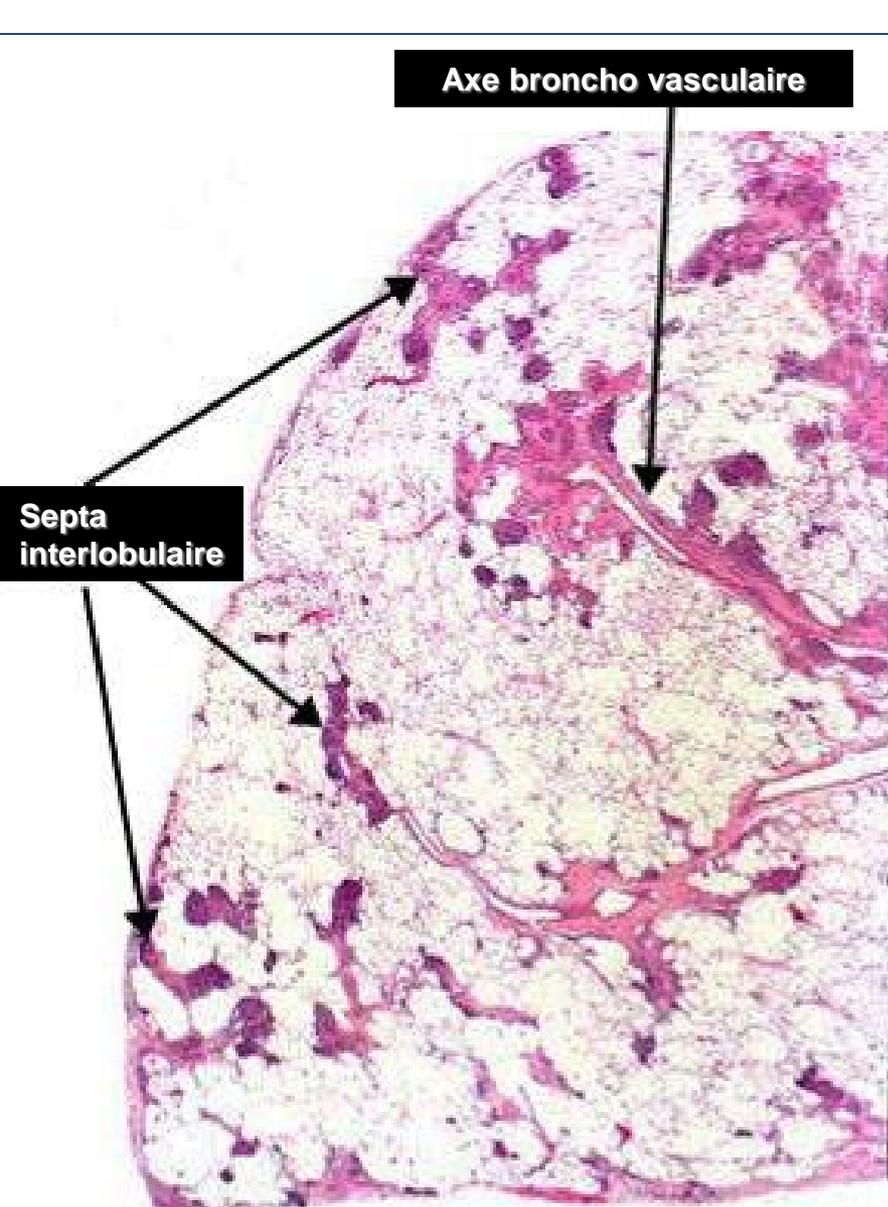
- lignes septales avec aspect perlé
- épaississement péri-broncho-vasculaire
- îlots de dépoli d'étendue variable
- hypertrophie ganglionnaire
- épanchement pleural

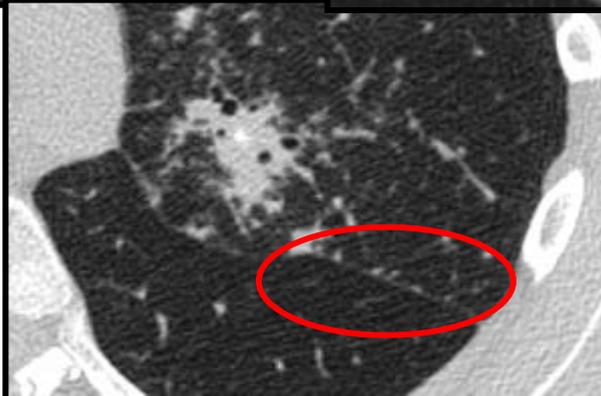
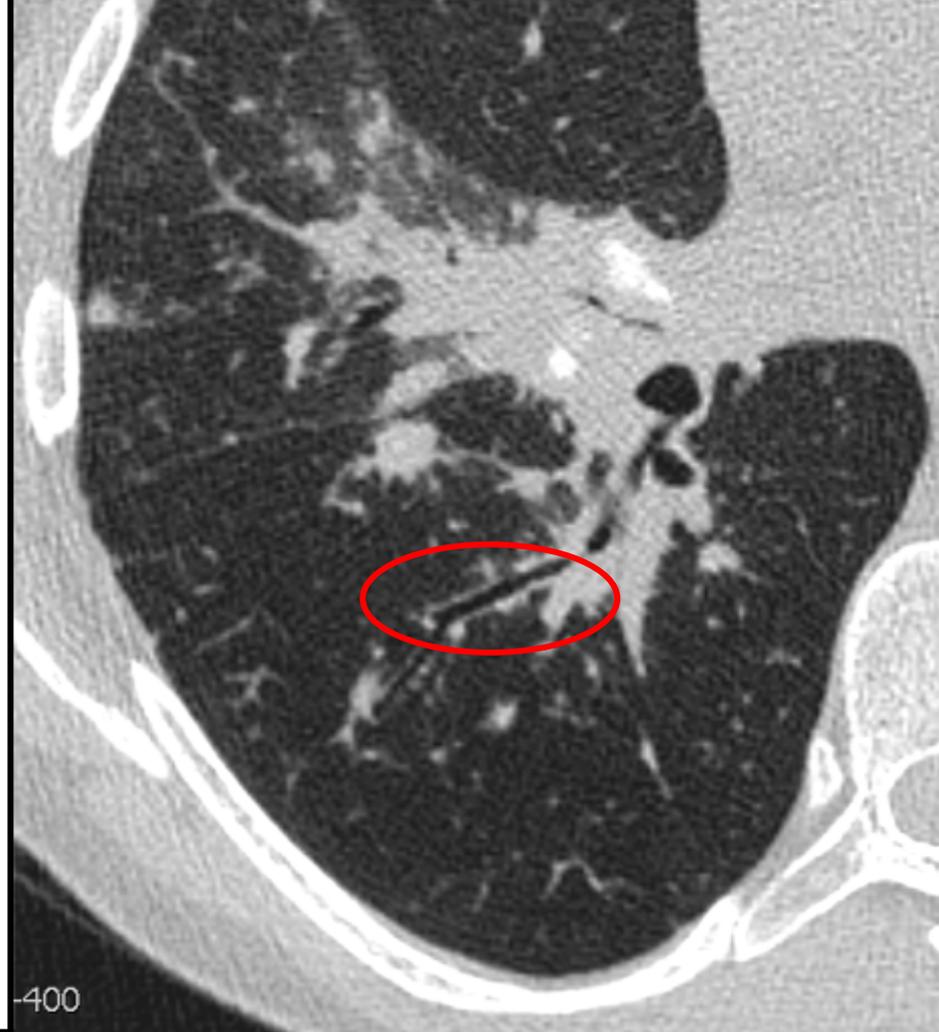
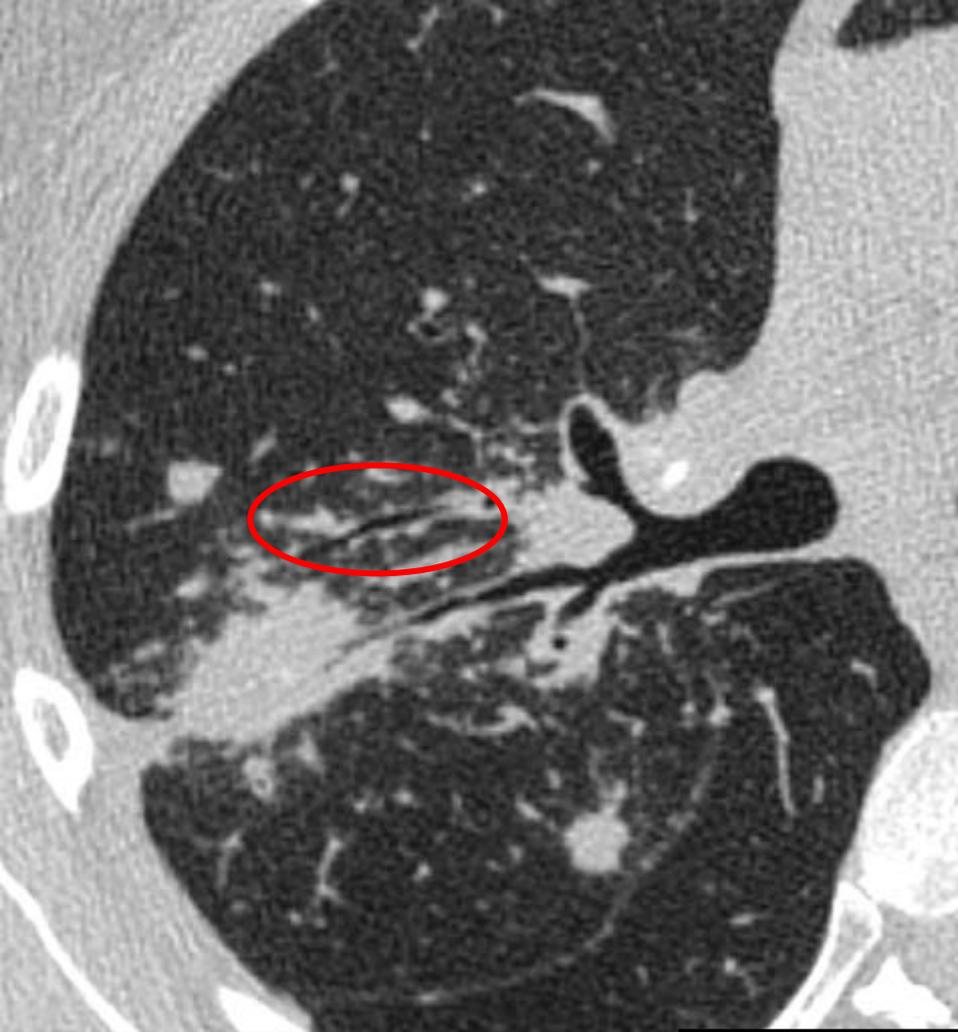


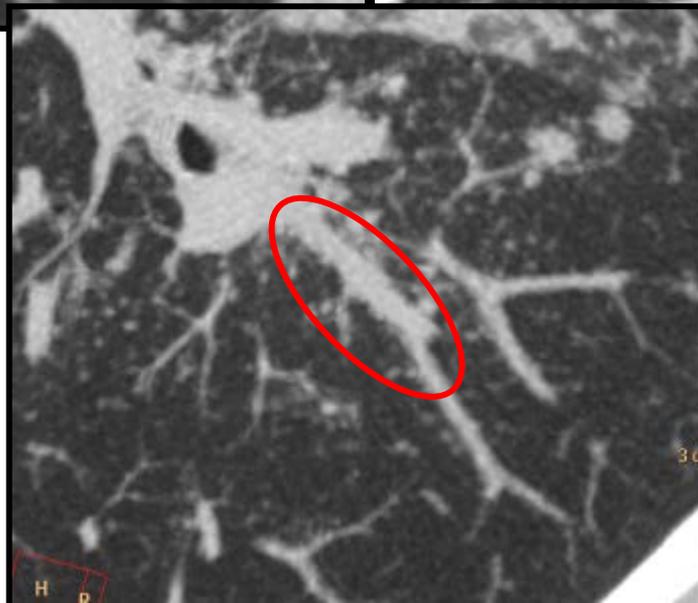
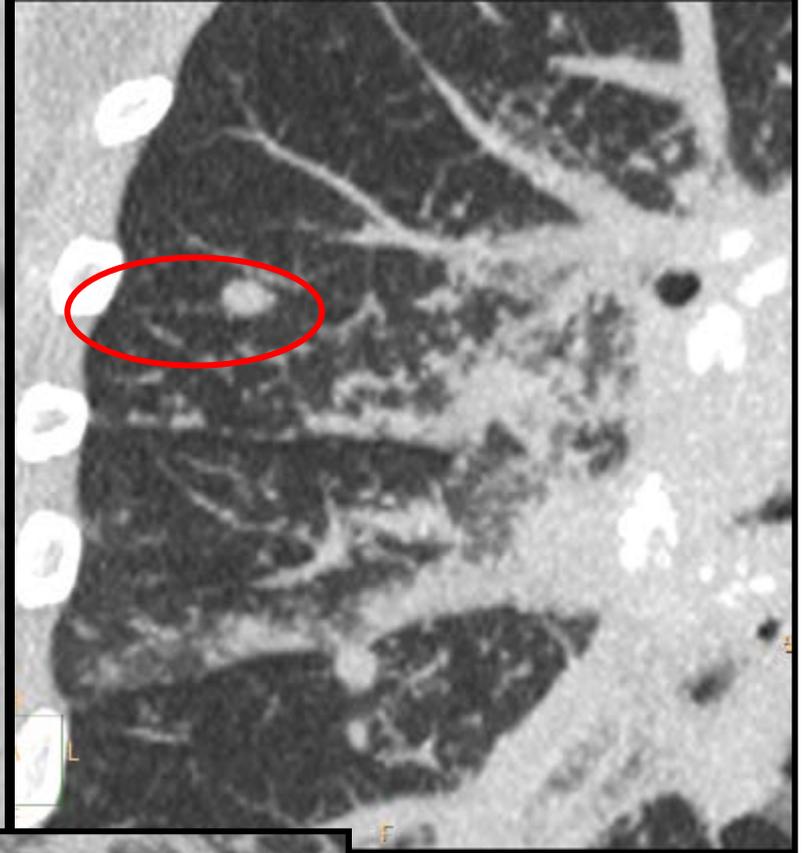


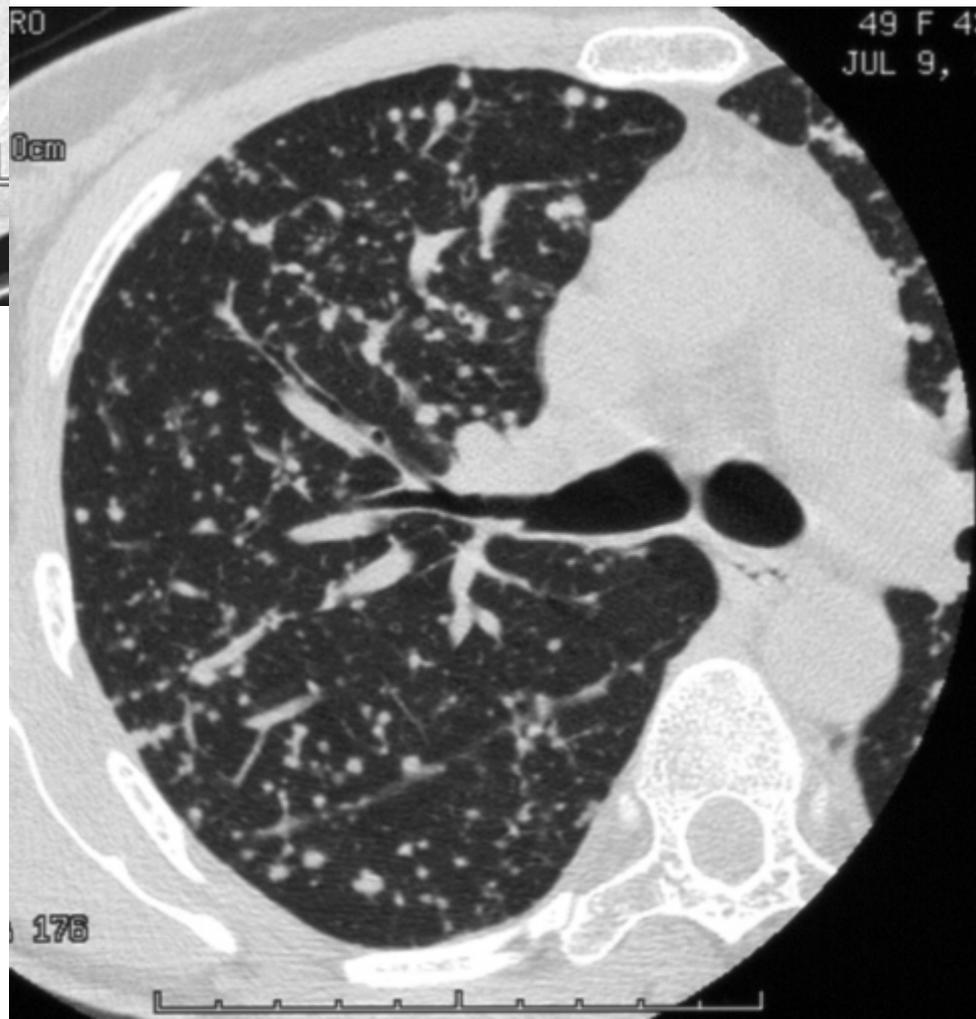
SARCOIDOSE



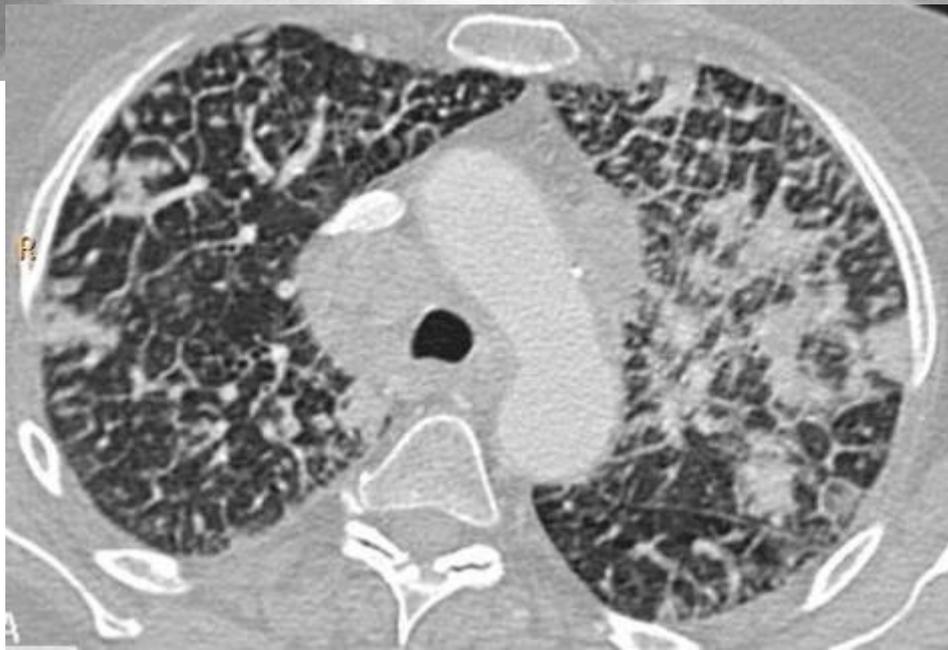
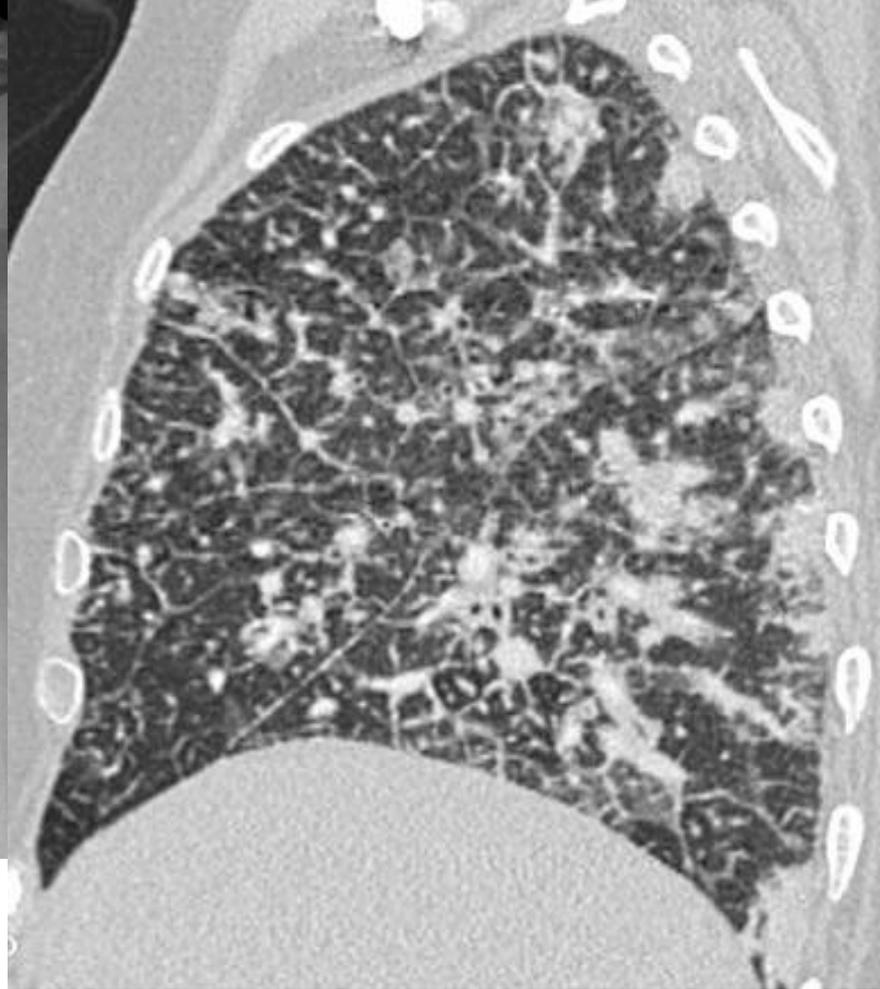
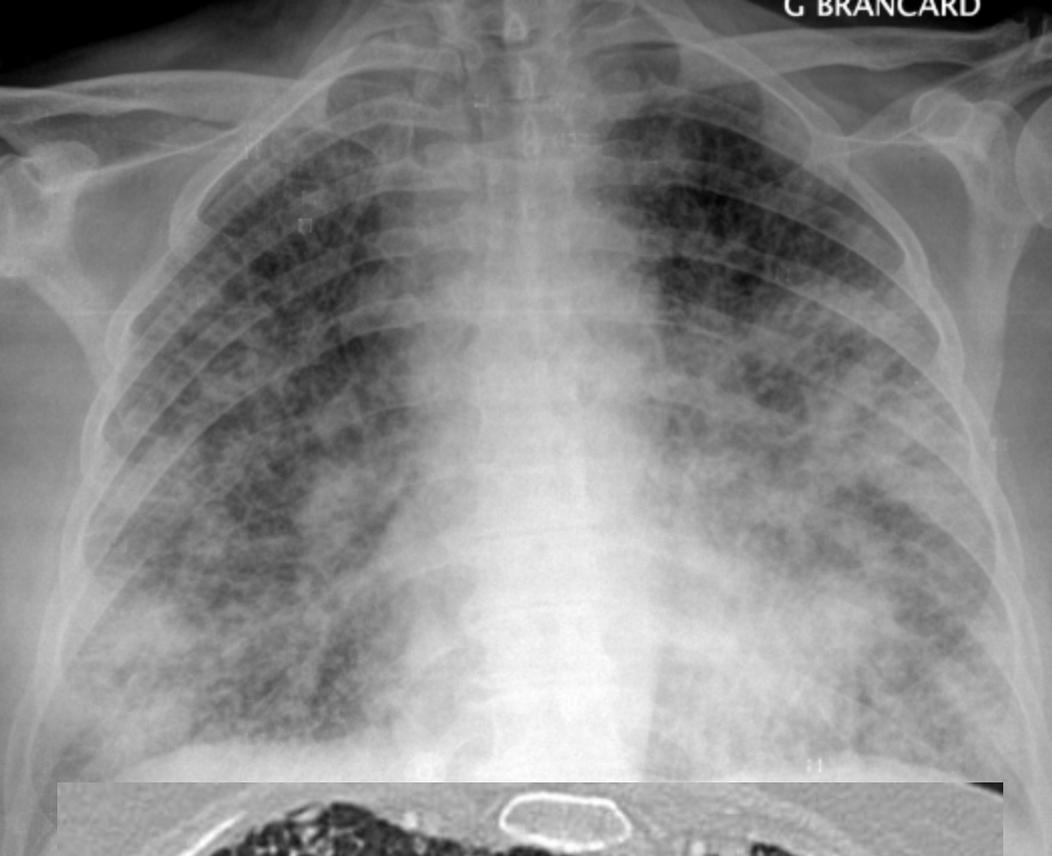








Lymphangite carcinomateuse.
L'infiltration micronodulaire est constituée par des amas de cellules tumorales situées autour ou dans les canaux lymphatiques dilatés.



**Cancer du rein droit avec
métastases pulmonaires et
hépatiques**



silicose





Principales étiologies

Sarcoïdose

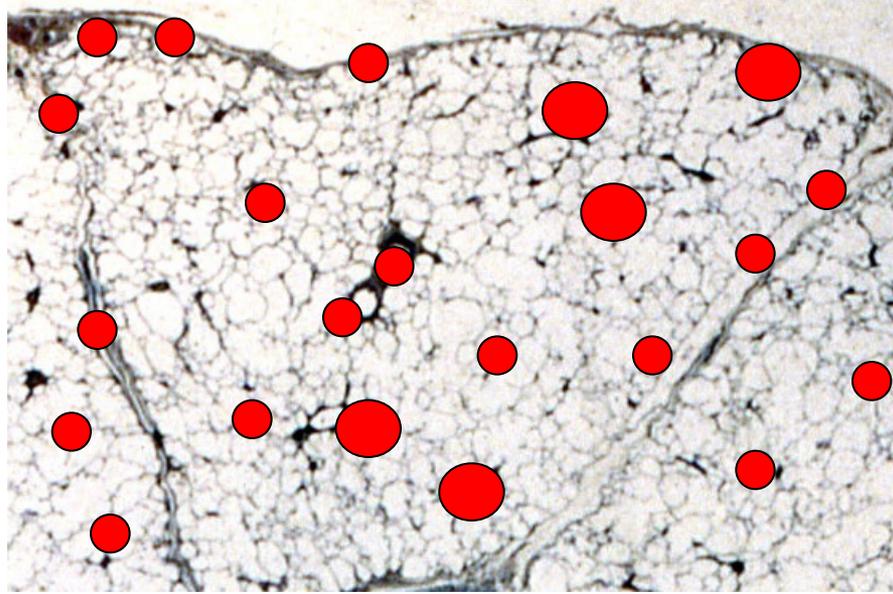
Lymphangite carcinomateuse

Lymphome pulmonaire

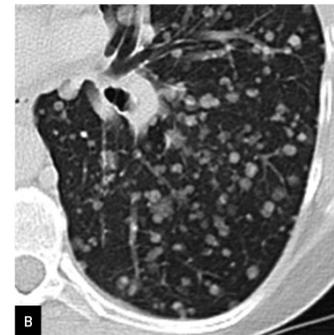
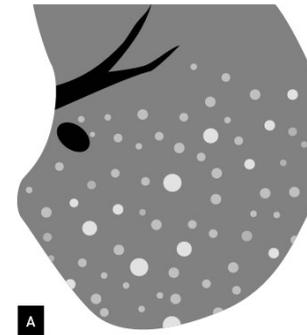
Bérylliose ; Silicose

Amylose

LIP (*syndrome sec ; maladie auto-immune ; VIH*)



MICRONODULATION UBIQUISTE



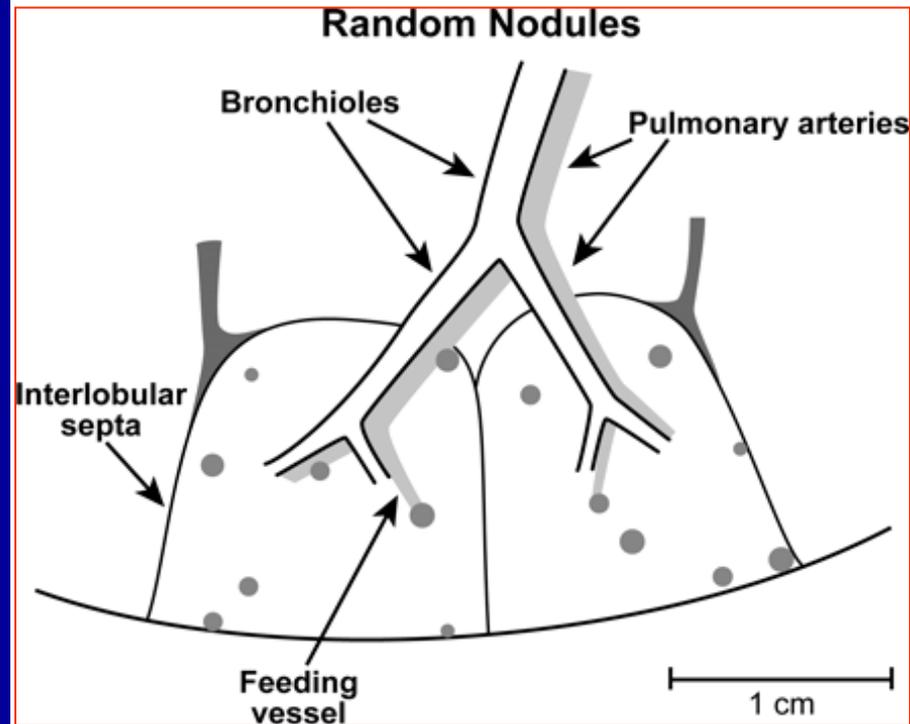
Modèle micronodulaire ubiquiste

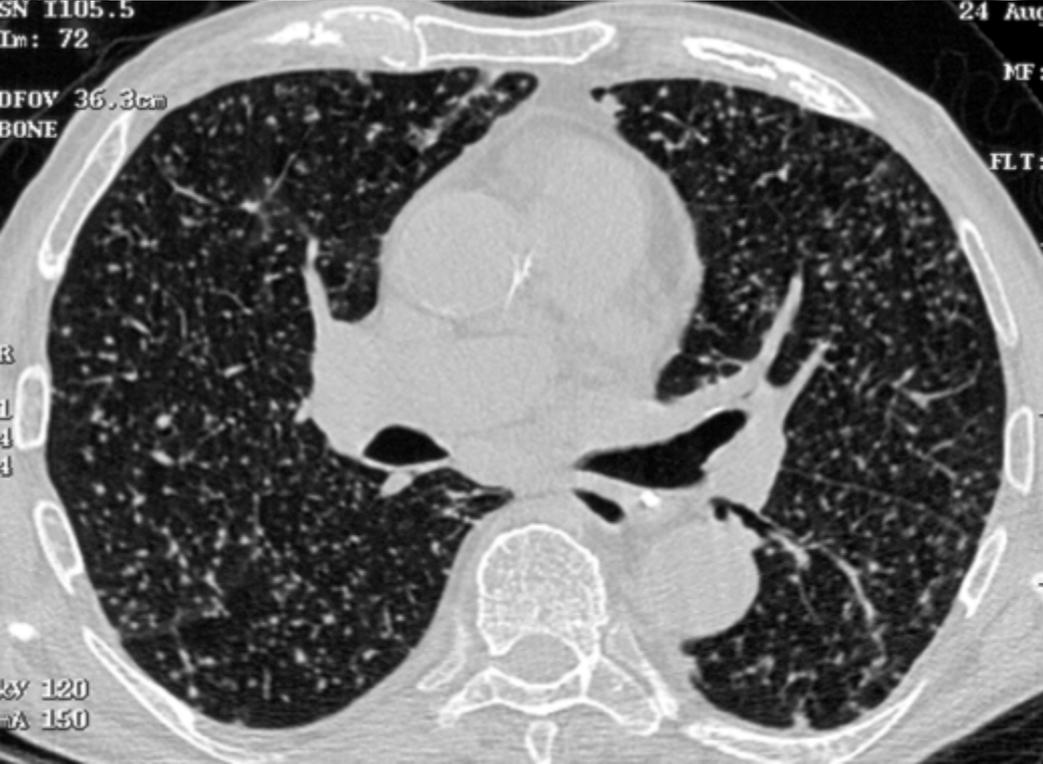
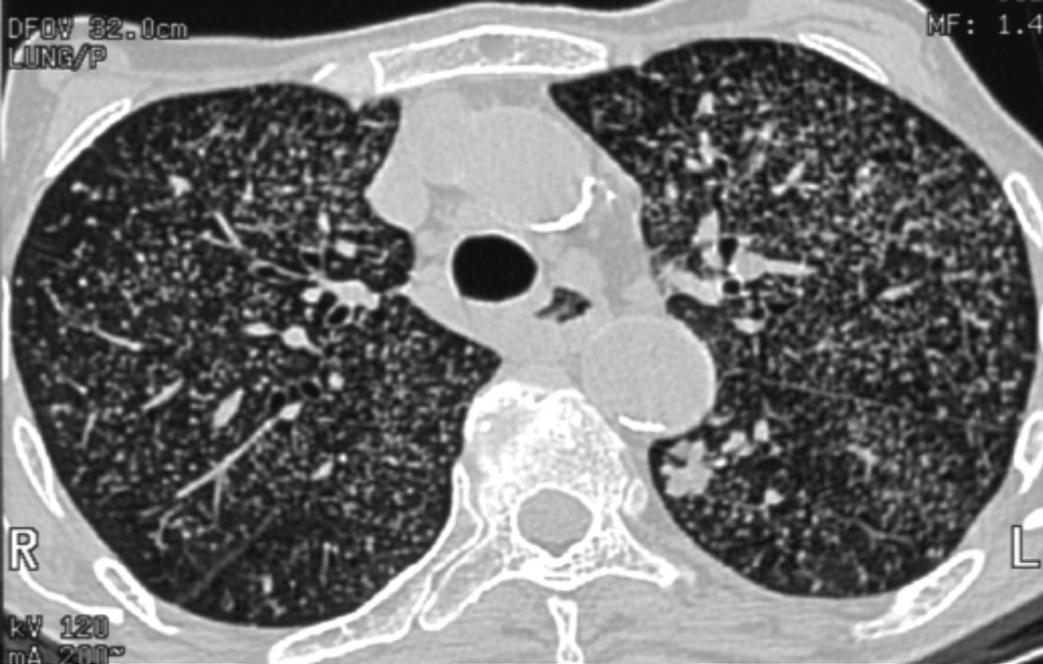
micronodules régulièrement répartis au niveau des deux plages pulmonaires, **sans prédominance éléments topographique par rapport à la surface pleurale, aux scissures, aux éléments bronchovasculaires, et aux limites du lobule.**

diamètre identique

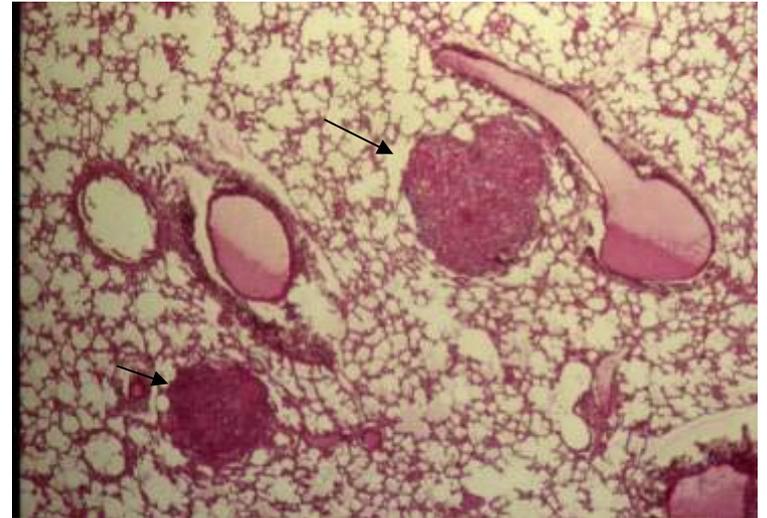
étiologies

- tuberculeuse
- métastatique
- **mycoses** (*aspergillose, candidose*),
- **viroses** (*herpès, CMV*).





