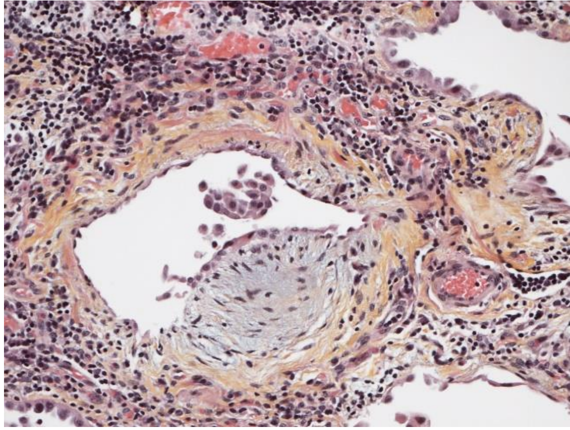
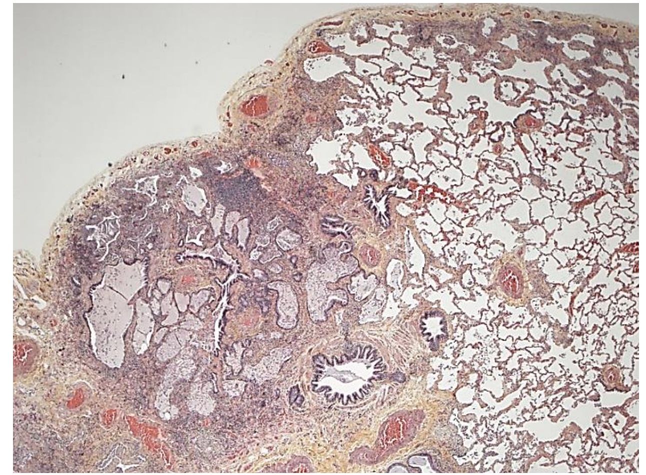




Stratégie diagnostique des Pneumopathies interstitielles diffuses PID

Carrefour de Pathologie 2016

Jeudi 10 novembre 2016

Dr C SAGAN
Service d'Anatomie et Cytologie Pathologiques
CHU Nantes
CENTRE HOSPITALIER UNIVERSITAIRE DE NANTES

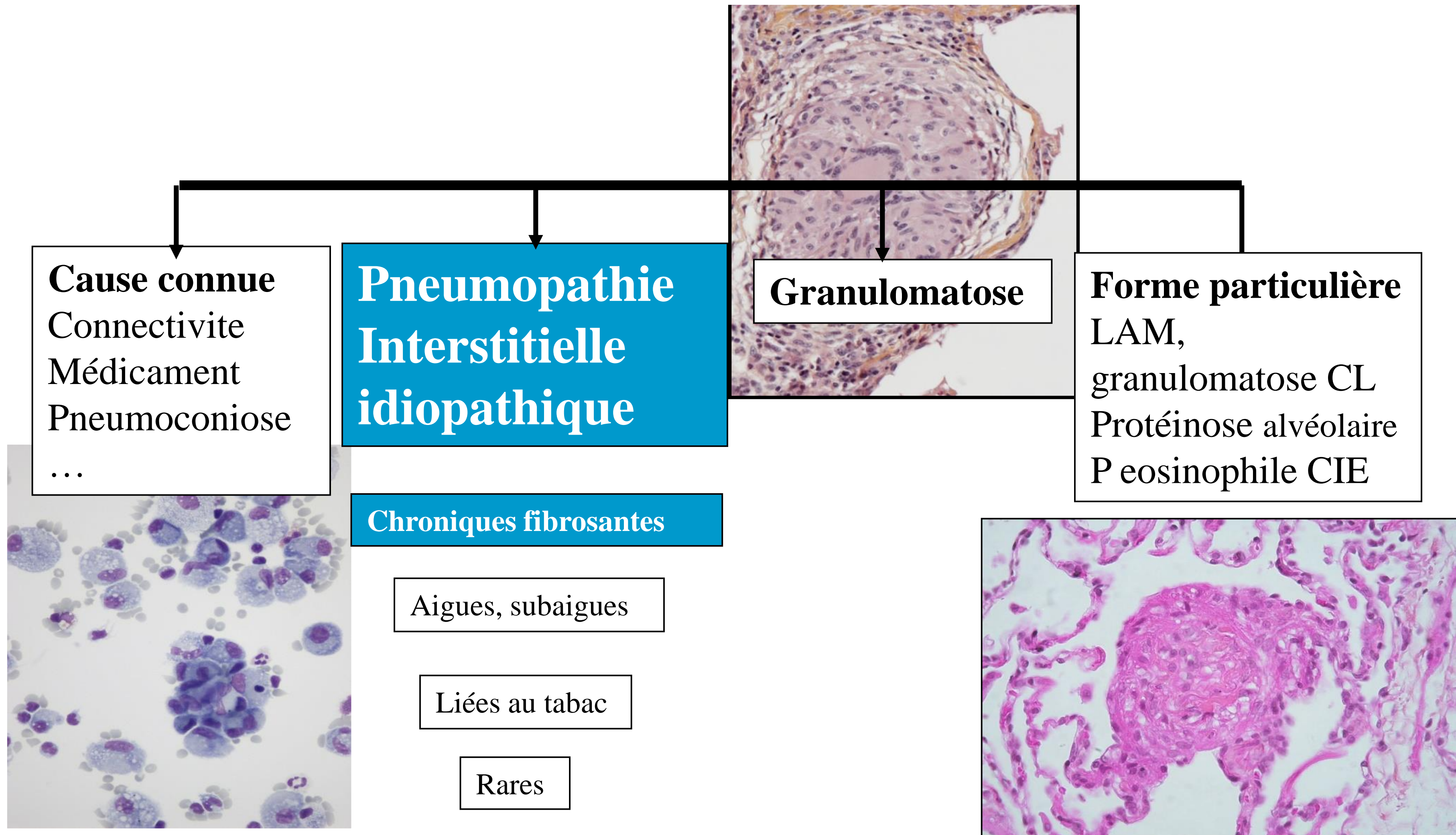
Déclarations d'intérêts

L'objectif de cette déclaration est d'exposer aux congressistes l'existence d'éventuels liens qui pourraient influencer, d'une façon ou d'une autre, votre intervention.

Je déclare ne pas avoir de conflits d'intérêts en rapport avec mon intervention

Illustration de la démarche du pathologiste dans les PID « idiopathiques » fibrosantes

Classification des Pneumopathies interstitielles diffuses 2012



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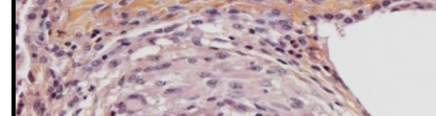
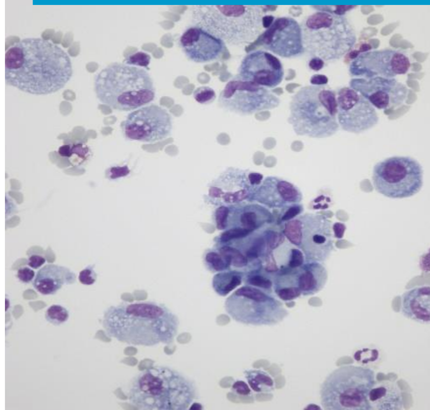


Lésions support histologique de la PFI

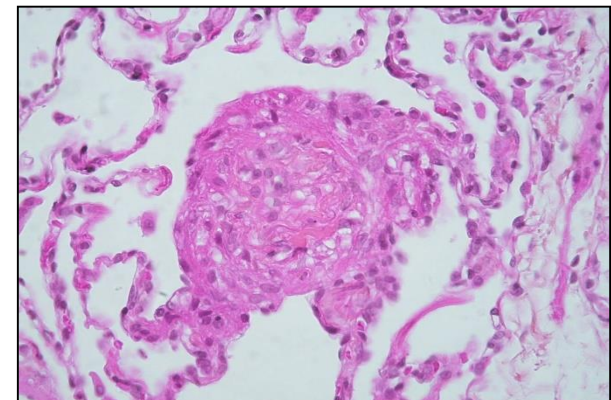
- **Forme la plus fréquente**
- **Pronostic sombre**
- **Nouvelles Thérapeutiques**



Chroniques fibrosantes

Aigues, subaigues

Liées au tabac



Démarche
du pathologiste

Prendre en charge des PID
Impose un respect de prérequis

American Thoracic Society/Eu
Society International Multidis
Classification of the Idiopathi

This Joint Statement of the American Thoracic Society (AT
adopted by the ATS Board of Directors, June 2001 and by T

RECOMMANDATIONS

Recommandations pratiques pour le diagnostic et la prise en charge de la fibrose pulmonaire idiopathique. Élaborées par le centre national de référence et les centres de compétence pour les maladies pulmonaires rares sous l'égide de la Société de pneumologie de langue française

Revue des maladies respiratoires 201 3

An Official American Thoracic Society/Eur
Respiratory Society Statement: Update of
International Multidisciplinary Classification of the
Idiopathic Interstitial Pneumonias

William D. Travis, Ulrich Costabel, David M. Hansell, Talmadge E. King, Jr., David A. Lynch, Andrew G. Nicholson, Christopher J. Ryerson, Jay H. Ryu, Moisés Selman, Athol U. Wells, Jurgen Behr, Demosthenes Bouros, Kevin K. Brown, Thomas V. Colby, Harold R. Collard, Carlos Robalo Cordeiro, Vincent Cottin, Bruno Crestani, Marjolein Drent, Rosalind F. Dudden, Jim Egan, Kevin Flaherty, Cory Hogaboam, Yoshikazu Inoue, Takeshi Johkoh, Dong Soon Kim, Masanori Kitaichi, James Loyd, Fernando J. Martinez, Jeffrey Myers, Shandra Protzko, Ganesh Raghu, Luca Richeldi, Nicola Sverzellati, Jeffrey Swigris, and Dominique Valeyre; on behalf of the ATS/ERS Committee on Idiopathic Interstitial Pneumonias

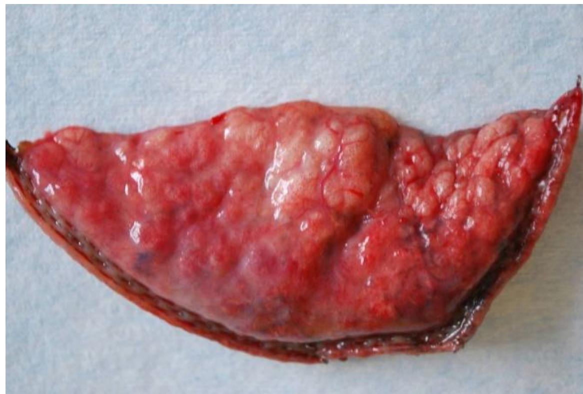
This official statement of the American Thoracic Society (ATS) and the European Respiratory Society (ERS) was approved by the ATS Board of Directors, June 2013, and by the ERS Steering Committee, March 2013

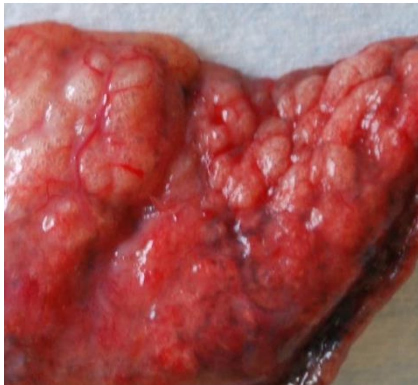
Diagnosis and management of idiopathic pulmonary fibrosis: French practical guidelines

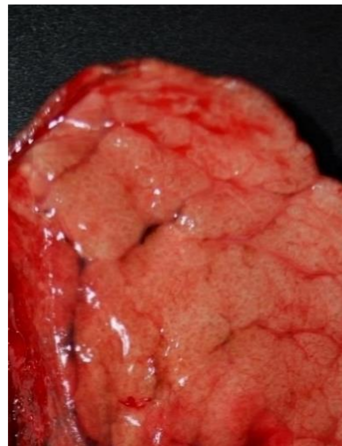
Vincent Cottin, Bruno Crestani, Dominique Valeyre, Benoit Wallaert, Jacques Cadranel, Jean-Charles Dalphin, Philippe Delaval, Dominique Israel-Biet, Romain Kessler, Martine Reynaud-Gaubert, Bernard Aguilaniu, Benoit Bouquillon, Philippe Carré, Claire Danel, Jean-Baptiste Faivre, Gilbert Ferretti, Nicolas Just, Serge Kouzan, François Lebargy, Sylvain Marchand-Adam, Bruno Philippe, Grégoire Prévot, Bruno Stach, Françoise Thivolet-Béjui, Jean-François Cordier and the French National Referer for Rare Lung Diseases

Eur Resp Rev 2014

1) Prendre en charge des prélèvements adaptés et de qualité







Biopsies chirurgicales de bonne qualité

- Adapter sites de biopsies aux lésions TDM
- Pas dans lingula et lobe moyen
- Sur plusieurs sites
- **En territoire lésionnel et non lésionnel**
- A l'état frais

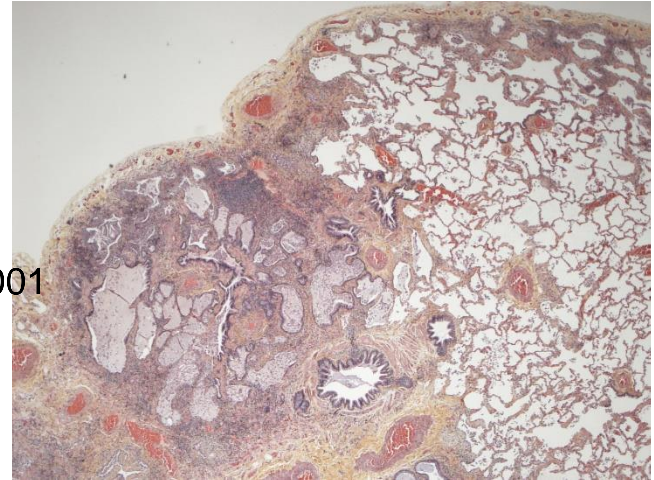
2) Connaitre les critères anapath définissant les profils lésionnels des PID

2001

American Thoracic Society/European Respirat Society International Multidisciplinary Conser Classification of the Idiopathic Interstitial Pneumonias

This Joint Statement of the American Thoracic Society (ATS), and the European Respiratory Society (ERS) was adopted by the ATS Board of Directors, June 2001 and by The ERS Executive Committee, June 2001

Am J Respir Crit Care Med Vol 2001



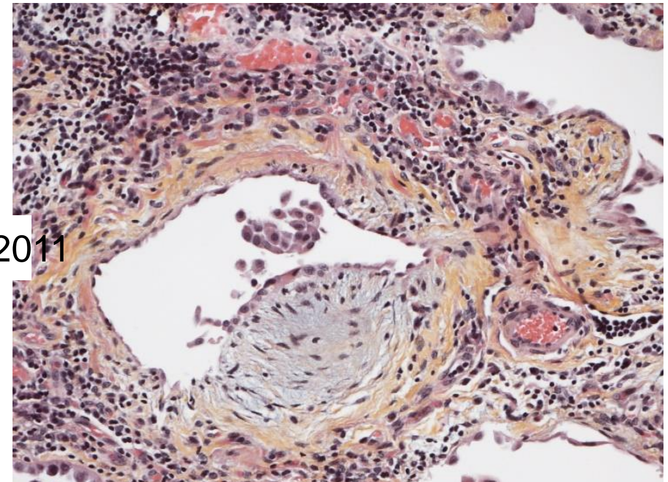
An Official ATS/ERS/JRS/ALAT Statement: Idiop Pulmonary Fibrosis: Evidence-based Guidelines Diagnosis and Management

2011

Ganesh Raghu, Harold R. Collard, Jim J. Egan, Fernando J. Martinez, Juergen Behr, Kevin K. Bro Thomas V. Colby, Jean-François Cordier, Kevin R. Flaherty, Joseph A. Lasky, David A. Lynch, Jay Jeffrey J. Swigris, Athol U. Wells, Julio Ancochea, Demosthenes Bouros, Carlos Carvalho, Ulrich Costabel, Masahito Ebina, David M. Hansell, Takeshi Johkoh, Dong Soon Kim, Talmadge E. King, Jr., Yasuhiro Kondoh, Jeffrey Myers, Nestor L. Müller, Andrew G. Nicholson, Luca Richeldi, Moisés Selman, Rosalind F. Dudden, Barbara S. Griss, Shandra L. Protzko, and Holger J. Schünemann, on behalf of the ATS/ERS/JRS/ALAT Committee on Idiopathic Pulmonary Fibrosis

This Official Statement of the American Thoracic Society (ATS), the European Respiratory Society (ERS), the Japanese Respiratory Society (JRS), and the Latin American Thoracic Association (ALAT) Was Approved by the ATS Board of Directors, November 2010, the ERS Executive Committee, September 2010, the JRS Board of Directors, December 2010, and the ALAT Executive Committee, November 2010

Am J Respir Crit Care Med Vol 2011



2) Connaitre les critères anapath définissant le profil lésionnel de PIC

PIC certaine (présence des 4 critères)	PIC probable (présence des 3 critères)	PIC possible (présence des 3 critères)	Signes incompatibles avec un aspect de PIC (au moins un des 6 critères)
• Fibrose marquée/remodelage architectural, +/- rayon de miel de distribution sous-pleurale/paraseptale prédominante • Atteinte disséminée du parenchyme par la fibrose • Présence de foyers fibroblastiques • Absence de signes suggérant un autre diagnostic	• Fibrose marquée/remodelage architecturale, +/ rayon de miel • Absence soit d'atteinte disséminée du parenchyme par la fibrose, soit de foyers fibroblastiques (mais pas d'absence des 2 critères) • Absence de signes suggérant un autre diagnostic	• Fibrose parenchymateuse disséminée ou diffuse, avec ou sans inflammation interstitielle • Absence d'autres critères de PIC • Absence de signes suggérant un autre diagnostic	• Membranes hyalines • Pneumopathie organisée (bourgeons fibro-inflammatoires alvéolaires) • Granulomes • Infiltration inflammatoire interstitielle marquée à distance du rayon de miel • Anomalies prédominantes centrées sur les voies aériennes • Autres signes suggérant un autre diagnostic

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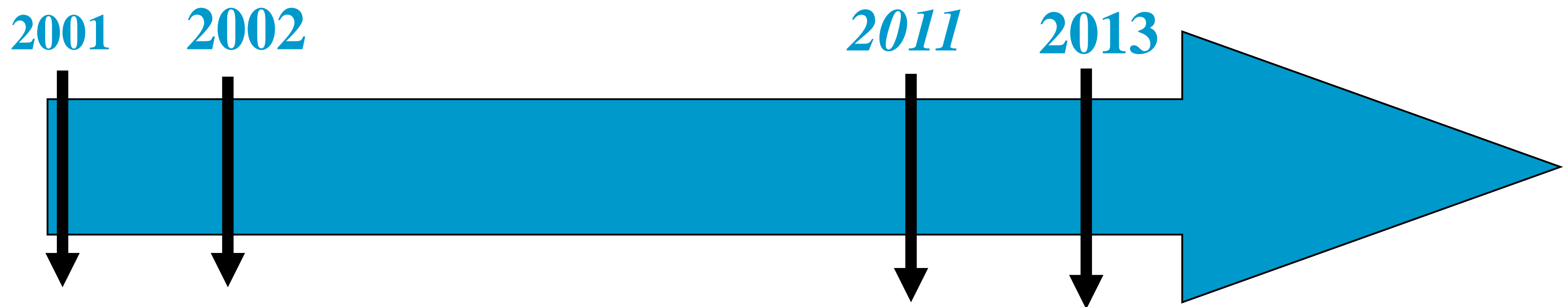
3) Place de la Discussion Multidisciplinaire

- **Évolution des critères diagnostiques**



ATS/ERS consensus
Am J respir Crit care Med 2000

ATS/ERS consensus
Am J respir Crit care Med 2011

ATS/ERS consensus
Am J respir Crit care Med 2002

ATS/ERS consensus
Am J respir Crit care Med 2013

3) Place de la Discussion Multidisciplinaire

- Évolution des critères d

2001 2002

GENERAL PROGRESS IN IIPS SINCE 2002

Multidisciplinary Approach

The process of achieving a multidisciplinary diagnosis in a patient with IIP is dynamic, requiring close communication between clinician, radiologist, and when appropriate, pathologist (1). Clin-

- *Question*: Should a multi-disciplinary discussion be used in the evaluation of suspected IPF?