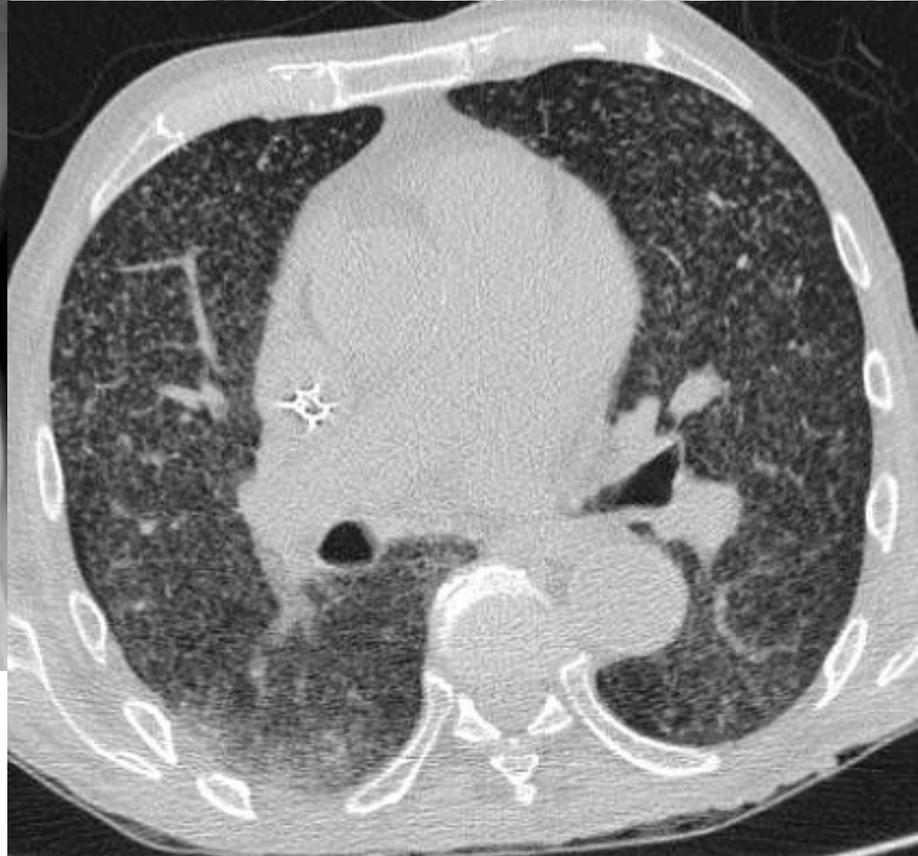
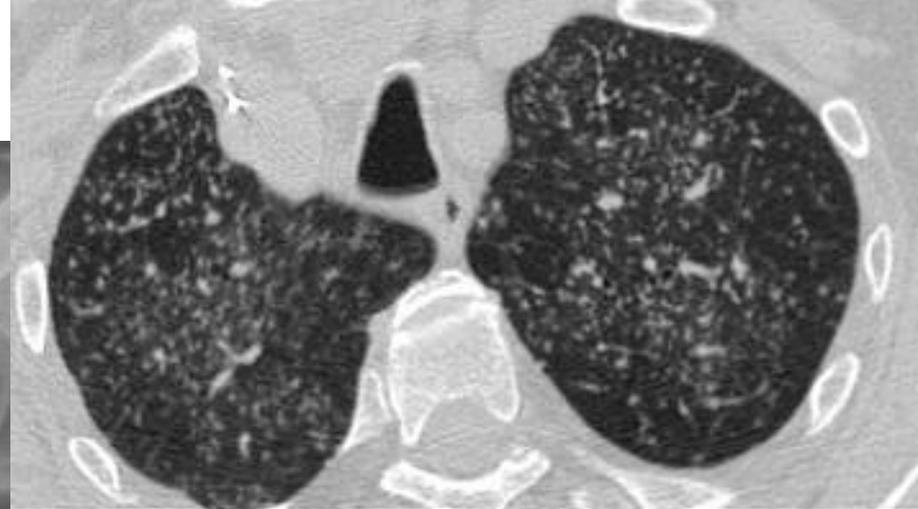
Miliaire tuberculeuse hémotogène

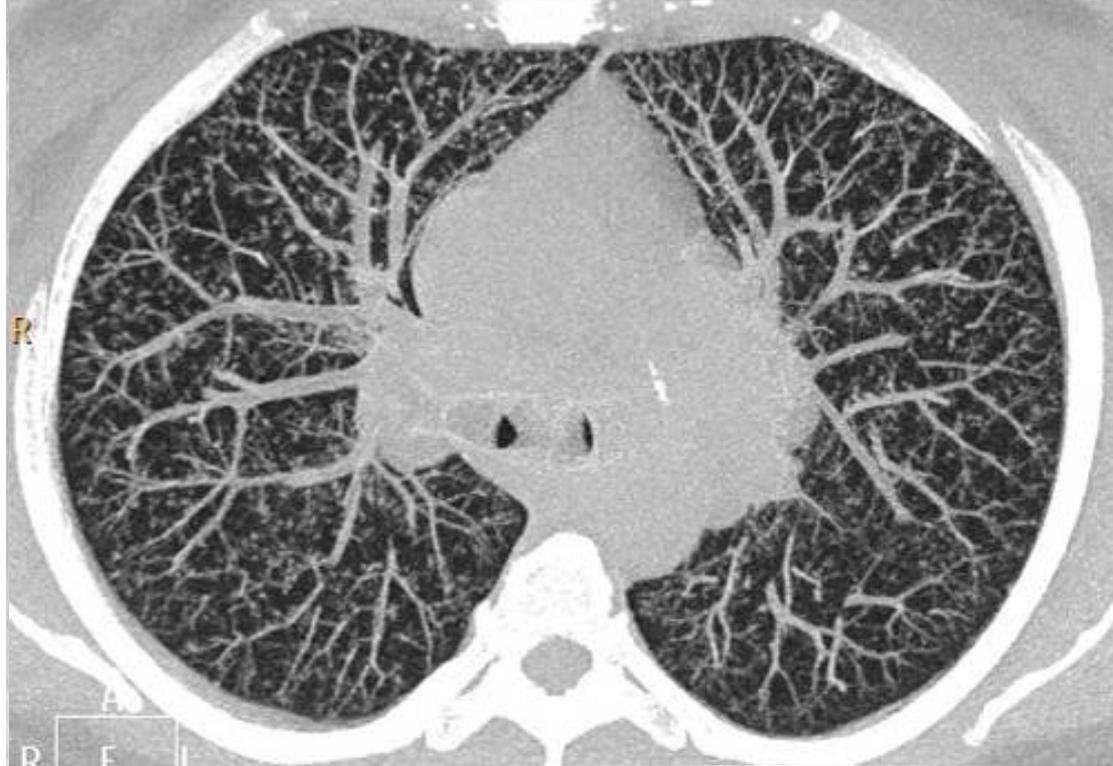
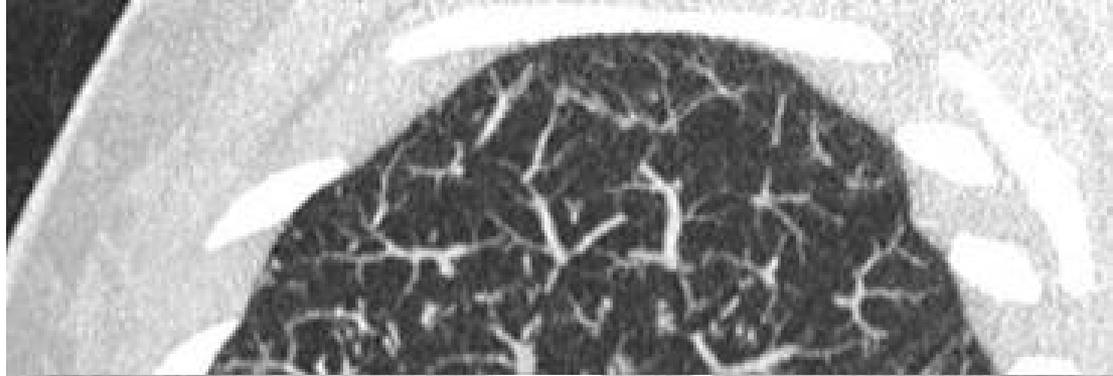


Histologie

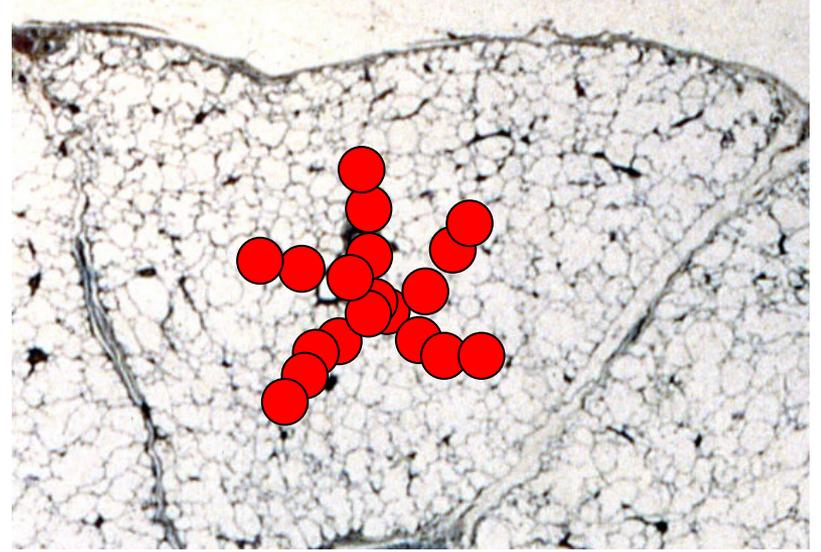
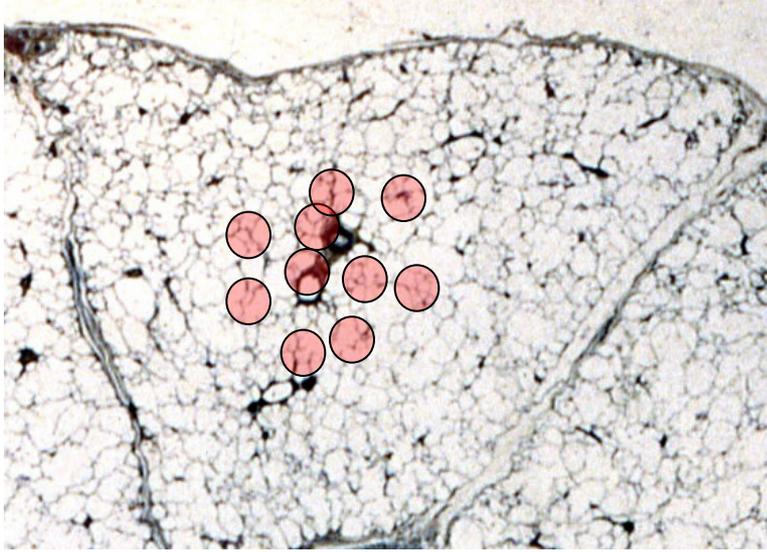
Agglomérats de follicules à distance des éléments broncho-artériels et veineux

Miliaire tuberculeuse hémato-gène





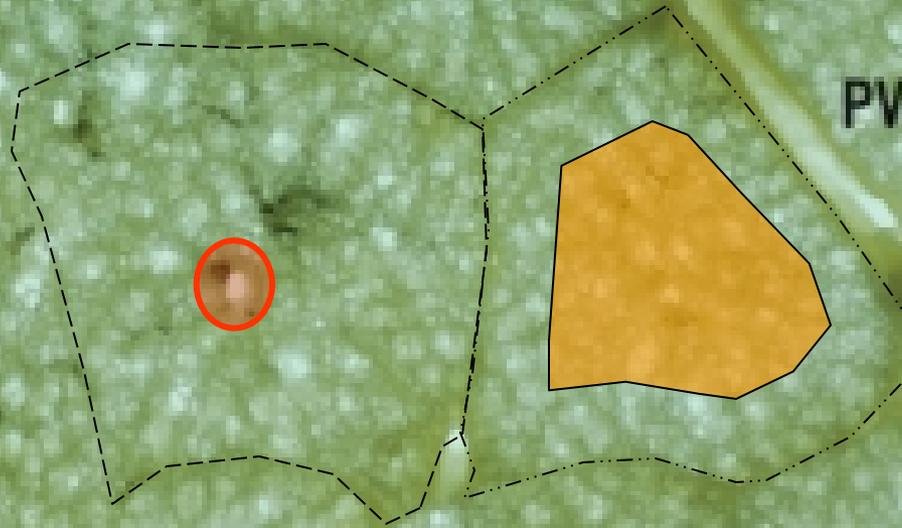
Miliaire tumorale sarcome



NODULES CENTROLOBULAIRES

Topographie centrolobulaire ?

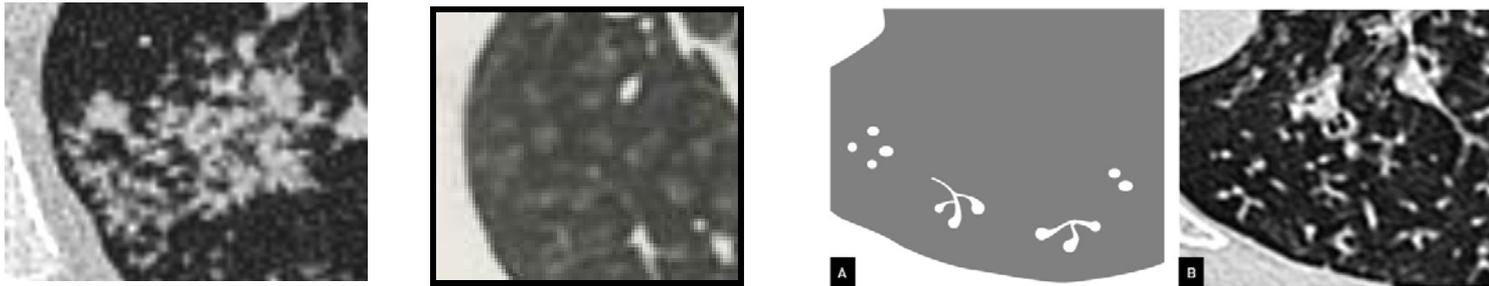
région du pédicule broncho vasculaire du LPS : B - A - L



typiquement à 3 mm de la plèvre et des septas

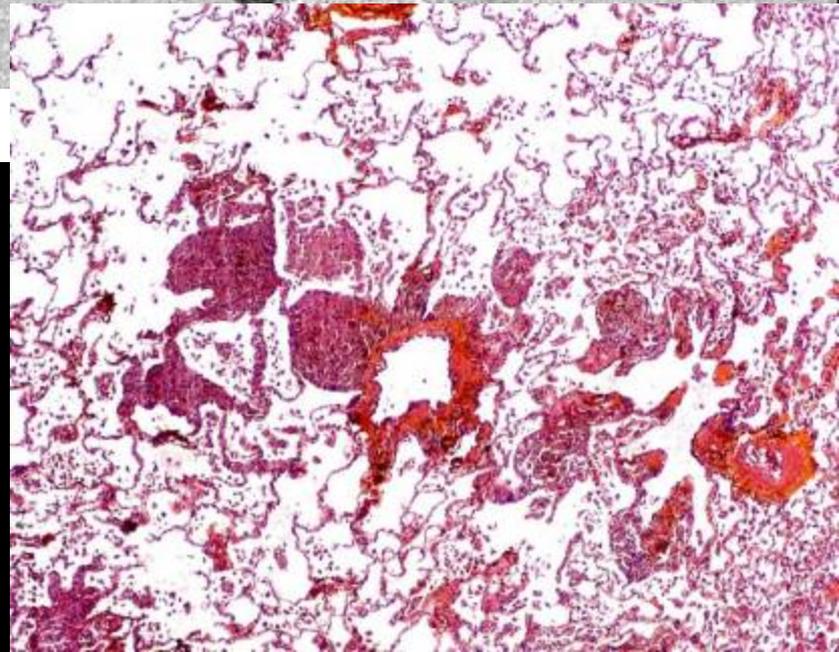
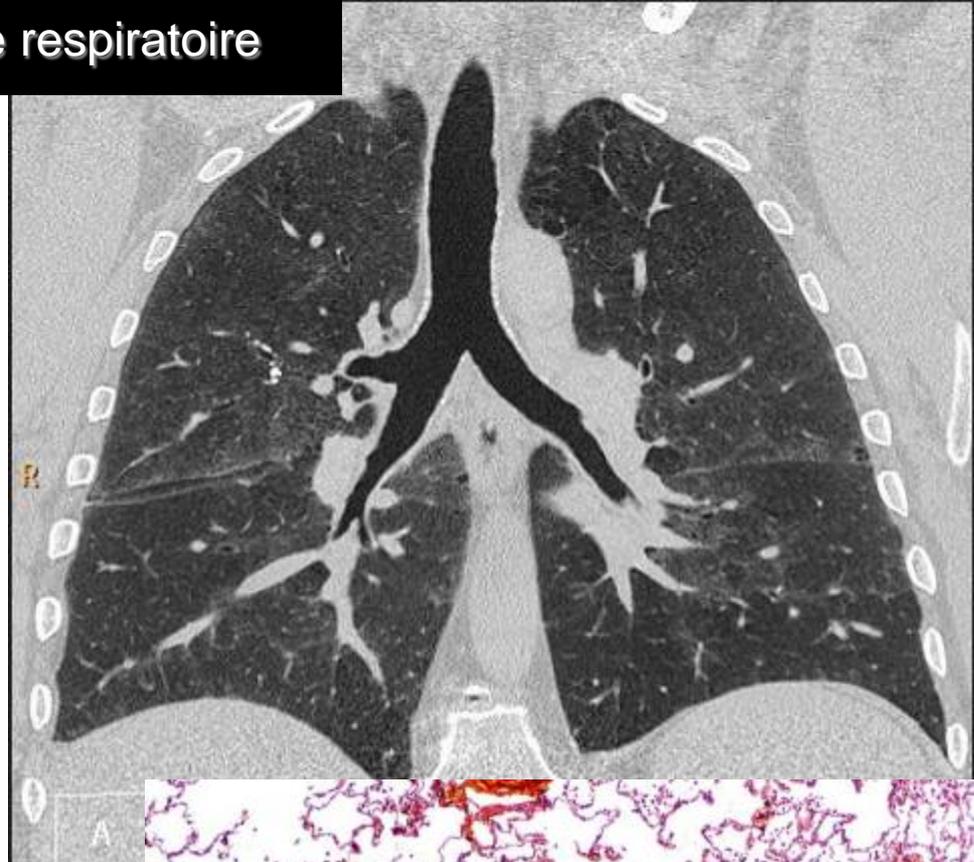
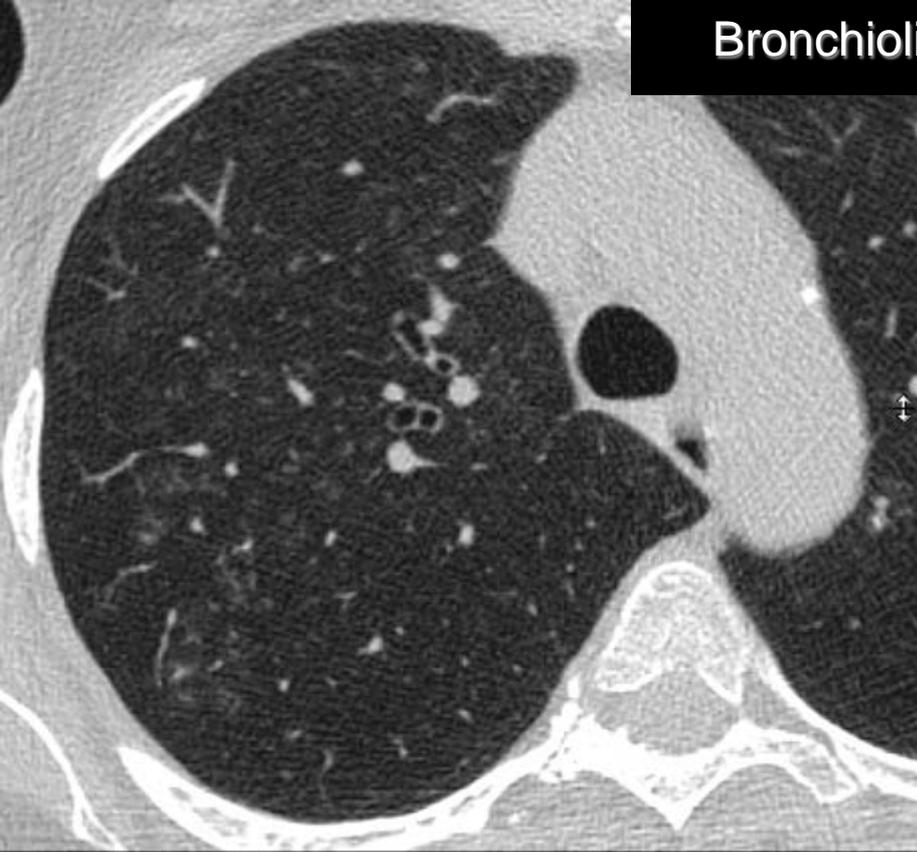
Nodules centrolobulaires

- Limites : nettes régulières / floues



- Densité : tissulaire / verre dépoli

Bronchiolite respiratoire



Pathologie tabagique : EXCES DE MACROPHAGES

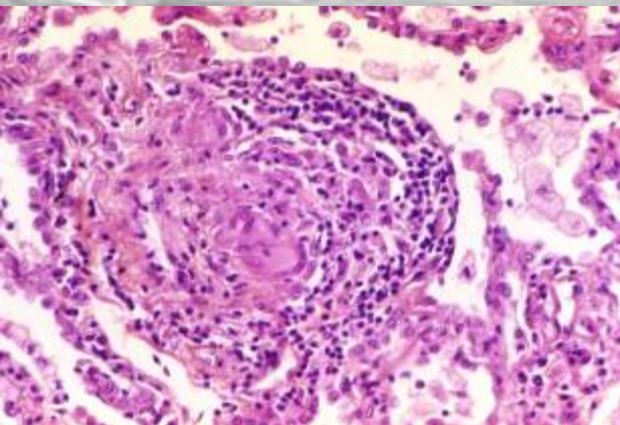
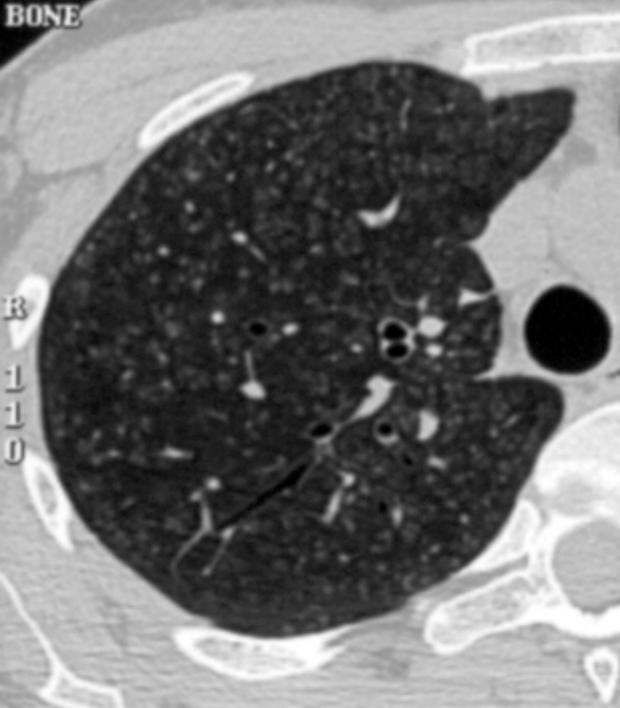
Bronchiolite respiratoire

- inflammation chronique bronchiolaire, macrophages tatoué

Bronchiolite respiratoire avec infiltration pulmonaire diffuse (RB-ILD)

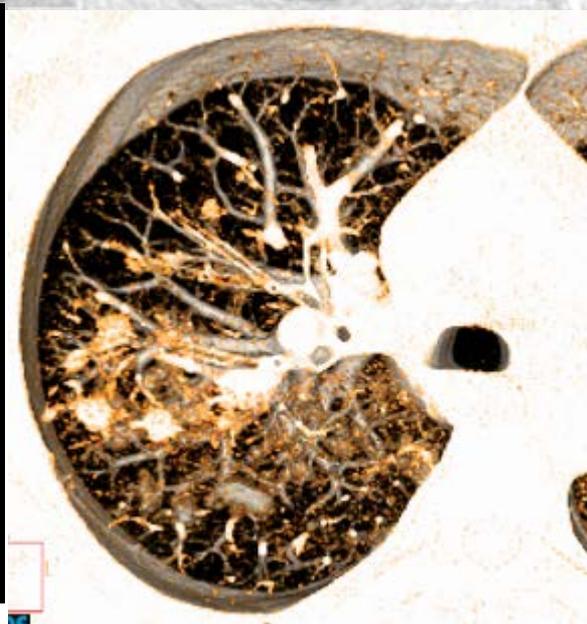
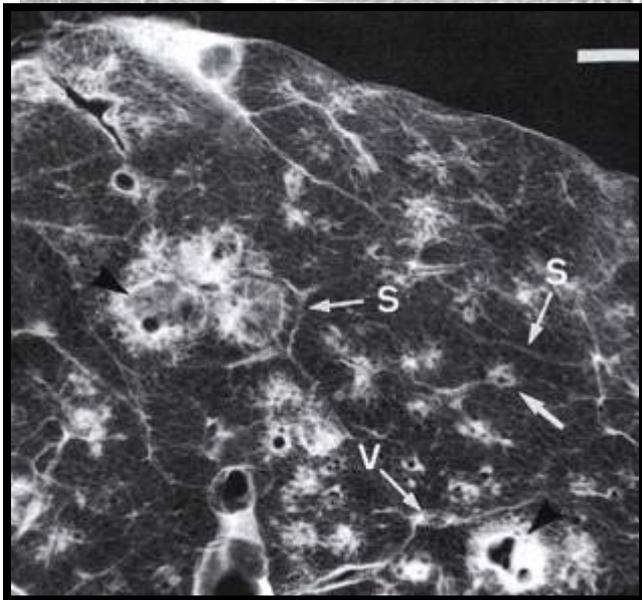
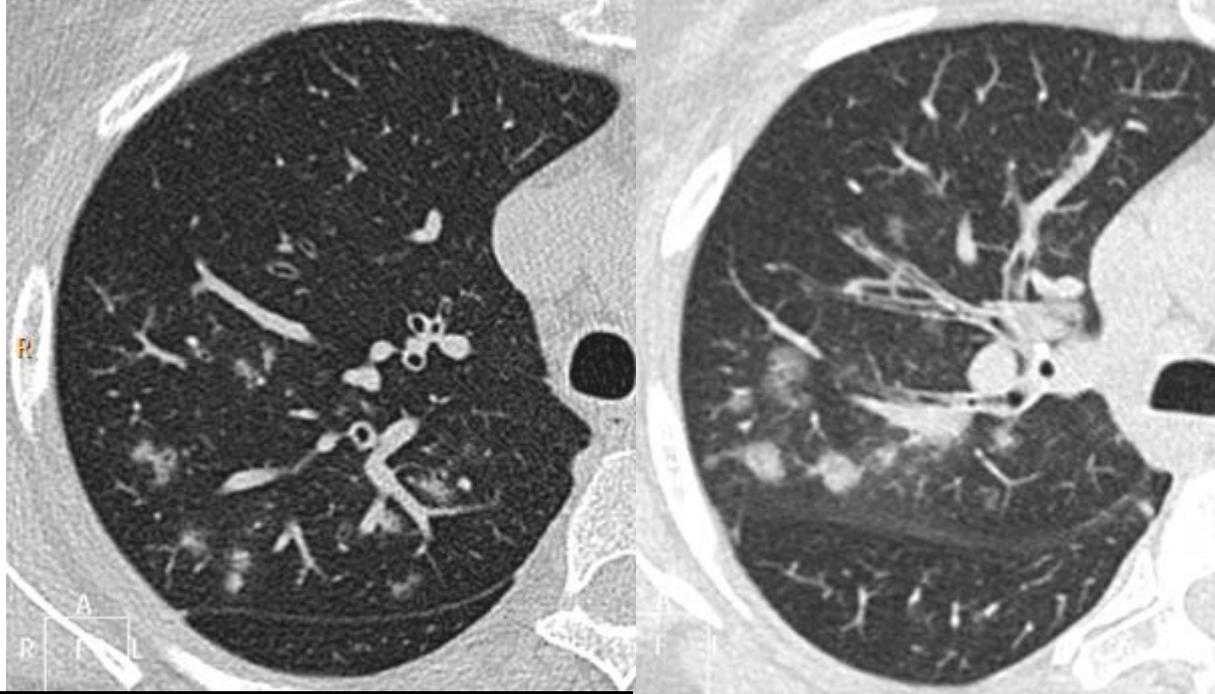
- Extension dans l'espace des anomalies TDM.

Pneumopathie interstitielle desquamative (DIP)



Alvéolite allergique extrinsèque (*forme aiguë*)

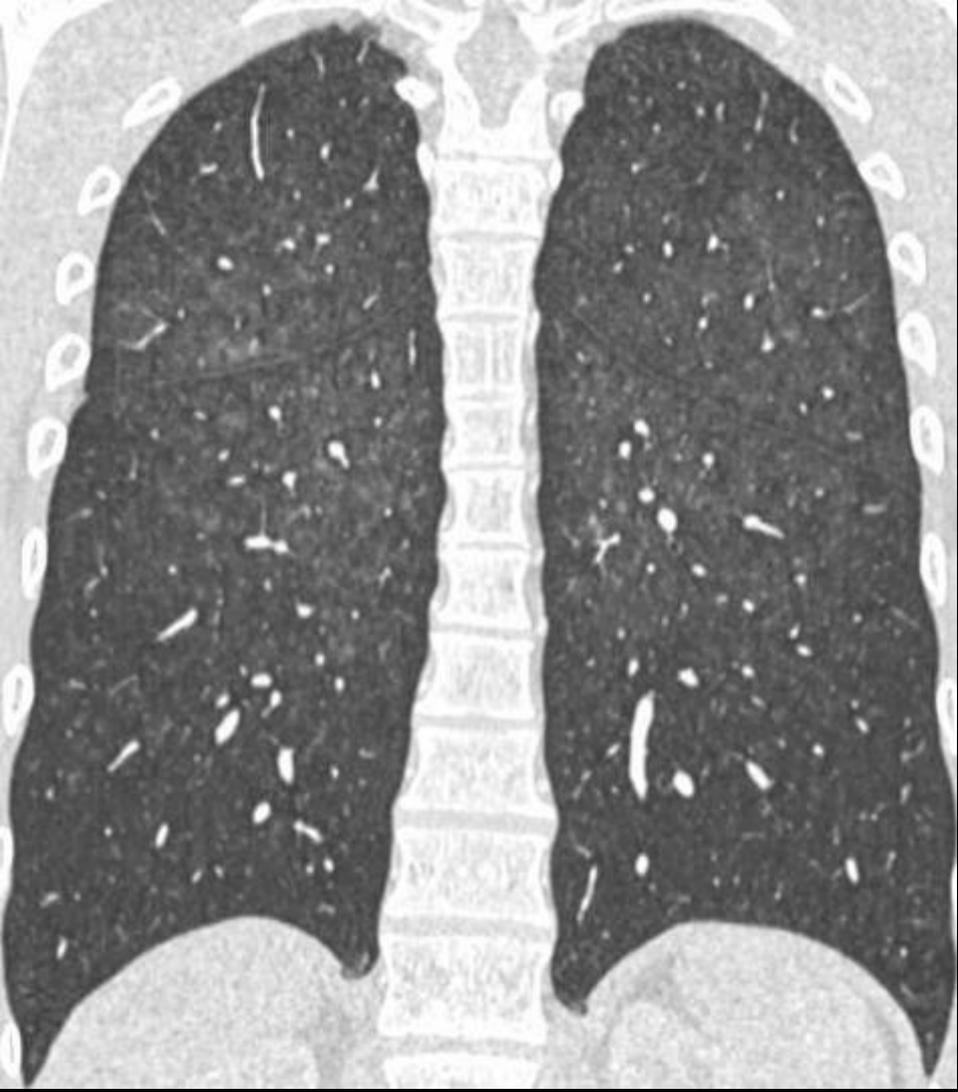
Opacités à contours flous de siège centro-lobulaire.



bronchopneumonie grippale

HX

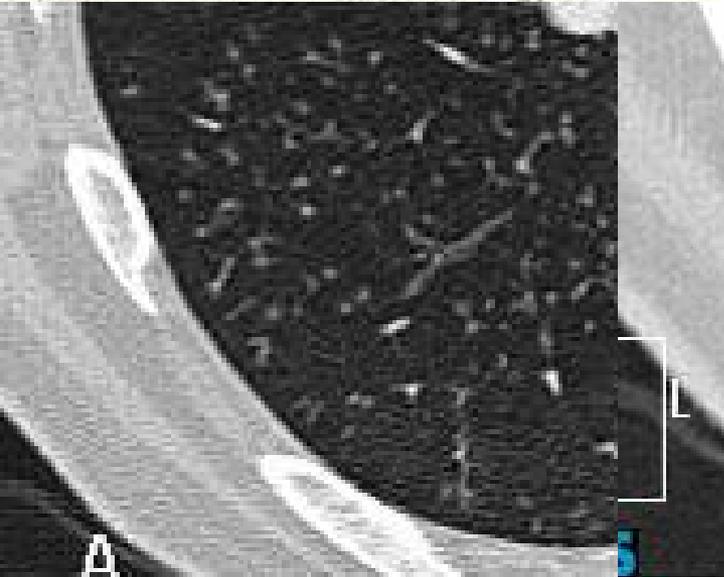


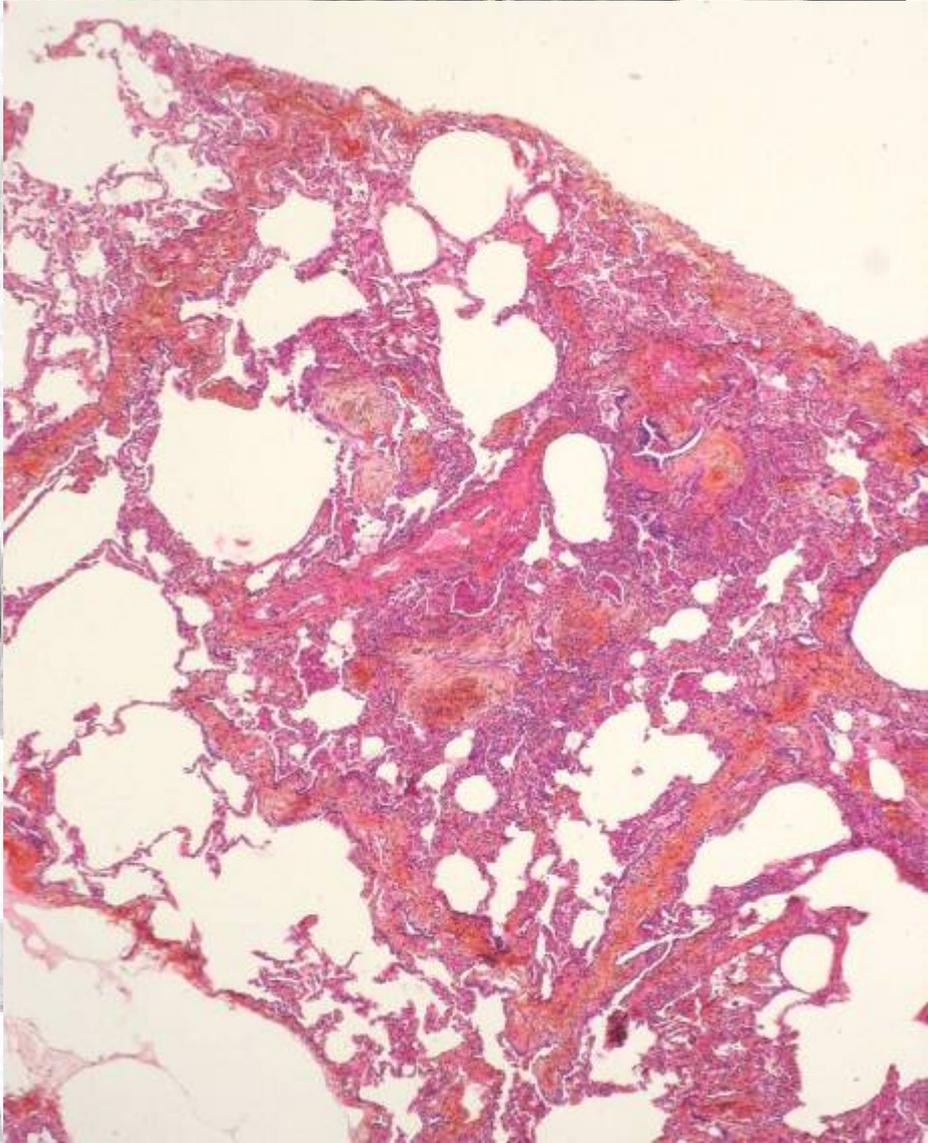
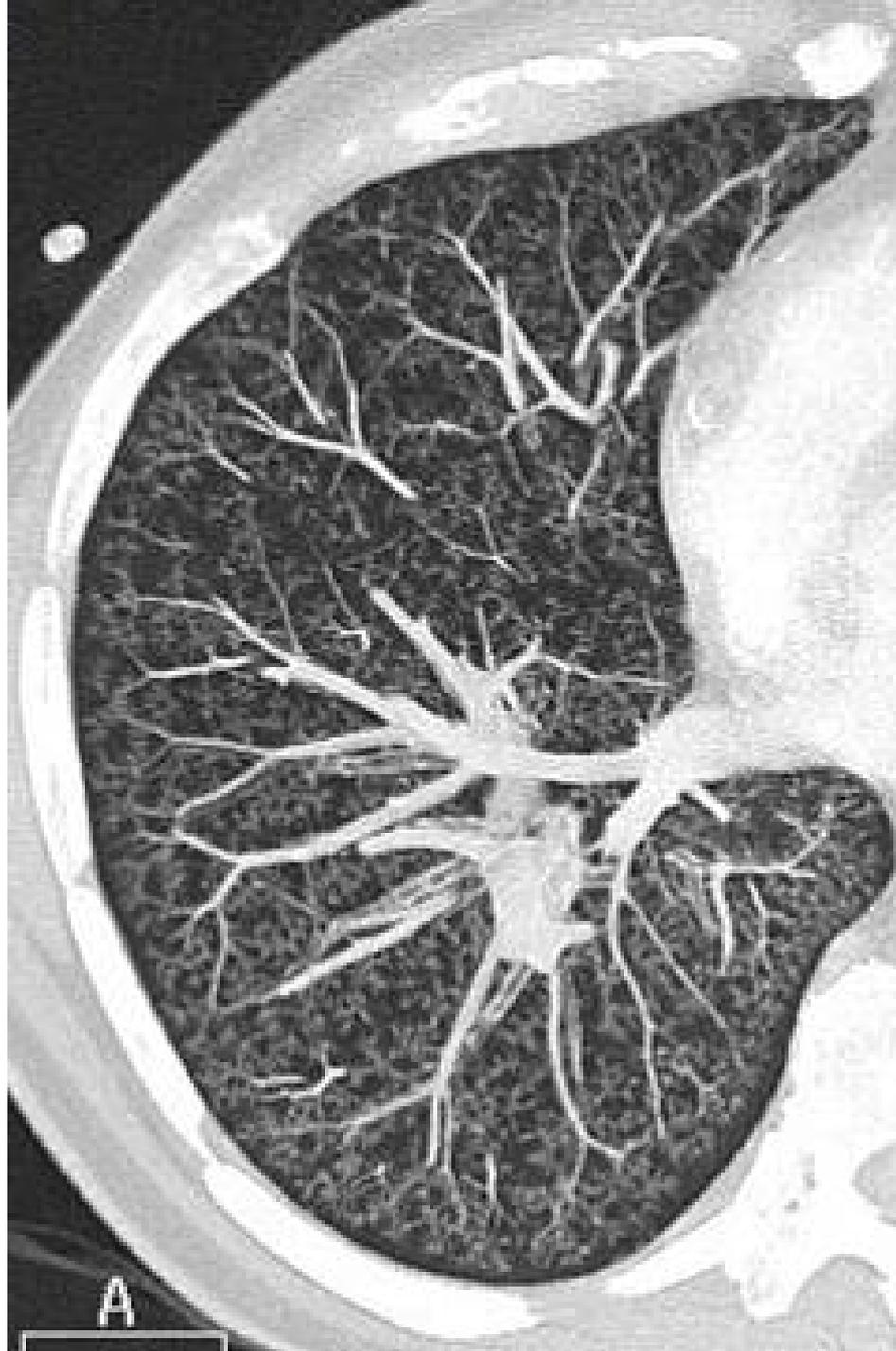