The diagnosis of IPF is, by definition, multidisciplinary, drawing on the expertise of experienced clinicians, radiologists, and pathologists. Proper communication between the various disciplines involved in the diagnosis of IPF (pulmonary, radiology, pathology) has been shown to improve inter-observer agreement among experienced clinical experts as to the ultimate diagnosis (111, 126).

Recommendation: We recommend that a multi-disciplinary discussion should be used in the evaluation of IPF (strong recommendation, low-quality evidence).

Values: This recommendation places a high value on the accurate diagnosis of IPF and a low value on the access to and availability of experts for multidisciplinary discussion.

Remarks: It is recognized that a formal multidisciplinary discussion (MDD) between the treating pulmonologist, radiologist, and pathologist is not possible for many practitioners. Effort should be made, however, to promote verbal communication between specialties during the evaluation of the case. There are data to suggest that the accuracy of diagnosis is improved through MDD among ILD experts compared with MDD among specialists in the community setting (126); timely referral to ILD experts is encouraged. (Vote: 23 for the use of multidisciplinary discussion, none against the use of multidisciplinary discussion, no abstentions, 8 absent.)

00

ed 2002

PRINCIPLES GUIDING THE ASSESSMENT OF PATIENTS WITH IDIOPATHIC INTERSTITIAL PNEUMONIAS

Before describing the features of each IIP, it is necessary to consider the principles that apply to the general assessment of patients with IIPs. The new classification of IIP is based on clinical, radiological, and pathological criteria.

The Diagnostic Process Is Dynamic

The process of achieving a diagnosis in a patient with IIP is dynamic, requiring close communication between clinician, radiologist, and pathologist. For example, a pathologist is at a disadvantage if asked to interpret a lung biopsy without a relevant history of clinical presentation, radiologic findings, occupational exposure, smoking status, and associated diseases. Also, once a pathologist has recognized a histologic pattern such as NSIP, the clinician needs to go back to the patient and check carefully for antigen exposure that could account for hypersensitivity pneumonitis, laboratory or clinical features of collagen

nsus

care Med 2011

consensus

pir Crit care Med 2013

3) Place de la Discussion Multidisciplinaire

- Évolution des critères d

20... 2002

GENERAL PROGRESS IN IIPS SINCE 2002

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It appears the current **gold standard** for the diagnosis of idiopathic interstitial pneumonia is a dynamic integrated process that requires direct interaction between clinicians and radiologists as well as pathologists when a surgical lung biopsy is available.

- *Question*: Should a ... the evaluation of su...

The diagnosis of ... drawing on the ... ogists, and path... the various dis... (pulmonary, ra... improve inter-... clinical experts as to the ultimate diagnosis (111, 126).

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...care Med 2011

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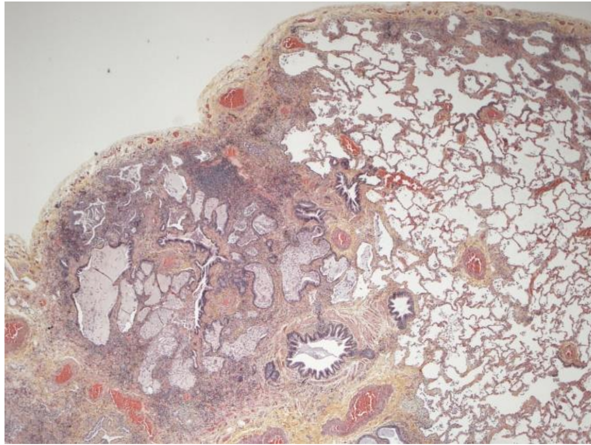
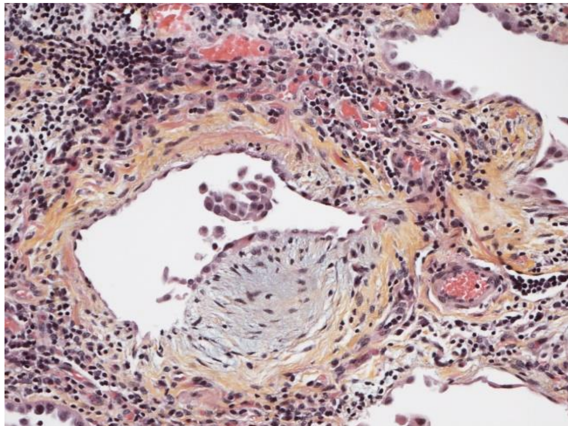
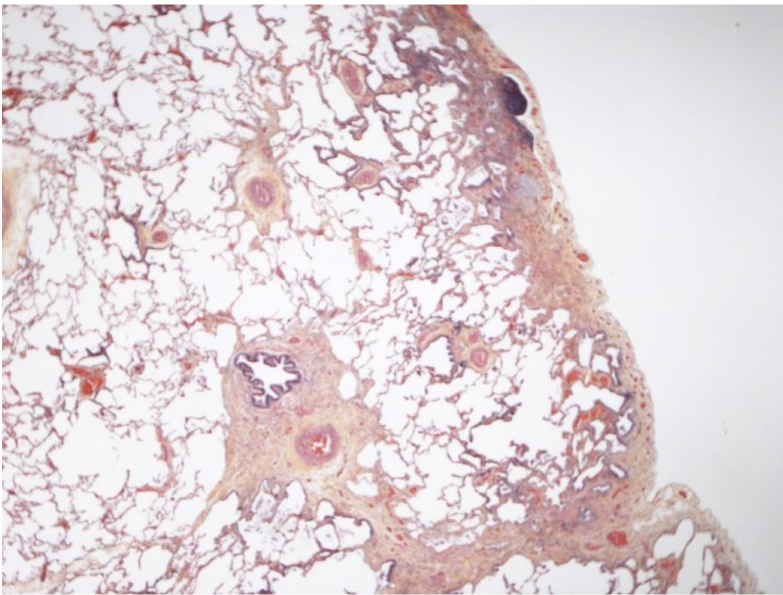
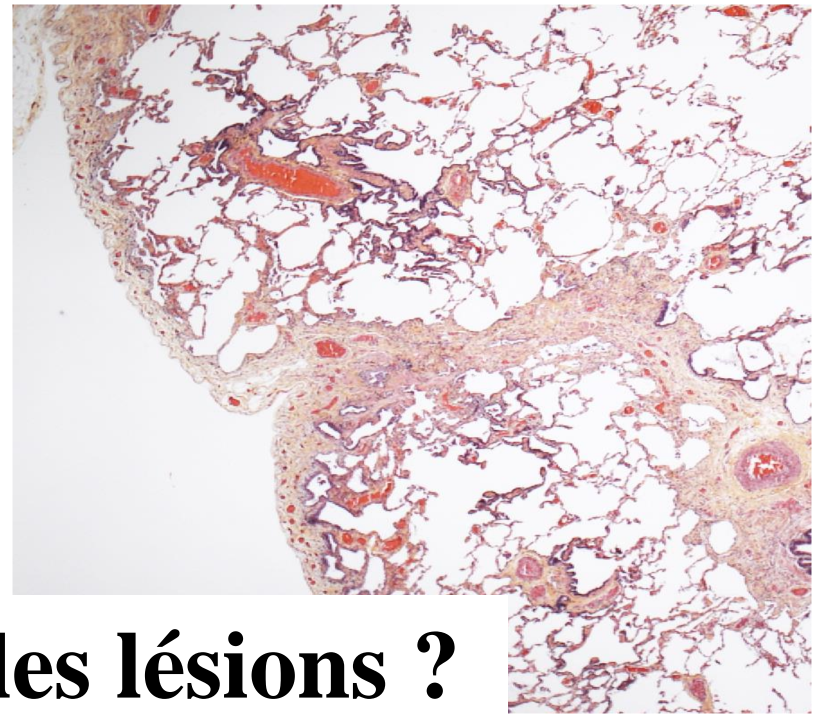
Démarche du pathologiste



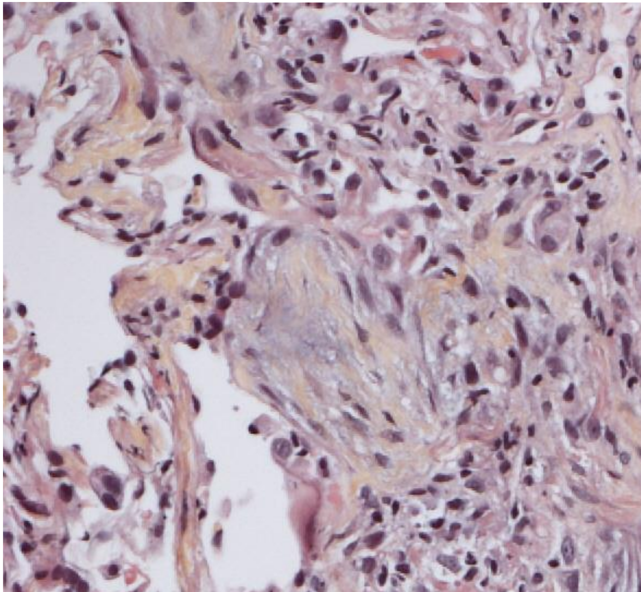


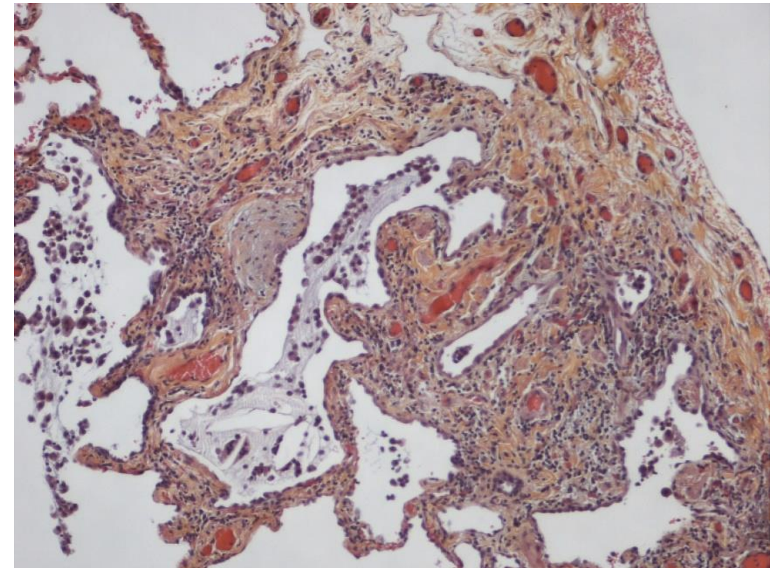
Analyse sémiologique

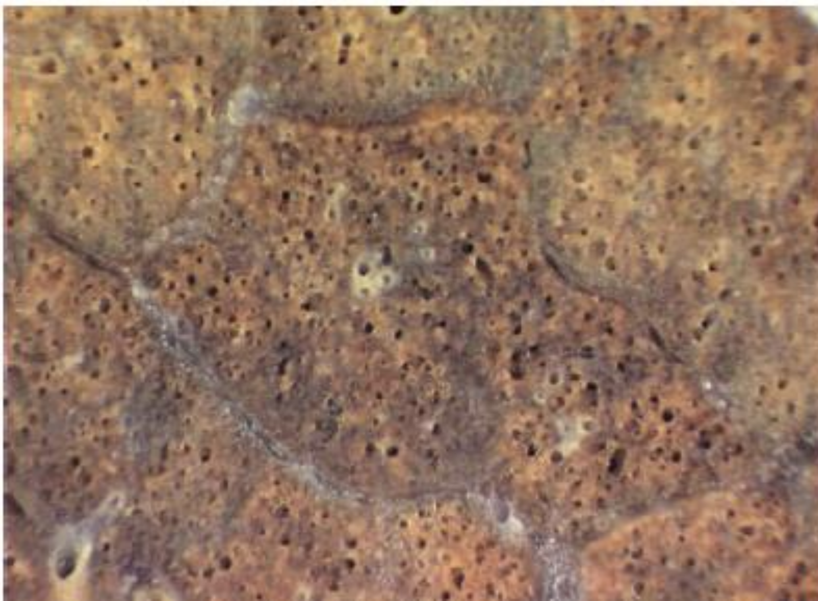




- **Où siègent les lésions ?**



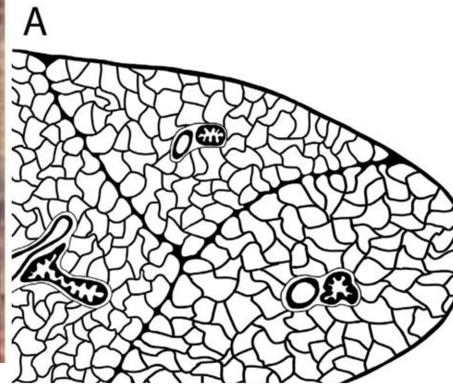


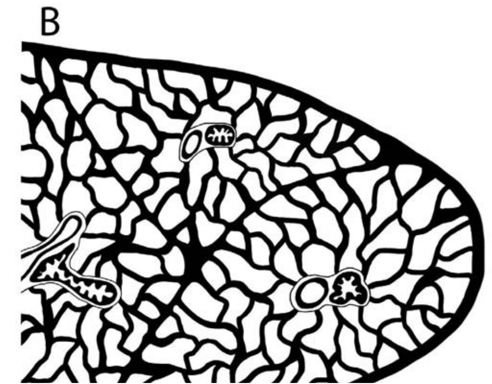


Update on Pulmonary Fibrosis

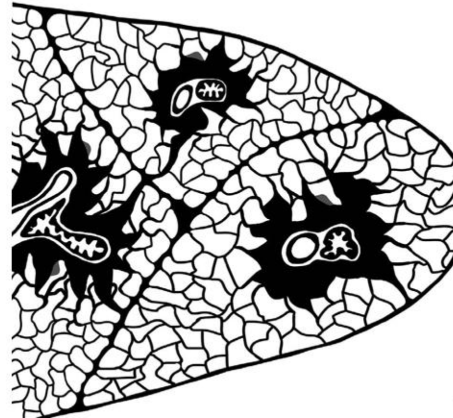
Not All Fibrosis Is Created Equally

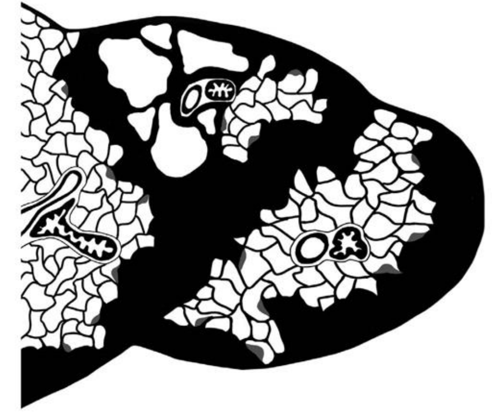
Maxwell L. Smith, MD



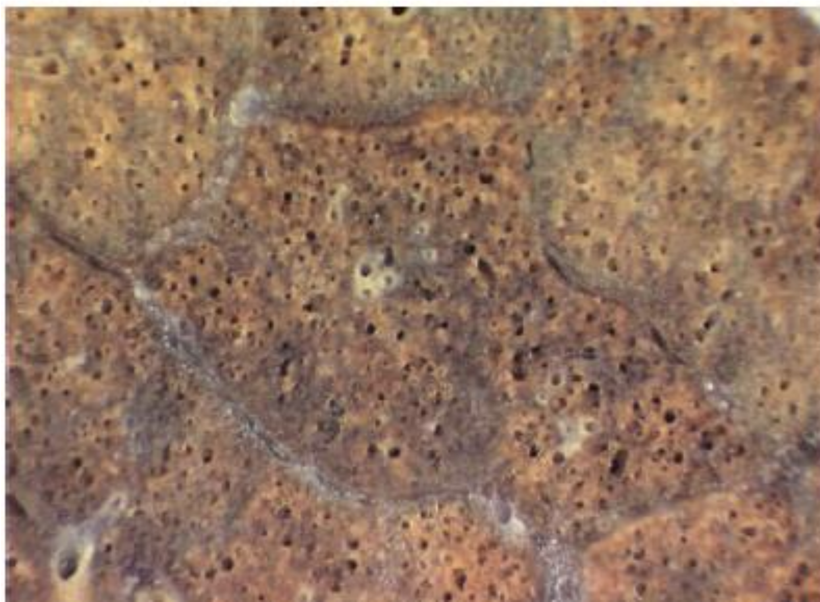


- **Où siegent les lésions dans le lobule ?**





Arch Pathol Lab Med 2016



Update on Pulmonary Fibrosis

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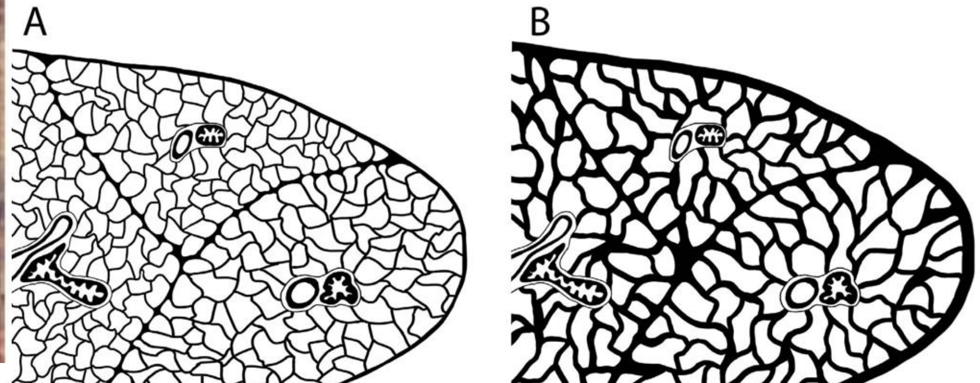
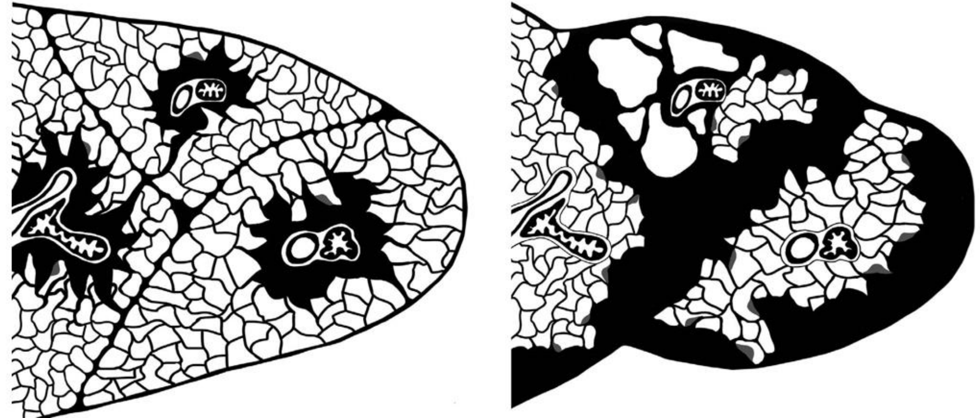