Hémorragie
alvéolaire



Hémosidérose





A

Micronodulation

Sous pleurale ?

oui

non

Péri
lymphatique

Diffuse

centrolobulaire

Arbre en
bourgeons

Sans AeB

Sarcoïdose
Lymphangite

Miliaire
Hématogène
(BK, Tumorale, candida)

BK, MBA
Muco

AAE,
RB-ILB, oedème

Les Images kystiques et hypodensités

Kystes à parois fines (fréquent)

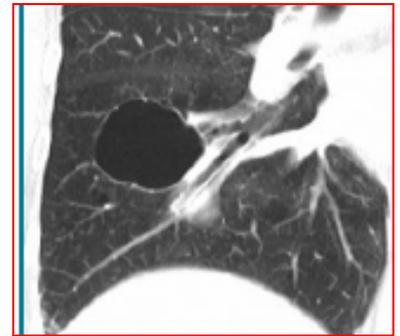
- Abscès
- Cancer bronchique
- Bulle, bleb
- Bronchectasies kystiques
- Infections fongiques (coccidiomycose, aspergillose)
- Rayon de miel
- Métastases
- Pneumatocèles
- Pneumocystose
- Tuberculose

Kystes à parois fines (peu fréquent)

- Amylose
- Behcet
- Kyste
- Malformation kystique adenomatoïde
- Mucoviscidose
- Histiocytose Langerhansienne
- Hernie diaphragmatique
- Kyste hydatique
- Hydropneumothorax, fistule bronchopleurale
- Pseudotumeur inflammatoire
- Lymphome
- Mélanoïdose
- Papillomatose
- Paragonemiase
- Plombage
- Périartérite noueuse, Lupus
- Infarctus pulmonaire
- Séquestation intralobaire
- Polyarthrite rhumatoïde
- Sarcoidose
- Poumon traumatique (lacération, hématome)
- Wegener

Fleischner Society: Glossary of Terms for Thoracic Imaging¹

- Kyste: espace de densité aérique circonscrit par une paroi habituellement fine, de qq mm à plusieurs cm
- Cavité: espace de densité aérique d'épaisseur variable dans une condensation, une masse ou un nodule



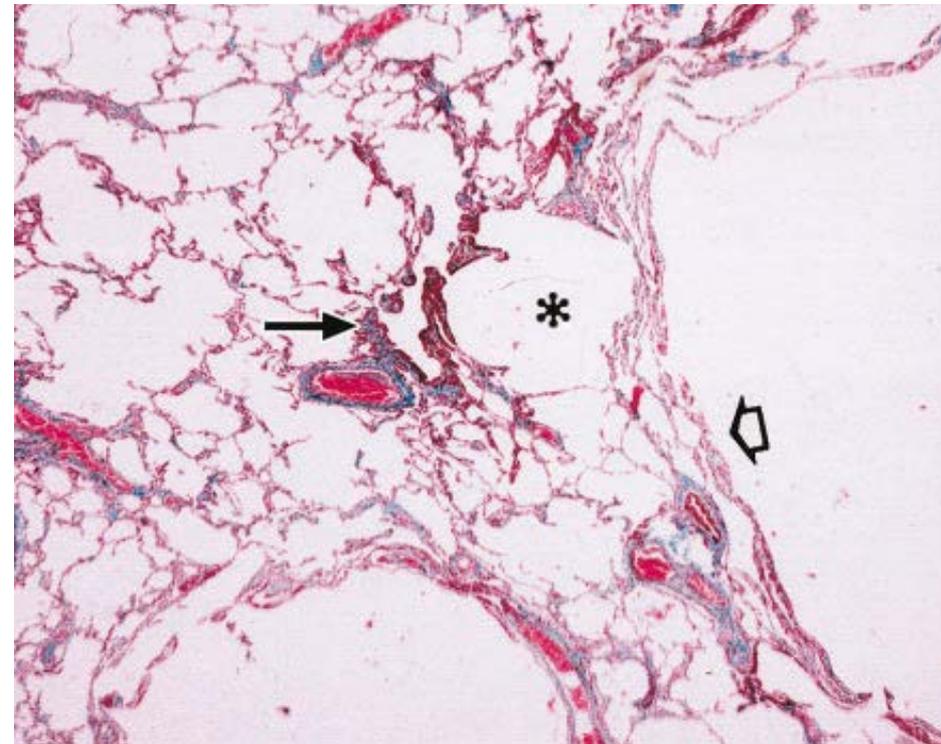
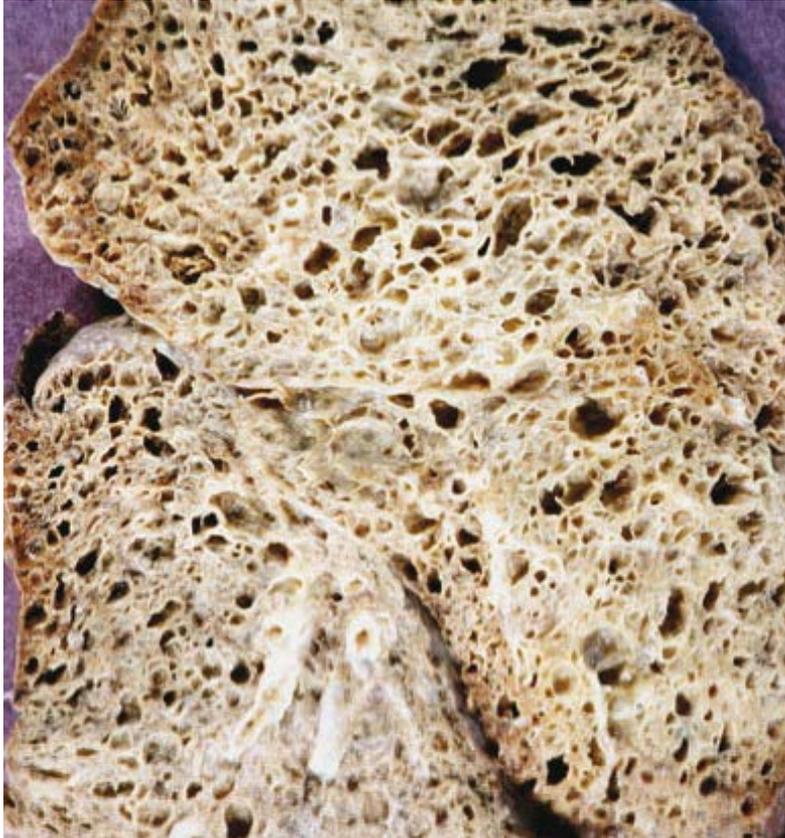
Images kystiques et hypodensités

- Kystes multiples à parois fines
- Kystes multiples à parois épaisses
- Cavités
 - Contexte de maladie systémique
 - Contexte infectieux
 - Contexte tumoral
 - Contexte traumatique
 - Voies aériennes
 - Congénital
- Hypodensités sans parois
 - Emphysème, bulles
 - Aspect en mosaïque et piégeage
- Rayon de miel

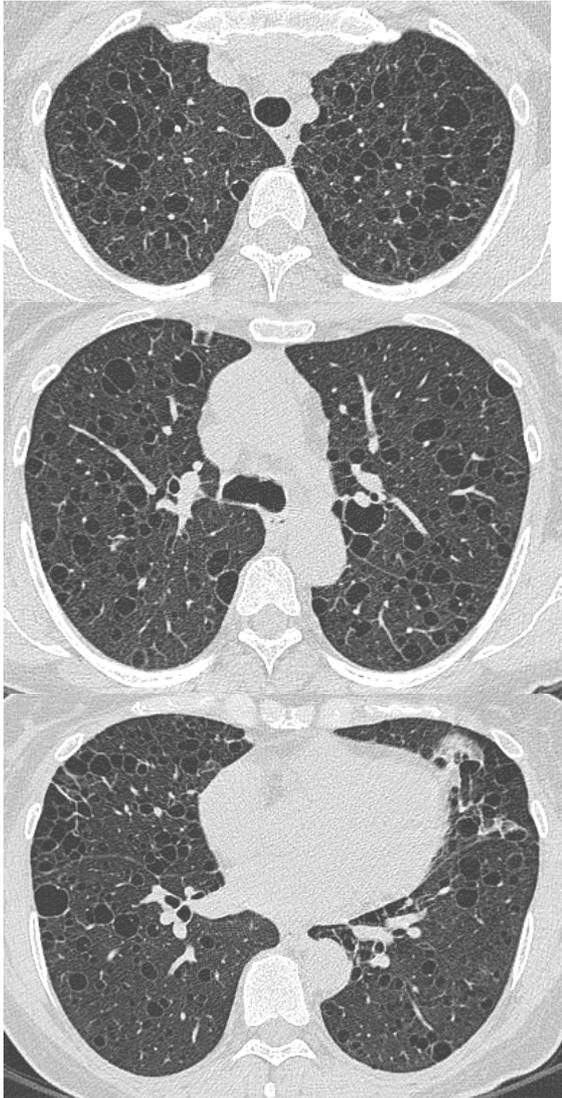
Etiologies des images kystiques

- Kystes multiples à parois fines
 - Lymphangioliomyomatose
 - Birtt-Hogg-Dubé
 - LIP
 - Amylose
- Kystes à parois fines et/ou épaisses
 - Histiocytose Langherhansienne
 - Pneumocystose (*Pneumocystis jirovecii*)

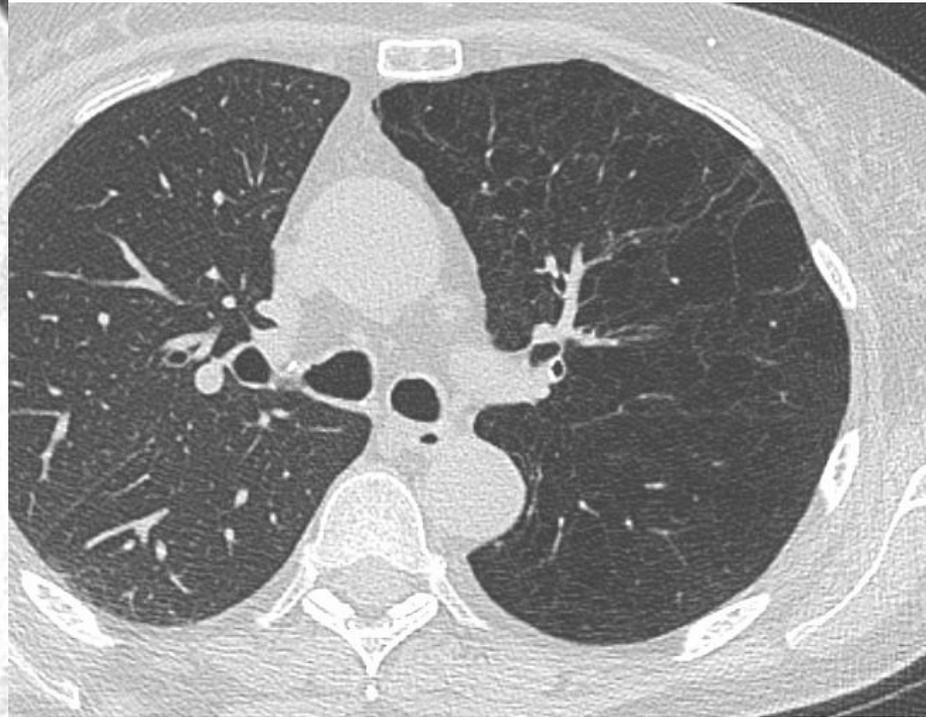
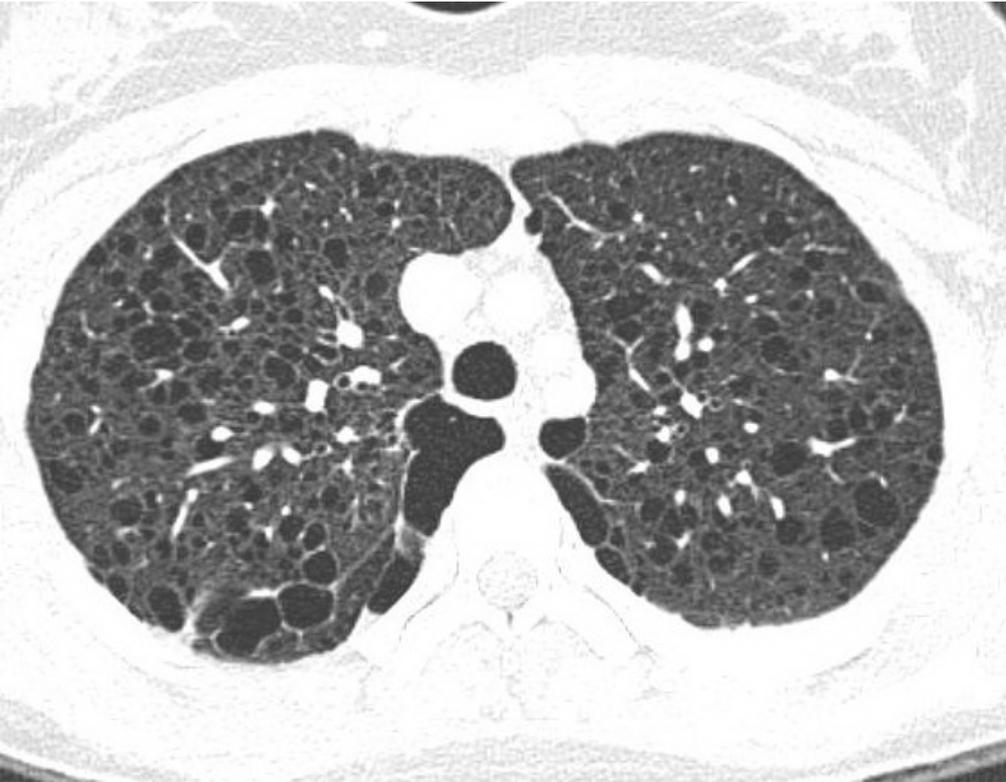
Lymphangiomyomatose (LAM)



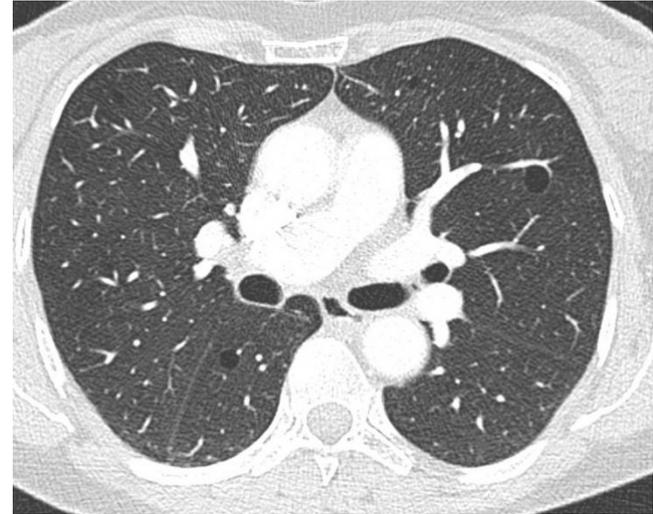
Lymphangioliomyomatose (LAM)



Lymphangioliomyomatose (LAM)

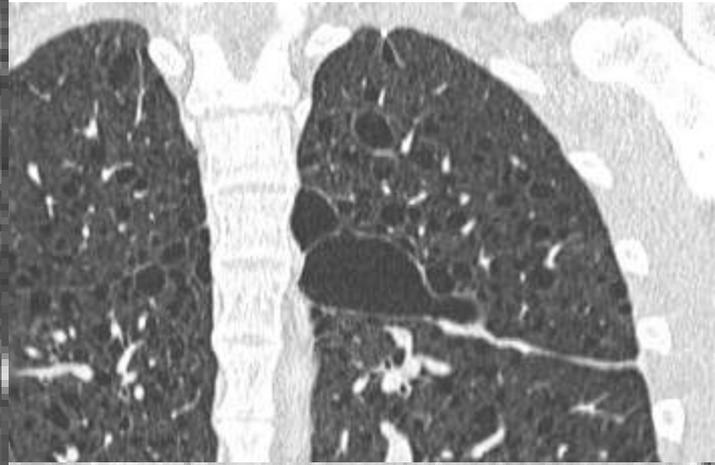
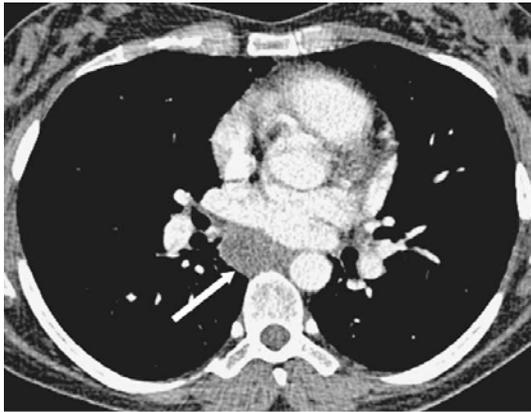


Lymphangiomeiomyomatose (LAM) Formes tardives



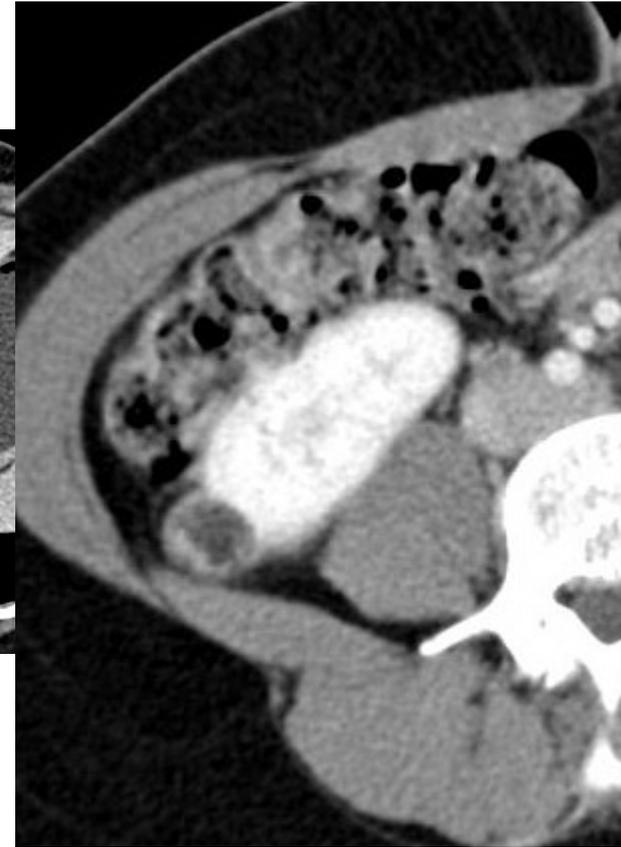
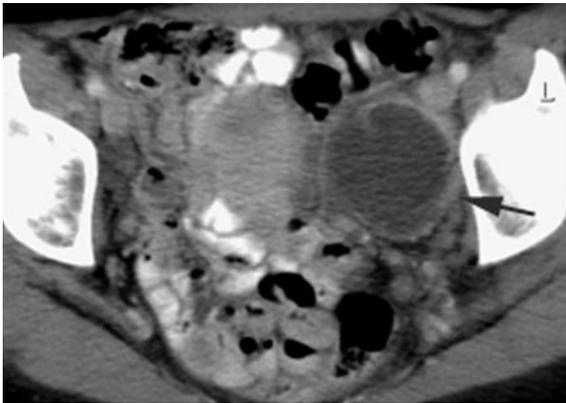
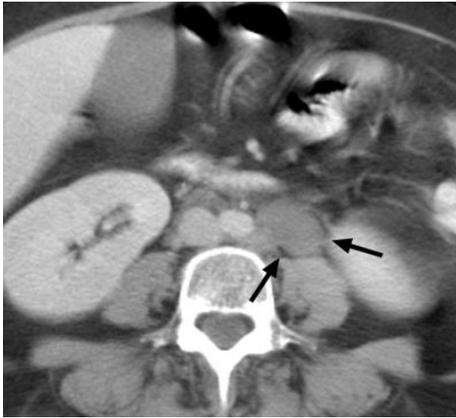
- Forme de découverte fortuite
- Femme après la ménopause
- Pas d'atteinte abdominale
- Bon pronostic
- Risque de pnx, hémoptysie, pleurésie

Lymphangiomeiomyomatose (LAM)

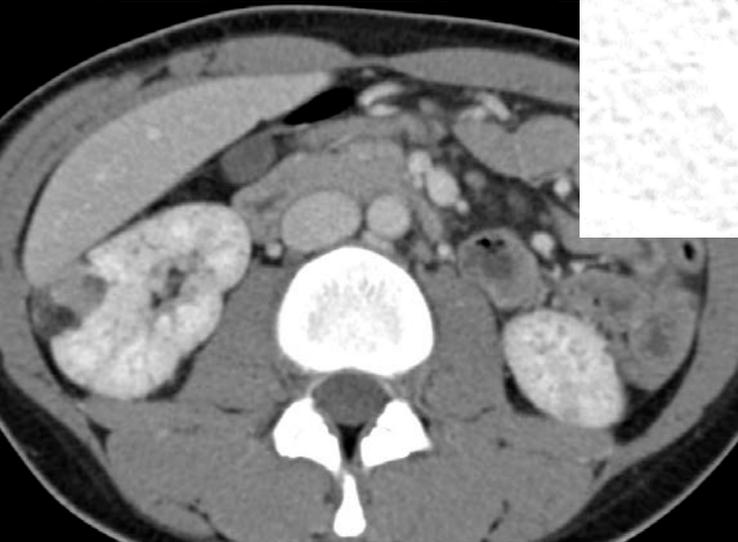
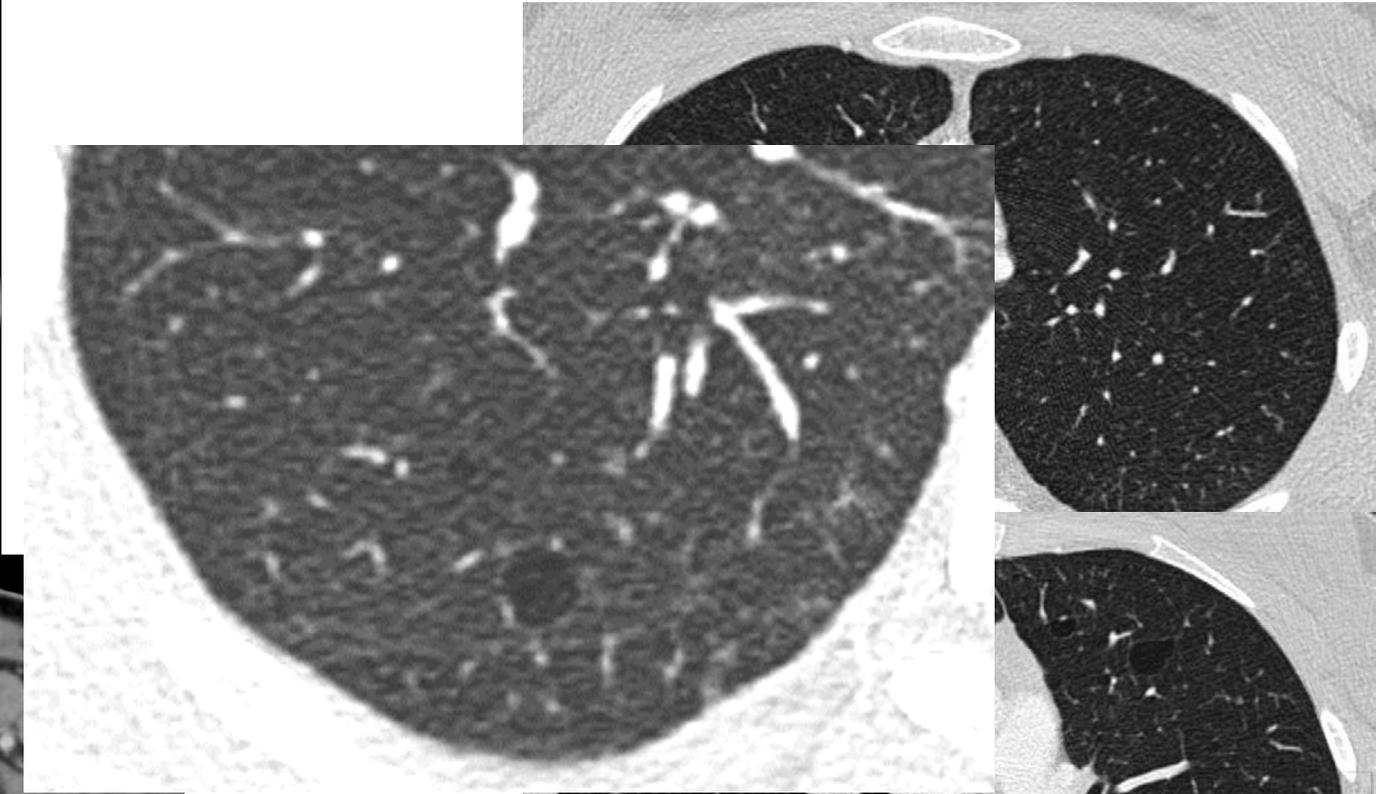


Canal thoracique
chylothorax

Lymphangiomyomatose an° abdominales

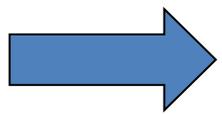


Sclérose tubéreuse et LAM



Histiocytose Langerhansienne

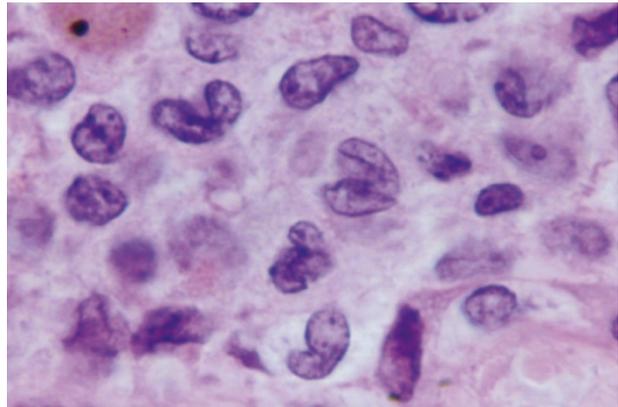
- Prolifération d'histiocytes CD1+ (cellules de Langerhans) formant des granulomes.
- Développement au niveau des parois des bronchioles



Bronchioectasies/cavitation

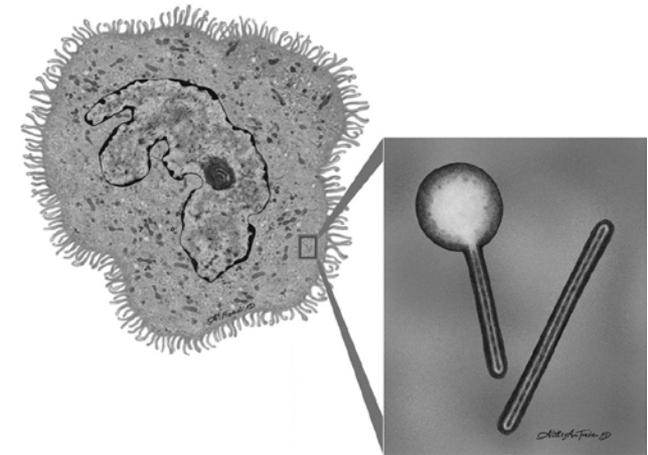
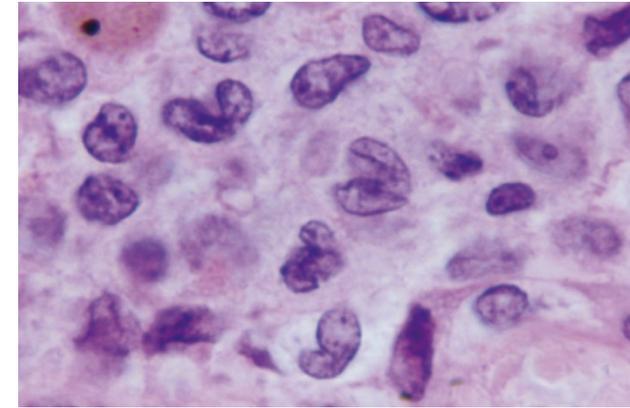
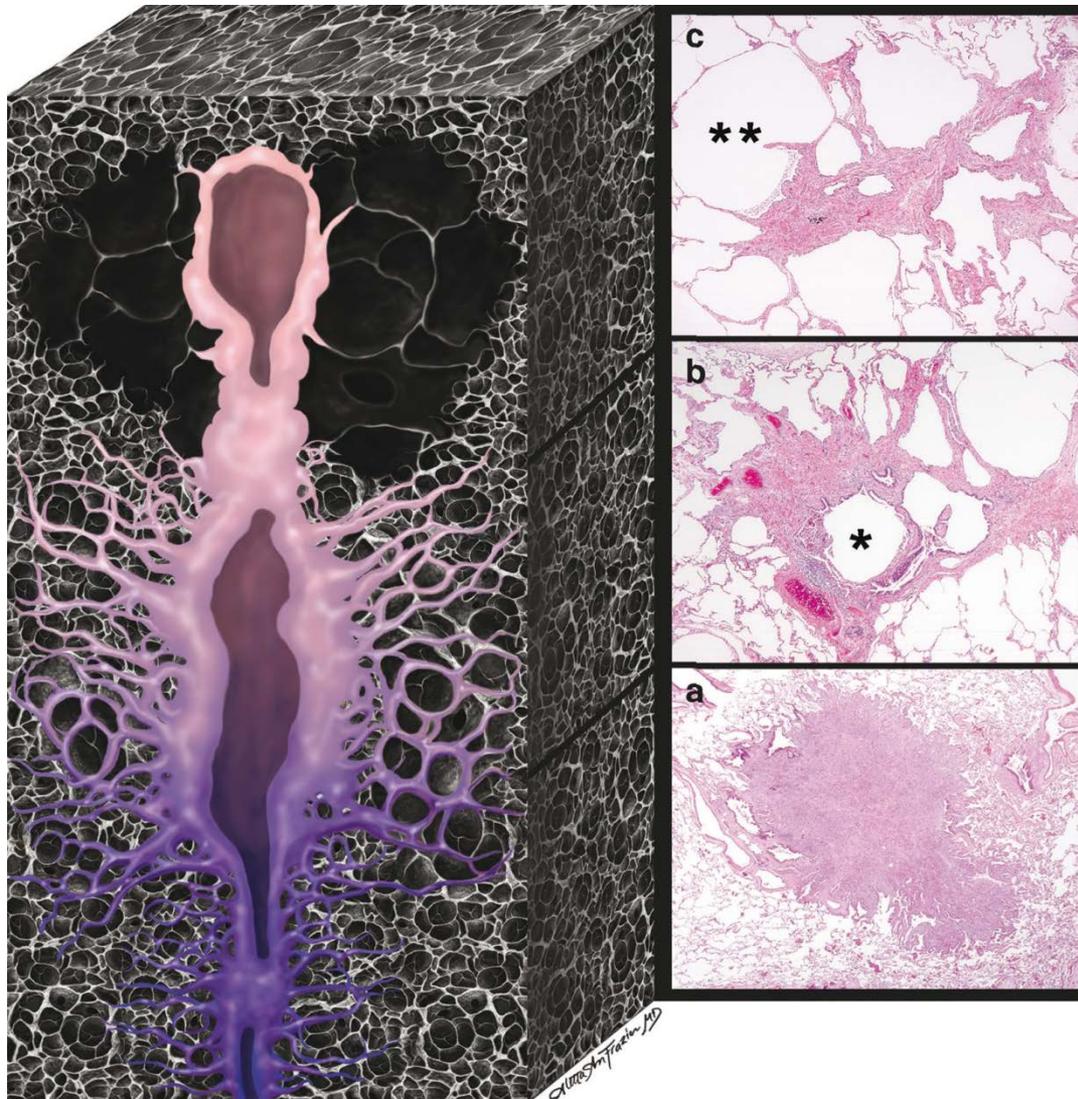


Kystes



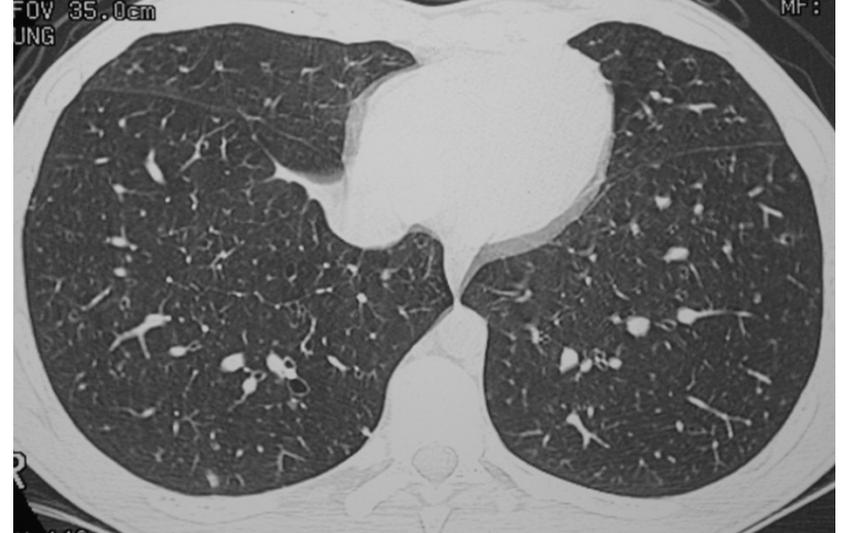
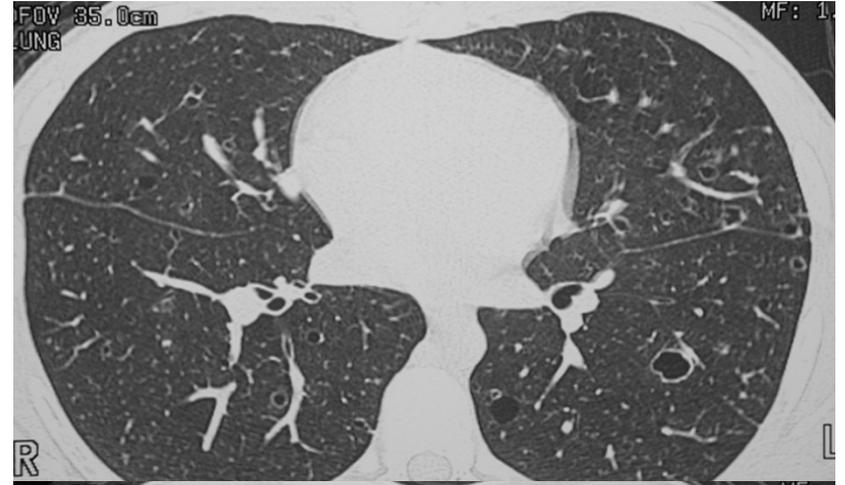
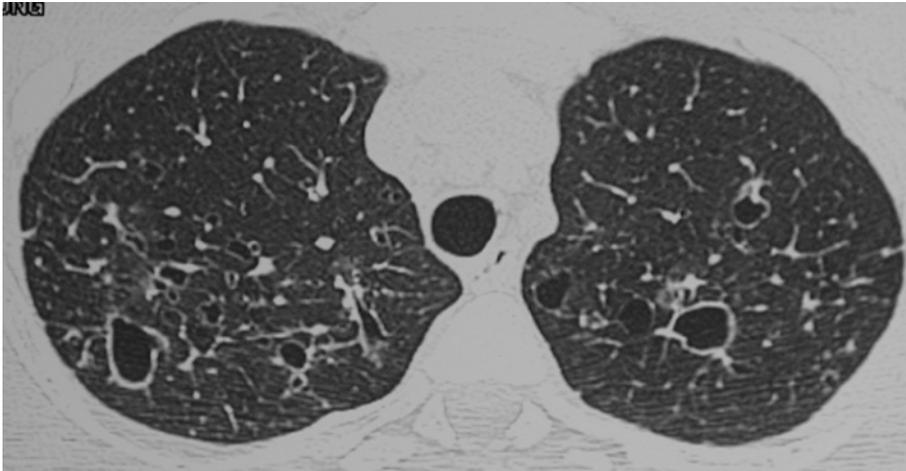
90 % des patients atteints sont fumeurs. Association fréquente avec l'emphysème ou des lésions de fibrose