- **Où siegent les lésions dans le lobule**

Disposition architecturale dans le lobule est un critère positif pour certains profils lésionnels
Tous les prélèvements ne sont pas adaptés pour répondre aux questions posées



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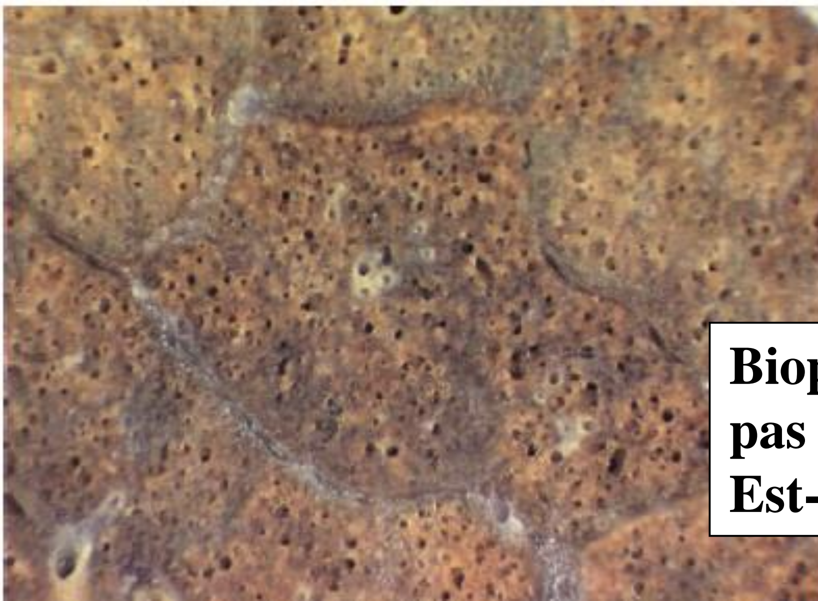

Update on Pulmonary Fibrosis

Not All Fibrosis Is Created Equally

Maxwell L. Smith, MD

A

B

Biopsies transbronchique standards
pas adaptées pour répondre à la question
Est-ce qu'il y a des lésions support de PII

- **Où siegent les lésions dans le lobule**

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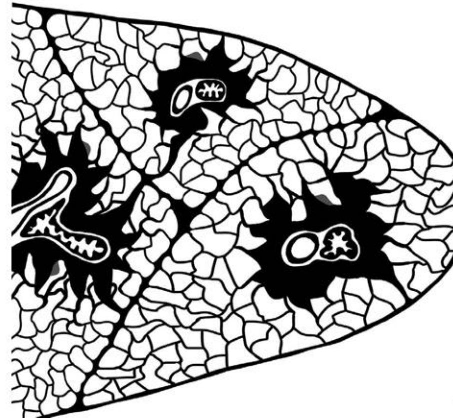
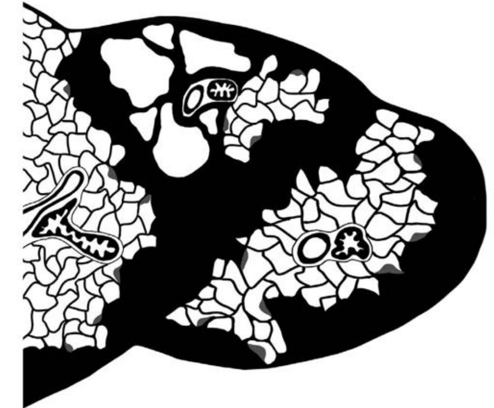
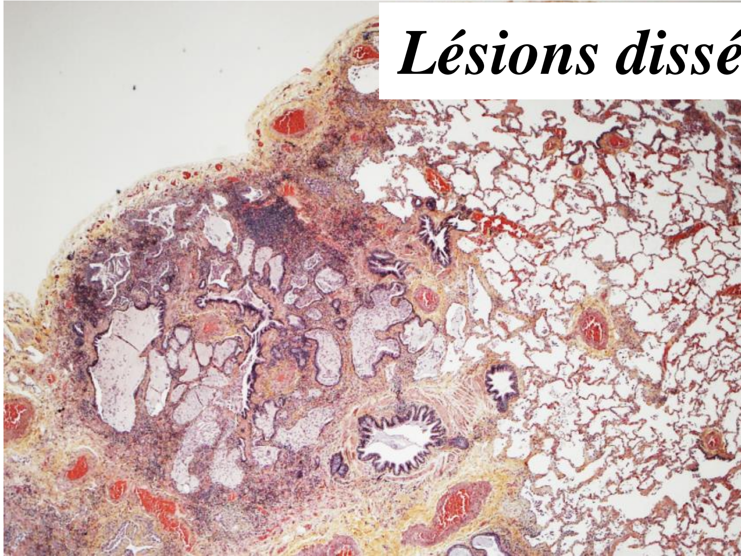
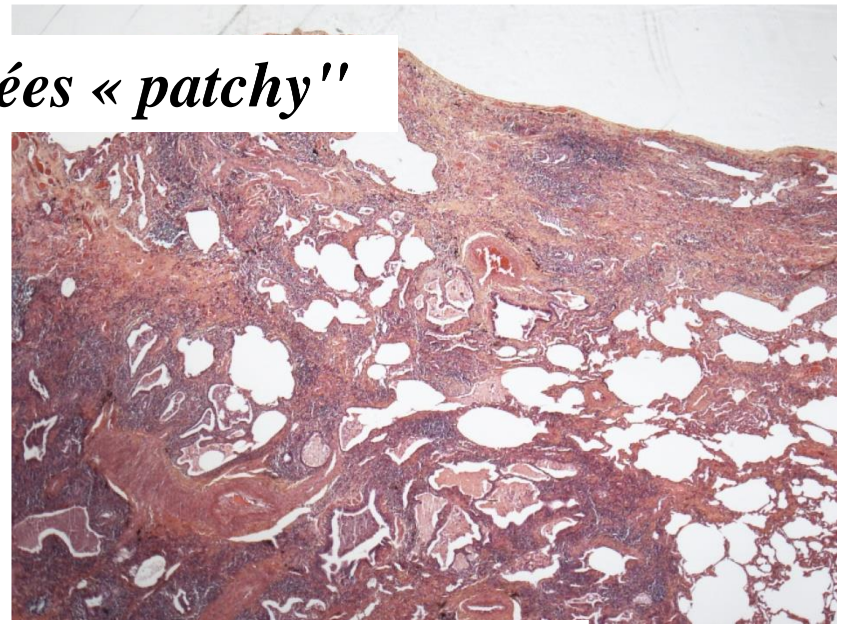




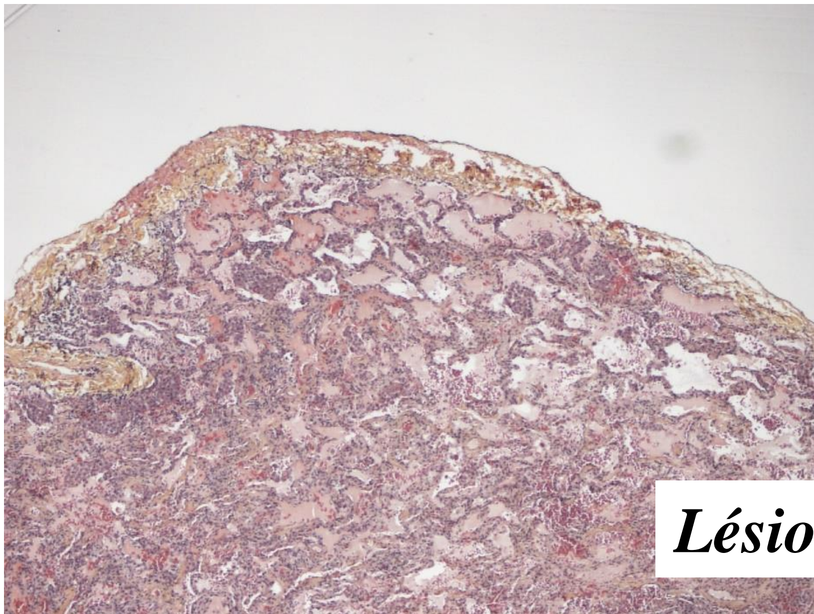
Arch Pathol Lab Med 2016

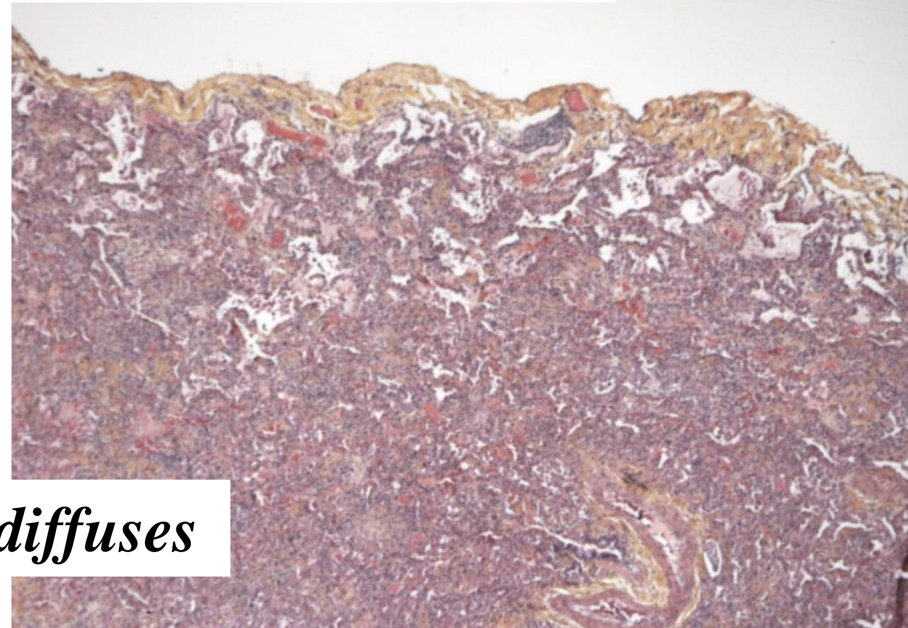
Lésions disséminées « patchy''



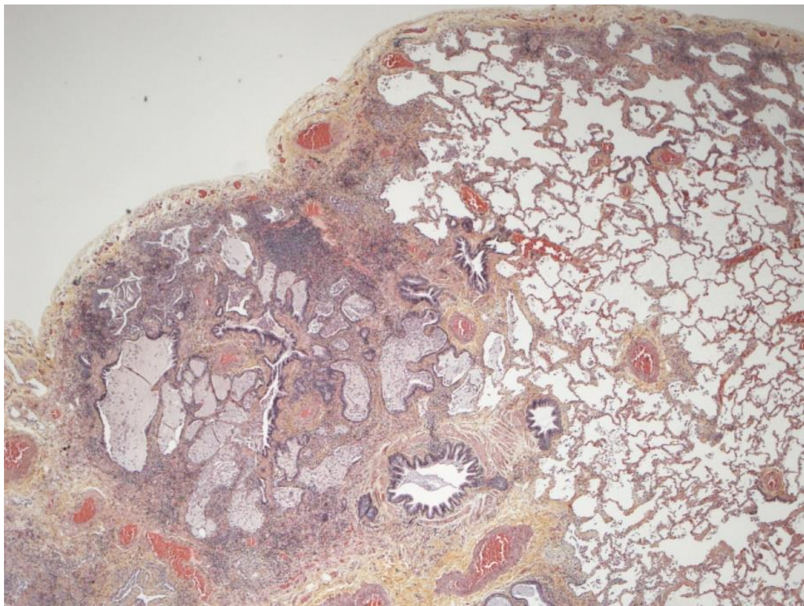
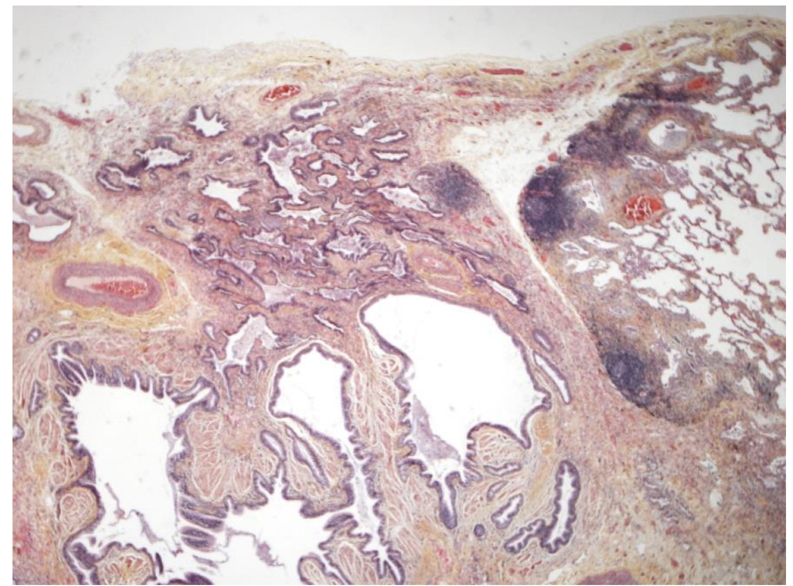


- **Quelle est la diffusion des lésions ?**

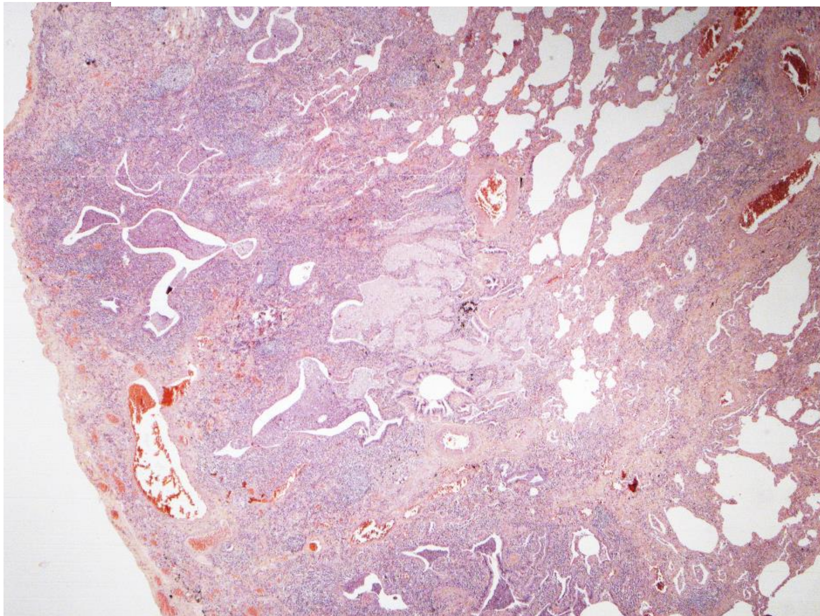
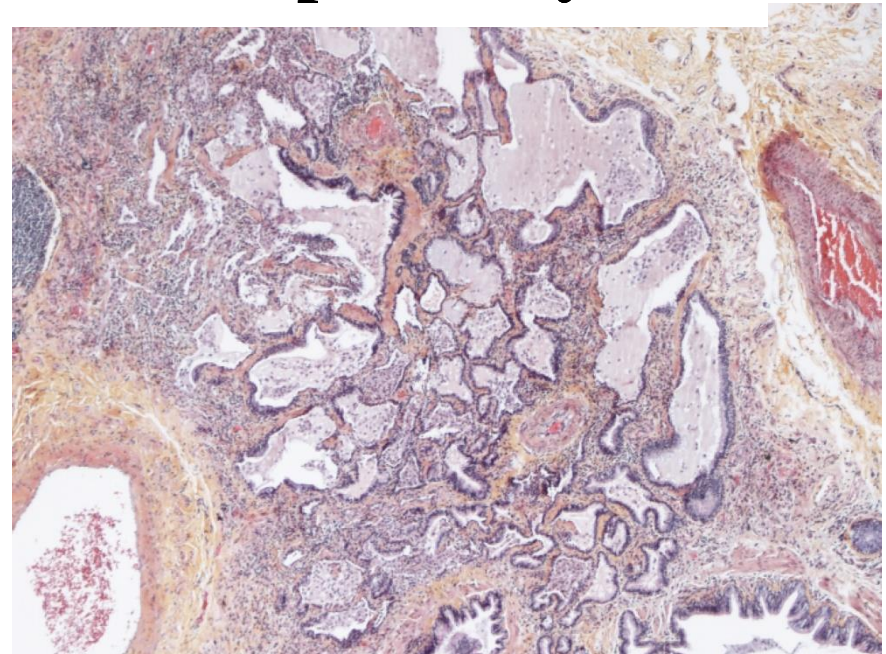