Histiocytose Langerhansienne

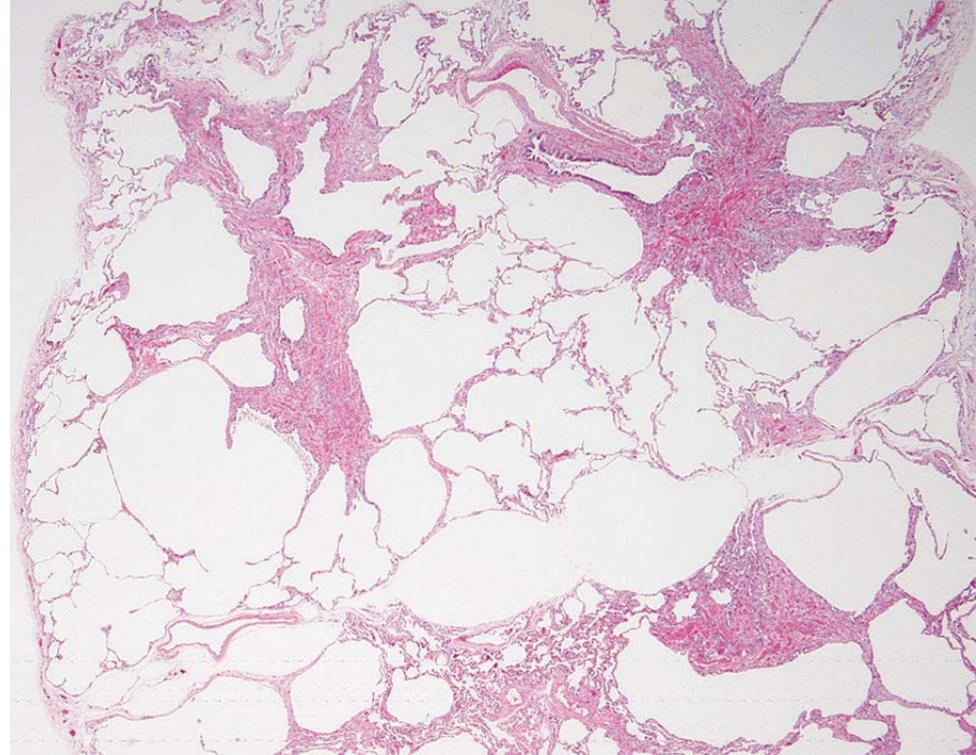
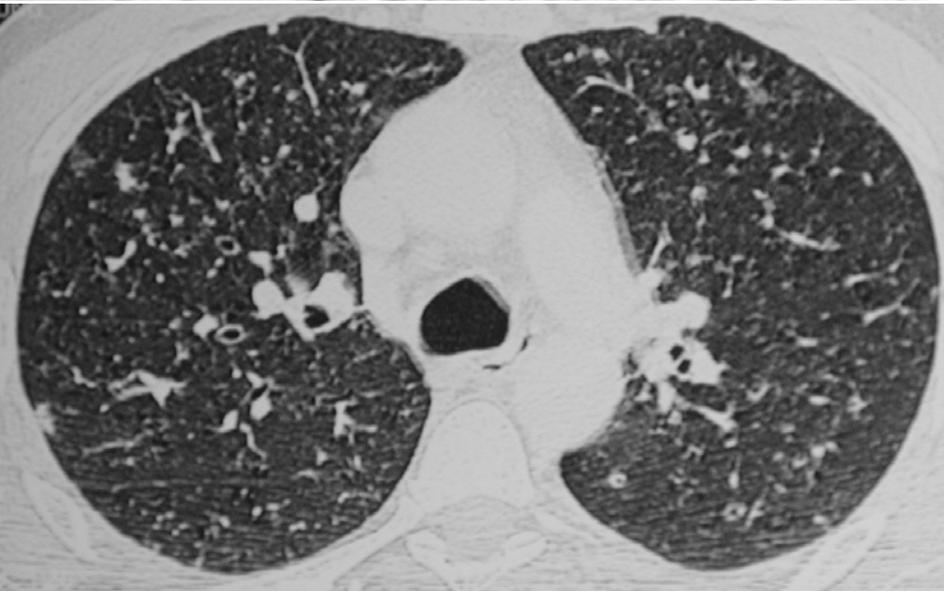
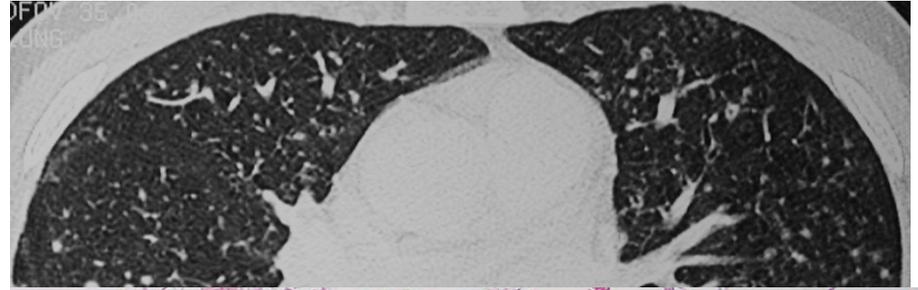
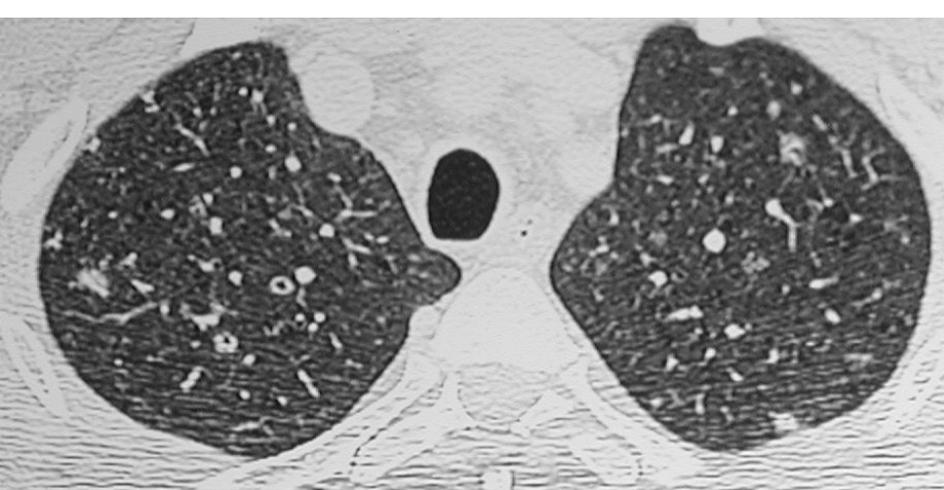


Abbot Radiographics 2004

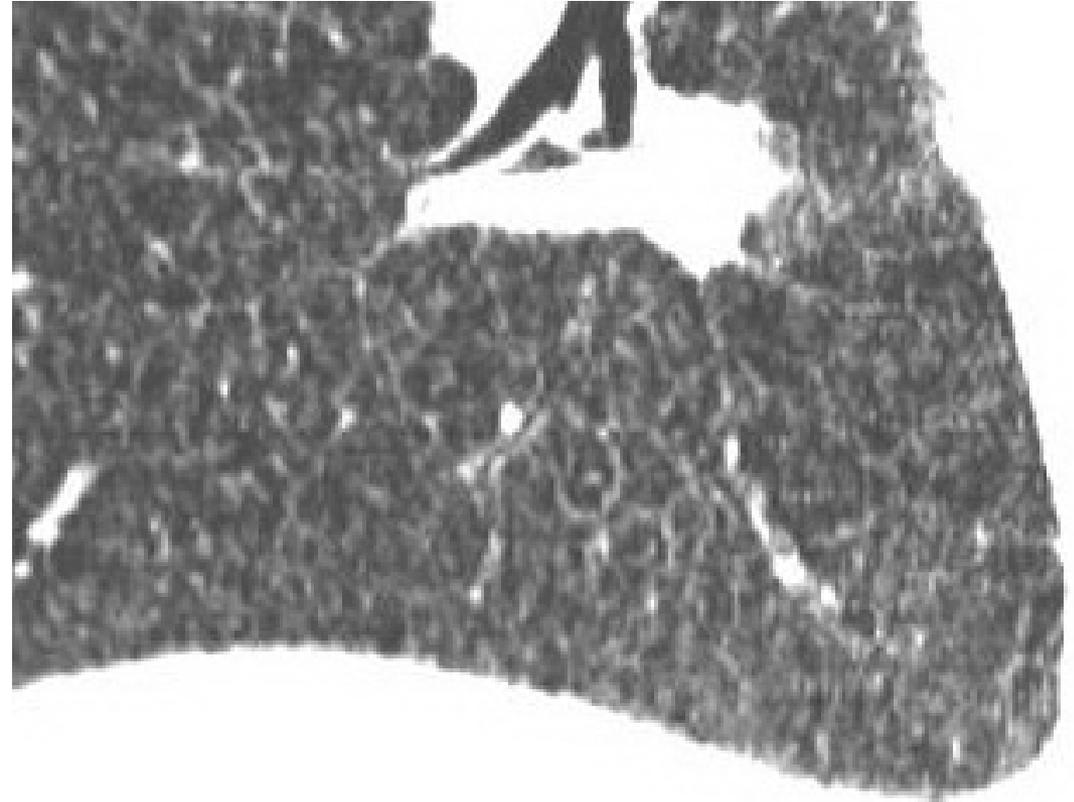
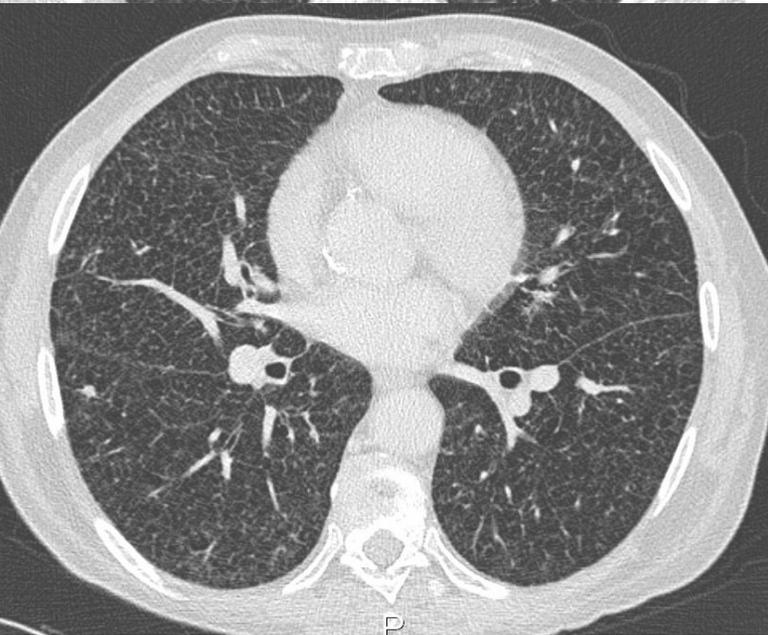
Histiocytose Langerhansienne



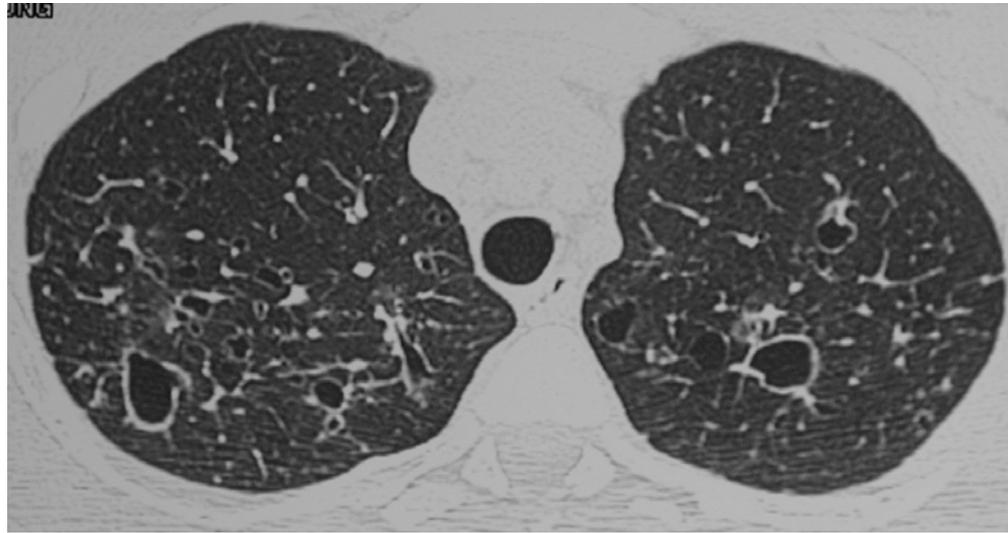
Histiocytose Langerhansienne



Histiocytose Langerhansienne

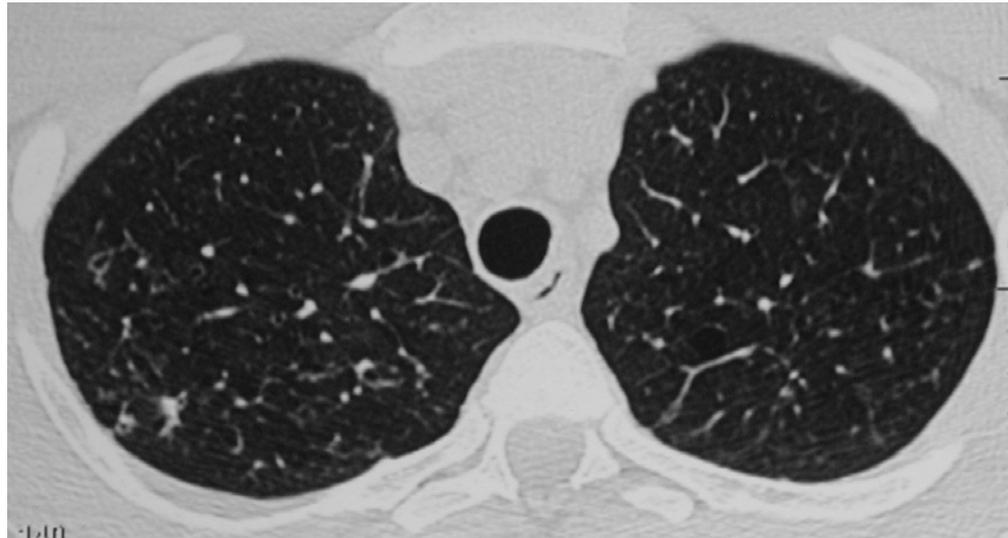


Histiocytose Langerhansienne



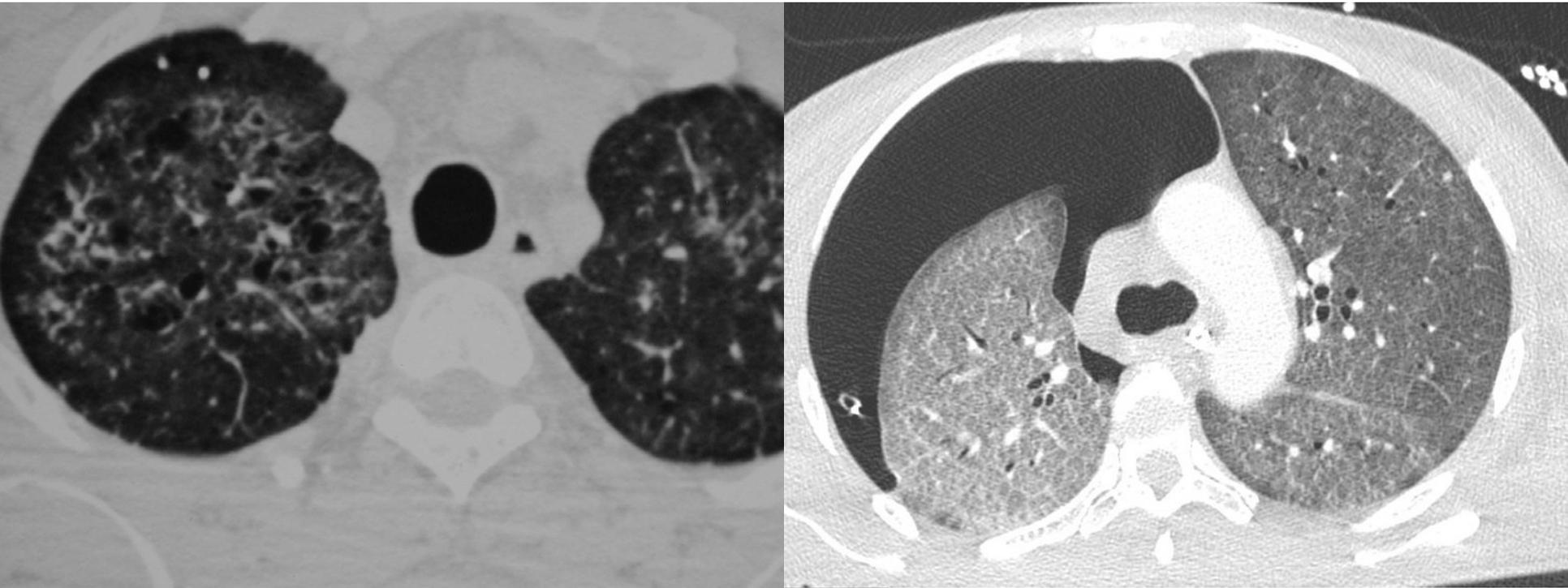
T0

ARRET
DU
TABAC



T1

Pneumocystose (pneumocystis jirovecii)



Images kystiques et hypodensités

- Kystes multiples à parois fines
- Kystes multiples à parois épaisses
- Cavités
 - Contexte de maladie systémique
 - Contexte infectieux
 - Contexte tumoral
 - Contexte traumatique
 - Voies aériennes
 - Congénital
- Hypodensités sans parois
 - Emphysème, bulles
 - Aspect en mosaïque et piégeage
- Rayon de miel

Cavités dans un contexte de maladie systémique

- Sarcoidose
- Wegener
- Polyarthrite rhumatoïde
- Sy de Sjögren

Sarcoidose

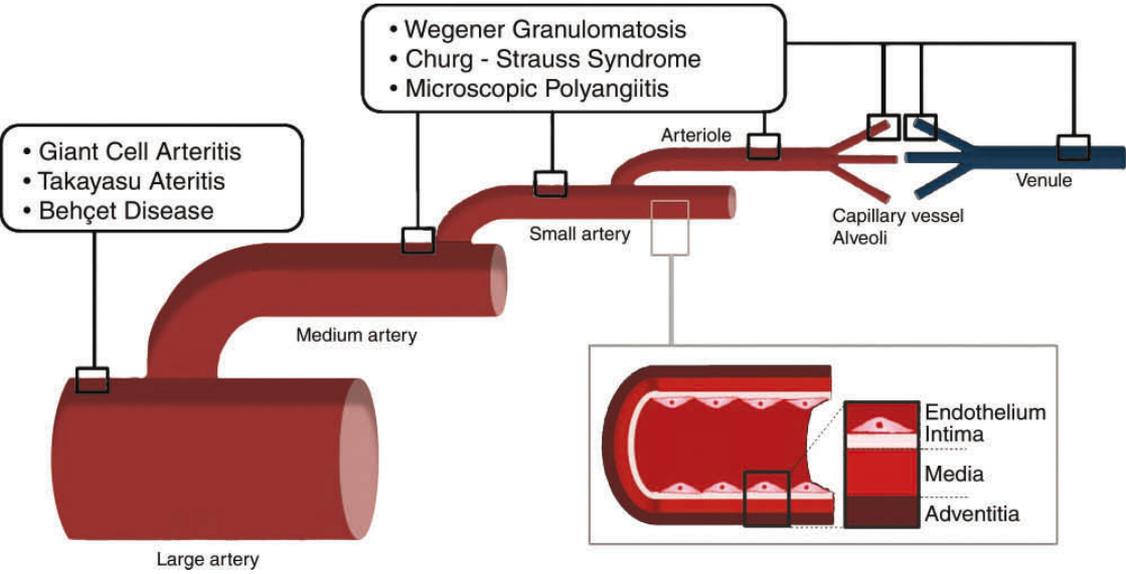


Polyarthrite rhumatoïde

- Nodules macrobiotiques
 - 0,5 à 5 cm
 - Prévalence 22% en HRCT
 - Risque de PNX
 - Sy de Caplan: nodules de PR chez mineur



Granulomatose avec polyangéite: Maladie de Wegener

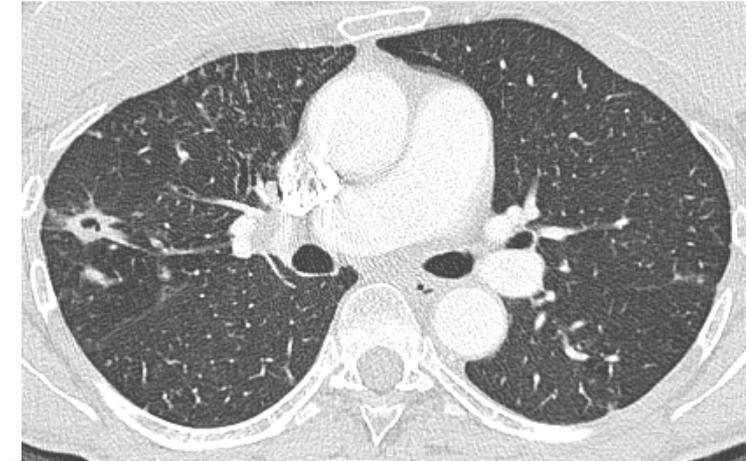
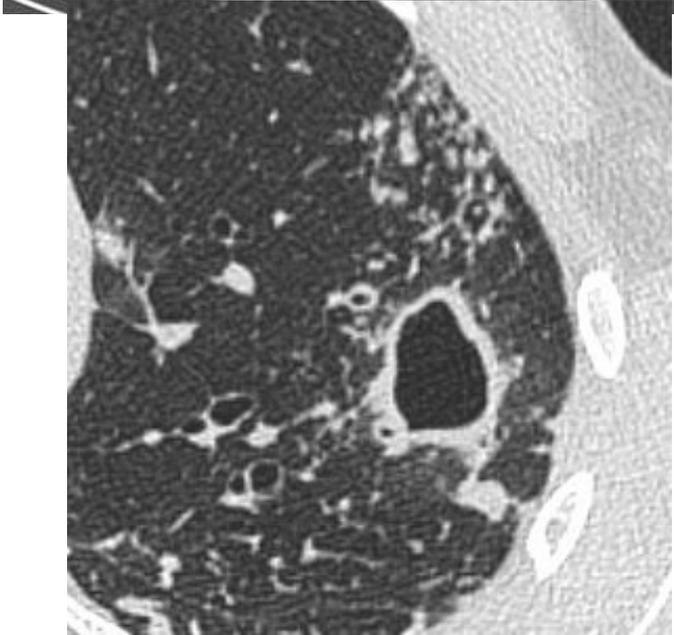


Cavités: Infections ou tumeurs?

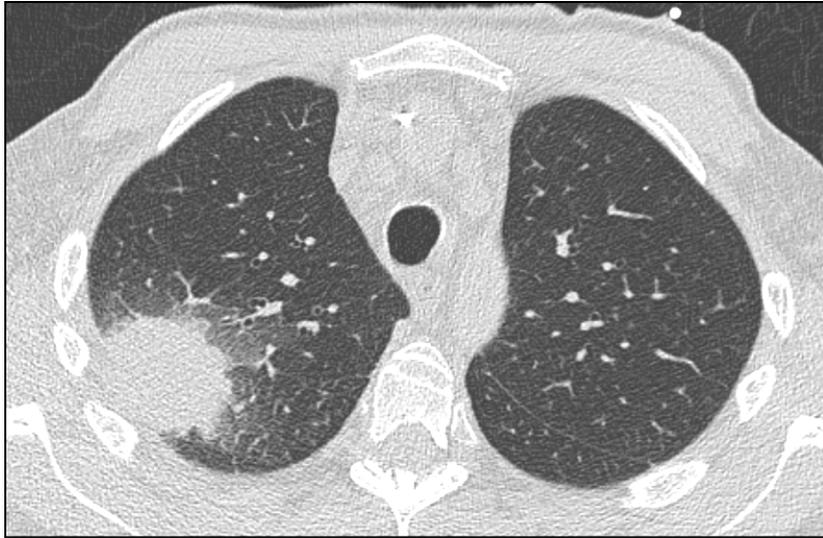
- Infections
 - Tuberculose
 - Abscès
 - Aspergillose invasive
 - Nocardiose
 - Coccidiomycose
- Tumeurs
 - Primitives pulmonaires
 - Secondaires
 - Lymphomes (récidives)



Cavités infectieuses



Aspergillose invasive



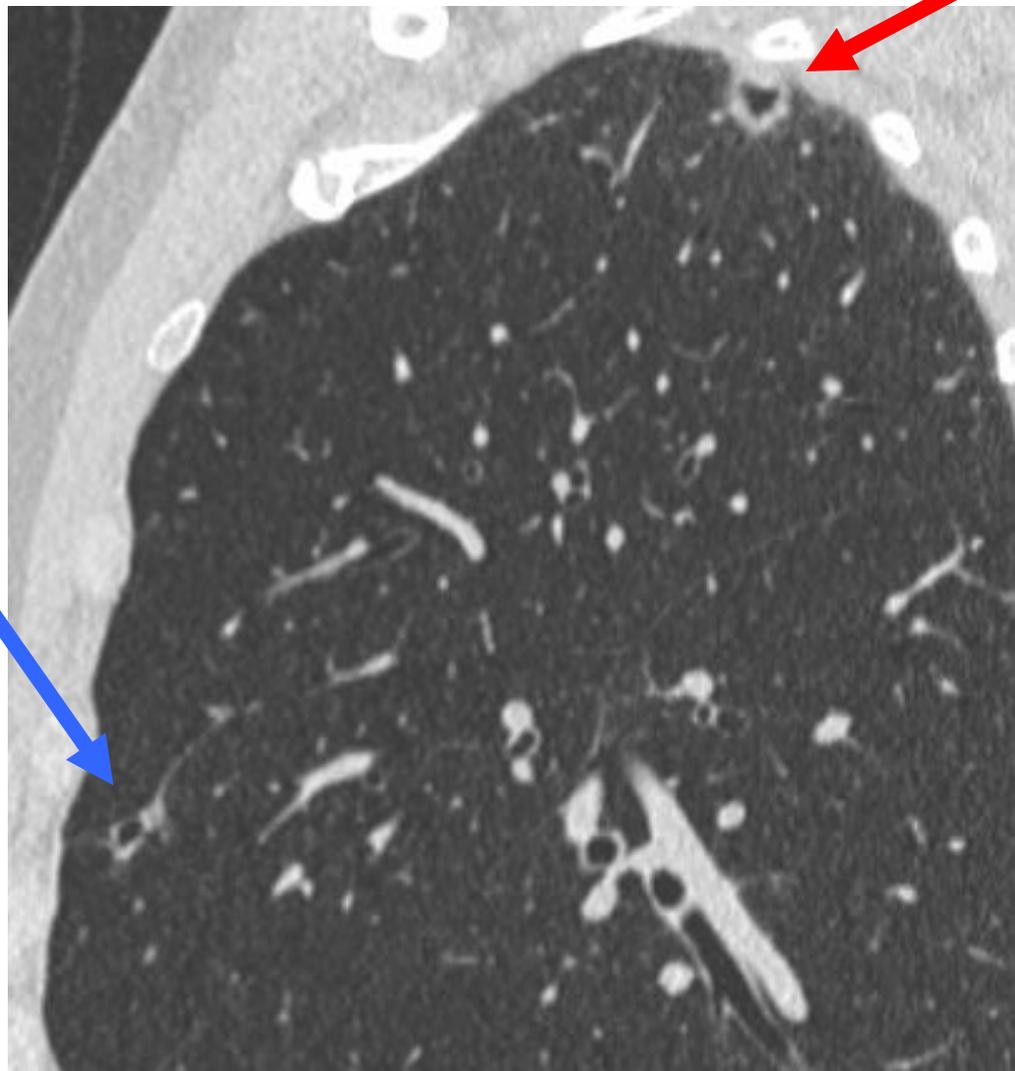
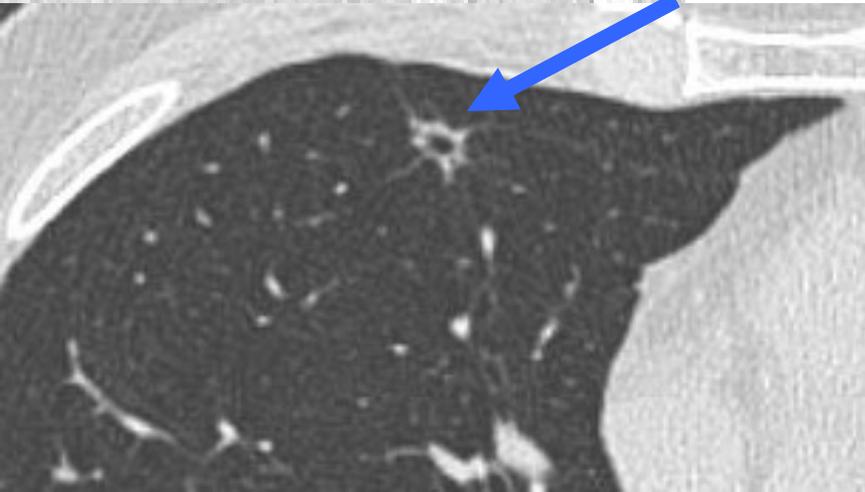
à J15



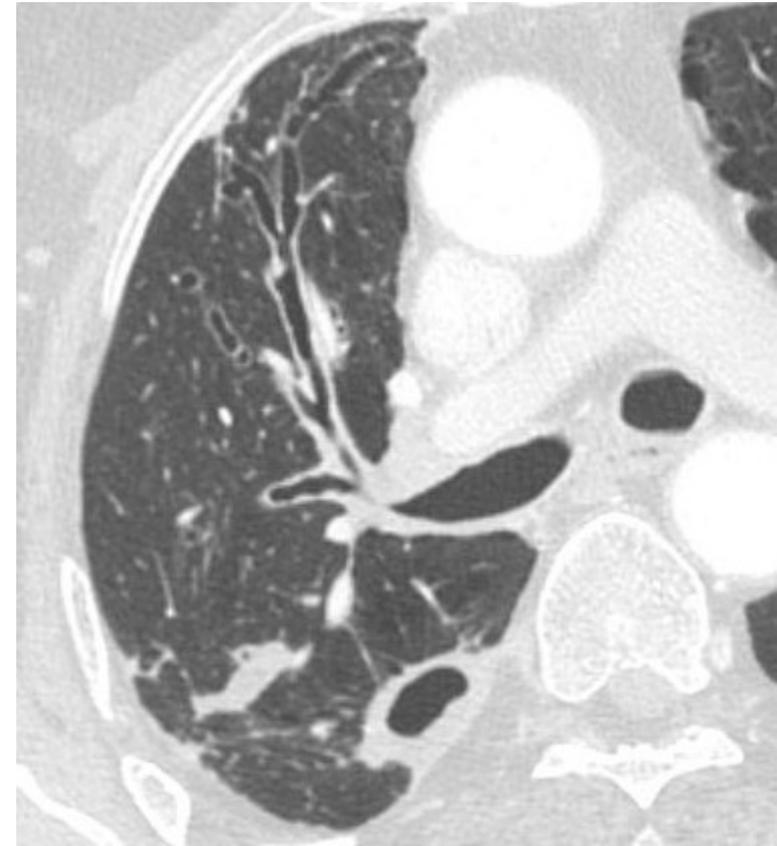
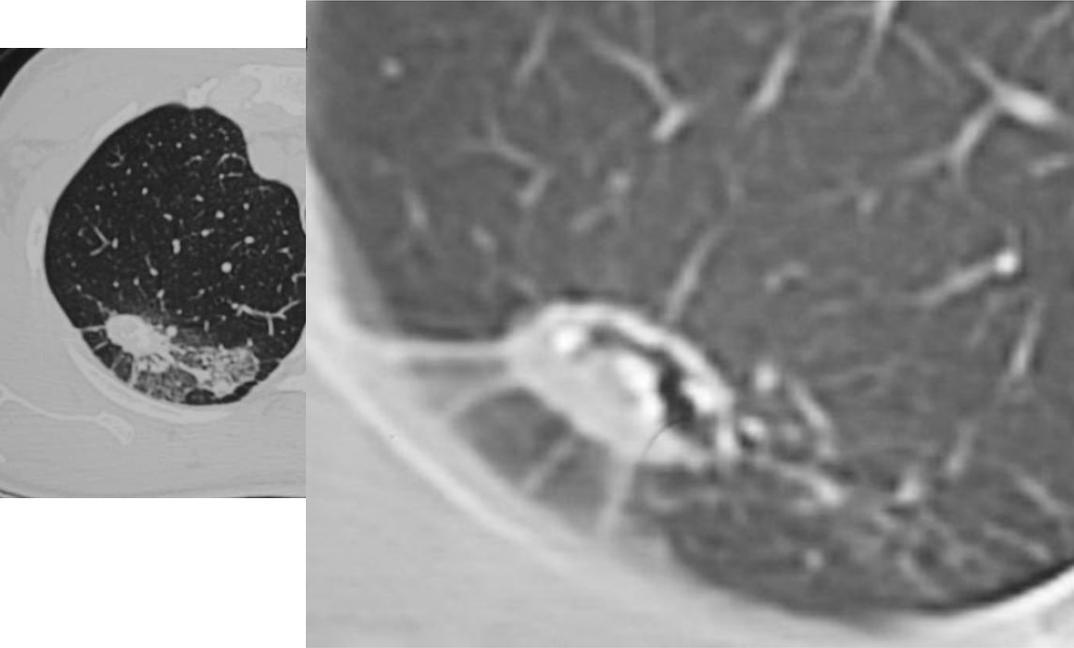
à J30

Tuberculose

Adenocarcinome
tuberculose



Mycobacteriose atypique

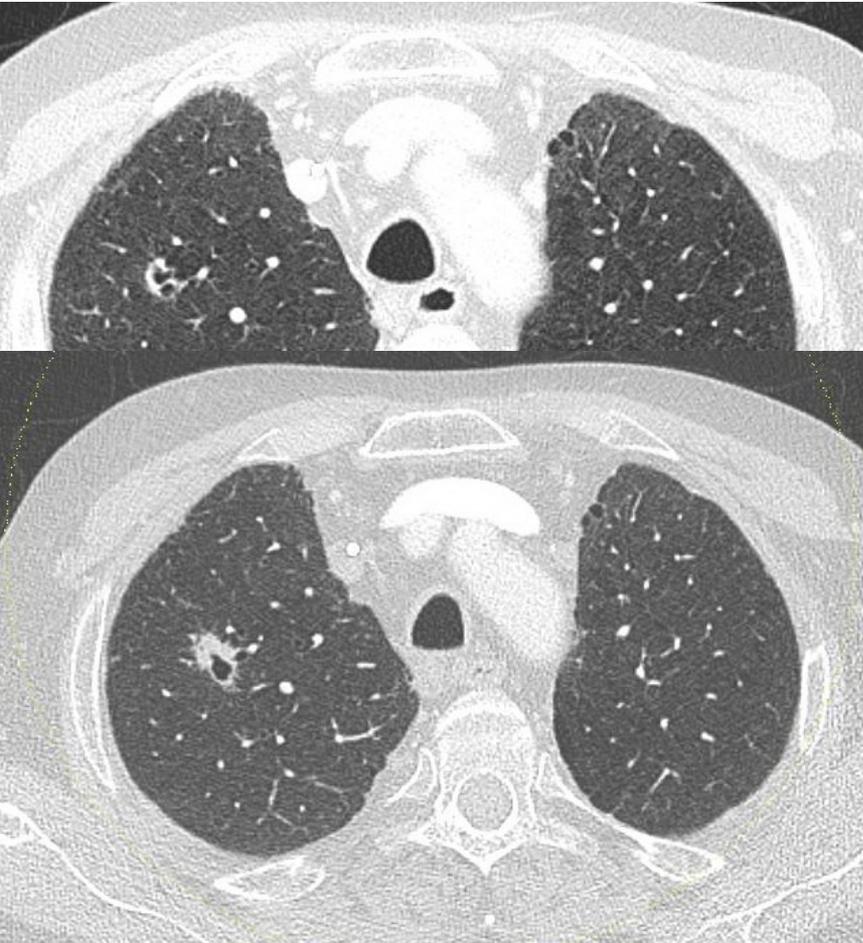


- Femme de 25 ans NF
- nodule connu depuis 5 ans
- hémoptysie

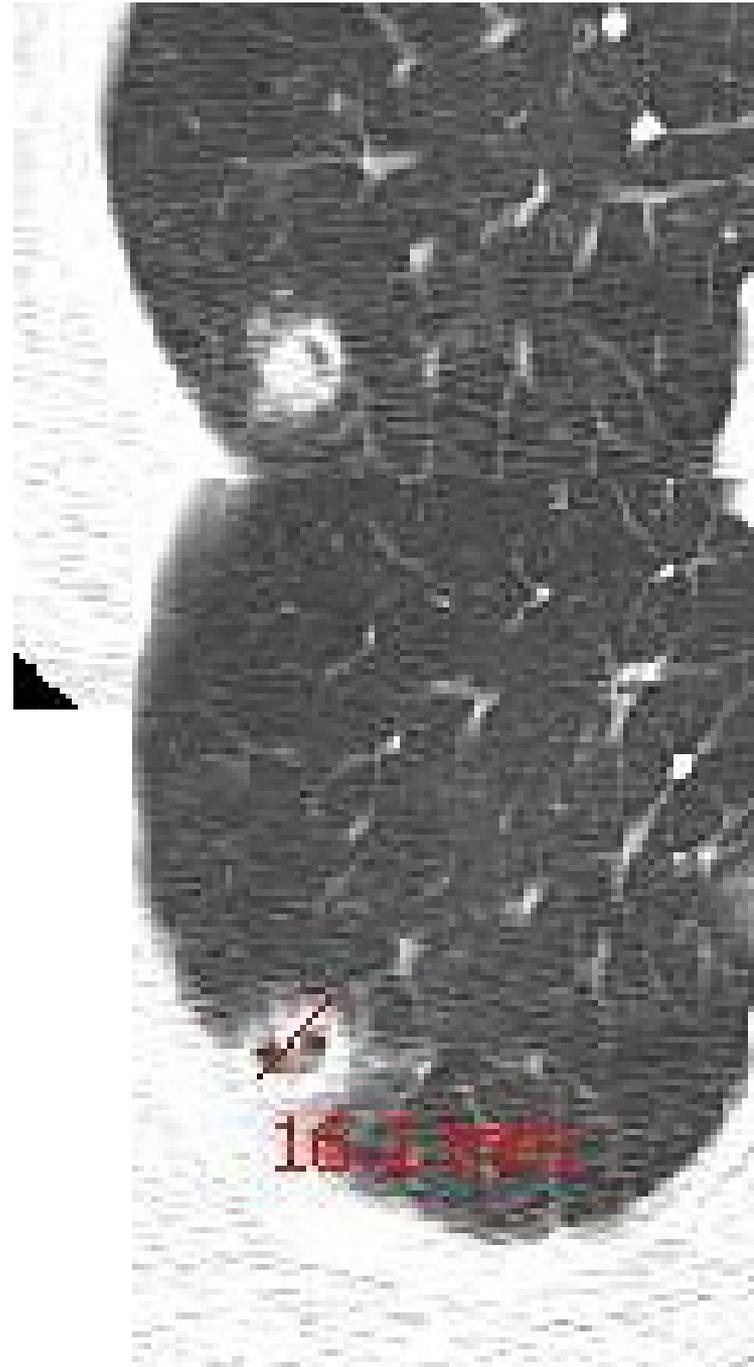
Mycobacterium Kansasii
Dg sur cultures ECBC
Confirmation par chirurgien