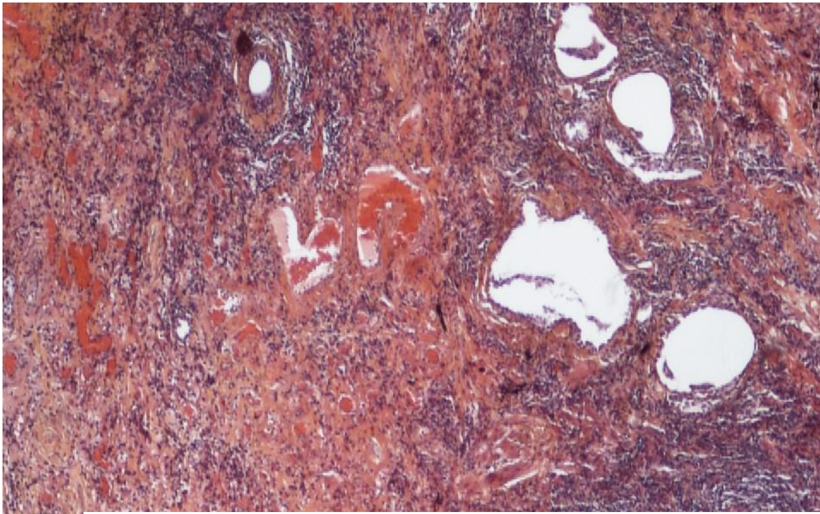


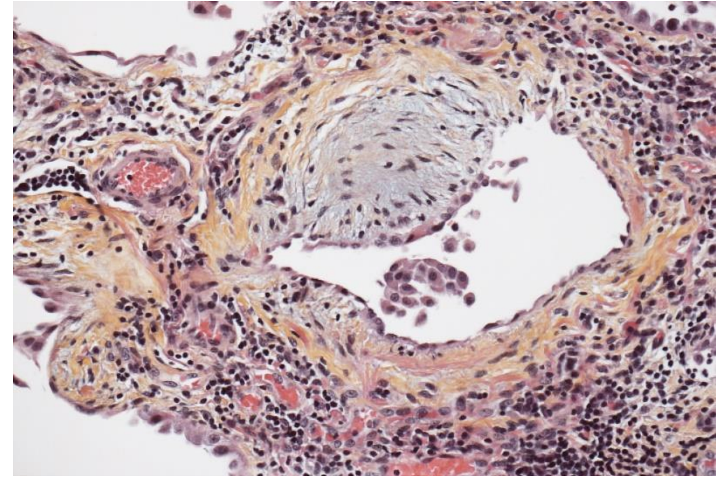
Lésions diffuses

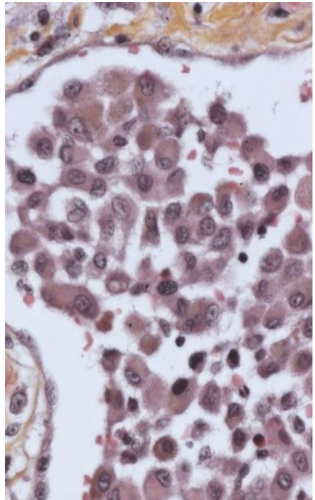
- **Quel est le retentissement sur le parenchyme ?**

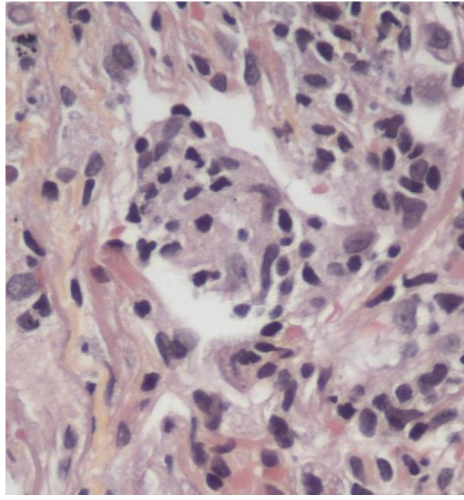



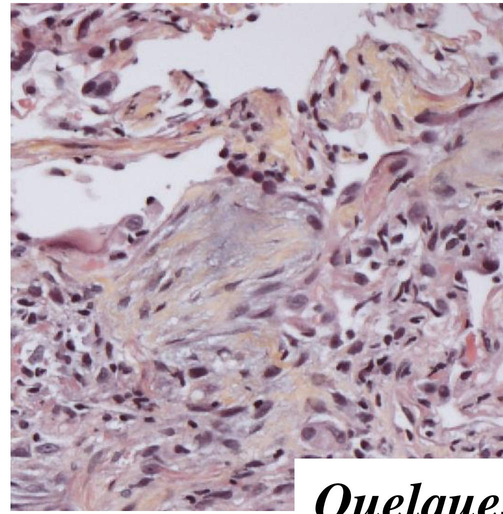


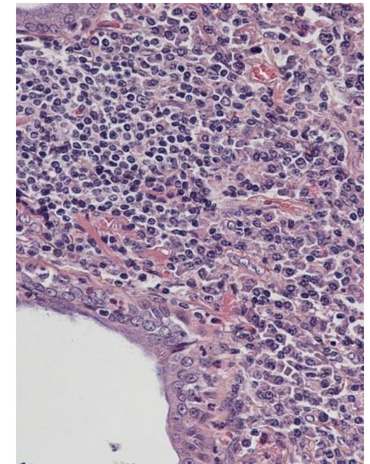


- **Quelles sont les lésions élémentaires ?**
- **Quelle est la lésion prédominante ?**





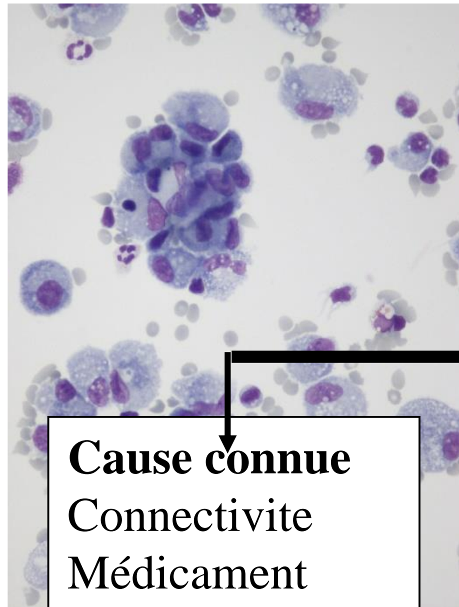


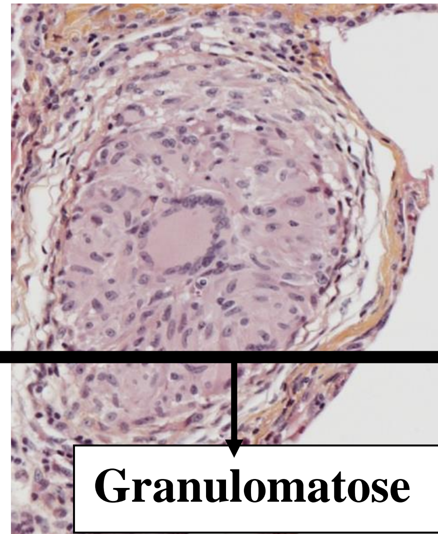


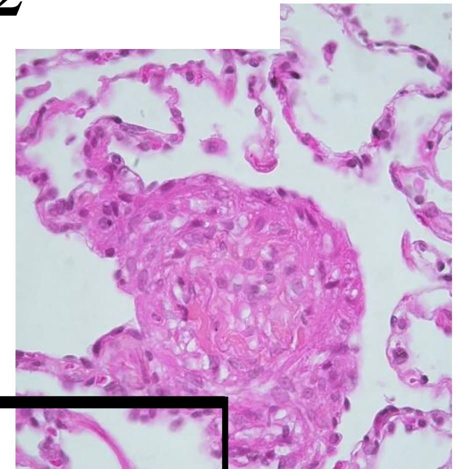
Quelques lésions élémentaires

Démarche du pathologiste identique à celle du clinicien, radiologue
Est-ce

- **Une infection**
- **Un cancer**
- **Une PID selon la *classification 2012***







Cause connue
Connectivite
Médicament
Pneumoconiose
..

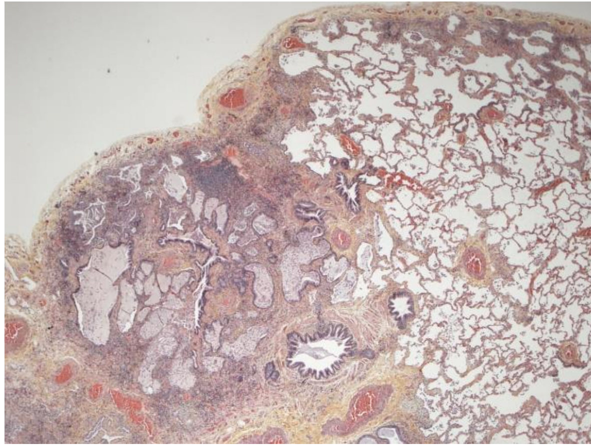
Pneumopathie Interstitielle idiopathique

Granulomatose

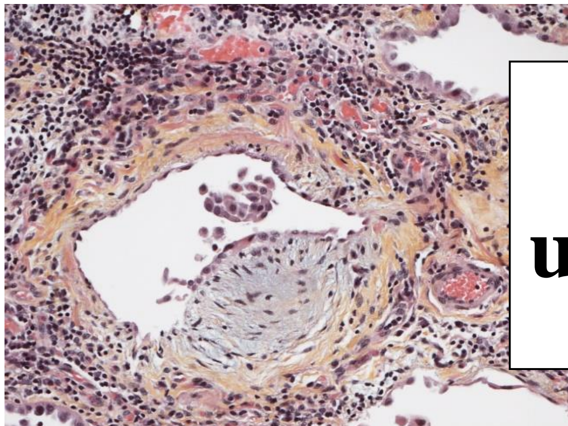
Forme particulière
LAM,
granulomatose CL
Protéinose alvéolaire
P eosinophile CIE

Classification des Pneumopathies interstitielles diffuses 2012

Démarche du pathologiste ?



Analyse sémiologique



Proposer
un profil ou « pattern » lésionnel

Observations

Observations

Femme de 65 ans tabagique
Toux dyspnée
pas d'hippocratisme digital
Pas de manifestation extra-thoracique
pas d'exposition aérocontaminant pas de médicament
Bilan biologique normal (pas d'anomalie auto-imunm)
EFR . Syndrome restrictif TVO
TDM : syndrome interstitiel bilatéral

Discussion Multidisciplinaire DMD :

- TDM : PIC possible
- décision de biopsie chirurgicale

BIOPSIES PULMONAIRES DU COTE GAUCHE SOUS VIDEO-THORACOSCOPIE ET LIBERATION D'UNE SYMPHYSE PLEURALE

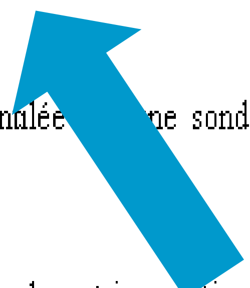
Données pré-opératoires :
Il s'agit d'une patiente qui présente une pneumopathie interstitielle diffuse dont le bilan étiologiqu[illegible] pas été contributif.
Le staff médico-chirurgical avait préconisé la réalisation de biopsies pulmonaires.

Préparation :
Patiente intubée et ventilée par une sonde trachéale sélective sous anesthésie générale et installation en décubitus latéral droit.

Intervention :
Mise en place des trocards en triangulation dans le 5ème et le 7ème espace inter-costal.
Il existe une symphyse pleurale apicale qui est libérée en totalité.
On réalise des biopsies pulmonaires par des résections atypiques sur 3 sites différents au niveau du lobe inférieur, de la lingula et de la jonction culmen-lingula.
Ces prélèvements sont adressés pour examen anatomopathologique.
Vérification de l'hémostase.
Mise en place d'un drain thoracique de taille 24.
Fermeture des orifices de trocard par des points de Blair.
Pansement.

BIOPSIES PULMONAIRES DU COTE GAUCHE SOUS VIDEO-THORACOSCOPIE ET LIBERATION D'UNE SYMPHYSE PLEURALE

<u>Données pré-opératoires</u> :
Il s'agit d'une patiente qui présente une pneumopathie interstitielle diffuse dont le bilan étiologique n'a pas été contributif.
Le staff médico-chirurgical avait préconisé la réalisation de biopsies pulmonaires.



<u>Préparation</u> :
Patiente intubée et ventilée [illegible]ne sonde trachéale sélective sous anesthésie générale et installation en décubitus latéral droit.

<u>Intervention</u> :
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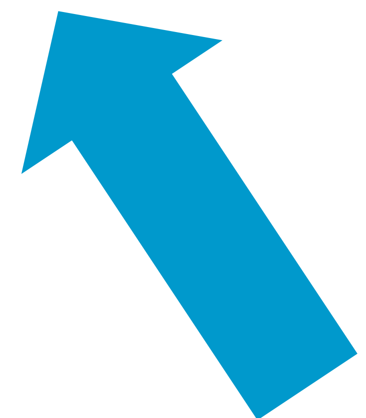
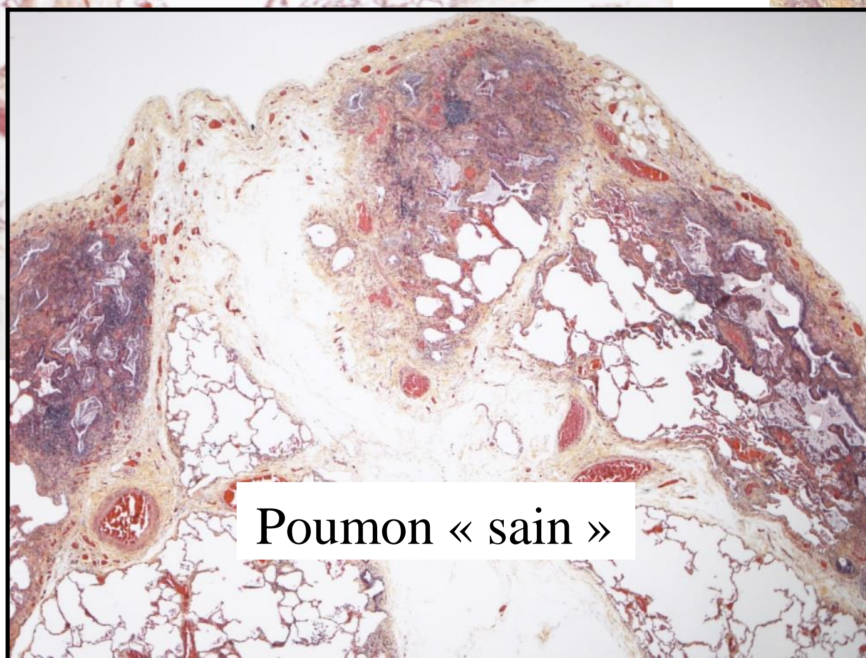


Sur les 3 biopsies

Poumon « sain »

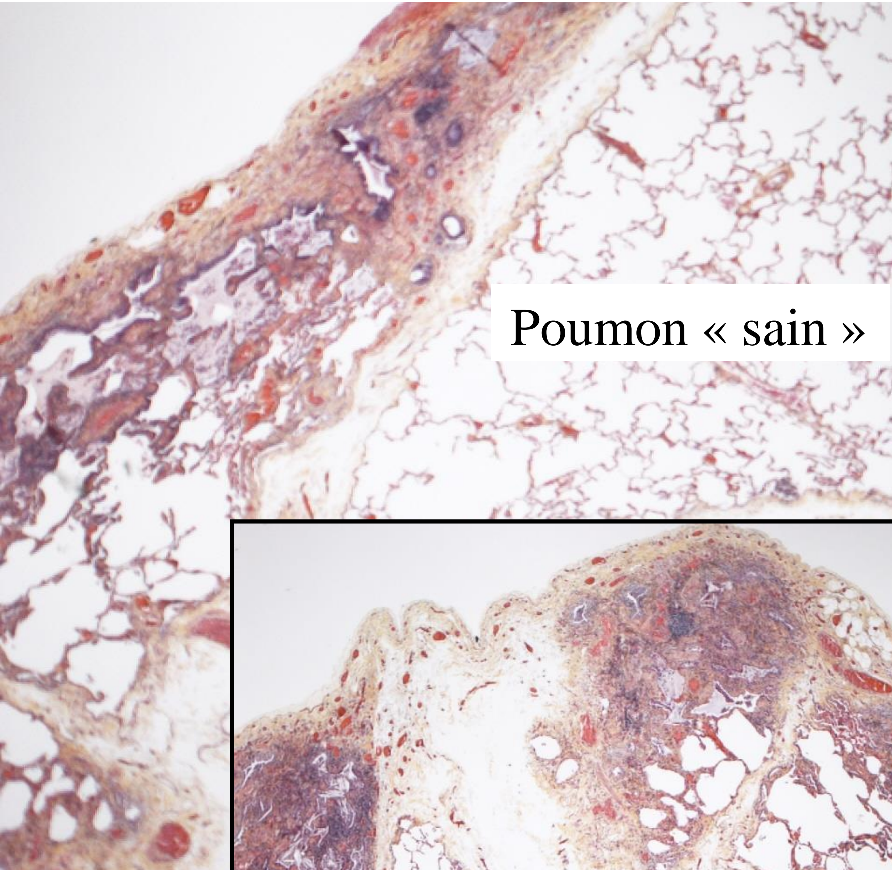


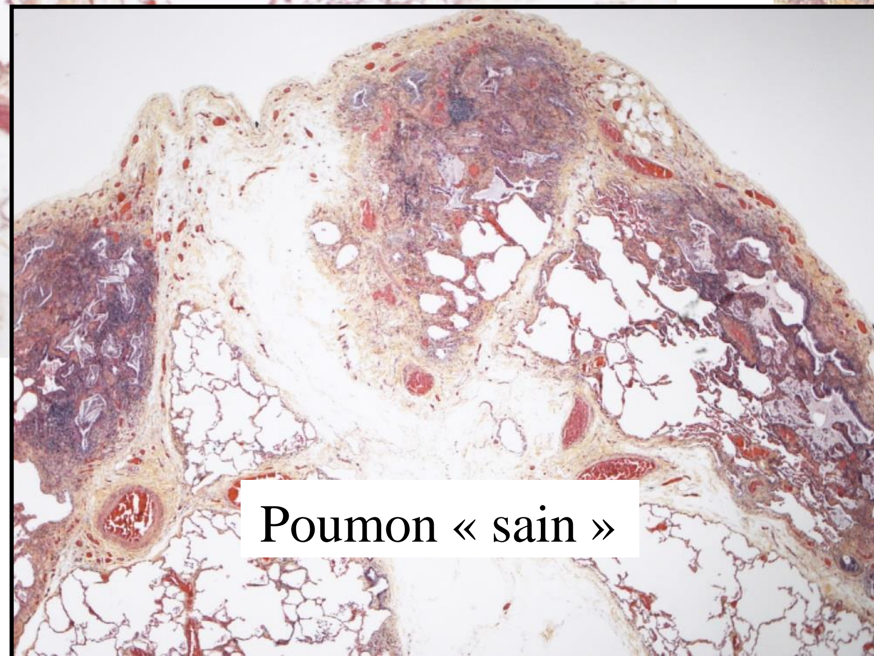
Poumon « sain »