Cavités et tumeurs

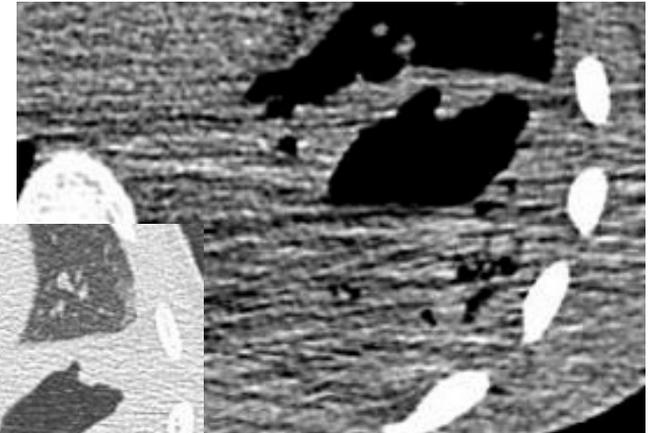
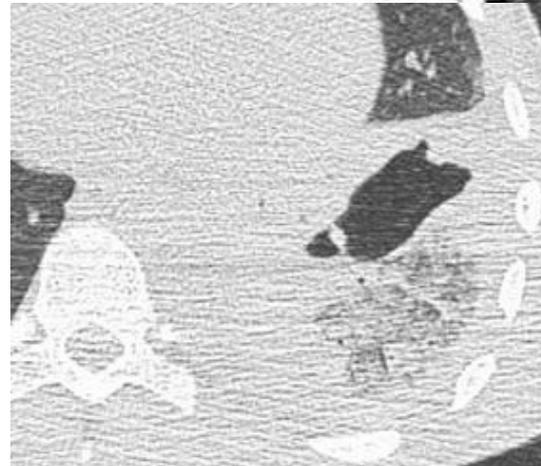
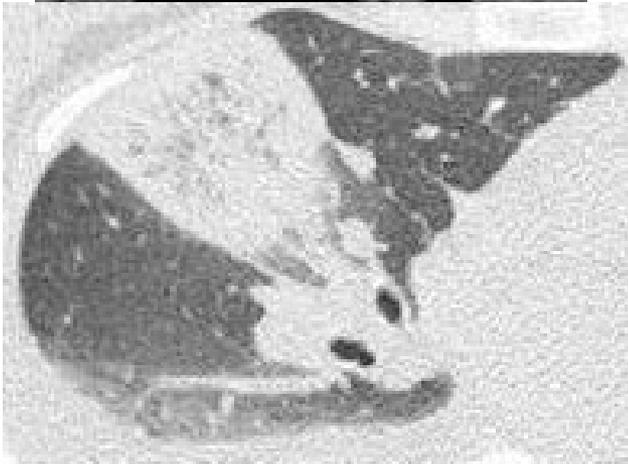
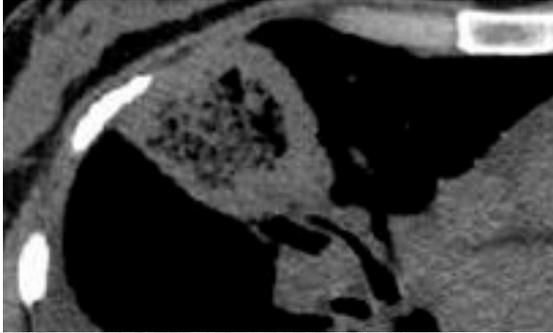
adenocarcinome



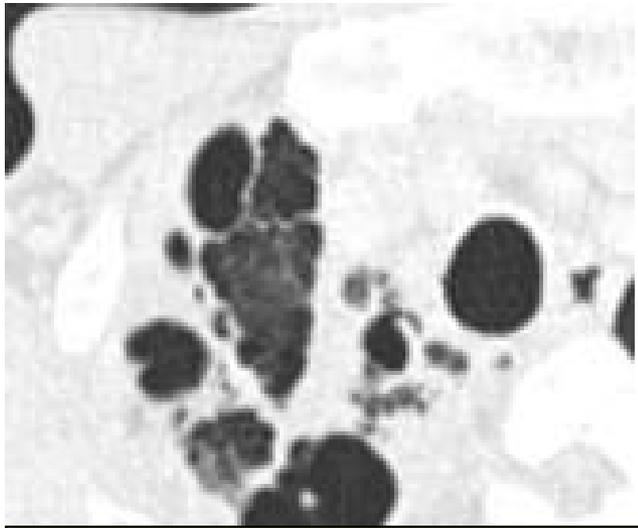
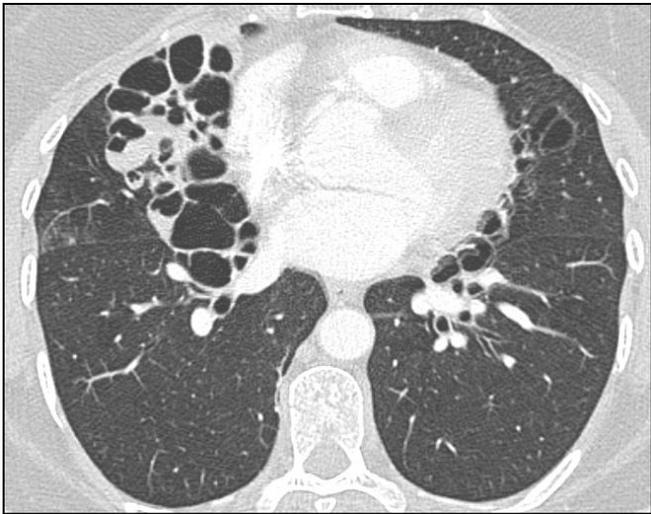
Transplantation pour sarcoidose



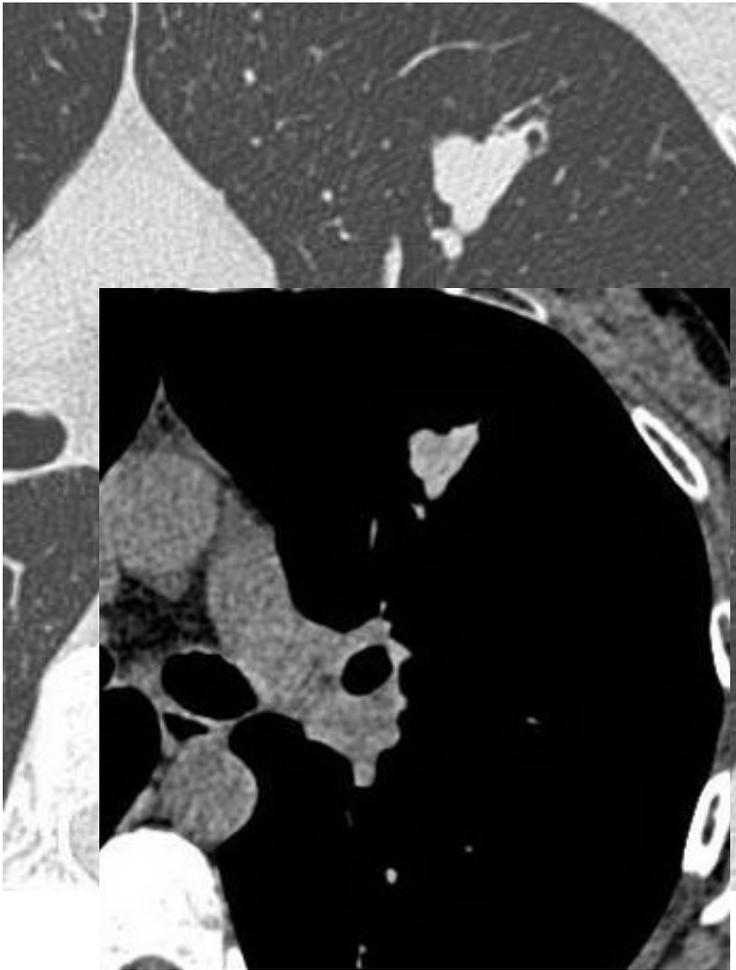
Infarctus pulmonaires et pneumatocèles



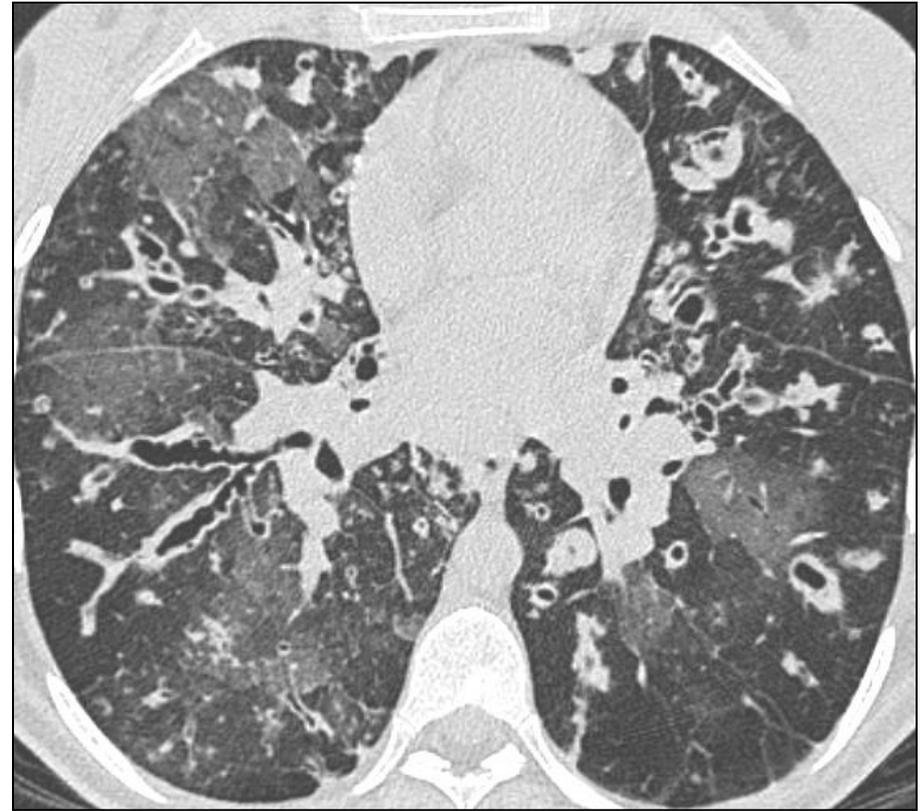
Dilatations des bronches



Dilations kystiques des bronches

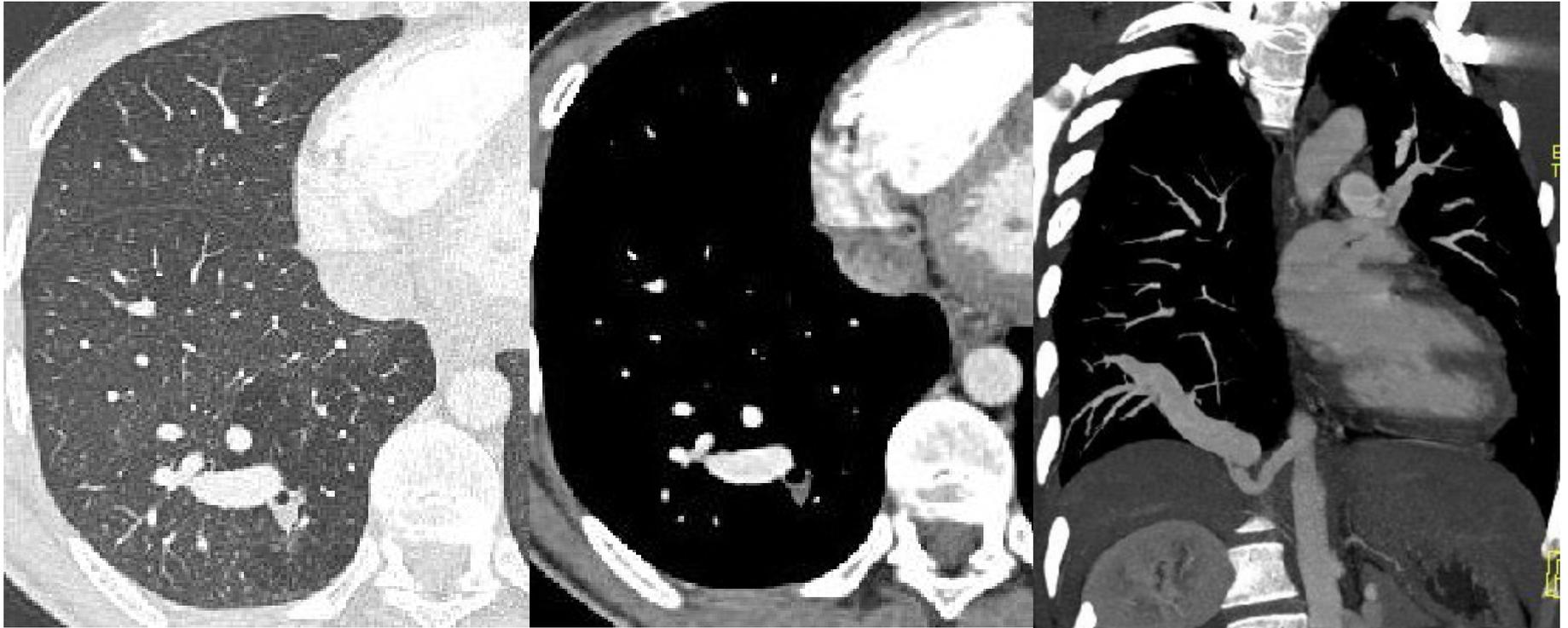


Aspergillose bronchopulmonaire allergique
(ABPA)



mucoviscidose

Séquestration

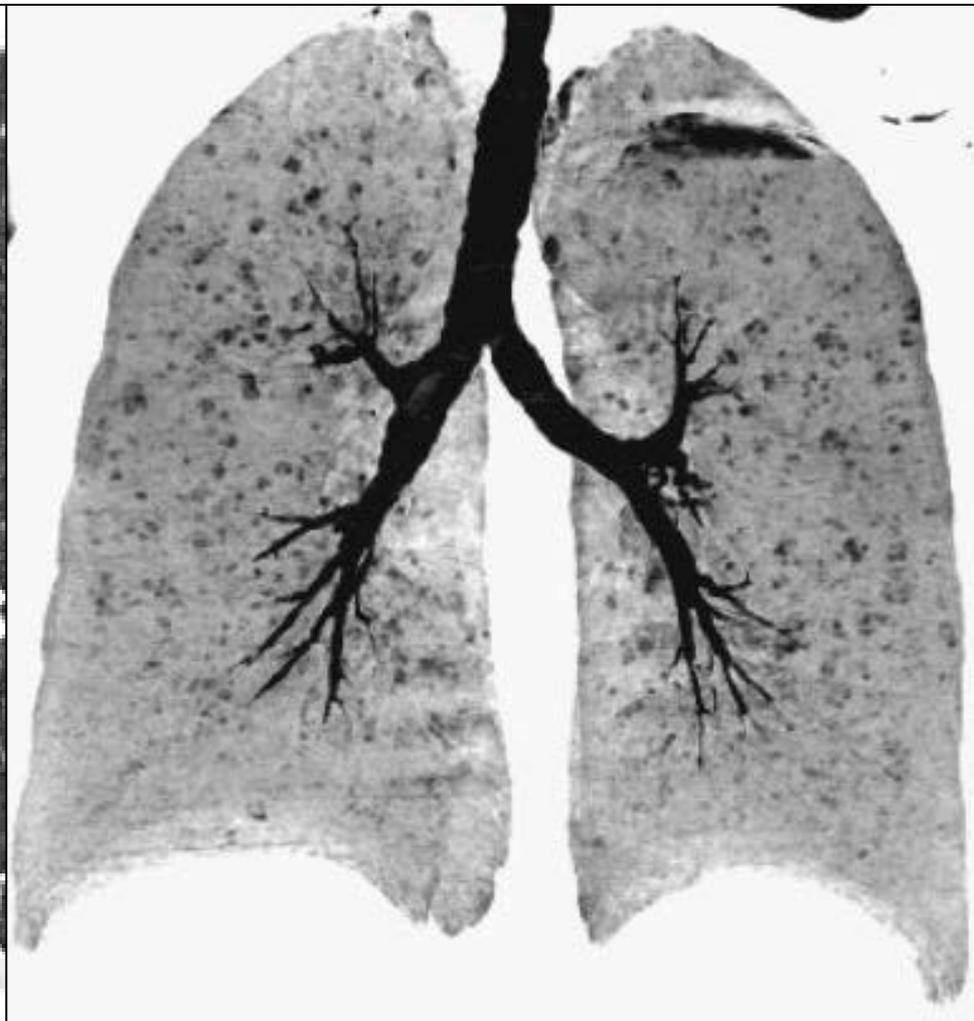


Séquestration broncho-vasculaire

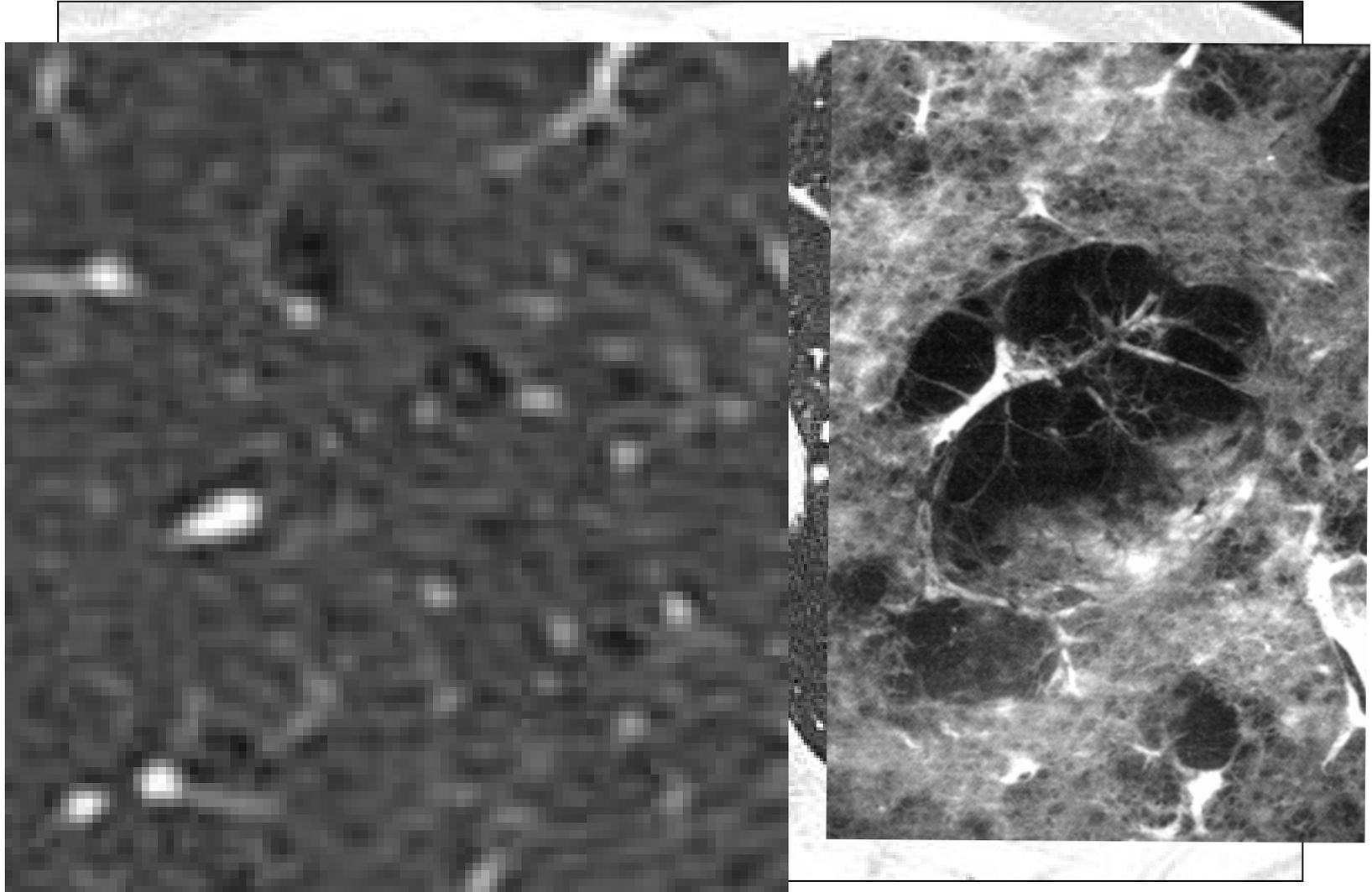
Images kystiques et hypodensités

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Emphysème centrolobulaire



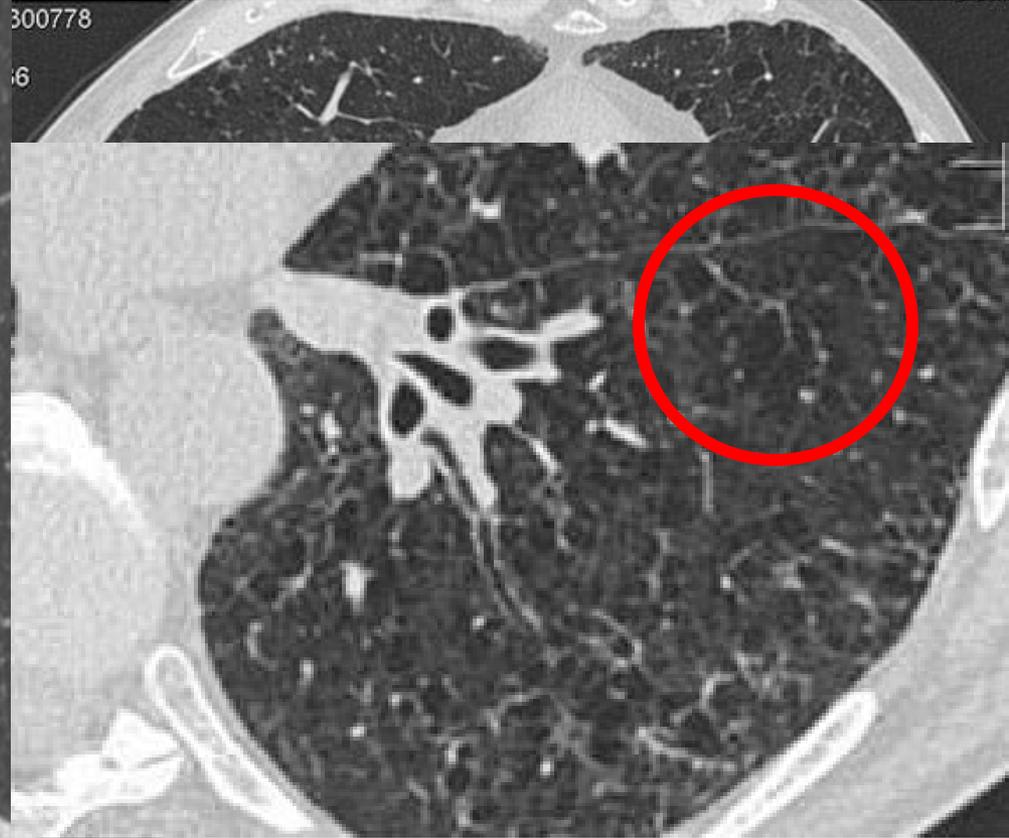
Emphysème centrolobulaire



Emphysème

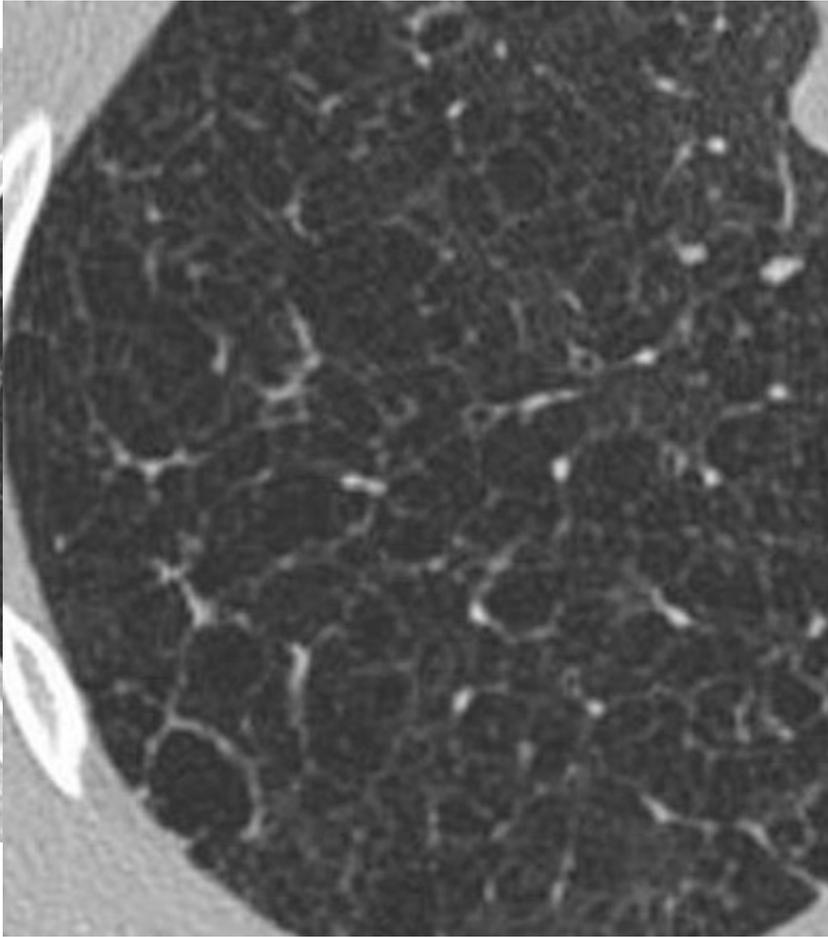


panlobulaire

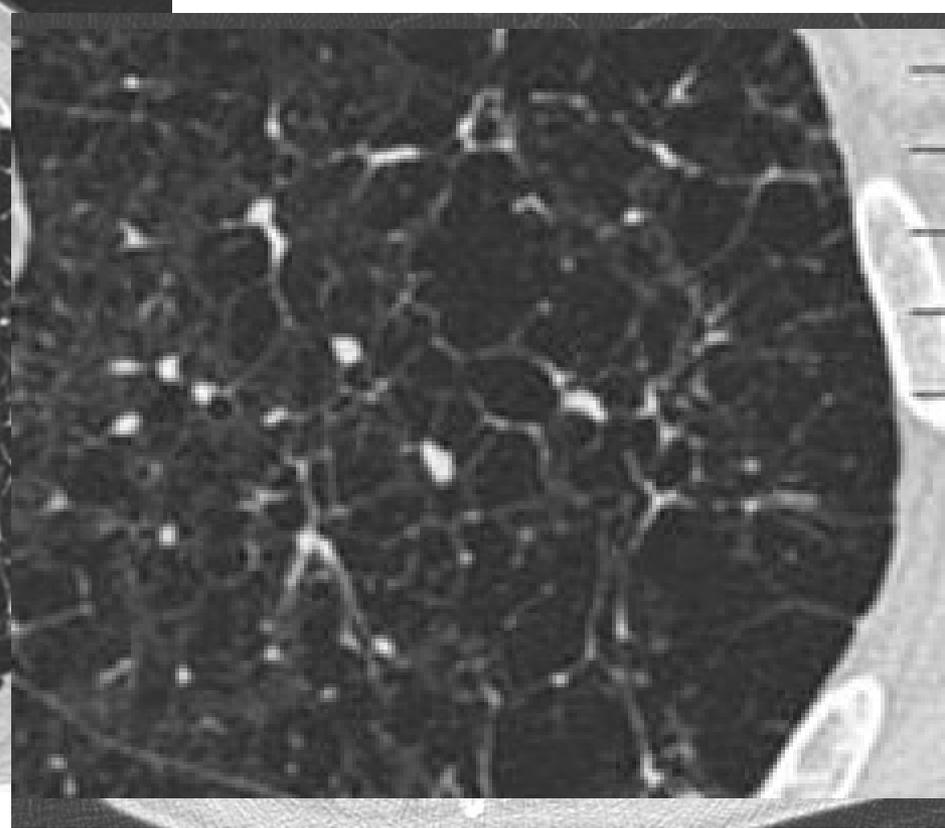


centrolobulaire

Histiocytose ou emphysème?

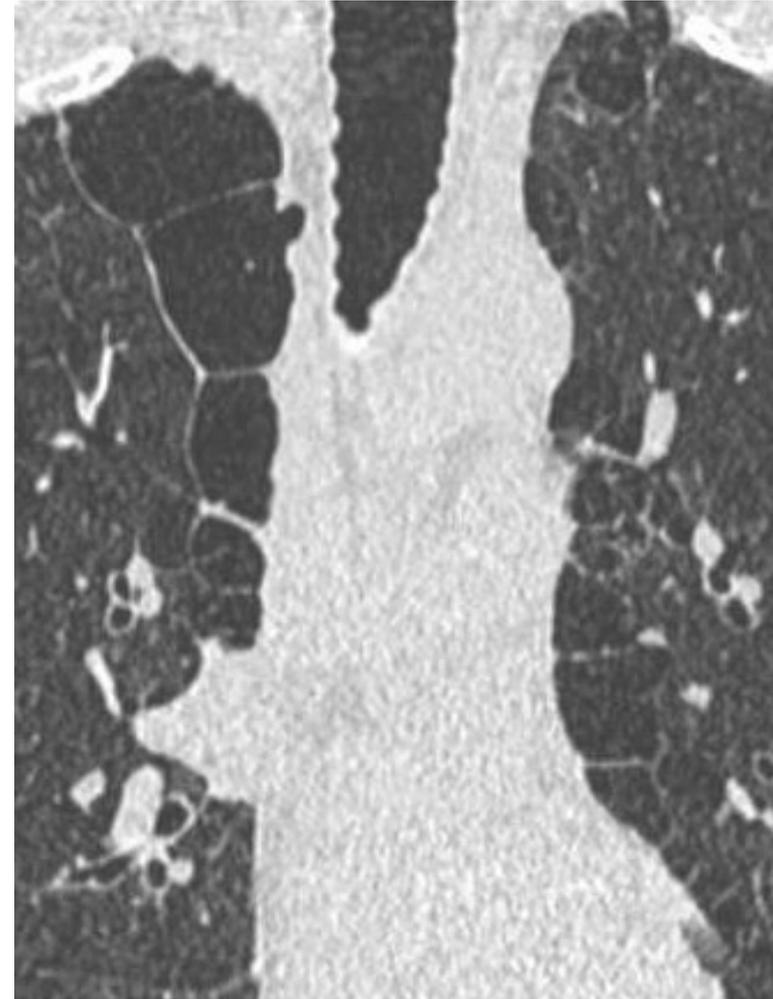
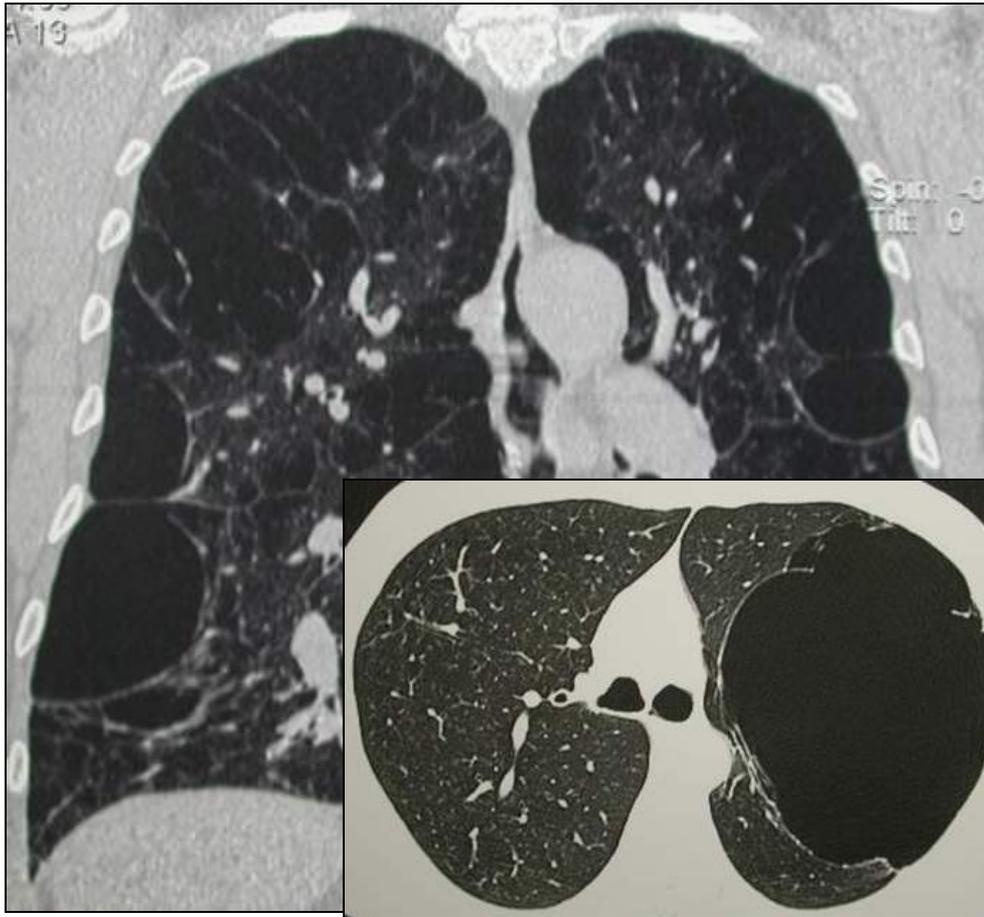


HX



Emphysème

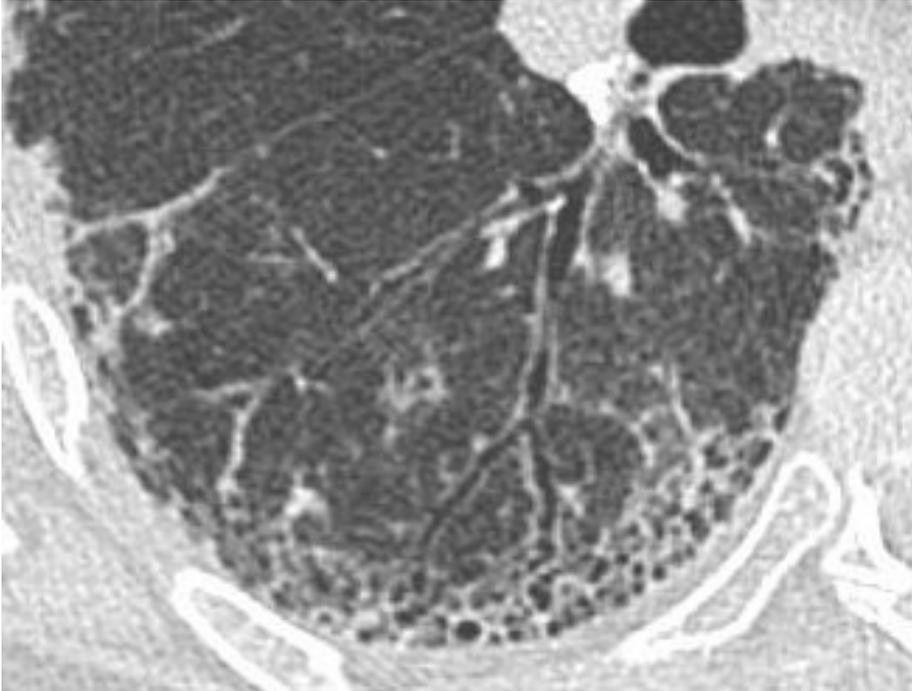
Bulles



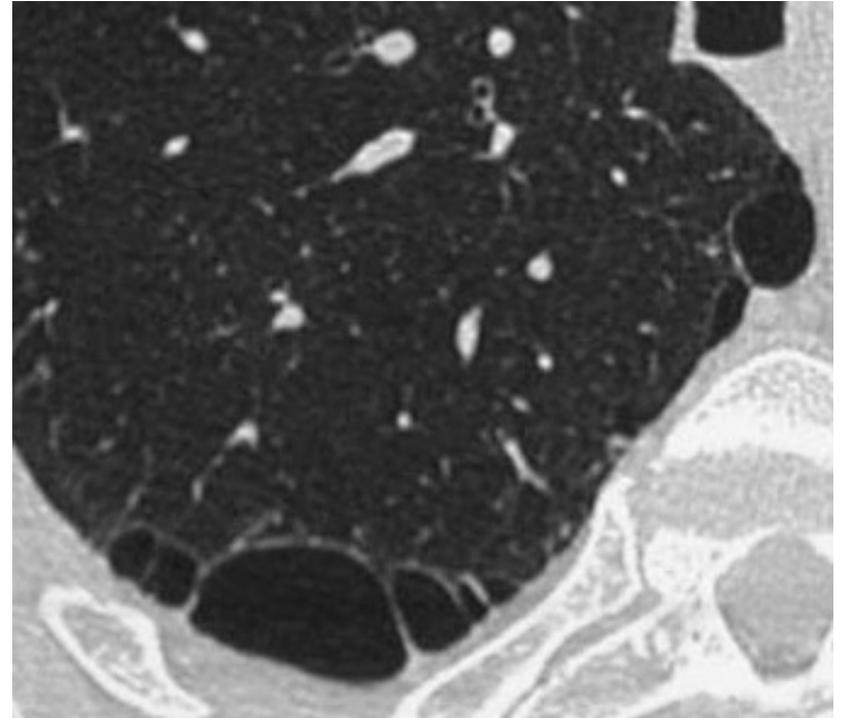
Images kystiques et hypodensités

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 - Congénital
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 - Aspect en mosaïque et piégeage
- Rayon de miel