Lésions de distribution hétérogène

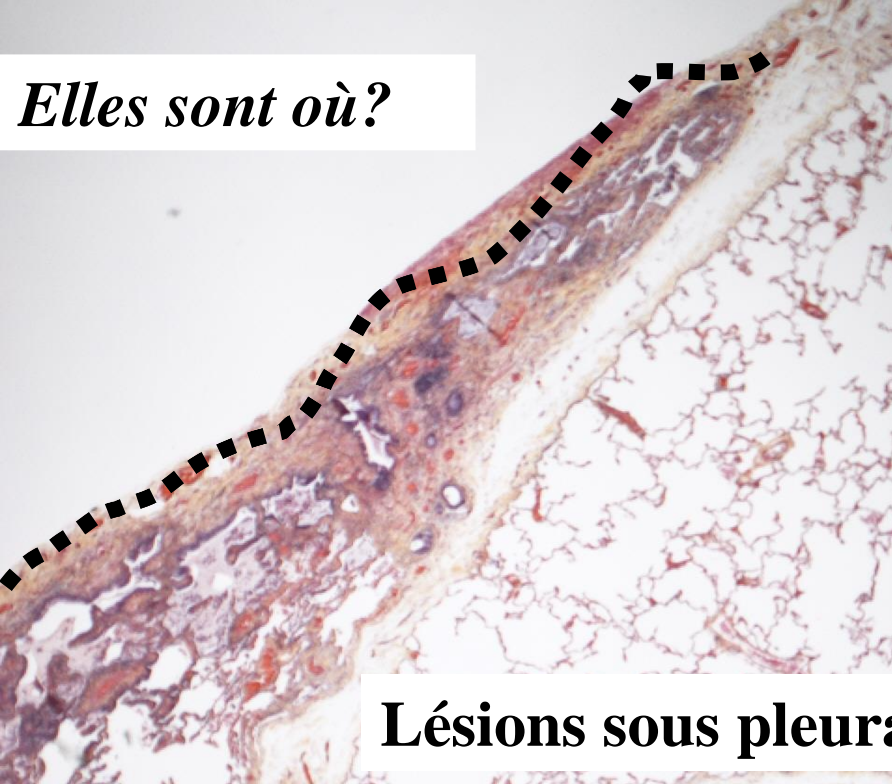
lésions disséminées

Hétérogénéité spatiale



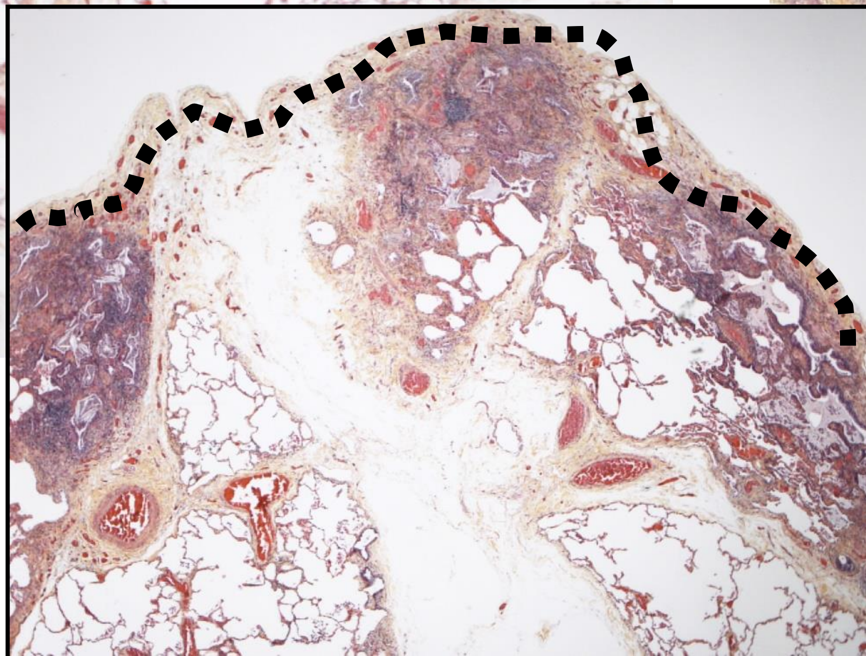


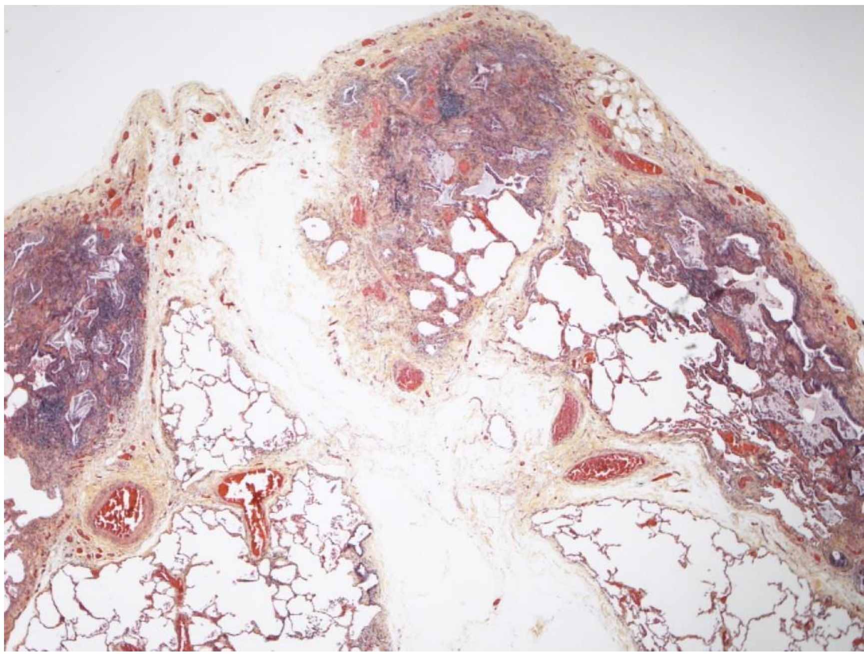
Poumon « sain »



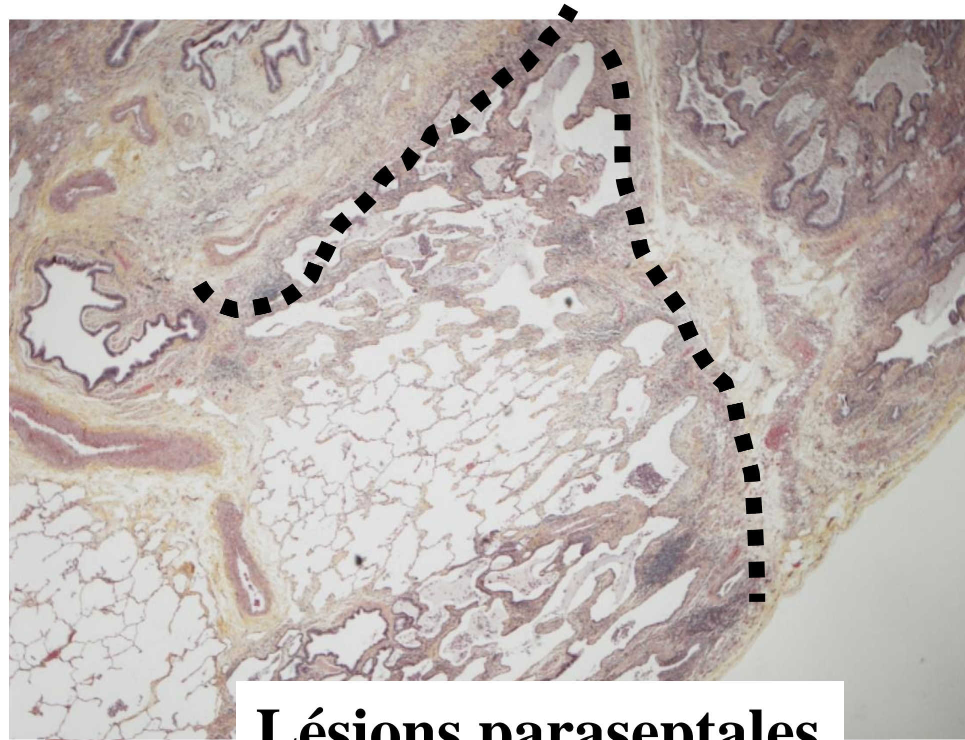
Plèvre viscérale

Lésions sous pleurales

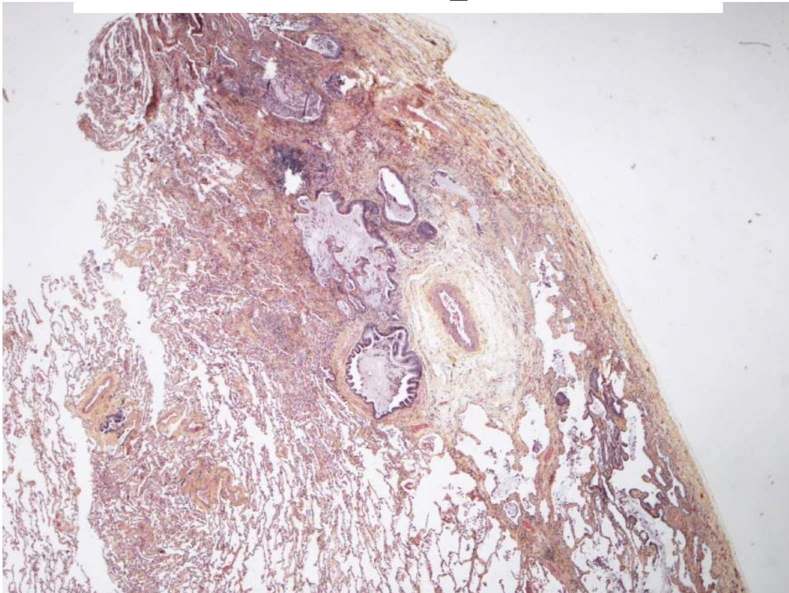


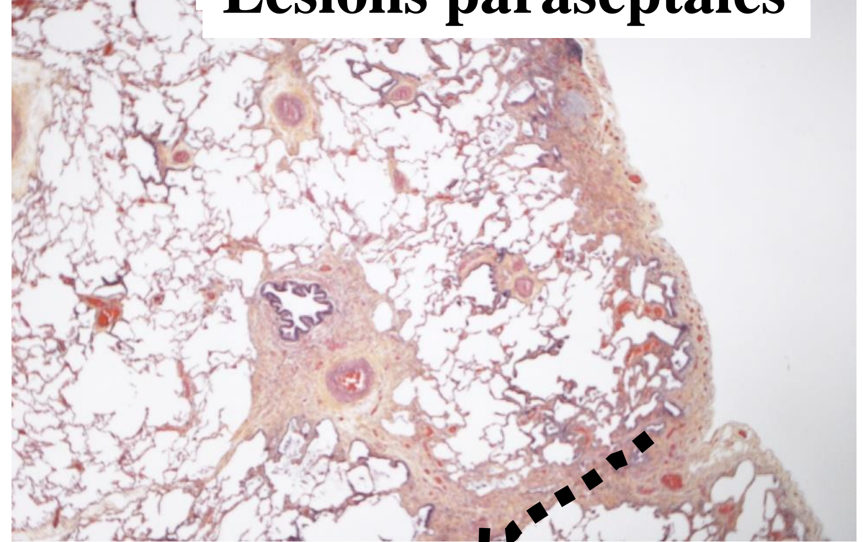


Lésions sous pleurales

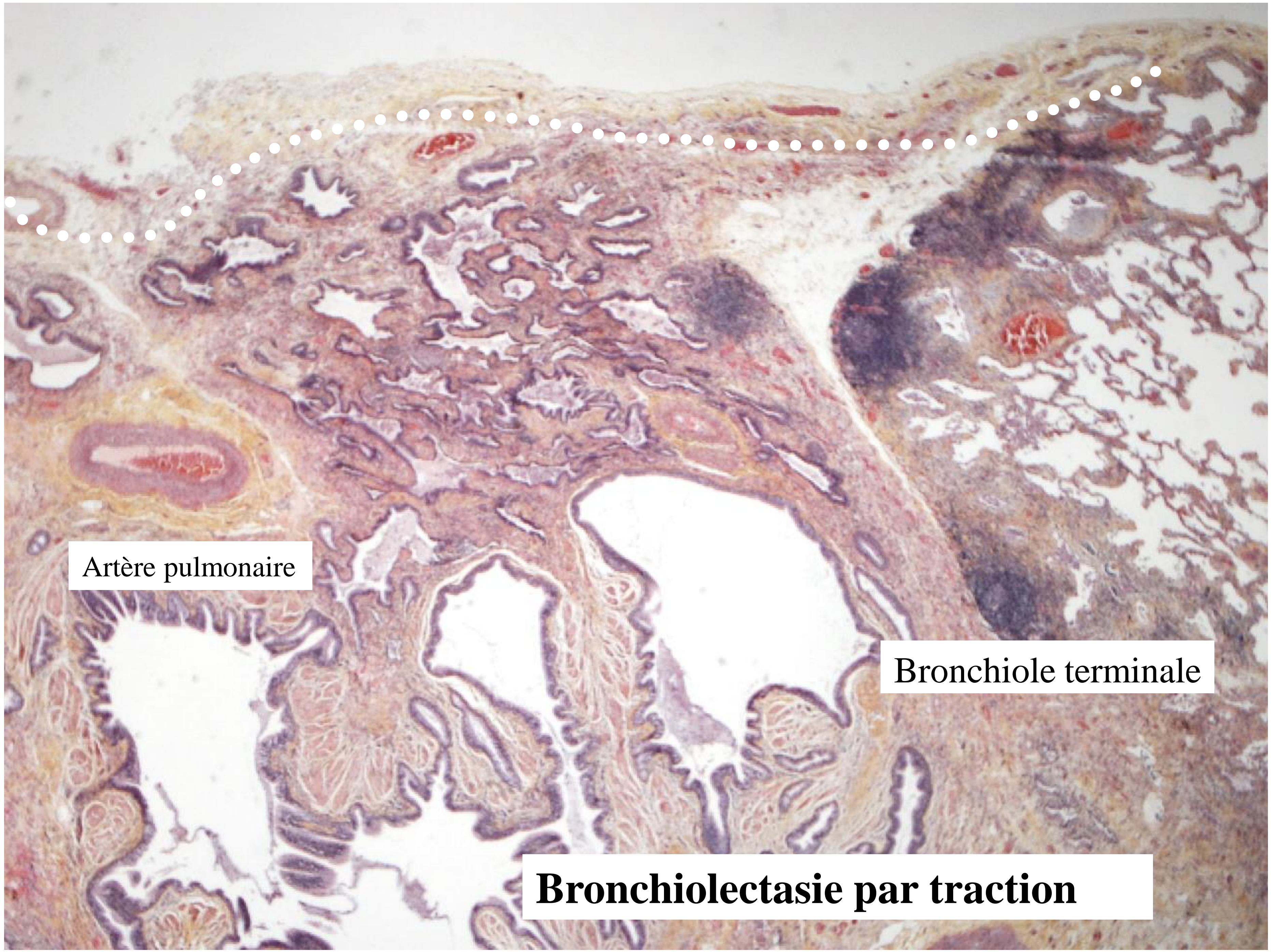


Lésions paraseptales



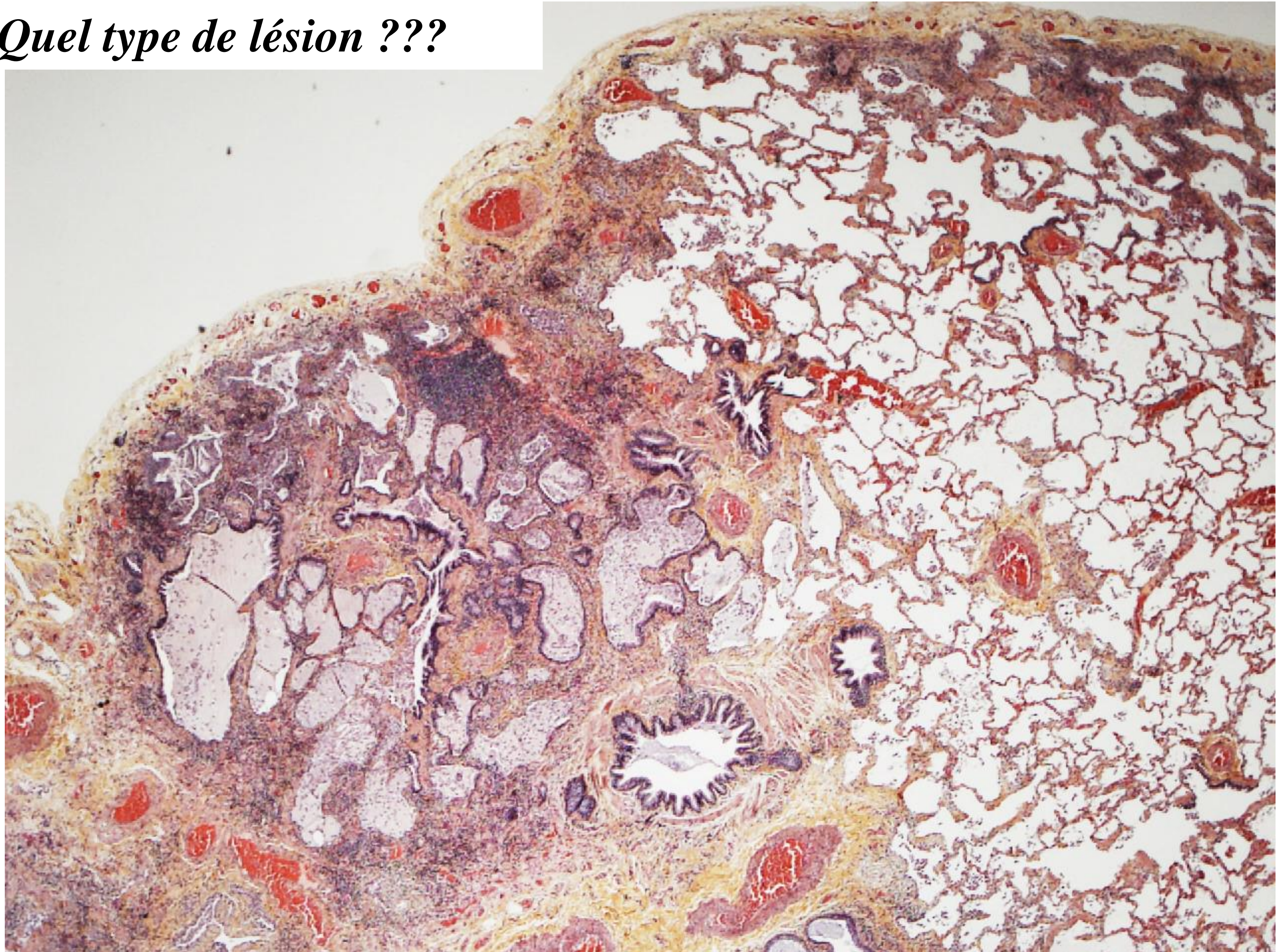


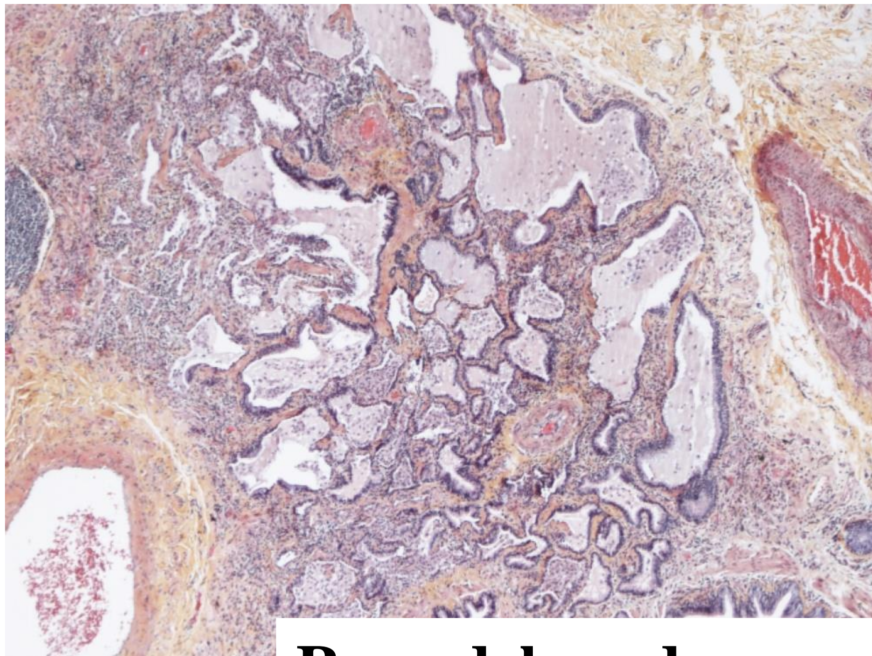
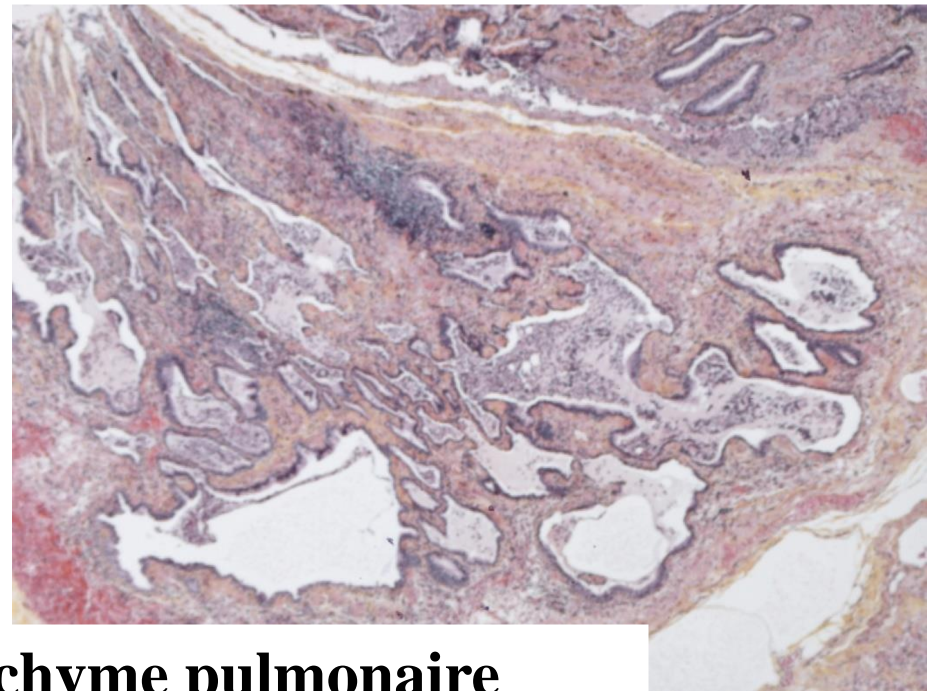
Septa interlobulaire



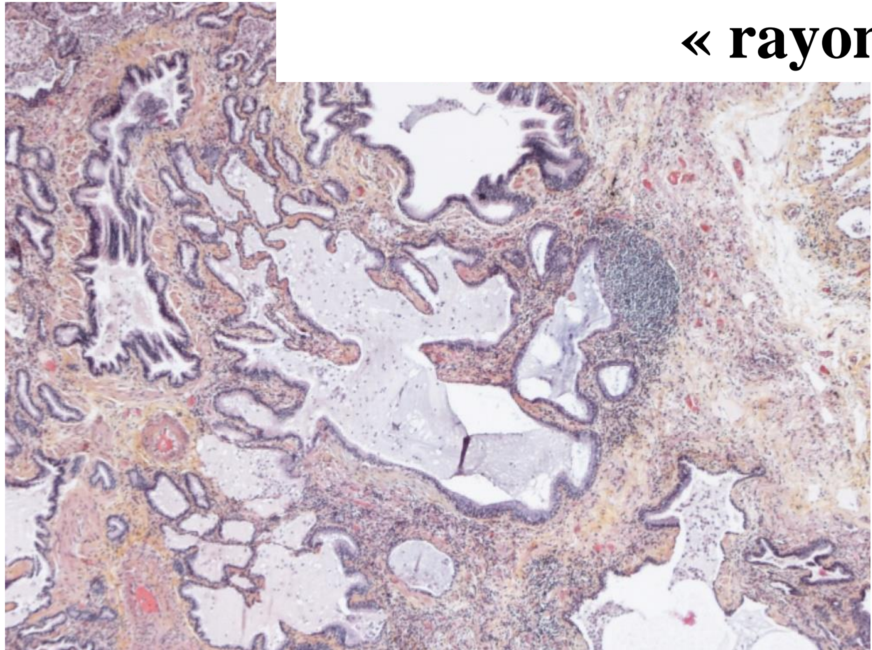
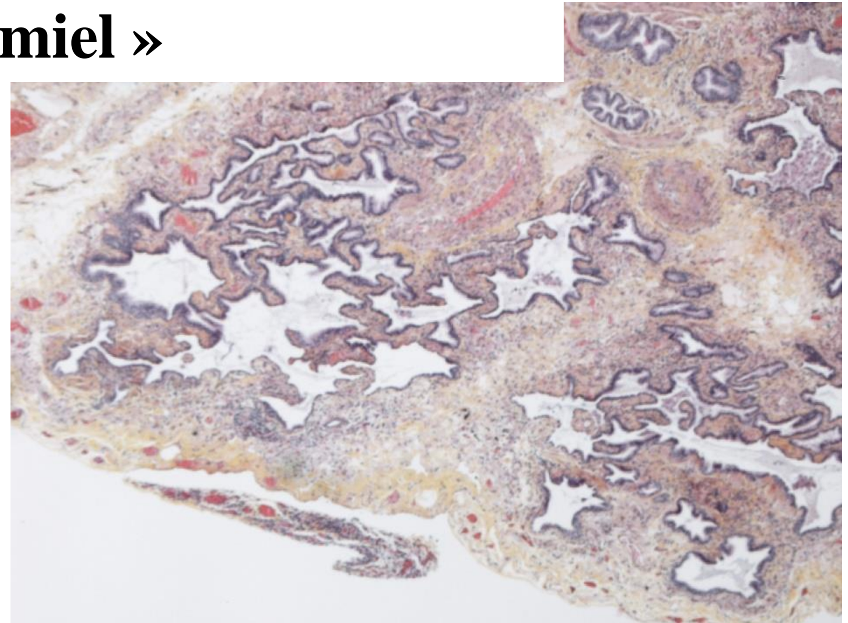
Artère pulmonaire
Bronchiole terminale
Bronchiolectasie par traction

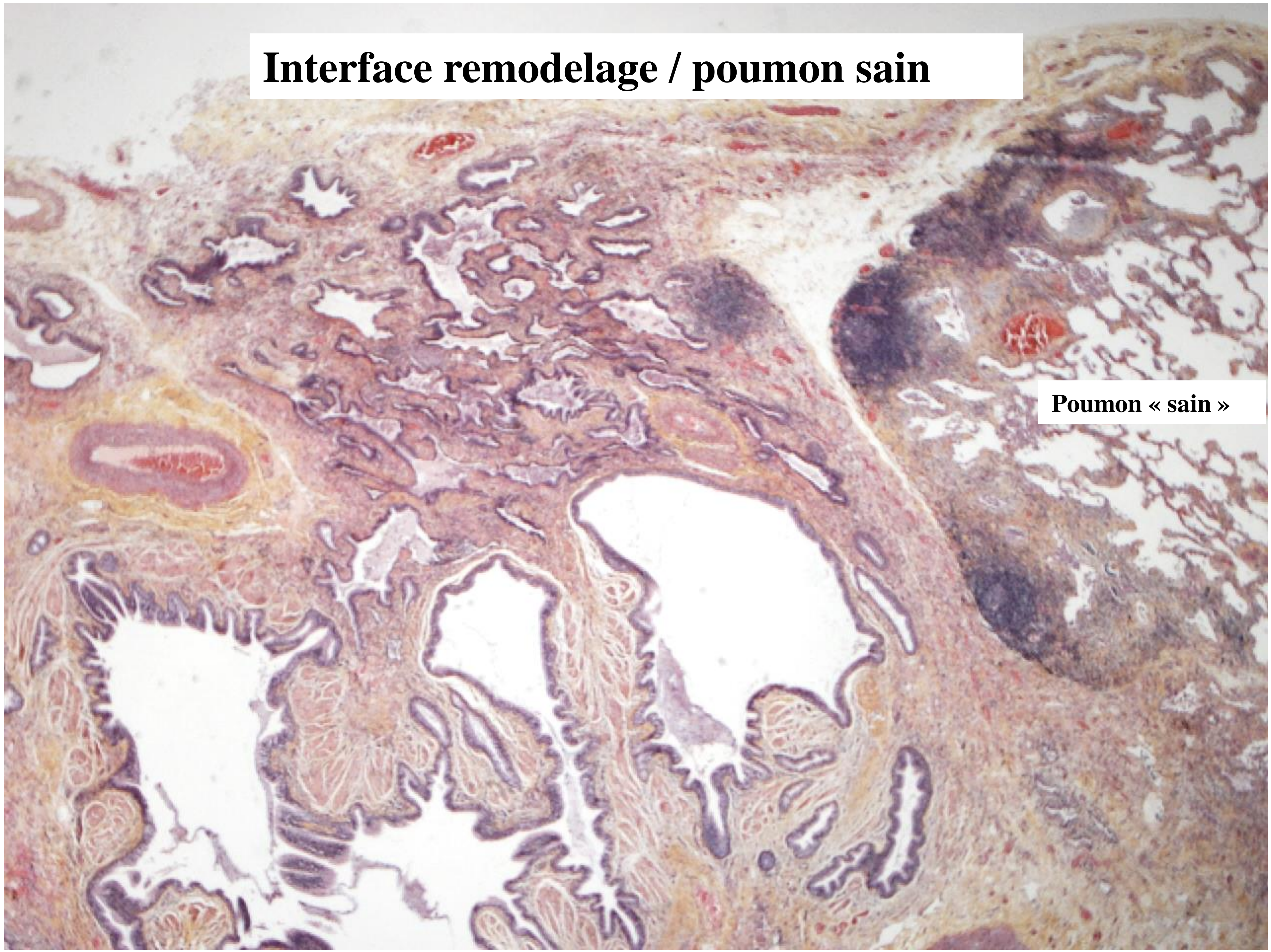
Quel type de lésion ???

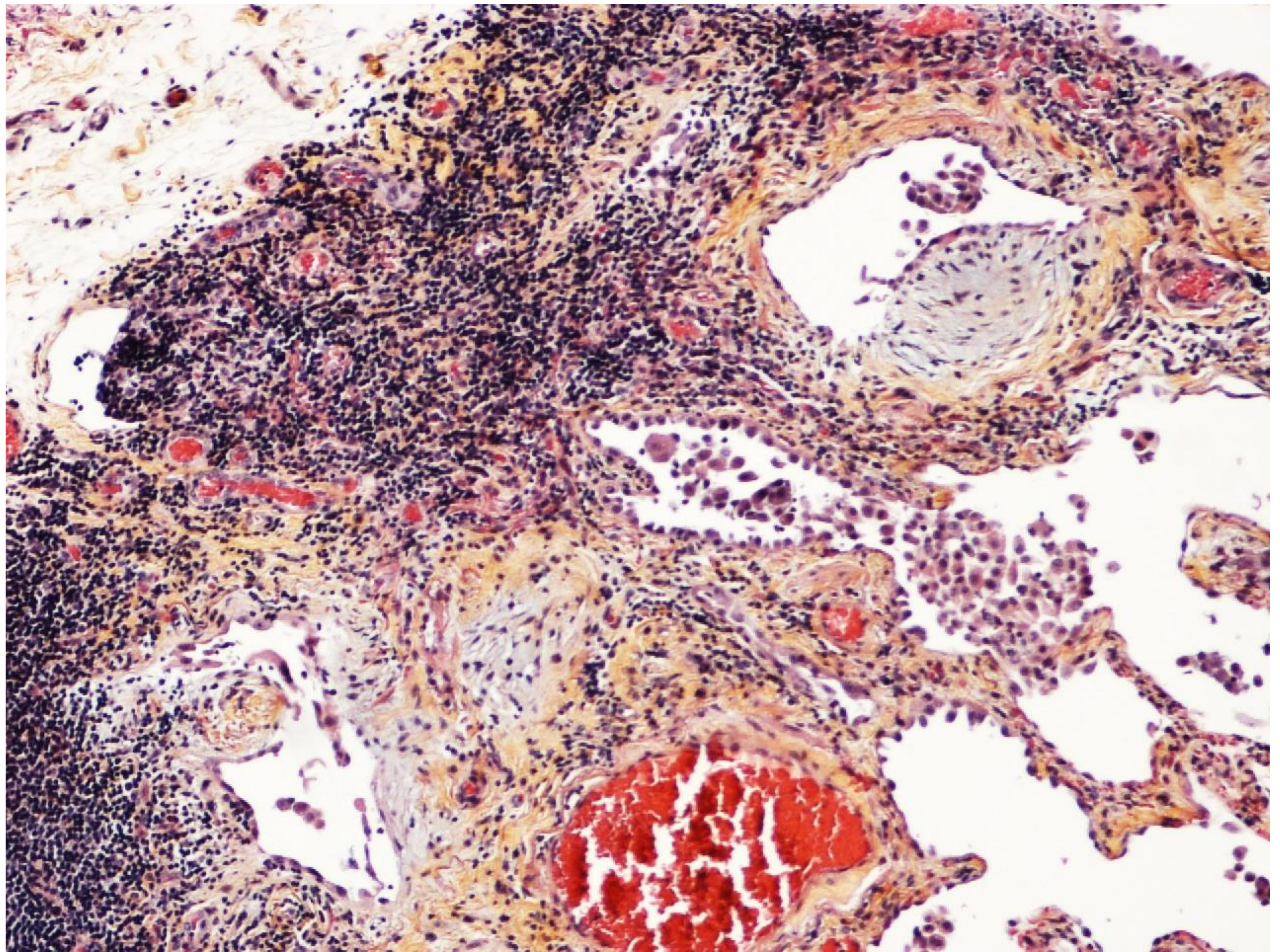


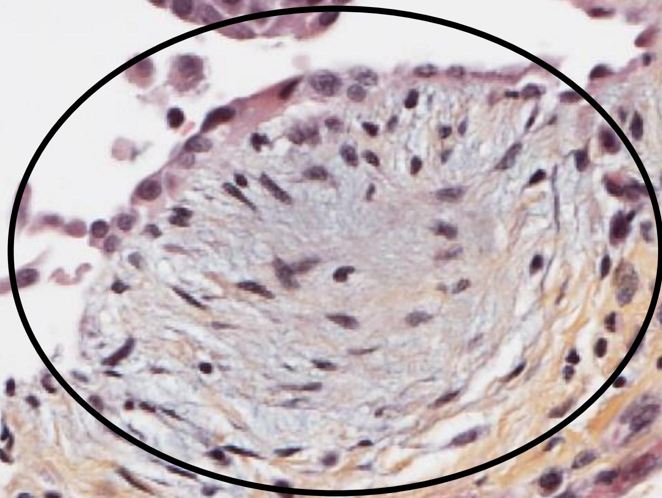



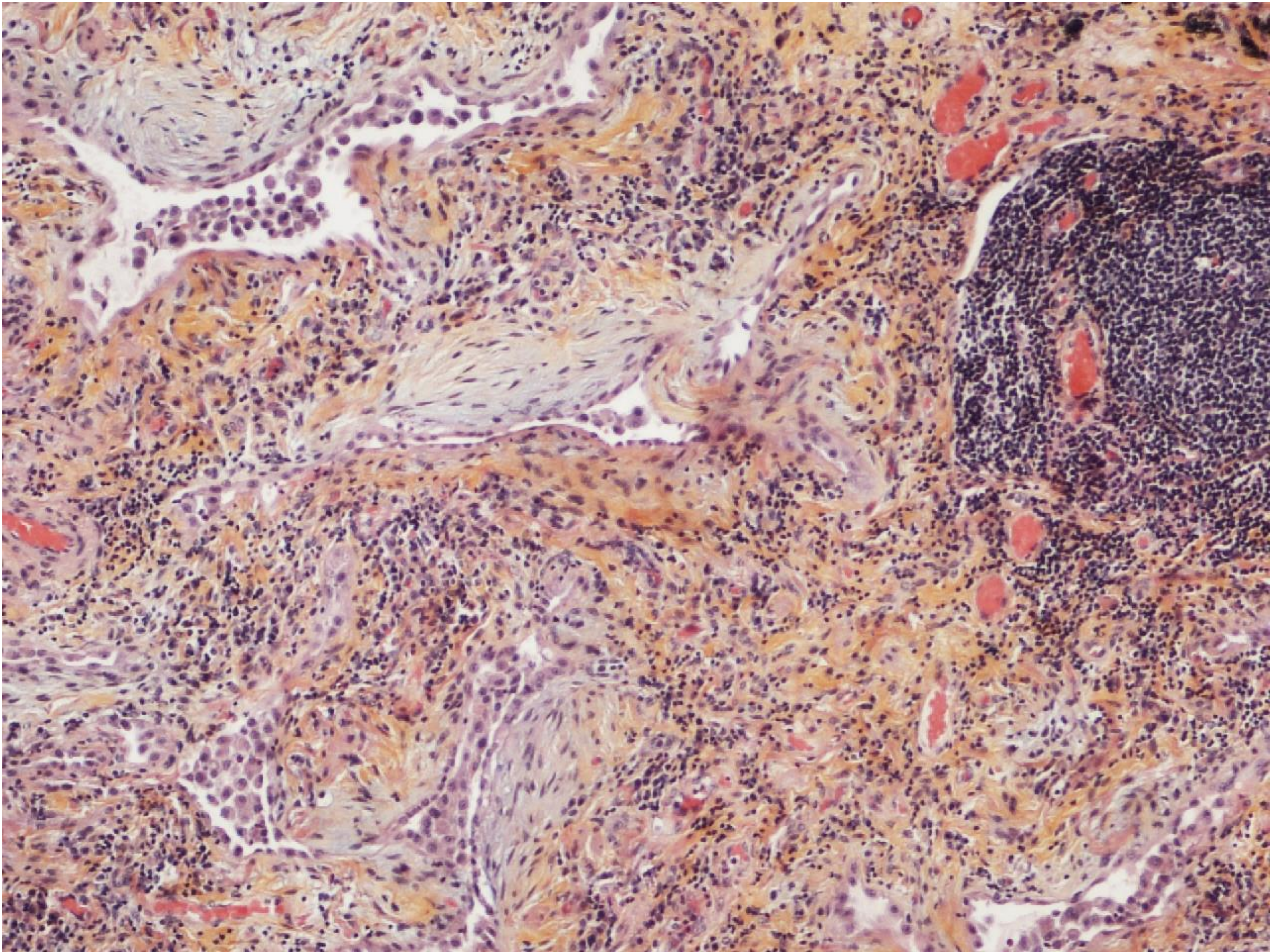
Remodelage du parenchyme pulmonaire
« rayon de miel »