Rayon de miel

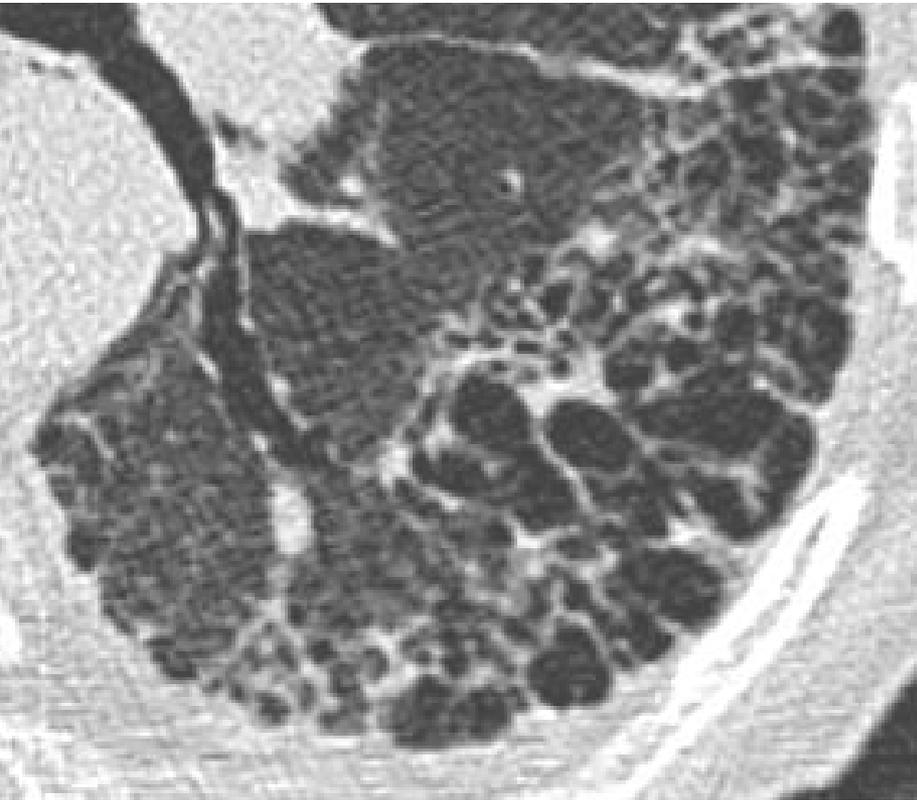


Rayon de miel
Nid d'abeille
Kystes en réseau

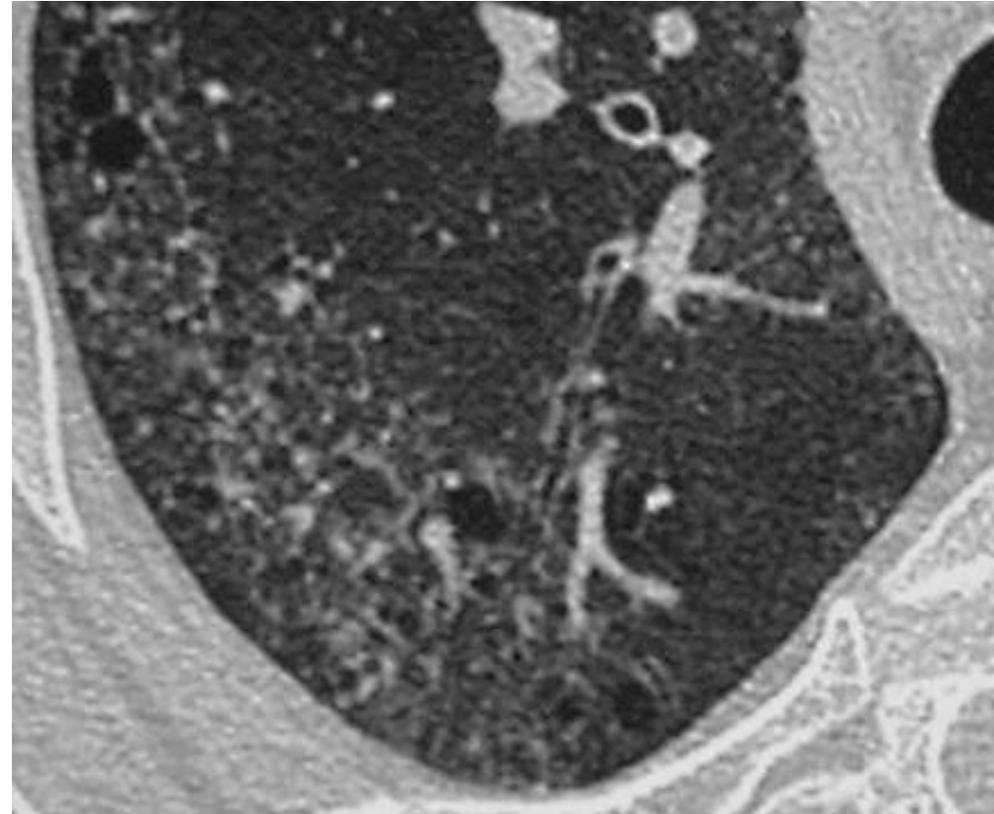


Emphysème sous pleural

Rayon de miel

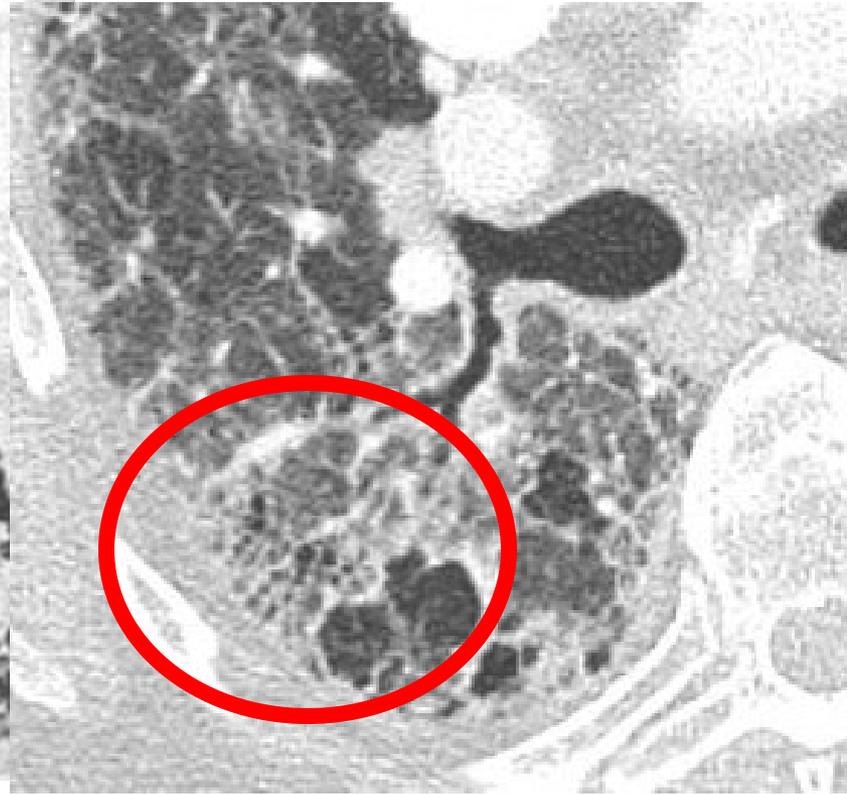
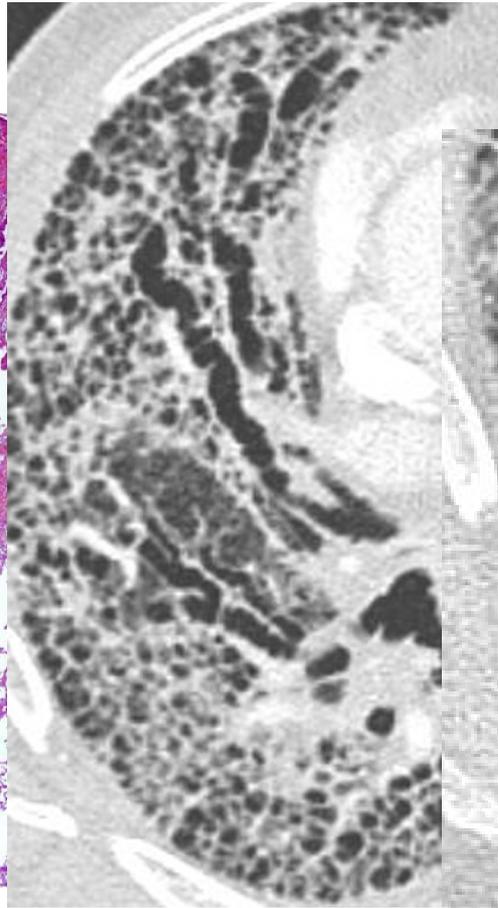
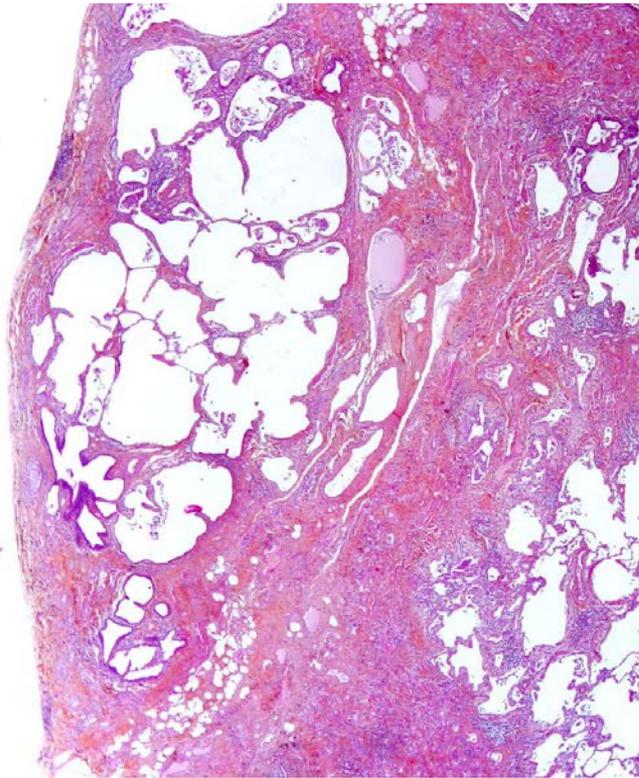


Rayon de miel
Kystes de tailles variables

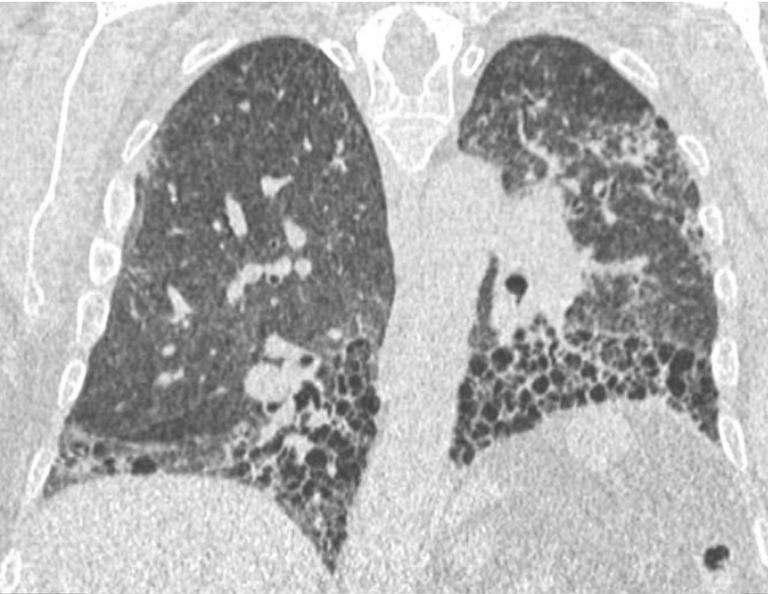


Rares kystes au sein de reticulations

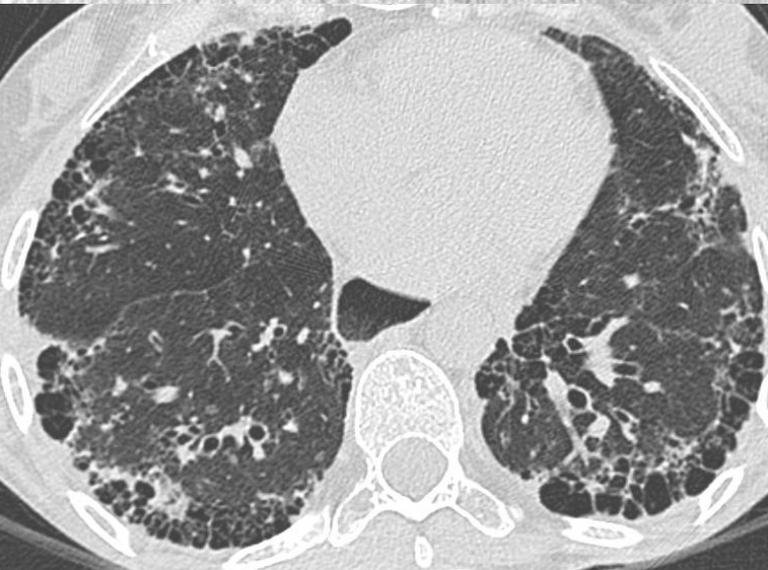
Fibrose pulmonaire idiopathique



Collagénoses



PR



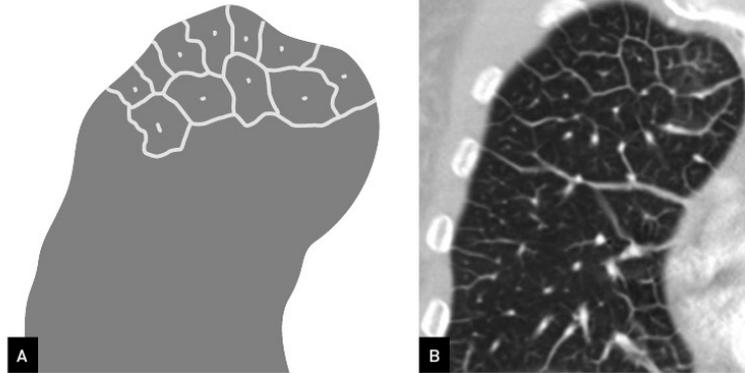
Dermatopolymyosite



Sclérodermie



Les infiltrations linéaires, le verre
dépoli, les condensations



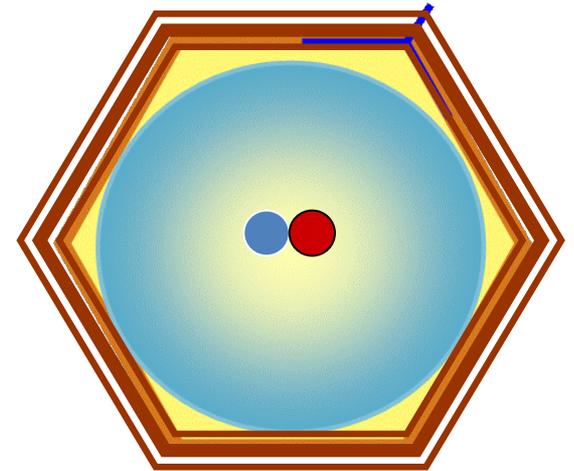
I. Les infiltrations linéaires

Epaississement anormal des septa interlobulaires: « lignes septales »
Peut être lisse/nodulaire/irrégulier

SUBSTRATUM ANATOMOPATHOLOGIQUE DES EPAISSISSEMENTS SEPTAUX

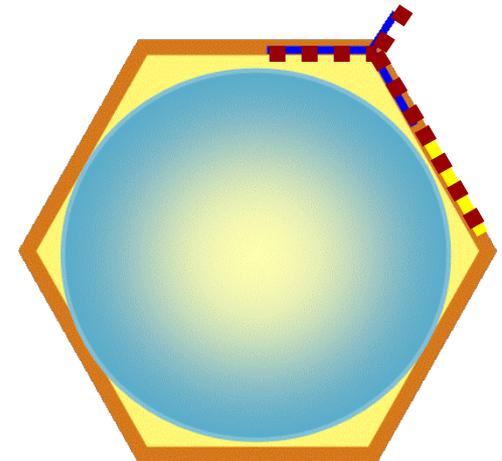
Atteinte de l'interstitium par:

- **Du tissu conjonctif:** fibrose interstitielle
- **Des liquides séreux:** œdème interstitiel
- **Des cellules ou protéines:** amylose, maladie de Gaucher et Nieman-Pick, maladie de Chester-Erdheim



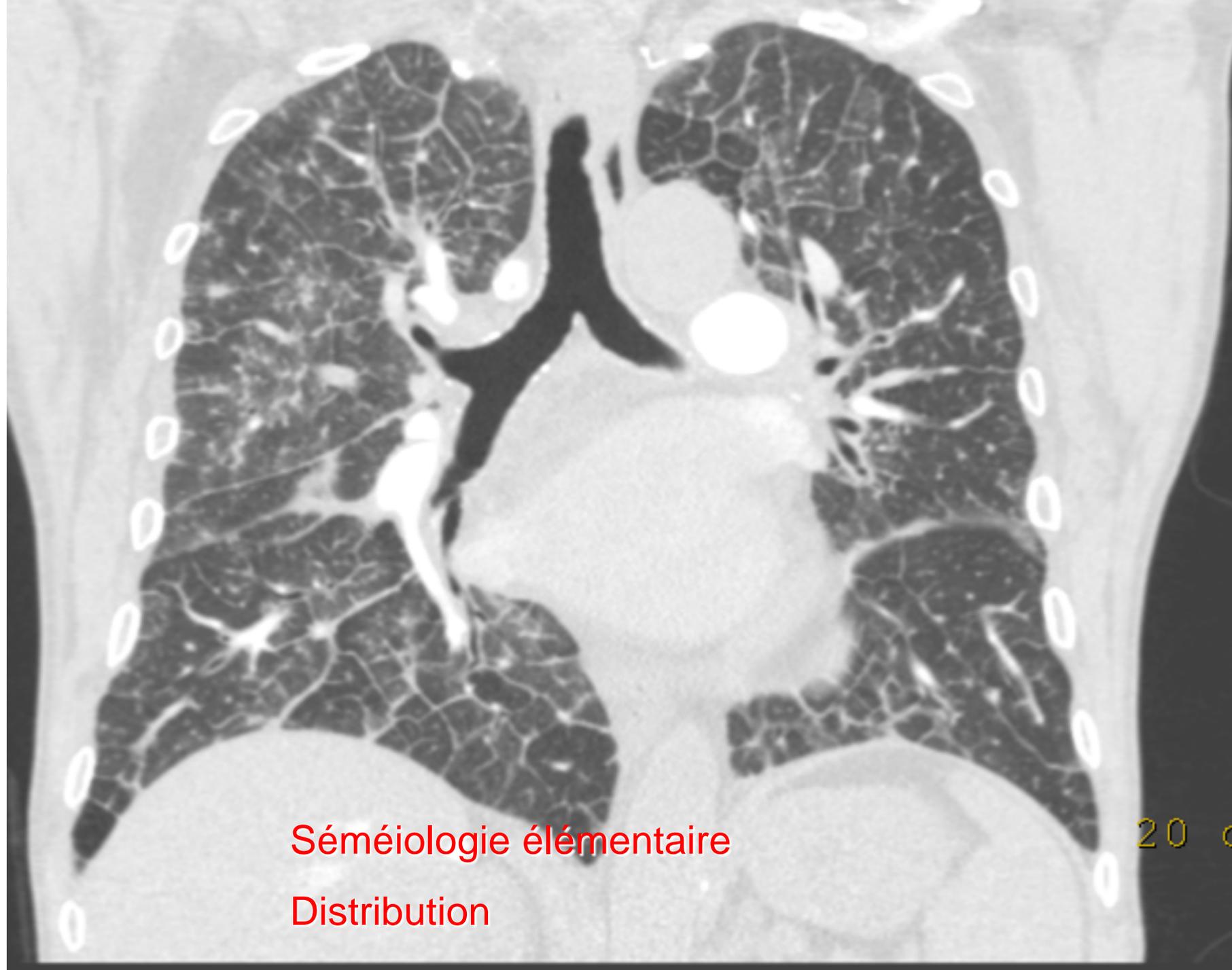
Atteinte des structures contenues dans les septa:

- **Veinules:** obstruction veineuse
- **Lymphatiques:** obstruction lymphatique



A. Epaissements septaux lisses

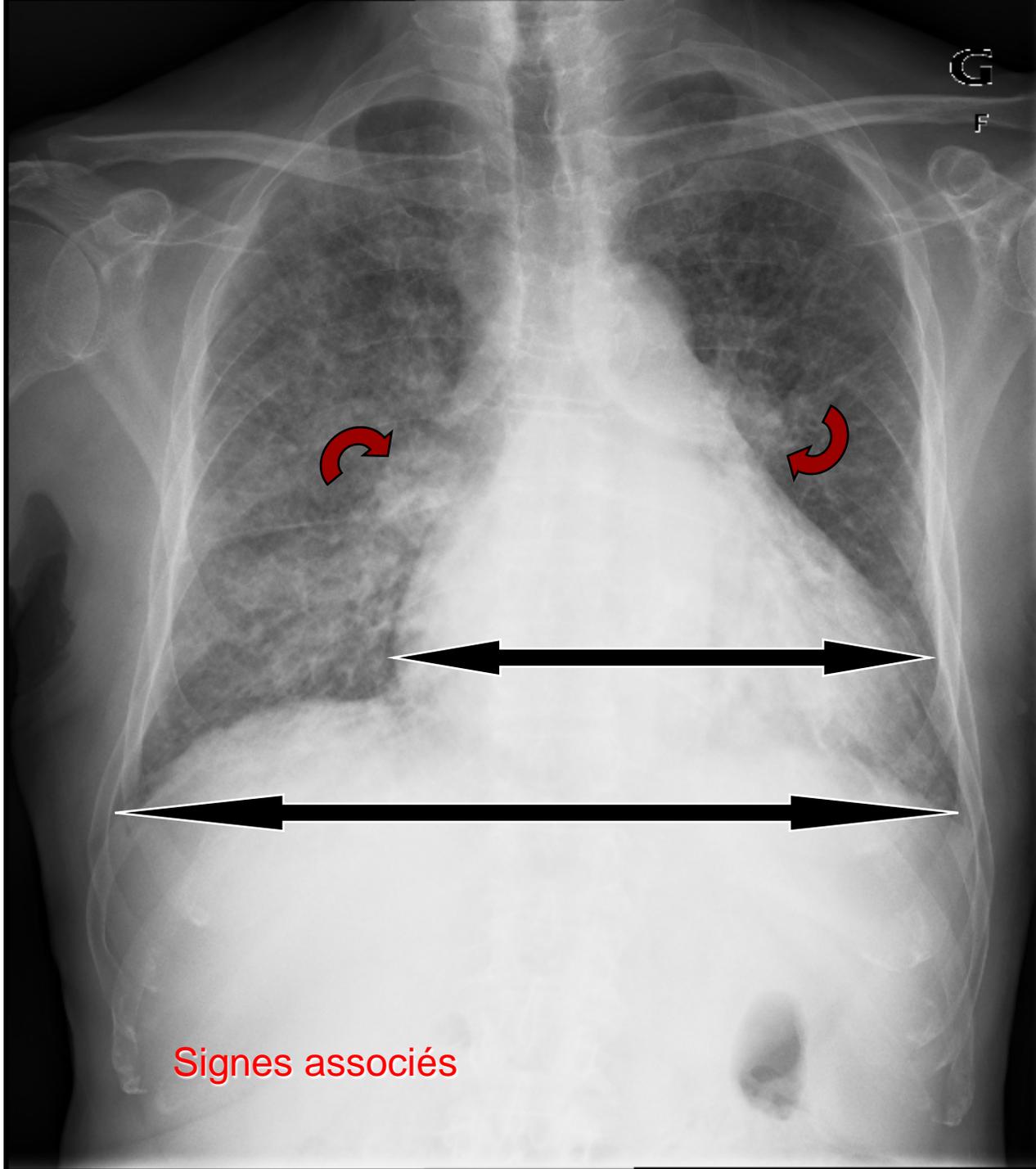
- Oedème pulmonaire
- Hémorragie pulmonaire, Lymphangite carcinomateuse, Maladie d'Erdheim- Chester, Pneumonie aiguë à éosinophiles, maladie veno-occlusive, lymphangiectasie pulmonaire congénitale, lymphome, leucémie, lymphangiomatose, amyloïdose.