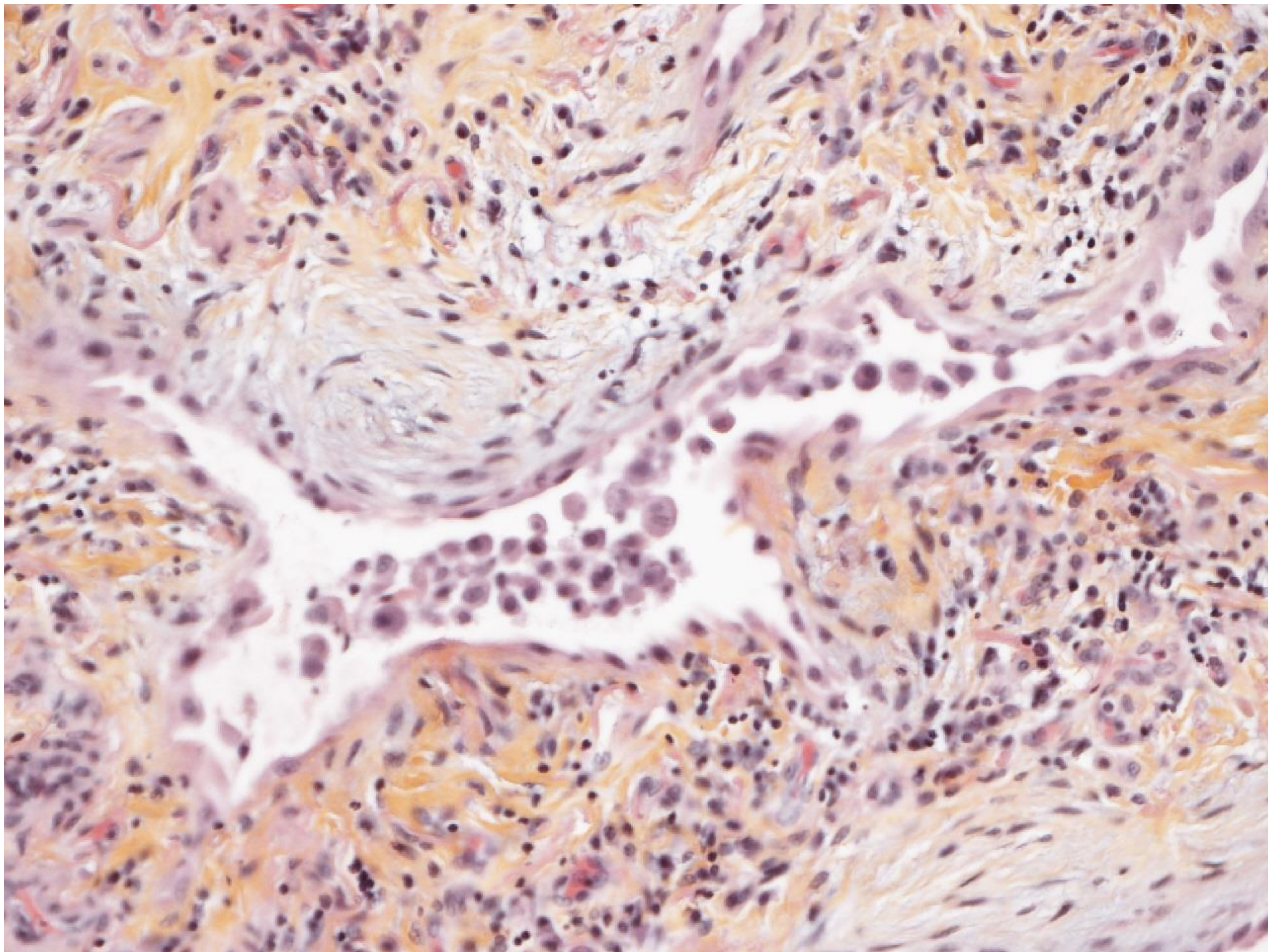



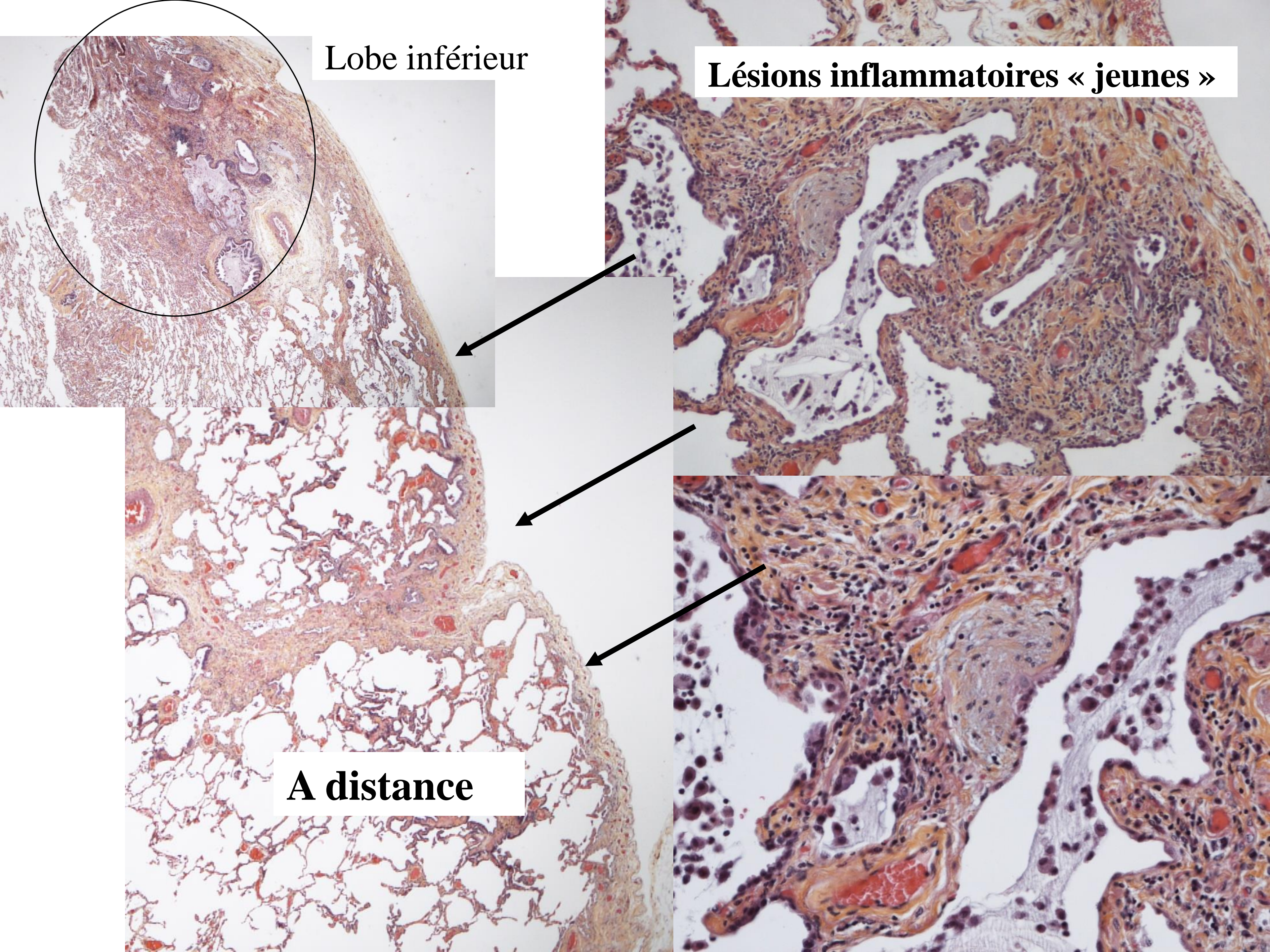

Interface remodelage / poumon sain
Poumon « sain »