Sémiologie élémentaire

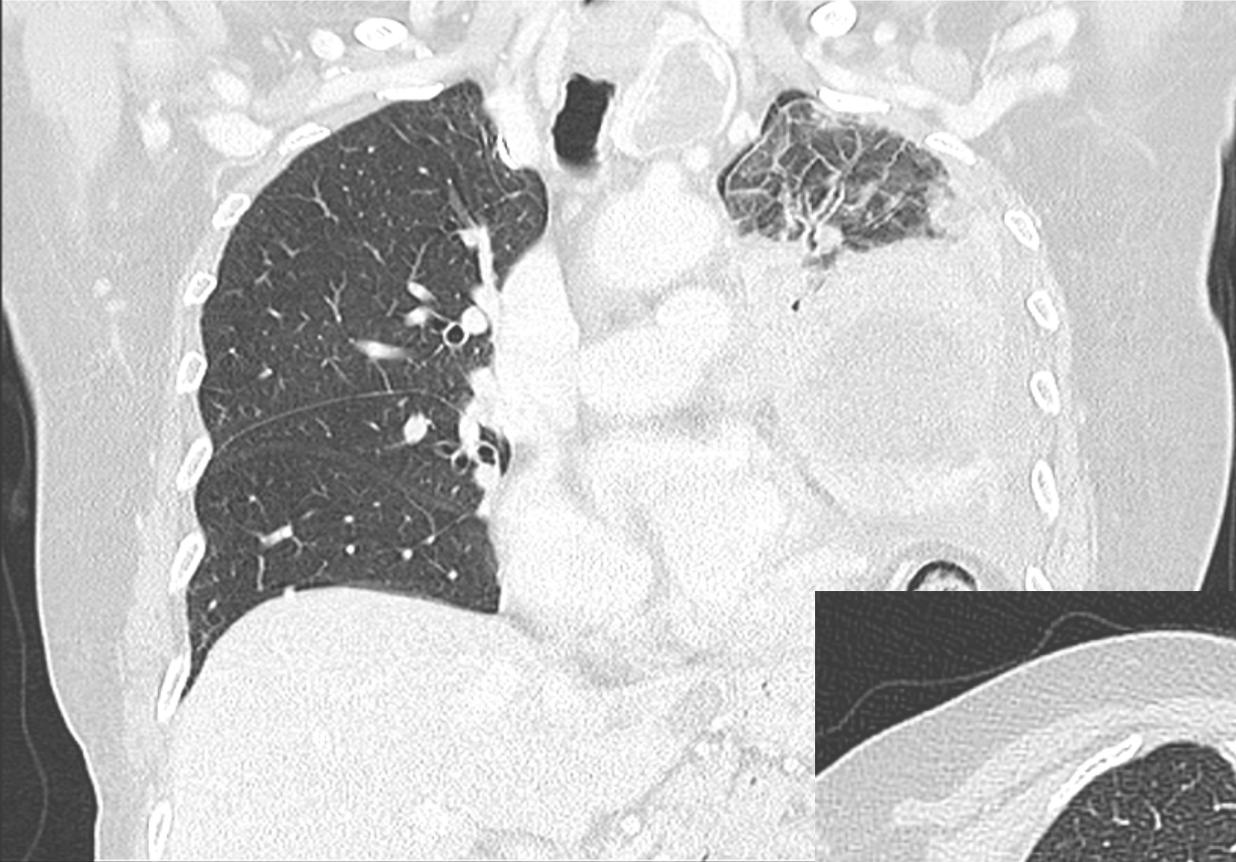
Distribution

20



Homme de 75 ans
Cardiopathie
valvulaire mitro-
Aortique
FA

Signes associés



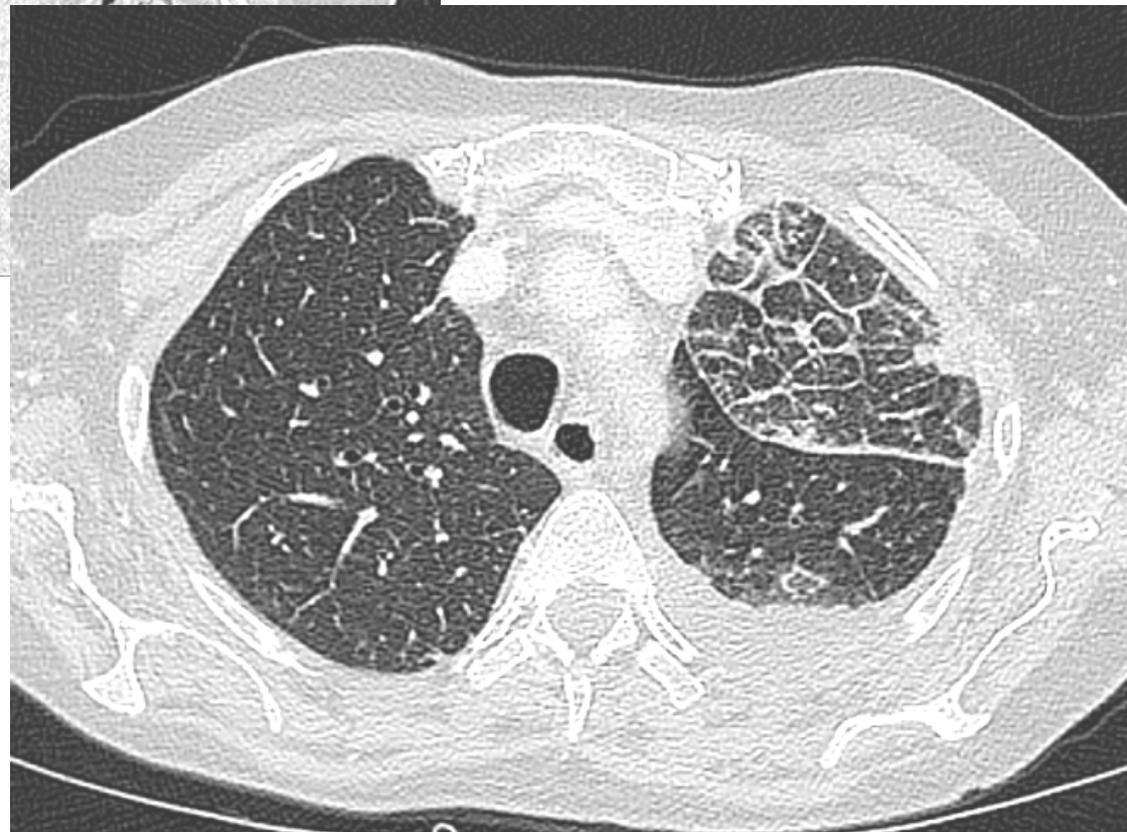
Femme de 83 ans

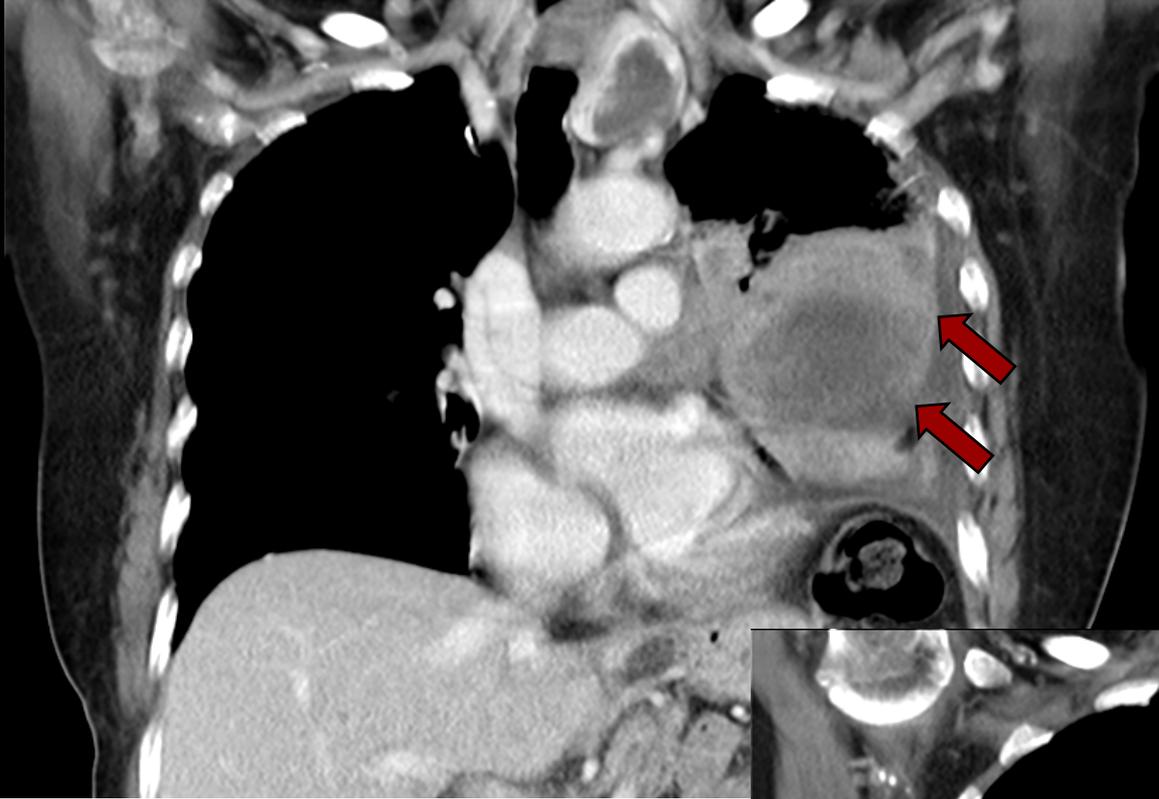
Dyspnée

Tabagisme

Séméiologie élémentaire

Distribution

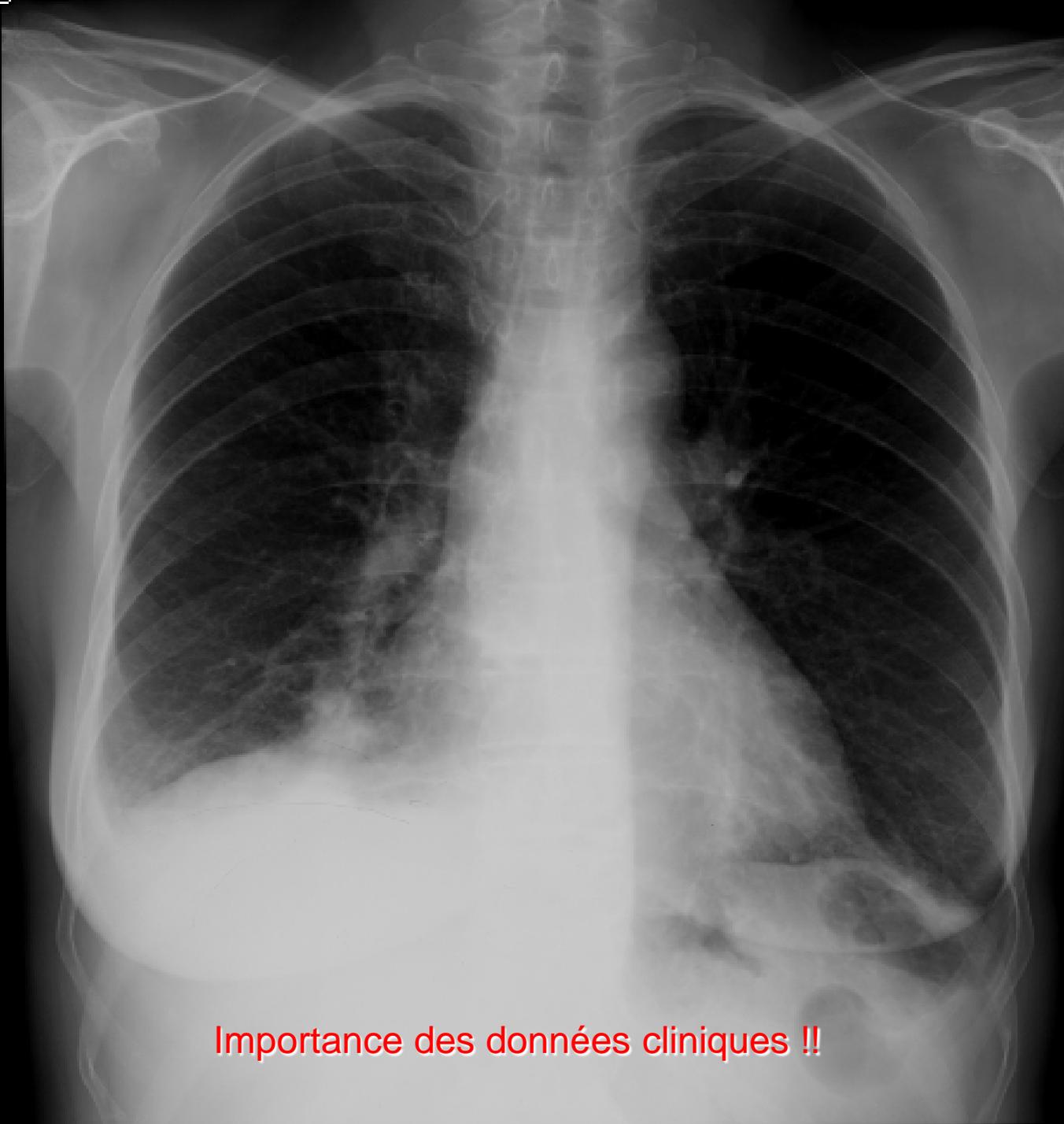




Anomalies associées

Obstruction veineuse et
lymphatique par masse
néoplasique





Femme

64 ans

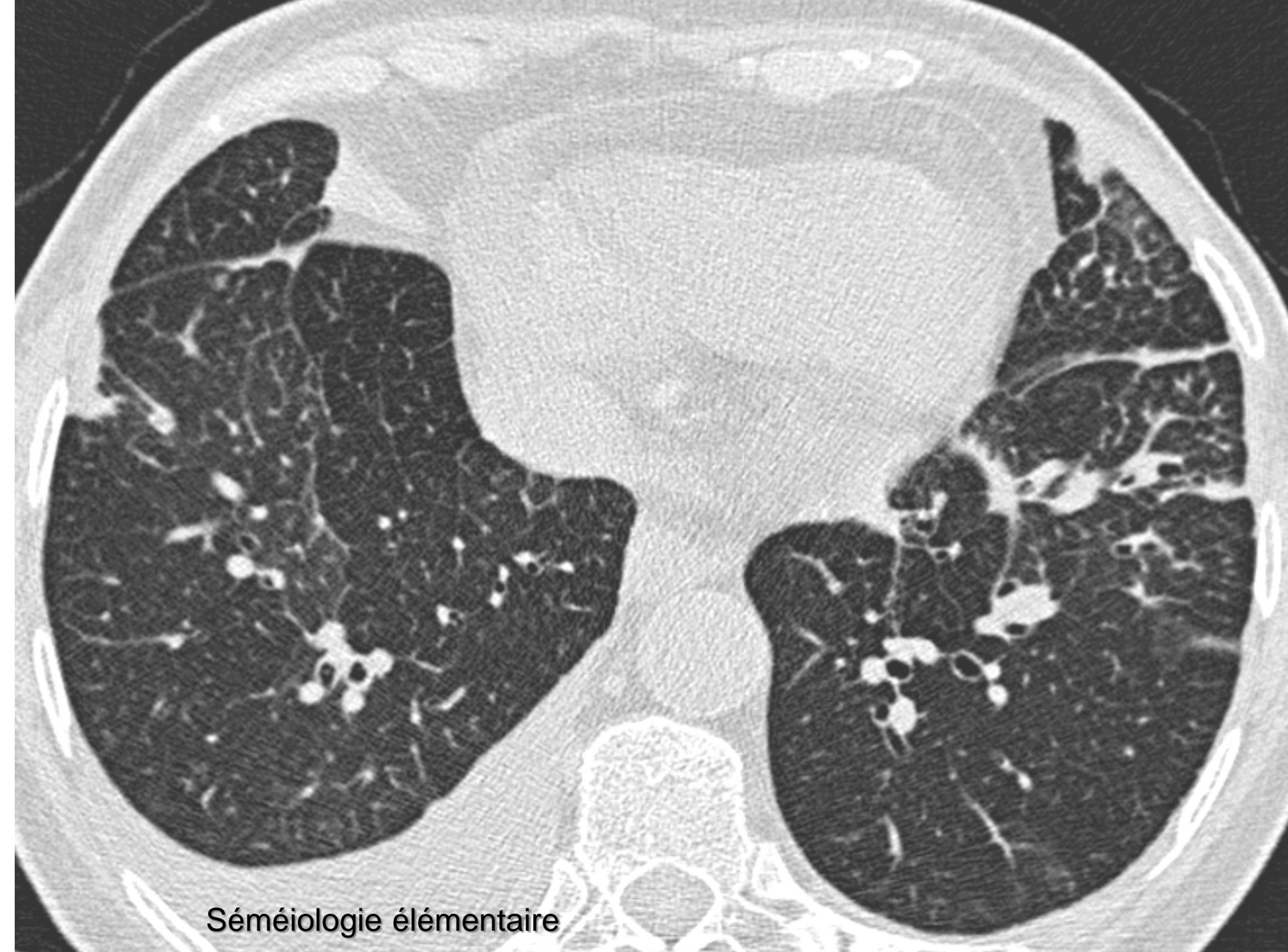
Toux

Episodes infectieux à répétition

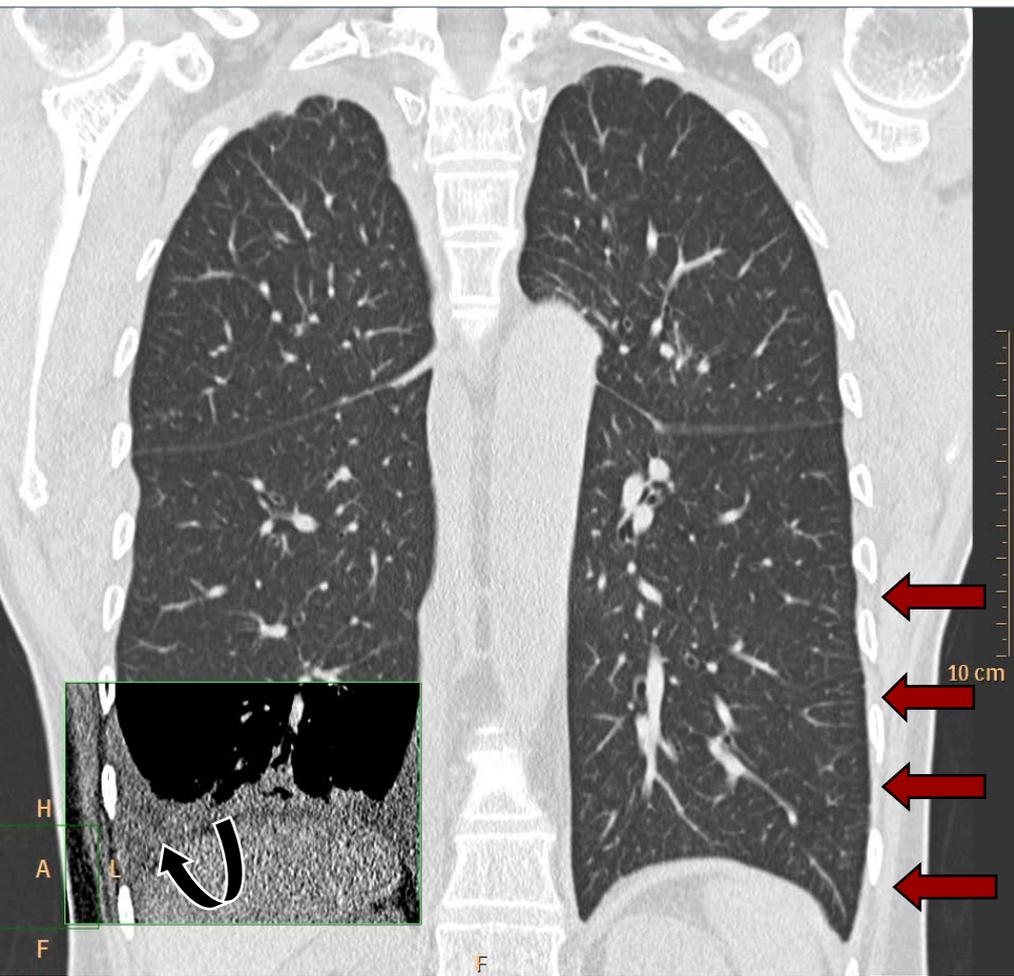
Anomalies des phanères

Quid?

Importance des données cliniques !!

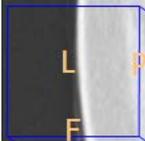


Séméiologie élémentaire



Distribution

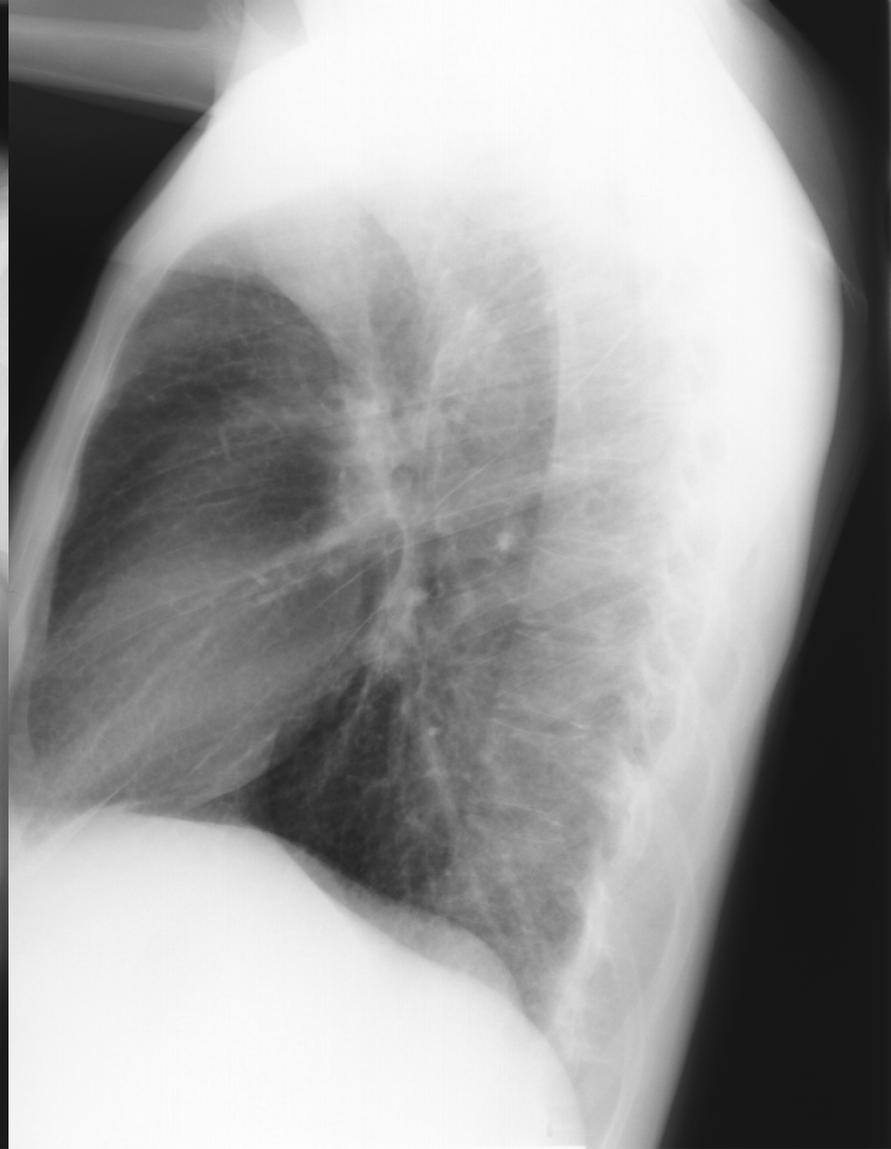
Anomalies associées



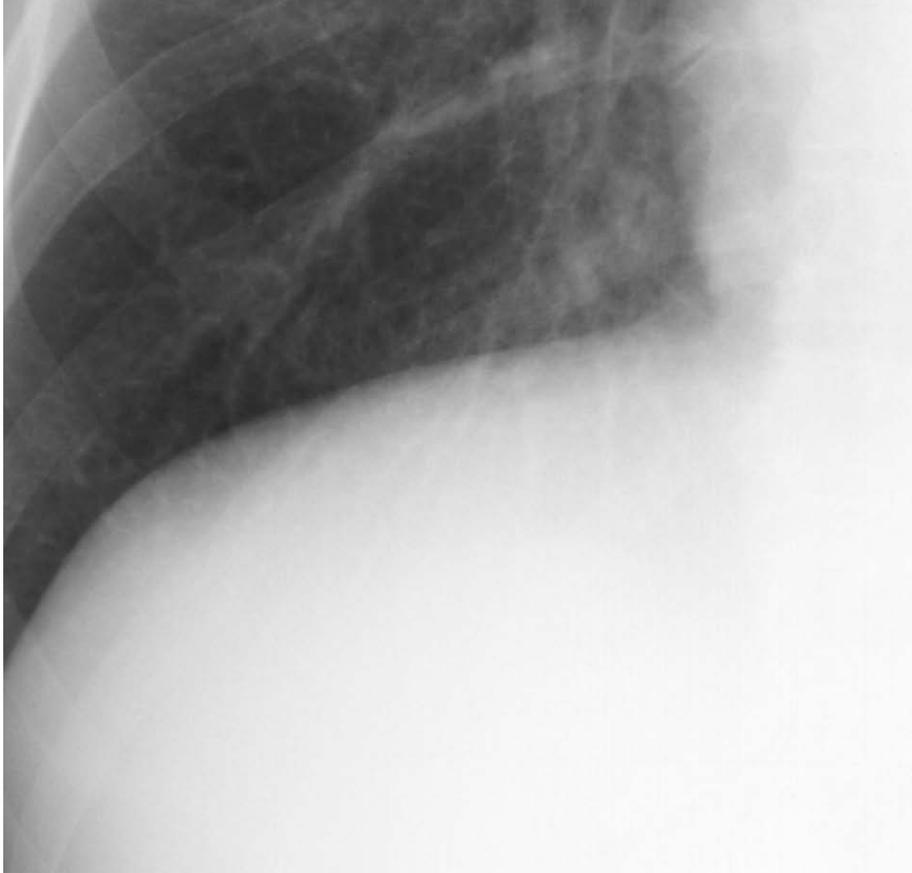
F



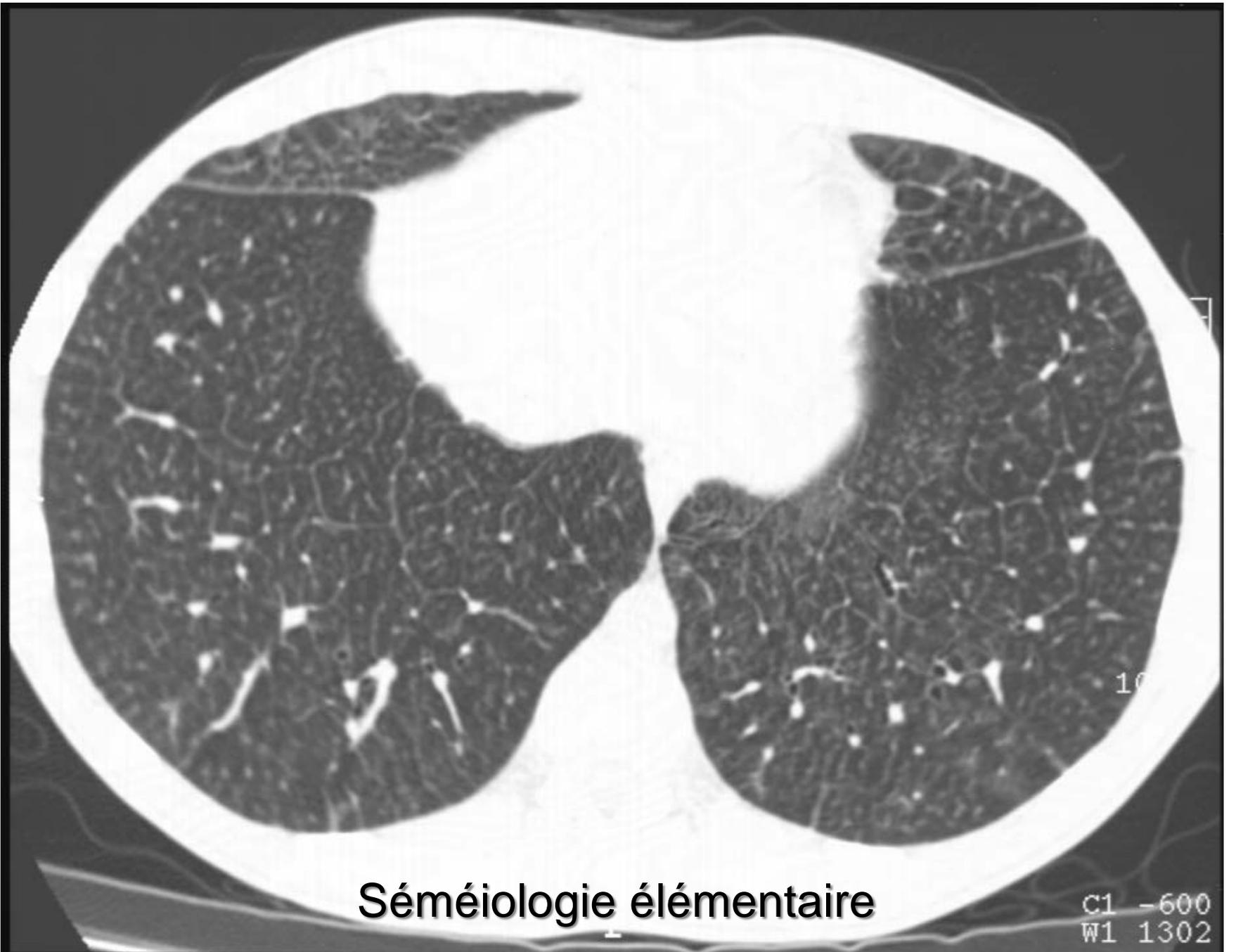
Syndrome des ongles jaunes



Homme de 22 ans admis pour stomatite herpétique. Pas de symptômes respiratoires. Examen clinique: splénomégalie-discrète hépatomégalie.

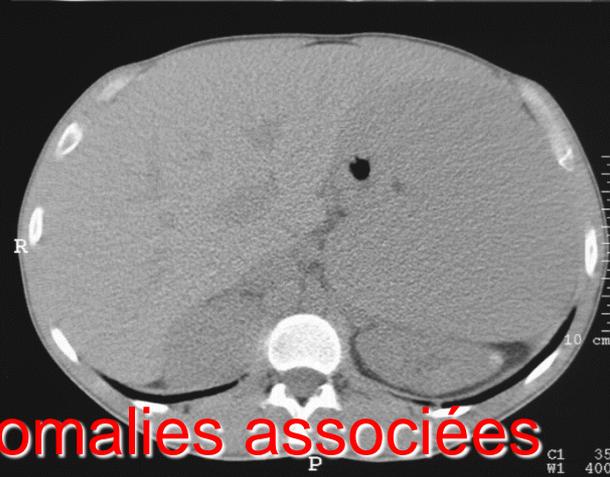
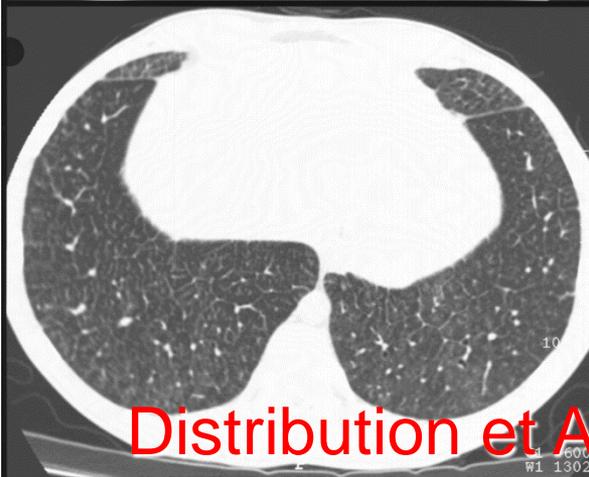
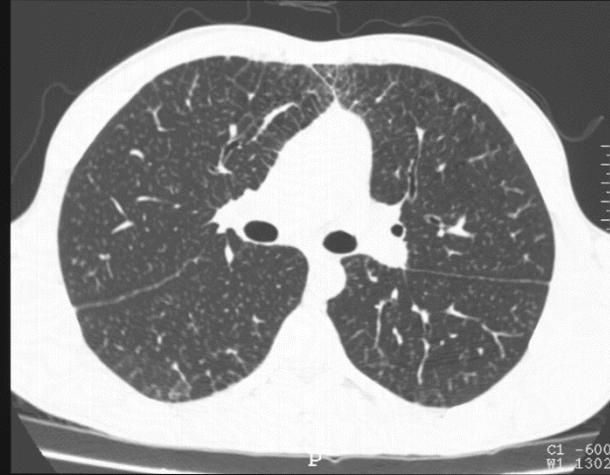
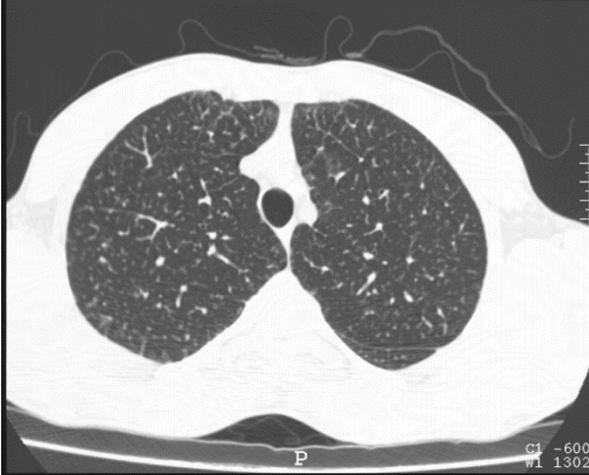


Sémiologie élémentaire et distribution

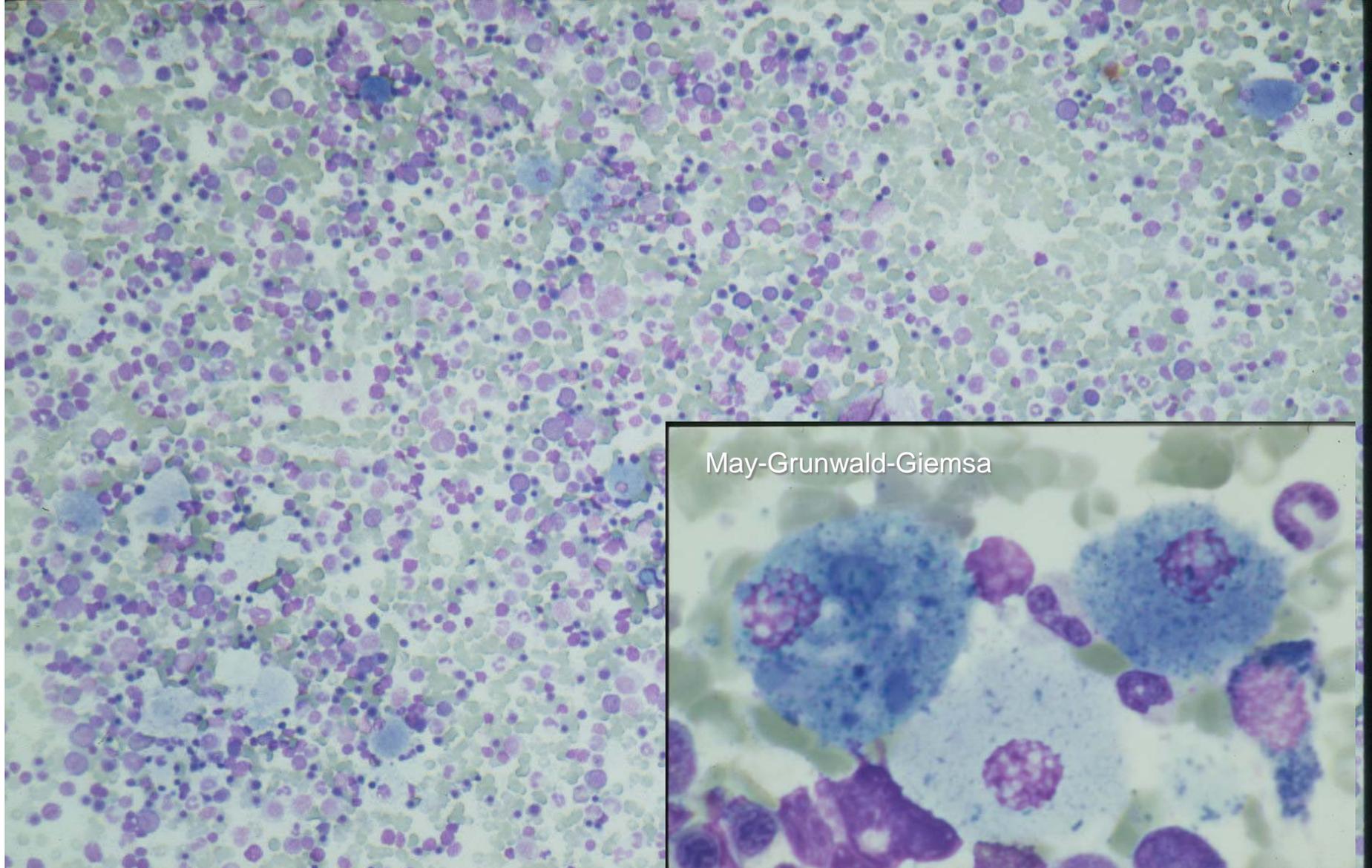


Séméiologie élémentaire

C1 -600
W1 1302

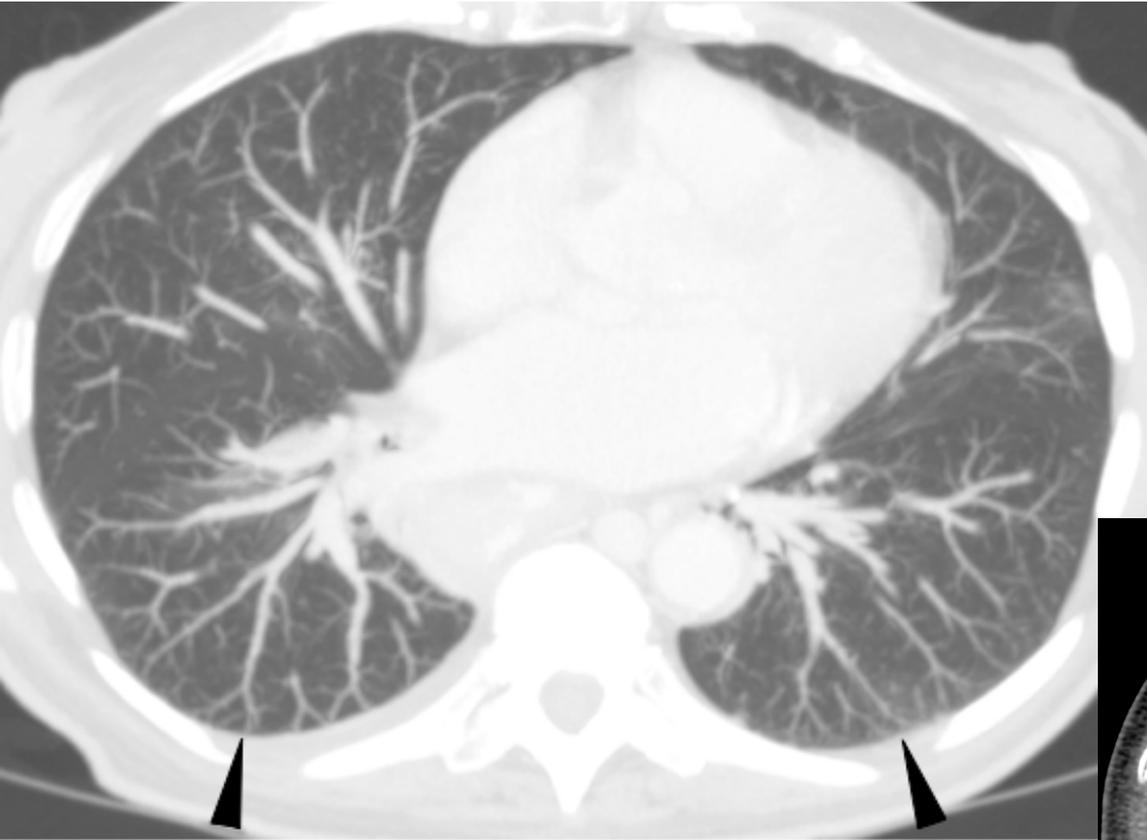


Distribution et Anomalies associées



Diagnostic: Niemann-Pick: déficit en sphingomyélinase

Pièges et variantes



Syndrôme hépato-pulmonaire



Pièges et variantes



Homme de 76 ans
Non fumeur



Homme de 84 ans
Non fumeur

B. Epaissements septaux nodulaires

- Lymphangite carcinomateuse
- Sarcoidose
- Lymphome
- LIP
- Amyloïdose (rare)
- Silicose
- Pneumocconiose

Femme de 60 ans: Bilan

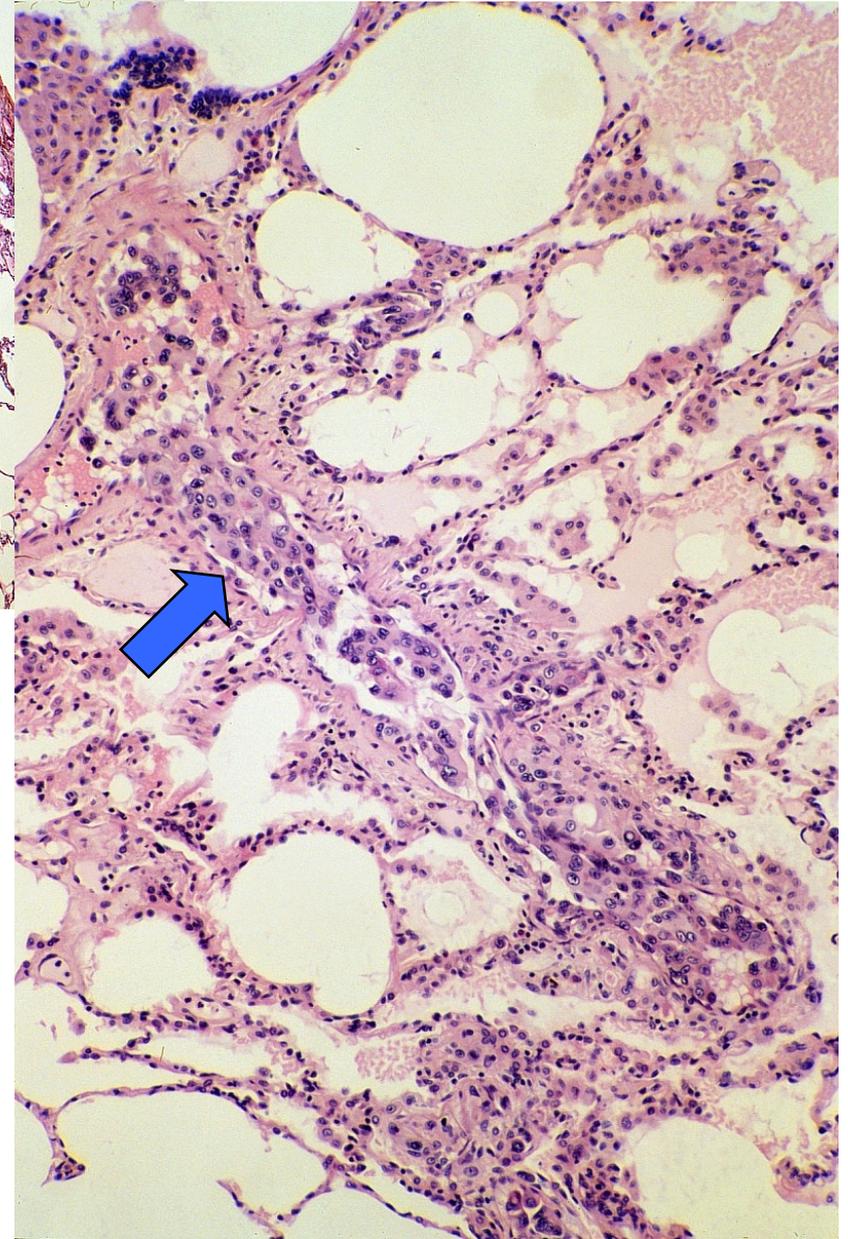
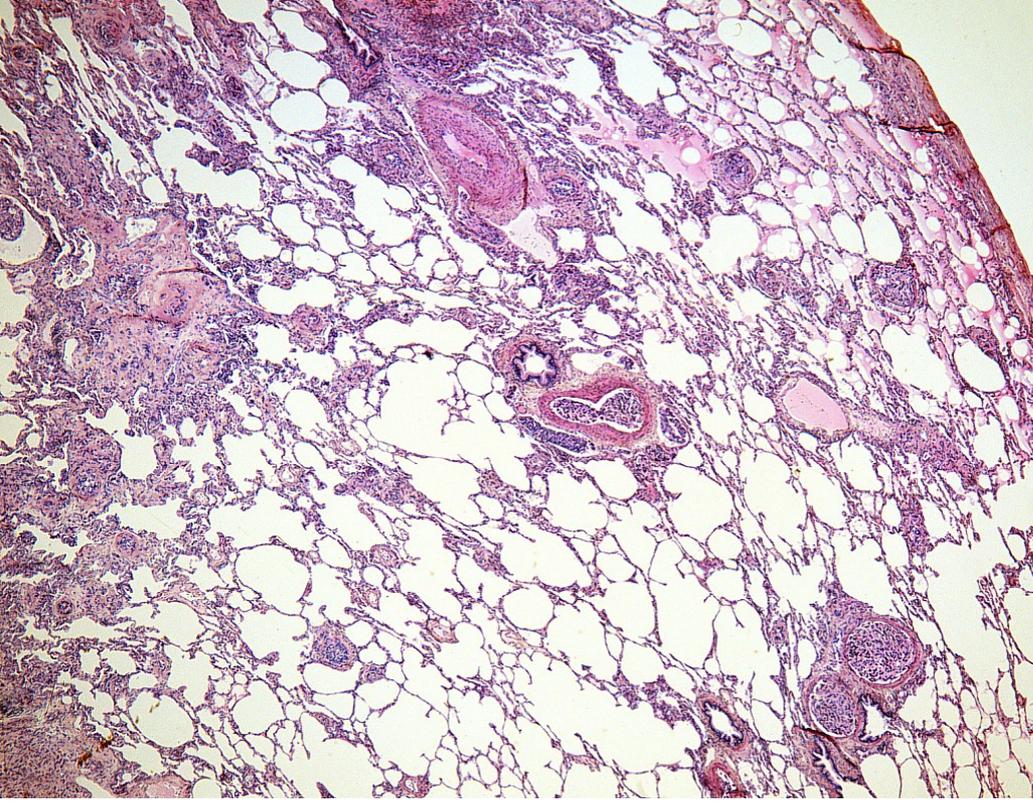


Séméiologie élémentaire





Signes associés



Diagnostic:

Lymphangite carcinomateuse

C. Epaissements septaux irréguliers

 Fibrose pulmonaire

 Sarcoïdose

 Asbestose



Sarcoïdose
Stade IV



H

A

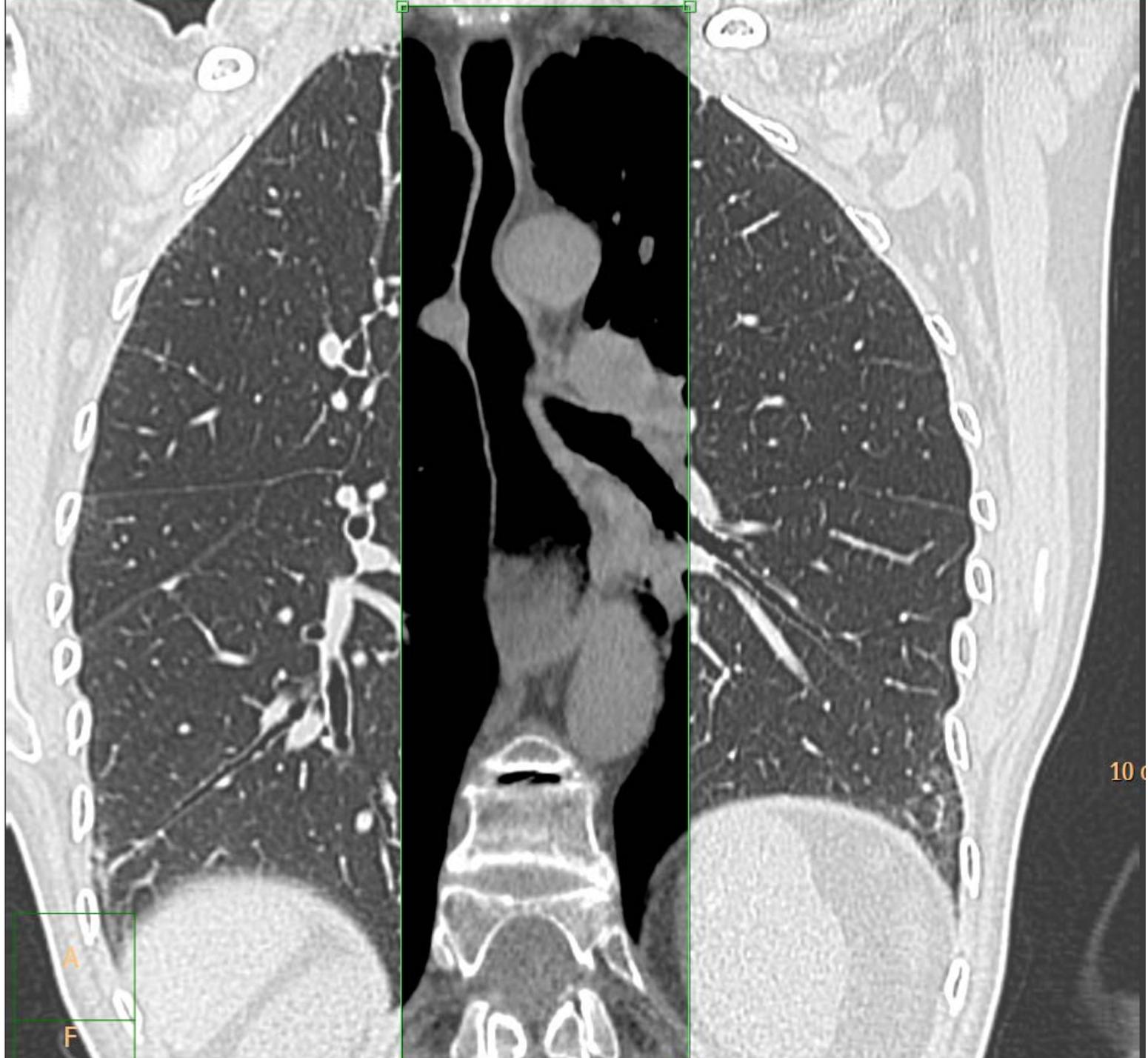
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Homme de 36 ans

Suivi



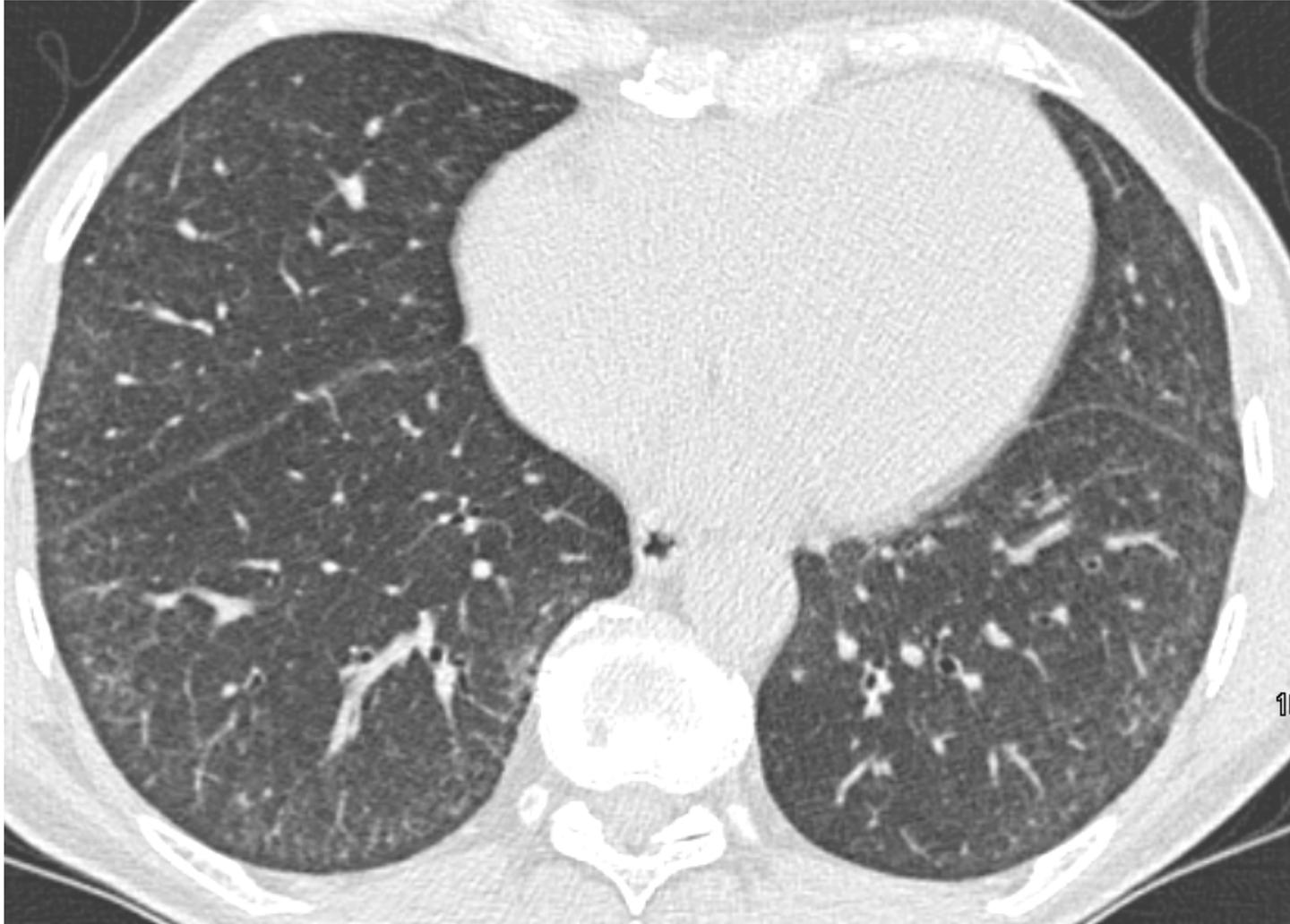


A

F

100

D. Réticulation intralobulaire



Distribution

Rayon de miel

```
graph TD; A[Rayon de miel] --> B[Sous-pleural, postérieur et prédominant aux lobes inférieurs]; A --> C[Distribution atypique (centrale ou lobes supérieurs)]; C --> D[▪ Sarcoïdose (fréquent)]; C --> E[▪ Pneumonie d'hypersensibilité]; C --> F[▪ Fibrose radique]; C --> G[▪ Séquelles de SDRA]; C --> H[▪ Fibrose pulmonaire idiopathique]; C --> I[▪ Collagénose]; C --> J[▪ Fibrose médicamenteuse];
```

Sous-pleural, postérieur et prédominant aux lobes inférieurs

- Fibrose pulmonaire idiopathique
- Collagénose
- Pneumonie d'hypersensibilité
- Pneumonie interstitielle non spécifique (PINS)
- Asbestose
- Fibrose médicamenteuse
- Sarcoïdose (rare)

Distribution atypique (centrale ou lobes supérieurs)

- Sarcoïdose (fréquent)
- Pneumonie d'hypersensibilité
- Fibrose radique
- Séquelles de SDRA
- Fibrose pulmonaire idiopathique
- Collagénose
- Fibrose médicamenteuse

Table 5. Histopathology Patterns and Features

UIP	Probable UIP	Indeterminate for UIP	Alternative Diagnosis
<ul style="list-style-type: none"> • Dense fibrosis with architectural distortion (i.e., destructive scarring and/or honeycombing) • Predominant subpleural and/or paraseptal distribution of fibrosis • Patchy involvement of lung parenchyma by fibrosis • Fibroblast foci • Absence of features to suggest an alternate diagnosis 	<ul style="list-style-type: none"> • Some histologic features from column 1 are present but to an extent that precludes a definite diagnosis of UIP/IPF <p style="text-align: center;"><i>And</i></p> <ul style="list-style-type: none"> • Absence of features to suggest an alternative diagnosis <p style="text-align: center;"><i>Or</i></p> <ul style="list-style-type: none"> • Honeycombing only 	<ul style="list-style-type: none"> • Fibrosis with or without architectural distortion, with features favoring either a pattern other than UIP or features favoring UIP secondary to another cause* • Some histologic features from column 1, but with other features suggesting an alternative diagnosis† 	<ul style="list-style-type: none"> • Features of other histologic patterns of IIPs (e.g., absence of fibroblast foci or loose fibrosis) in all biopsies • Histologic findings indicative of other diseases (e.g., hypersensitivity pneumonitis, Langerhans cell histiocytosis, sarcoidosis, LAM)

Definition of abbreviations: IIP = idiopathic interstitial pneumonia; IPF = idiopathic pulmonary fibrosis; LAM = lymphangioleiomyomatosis; UIP = usual interstitial pneumonia.

*Granulomas, hyaline membranes (other than when associated with acute exacerbation of IPF, which may be the presenting manifestation in some patients), prominent airway-centered changes, areas of interstitial inflammation lacking associated fibrosis, marked chronic fibrous pleuritis, organizing pneumonia. Such features may not be overt or easily seen to the untrained eye and often need to be specifically sought.

†Features that should raise concerns about the likelihood of an alternative diagnosis include a cellular inflammatory infiltrate away from areas of honeycombing, prominent lymphoid hyperplasia including secondary germinal centers, and a distinctly bronchiocentric distribution that could include extensive peribroncholar metaplasia.

AMERICAN THORACIC SOCIETY DOCUMENTS

Diagnosis of Idiopathic Pulmonary Fibrosis

An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline

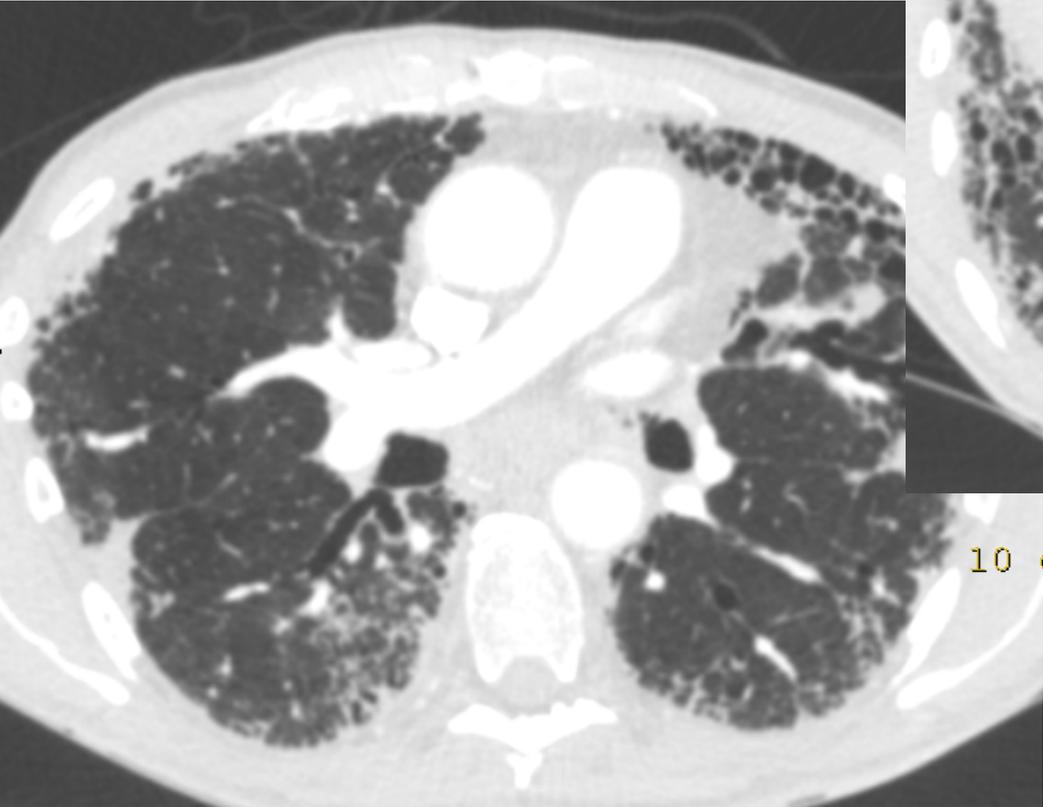
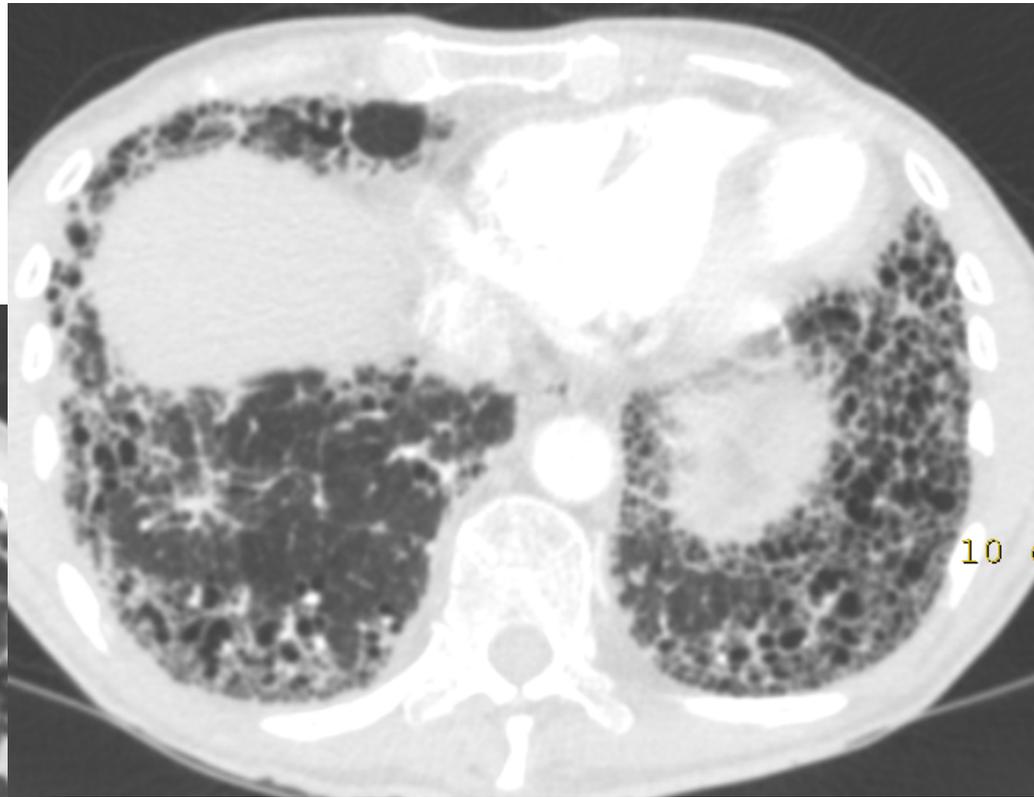
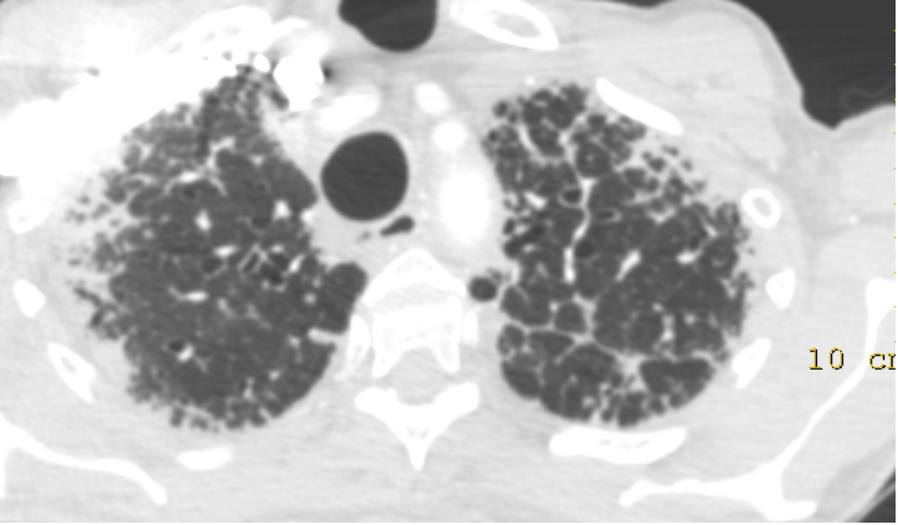
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THIS OFFICIAL CLINICAL PRACTICE GUIDELINE OF THE AMERICAN THORACIC SOCIETY (ATS), EUROPEAN RESPIRATORY SOCIETY (ERS), JAPANESE RESPIRATORY SOCIETY (JRS), AND LATIN AMERICAN THORACIC SOCIETY (ALAT) WAS APPROVED BY THE ATS, JRS, AND ALAT MAY 2018, AND THE ERS JUNE 2018

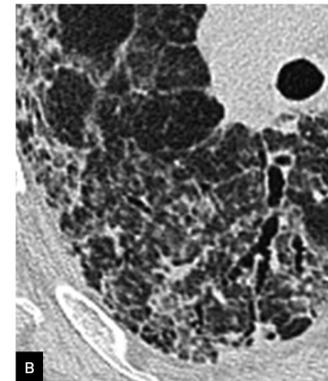
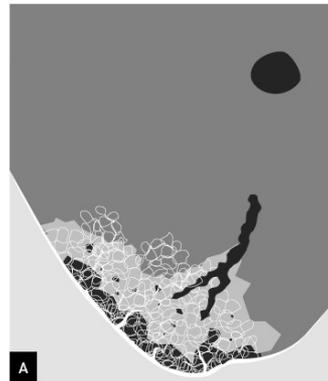
This official ATS/ERS/JRS/ALAT Clinical Practice Guideline was endorsed by the Pulmonary Pathology Society October 2018

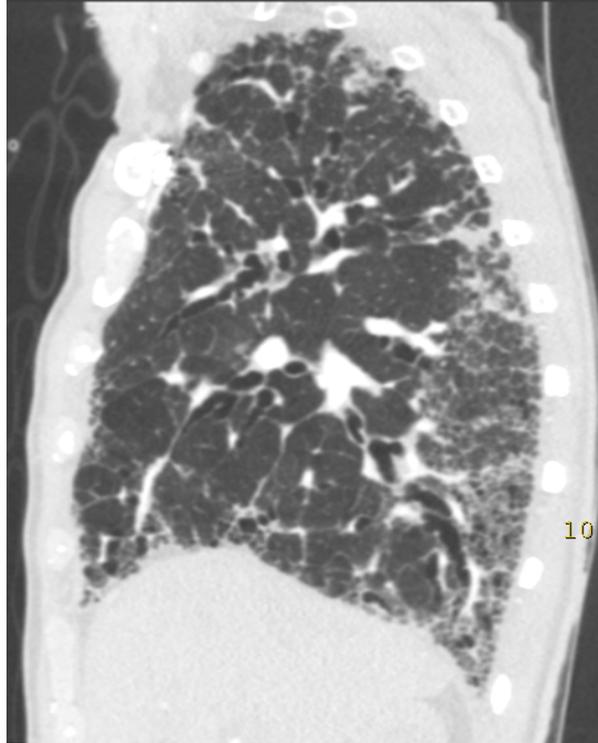
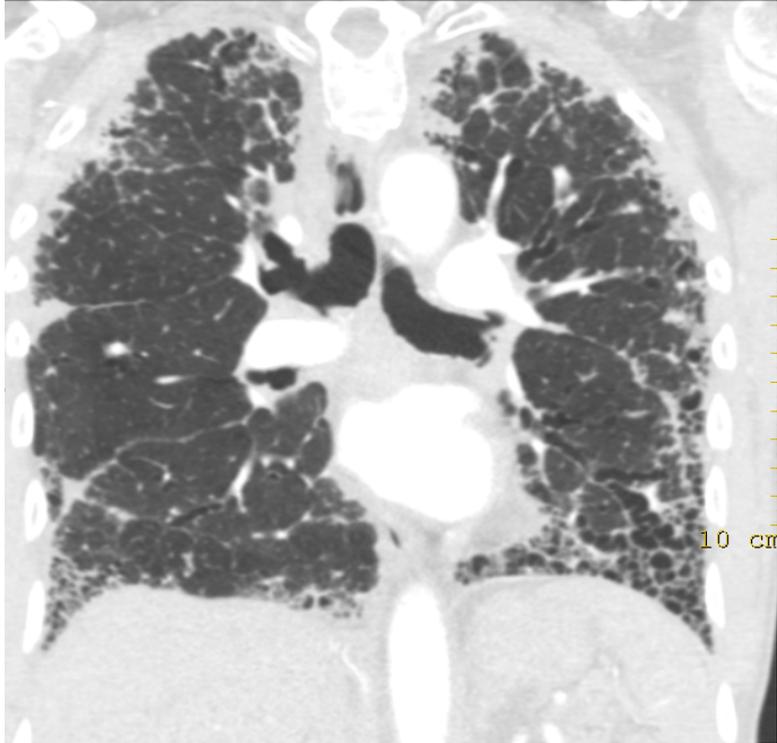


Homme de 77 ans, dyspnée

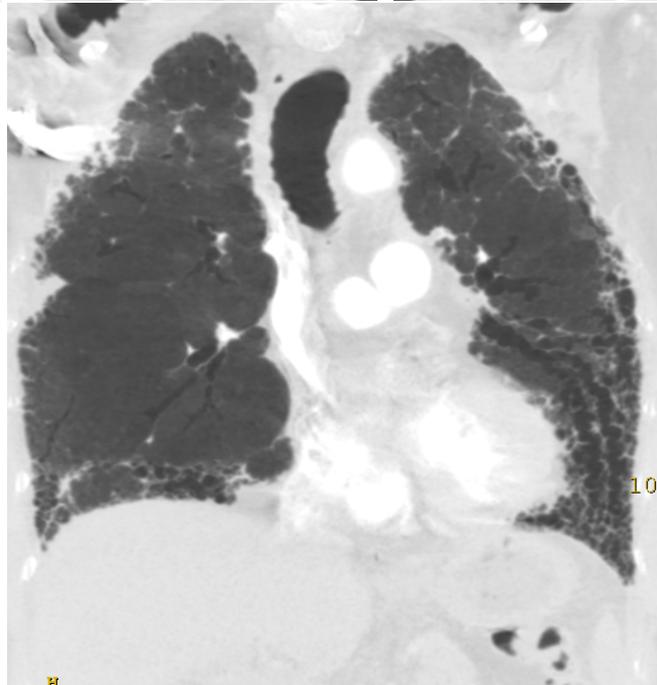


Evolution vers le rayon de miel

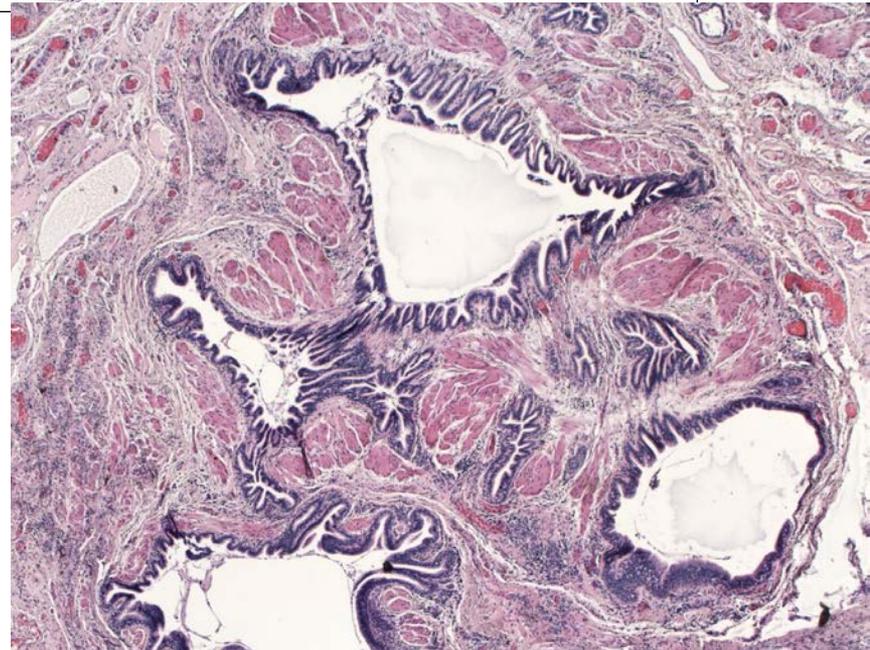
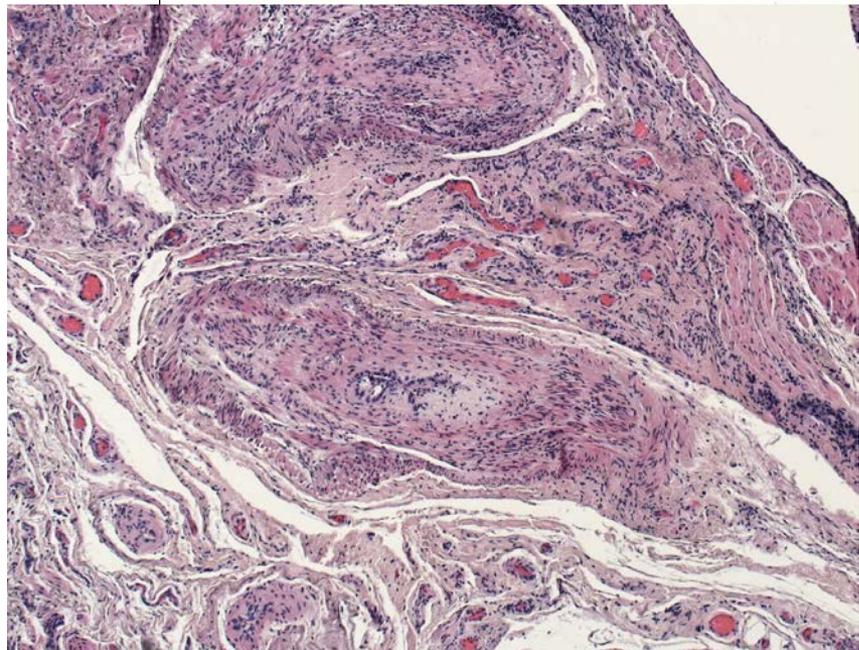
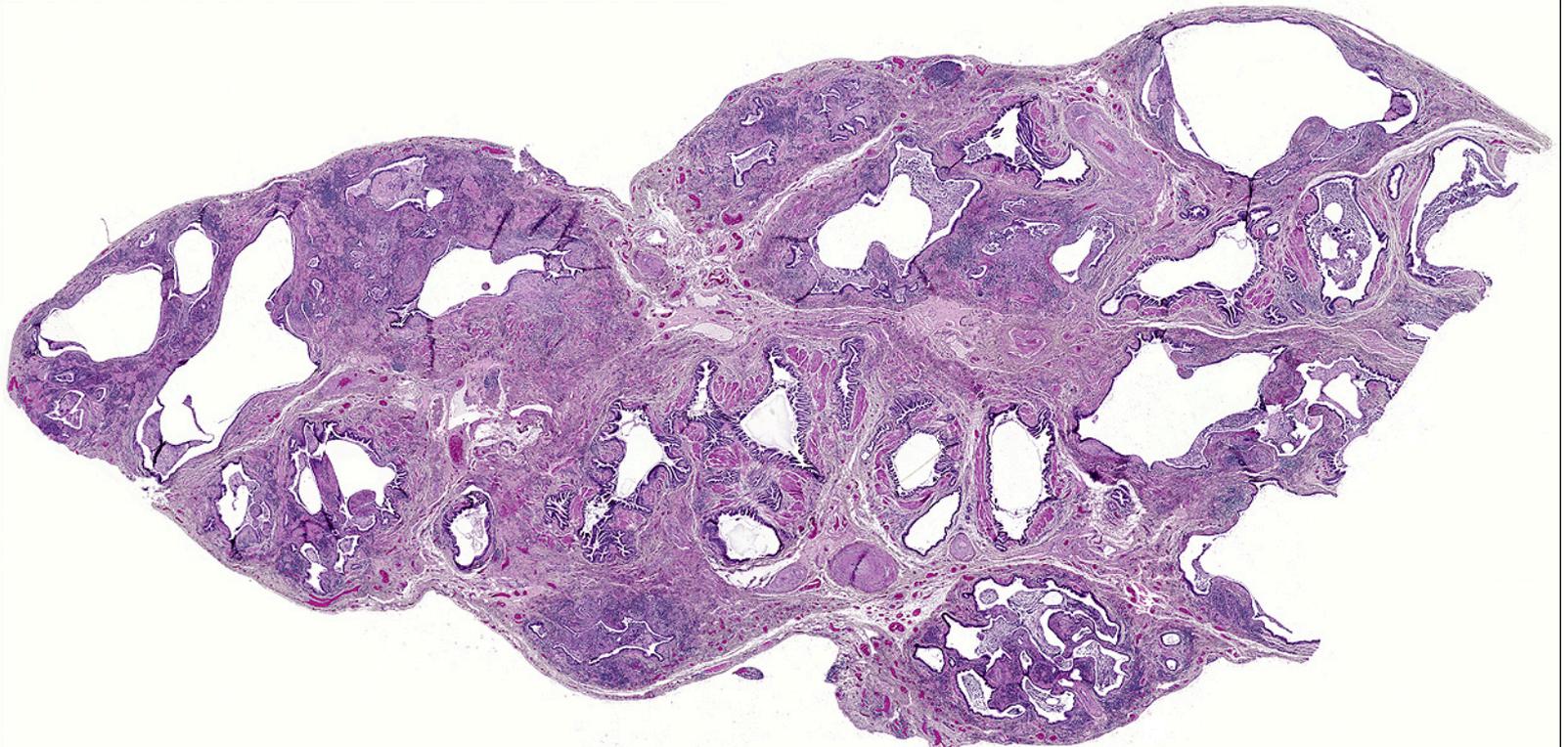




MPR



Minip



MULTIDISCIPLINARY MANAGEMENT OF INTERSTITIAL LUNG DISEASES: A REAL-LIFE STUDY

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Favah Tamirou⁵, Delphine Hoton⁶, Thierry Pieters⁷, Antoine Froidure^{1,2}

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ABSTRACT. Background: The guidelines on idiopathic pulmonary fibrosis (IPF) diagnosis established the crucial role of multidisciplinary discussion (MDD) in the diagnosis of interstitial lung diseases (ILD). However, real-life evaluation of MDD remains scarce. Our aim was to study the impact of a well-structured MDD on etiological assessment, diagnosis, and management of ILD. **Methods:** We collected and analysed all relevant data on patients concerning diagnosis and treatment before and after MDD during the year 2017. **Results:** One hundred fifty patients were included in the analysis. MDD had a significant impact on management: 42% of diagnoses were revised and the number of unclassifiable ILD was significantly reduced. Lung biopsy was performed in 26 patients (12 cryobiopsies and 14 surgical biopsies). The most prevalent diagnoses were connective-tissue disease associated ILD (32%), idiopathic pulmonary fibrosis (23%), hypersensitivity pneumonitis (13%) and granulomatous ILD (7%). MDD led to a change or initiation of treatment in 55% of cases. Nine patients were evaluated for transplantation, 23 patients were screened for academic or sponsored clinical trials and an 8-fold increase in rehabilitation inclusion was observed. **Conclusion:** Our results confirm the benefits of MDD on ILD management and diagnosis. MDD also facilitates access to non-pharmacological therapies and clinical trials. (*Sarcoidosis Vasc Diffuse Lung Dis* 2019; 36 (2): 108-115)

KEY WORDS: interstitial lung diseases, multidisciplinary management

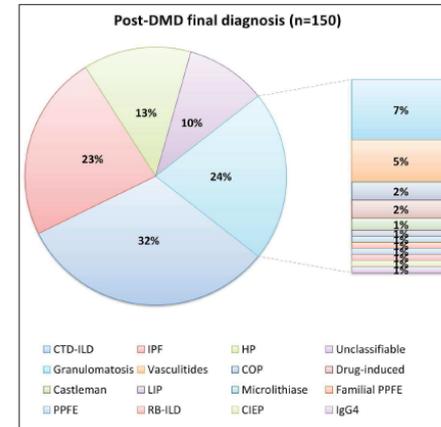
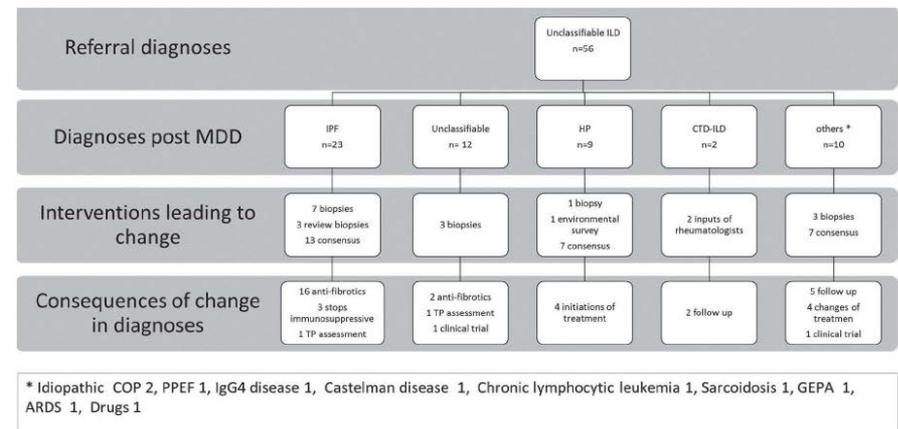


Fig. 1. Proportion of MDD final diagnoses



Cas Clinique

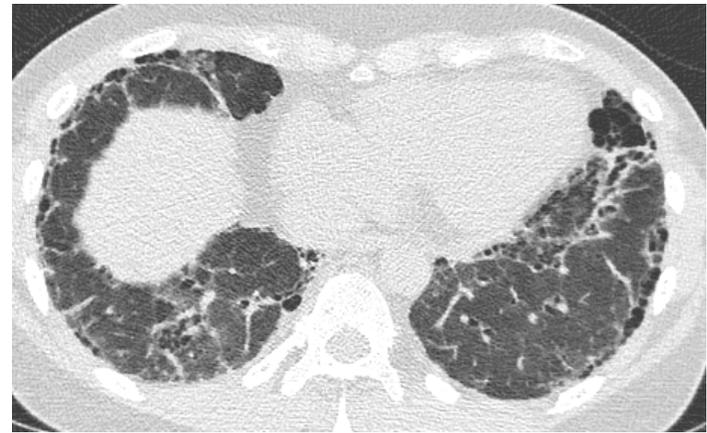
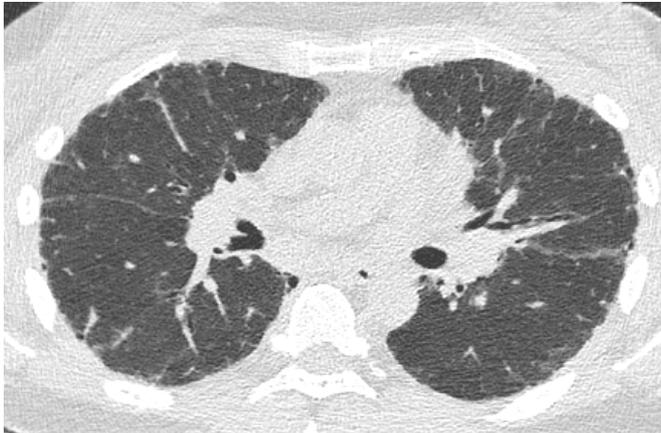
- Femme de 54 ans, se plaignant d'un essoufflement progressif et d'une toux sèche depuis un an
- Non-fumeur, pas d'exposition domestique ni professionnelle
- Aucun symptôme extra-respiratoire ou signe clinique.

- Auscultation : crépitements basaux bilatéraux

- Un scanner du thorax est effectué et comparé à un scanner plus ancien (2015).
- Le dosage des auto-AC et le lavage broncho-alvéolaire n'ont pas contribué à un diagnostic spécifique



17-08-2015



13-08-2018

Type d'atteinte?

1. UIP
2. Possible UIP
3. Fibrose de type Indéterminée
4. Diagnostic alternatif

Table 2 – Diagnostic Categories of UIP based on CT Patterns¹

	Typical UIP CT Pattern	Probable UIP CT Pattern	CT Pattern Indeterminate for UIP	CT features most consistent with non-IPF diagnosis
Distribution	Basal predominant (occasionally diffuse), and <u>subpleural</u> predominant; distribution is often heterogeneous	Basal and <u>subpleural</u> predominant; distribution is often heterogeneous	Variable or diffuse	Upper lung or mid-lung predominant fibrosis; <u>peribronchovascular</u> predominance with <u>subpleural</u> sparing
Features	Honeycombing; reticular pattern with peripheral traction bronchiectasis or <u>bronchiolectasis*</u> ; absence of features to suggest an alternative diagnosis	Reticular pattern with peripheral traction bronchiectasis or <u>bronchiolectasis*</u> ; honeycombing is absent;	Evidence of fibrosis with some inconspicuous features suggestive of non-UIP pattern*	Any of the following: predominant consolidation, extensive pure ground glass opacity (without acute exacerbation), extensive mosaic attenuation with extensive sharply defined lobular air trapping on expiration, diffuse nodules or cysts.

* Reticular pattern is superimposed on ground glass opacity, and in these cases it is usually fibrotic. Pure ground glass opacity, however, would be against the diagnosis of UIP or IPF and would suggest acute exacerbation, hypersensitivity pneumonitis or other conditions.

Diagnostic criteria for idiopathic pulmonary fibrosis: a Fleischner Society White Paper: The Lancet respiratory medicine 2018: 138-153



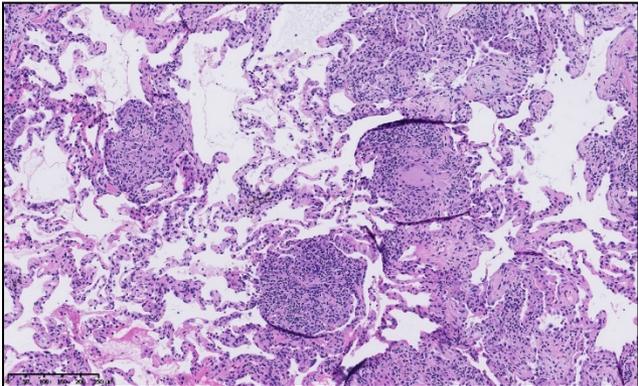
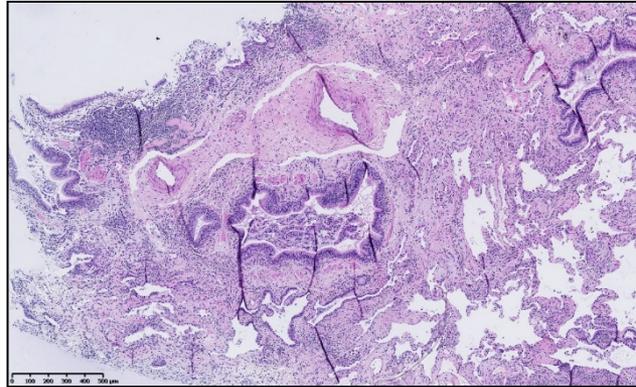
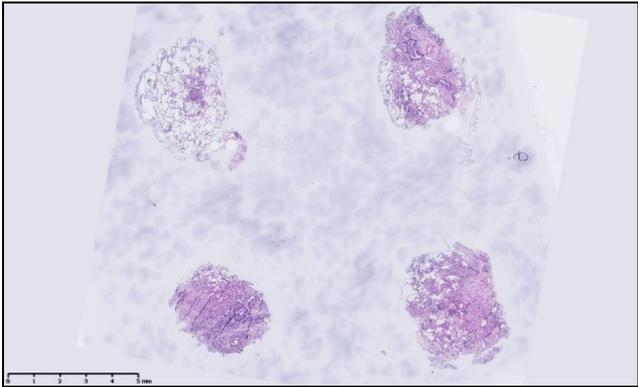
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Décision de la reunion multidisciplinaire

Perform a lung biopsy (surgical or cryos)

Lung cryobiopsies



- ✓ Extensive fibrosis
- ✓ Multiple non-necrotizing granuloma

➔ SARCOIDOSIS

Table 1
Typical and Atypical Features of Pulmonary Sarcoidosis at High-Resolution CT

Typical features

- Lymphadenopathy: hilar, mediastinal (right paratracheal), bilateral, symmetric, and well defined
- Nodules: micronodules (2–4 mm in diameter; well defined, bilateral); macronodules (≥ 5 mm in diameter, coalescing)
- Lymphangitic spread: peribronchovascular, subpleural, interlobular septal
- Fibrotic changes: reticular opacities, architectural distortion, traction bronchiectasis, bronchiolectasis, volume loss
- Bilateral perihilar opacities
- Predominant upper- and middle-zone locations of parenchymal abnormalities

Atypical features

- Lymphadenopathy: unilateral, isolated, anterior and posterior mediastinal
- Airspace consolidation: masslike opacities, conglomerate masses, solitary pulmonary nodules, confluent alveolar opacities (alveolar sarcoid pattern)
- Ground-glass opacities
- Linear opacities: interlobular septal thickening, intralobular linear opacities
- Fibrocystic changes: cysts, bullae, blebs, emphysema, honeycomb-like opacities with upper- and middle-zone predominance
- Miliary opacities
- Airway involvement: mosaic attenuation pattern, tracheobronchial abnormalities, atelectasis
- Pleural disease: effusion, chylothorax, hemothorax, pneumothorax, pleural thickening, calcification
- Pleural plaquelike opacities
- Mycetoma, aspergilloma

Criado E, Sanchez M, Ramirez J, et al. Pulmonary sarcoidosis: typical and atypical manifestations at High resolution CT with pathological correlations. *Radiographics* (2010); 30:1567-1586