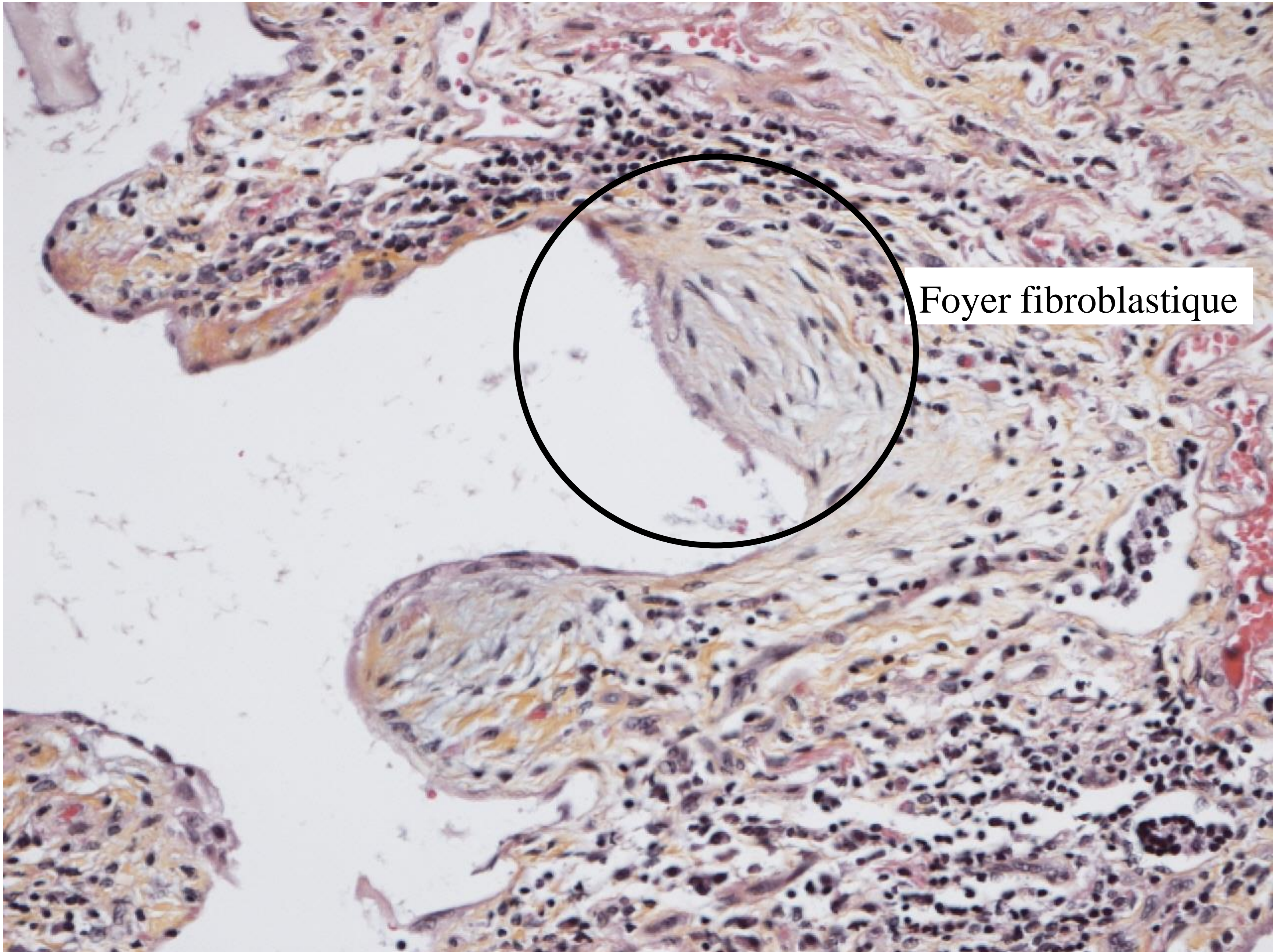


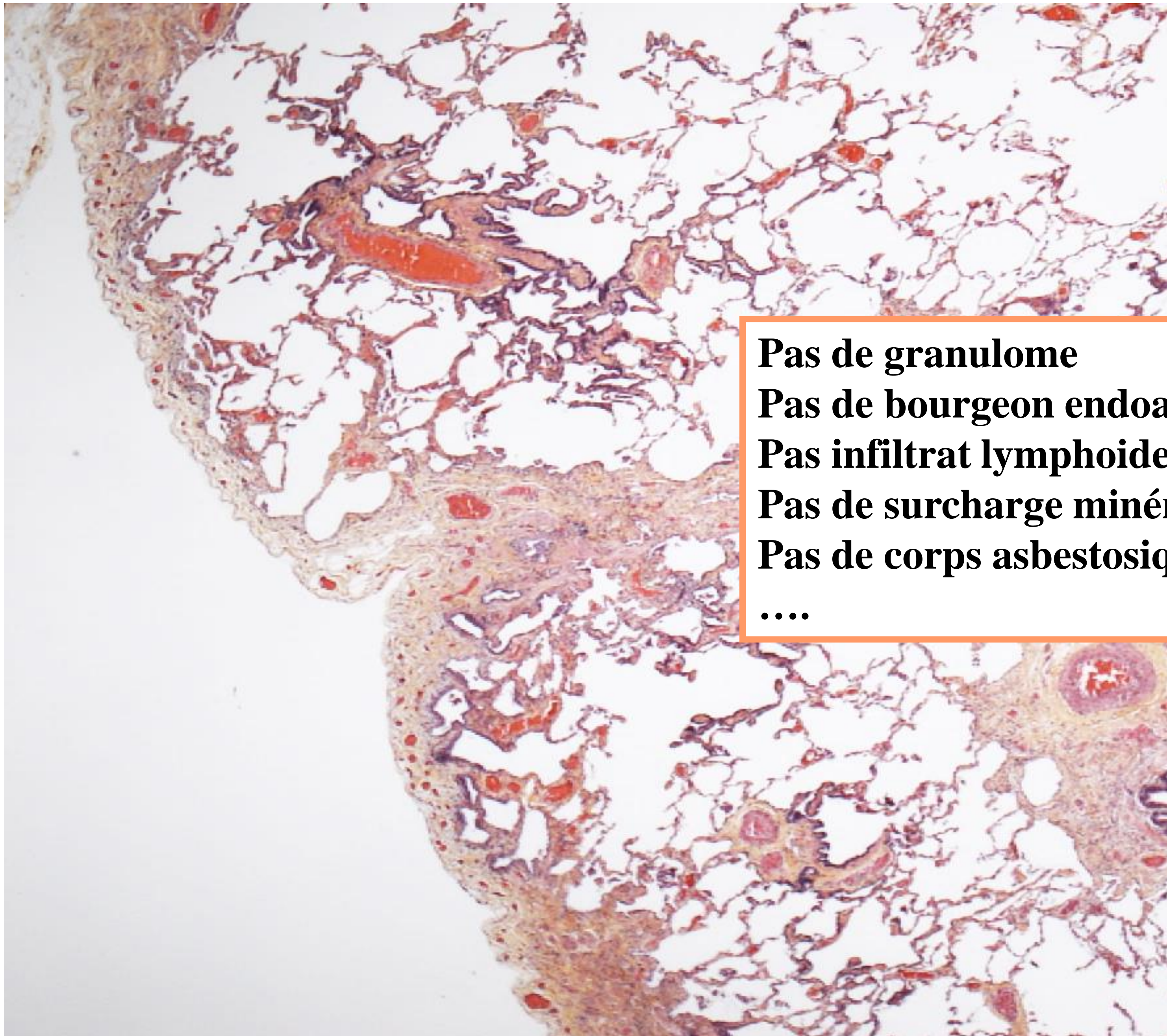

Foyer fibroblastique



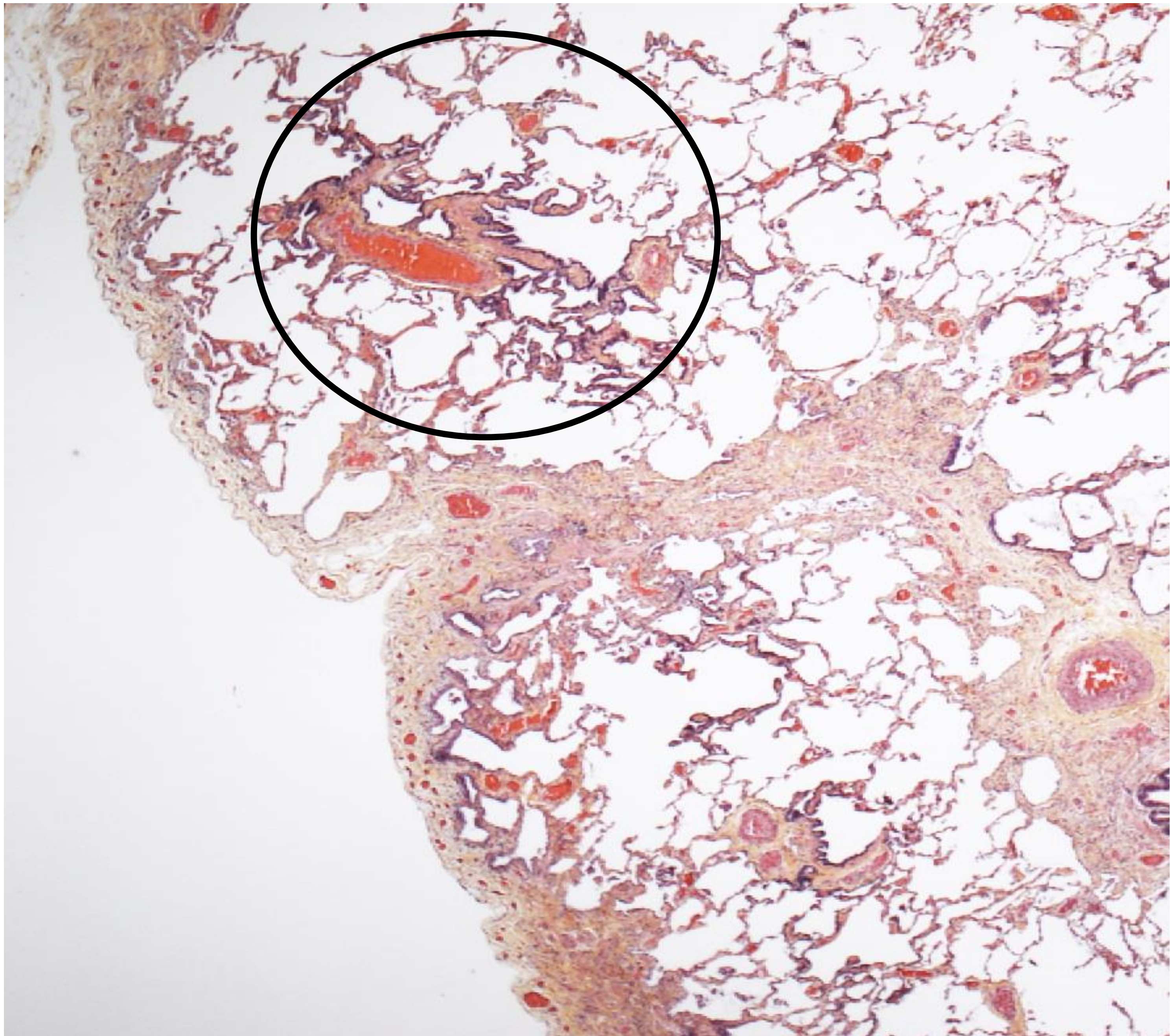



Lobe inférieur
Lésions inflammatoires « jeunes »
A distance

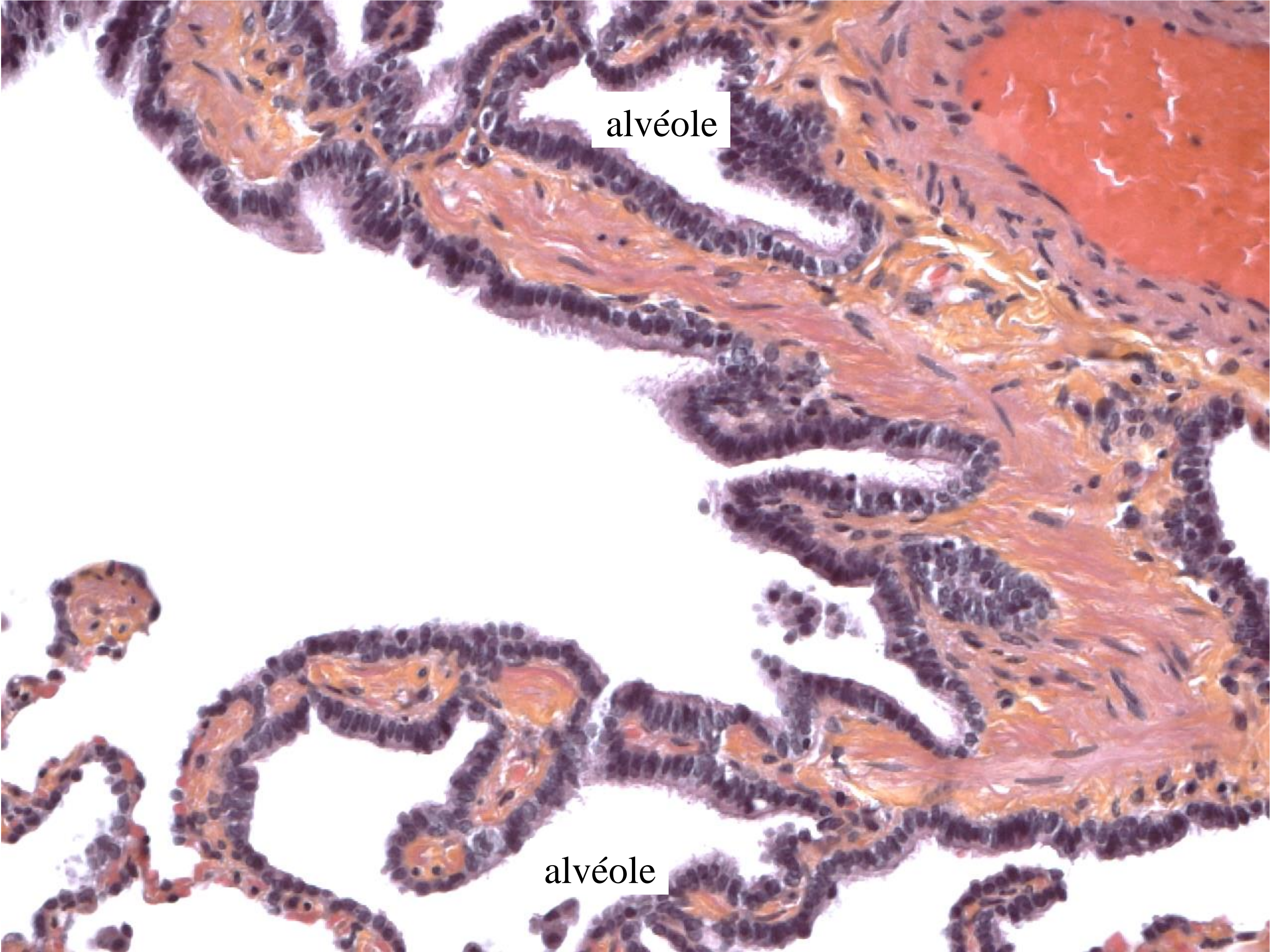

Foyer fibroblastique



Pas de granulome
Pas de bourgeon endoalvéolaire
Pas infiltrat lymphoide interstitiel
Pas de surcharge minérale
Pas de corps asbestosique
….



alvéole
alvéole
Bronchiole respiratoire


alvéole
alvéole

Peribronchiolar Metaplasia: A Common Histologic Lesion in Diffuse Lung Disease and a Rare Cause of Interstitial Lung Disease

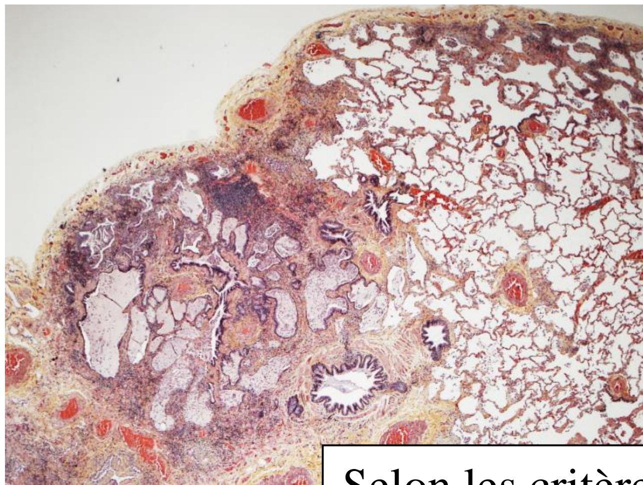
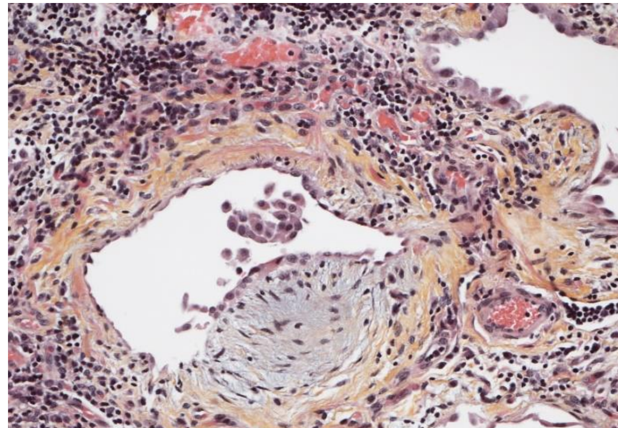
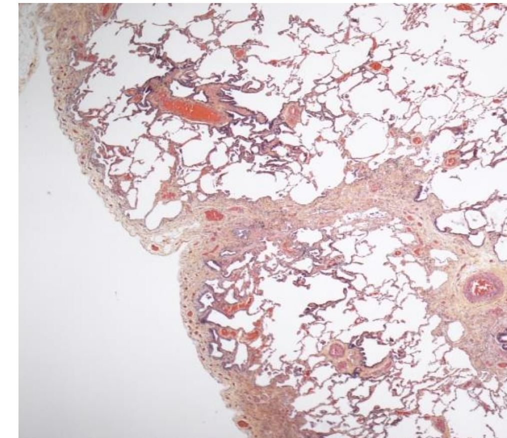
Clinicopathologic Features of 15 Cases

Junya Fukuoka, MD, PhD, Teri J. Franks, MD,† Thomas V. Colby, MD,‡ Kevin R. Flaherty, MD,§ Jeffrey R. Galvin, MD,‖ Dennis Hayden, DO,† Bernadette R. Gochuico, MD,¶ Ella A. Kazerooni, MD,# Fernando Martinez, MD,§ and William D. Travis, MD**

TABLE 3. Presence of Peribronchiolar Metaplasia (PBM) in Chronic Interstitial Lung Disease

	No. of Cases	No. (%) of Cases With PBM	Bronchiole Per Case* (%)
UIP	29	17 (59)	17.8
NSIP	20	10 (50)	22.5
DIP	6	3 (50)	11.1
RB	18	2 (11)	18.1
HSP	18	9 (50)	16.7
PBM-ILD	15	15 (100)	71.0

(Am J Surg Pathol 2005;29:948–954)

Selon les critères ATS ERS 2001 - **Profil lésionnel de PIC**
Selon les critères ATS ERS 2011 - **PIC certaine**
Foyers de métaplasie bronchiolaires

DMD discussion multidisciplinaire

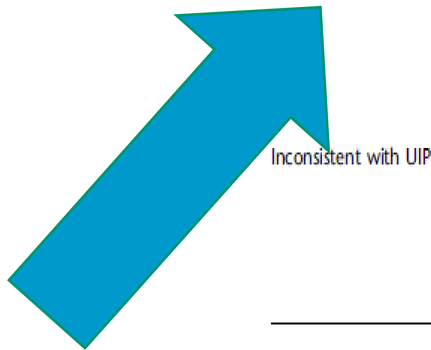
Proposer un diagnostic de FPI ou non sur une combinaison de critères

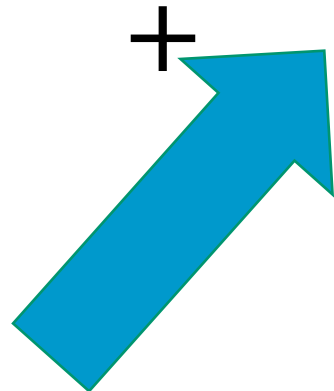
selon les critères ATS ERS 2011 - FIC certaine

TABLE 6. COMBINATION OF HIGH-RESOLUTION COMPUTED TOMOGRAPHY AND SURGICAL LUNG BIOPSY FOR THE DIAGNOSIS OF IPF (REQUIRES MULTIDISCIPLINARY DISCUSSION)

HRCT Pattern*	Surgical Lung Biopsy Pattern* (When Performed)	Diagnosis of IPF?[†]
UIP	**UIP** **Probable UIP** **Possible UIP** **Nonclassifiable fibrosis**[‡]	**YES**
	Not UIP	No
Possible UIP	**UIP** **Probable UIP**	**YES**
	Possible UIP Nonclassifiable fibrosis	Probable[§]
	Not UIP	No
Inconsistent with UIP	UIP	Possible[§]
	Probable UIP Possible UIP Nonclassifiable fibrosis Not UIP	No

+





An Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management

Observation N°2

patient de 65 ans

antécédents

- Ancien ajusteur
- Asthme apparu en 1989, sévère - actuellement de palier 2 du GINA contrôlé par SERETIDE 250X2
- Rhinite inflammatoire et obstructive – 1993: septoplastie, ethmoidectomie antérieure, méatomie moyenne bilatérale
- Diabète non compliqué sous insuline LANTUS
- Non fumeur

biologie

- Hb : 15,6 – leucocytes : 5 540 – éosinos 4,7%- plaquettes : 266 000 – CRP 1
- Clearance créatinine : 75 ml/mn, iono Normal
- SGOT 28 SGPT 28 Ph alcaline 105
- Pro BNP : 60
- Électrophorèse des protides normale :augmentation polyclonale sans pic
- Enzyme de conversion de l'angiotensine : 29 UI
- VIH 1 et 2 : négatif
- Bilan d'auto immunité : AAN positifs au 1/320, fluorescence mouchetée – anti DNA négatifs – ANCAS négatifs - Latex Waaler Rose négatif- anti SSA et anti RNP négatifs

Histoire de la maladie

Dyspnée d'effort modérée

Découverte de crépitants velcro

Bon EG – 75 kg pour 1m72

Pas d'hippocratisme digital

Bilan fonctionnel

- **EFR**
  CV 48 % (1L750) – VEMS 61,7% - CPT 43,7
- **GDS sous air**
  PaO2 77 – PaCO2 46 – SaO2 95,4%
- **TCO**
  73,8%

Dossier clinique Dr T.Pigeanne pneumologue Sables d'Olonnes

antécédents

- Ancien ajusteur
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  73,8%

Dossier clinique Dr T.Pigeanne pneumologue Sables d'Olonnes

Vidéo endoscopie

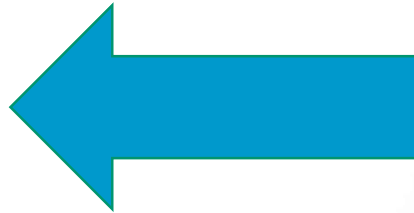
- Examen macroscopiquement **normal**
- **LBA lingula – cellularité 135/ml**

  macrophages 63%

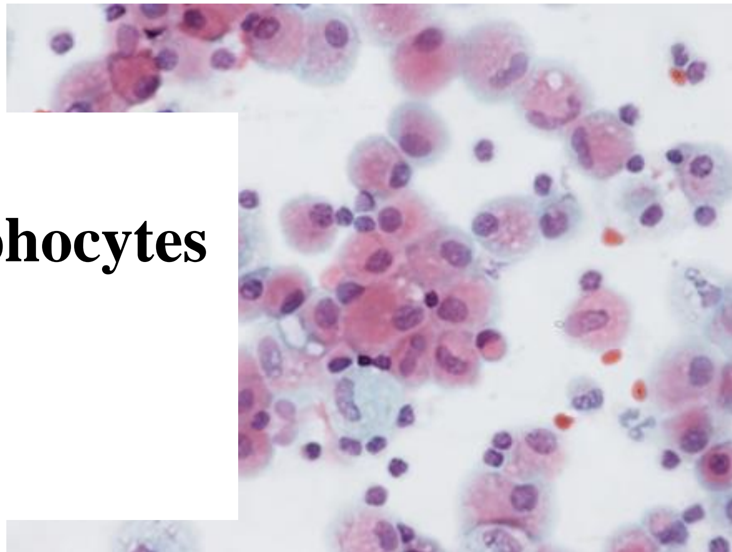
  lymphocytes 18%

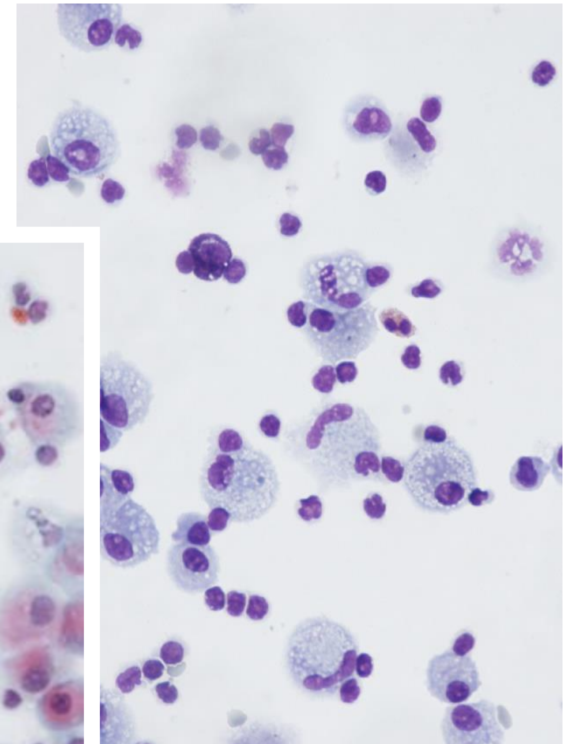
  PN 11%

  PE 6 %

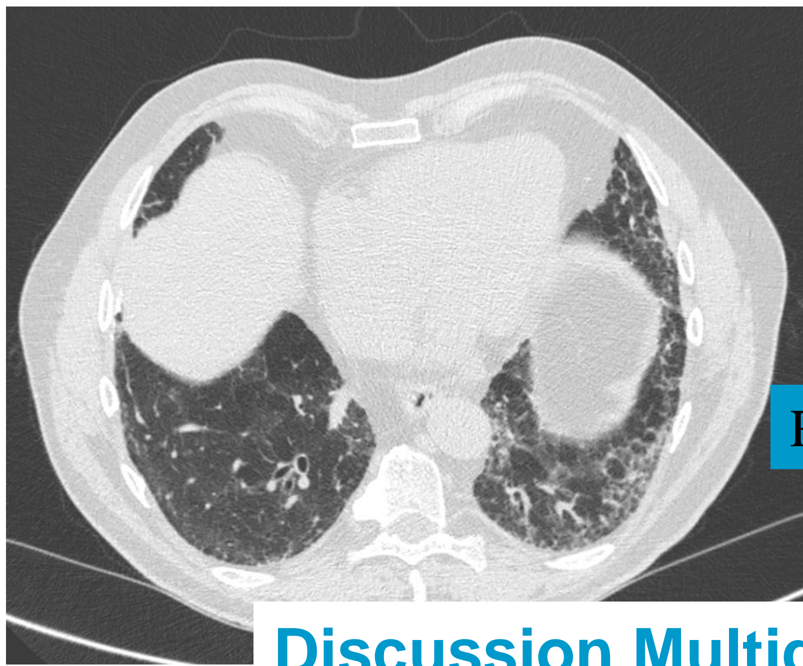


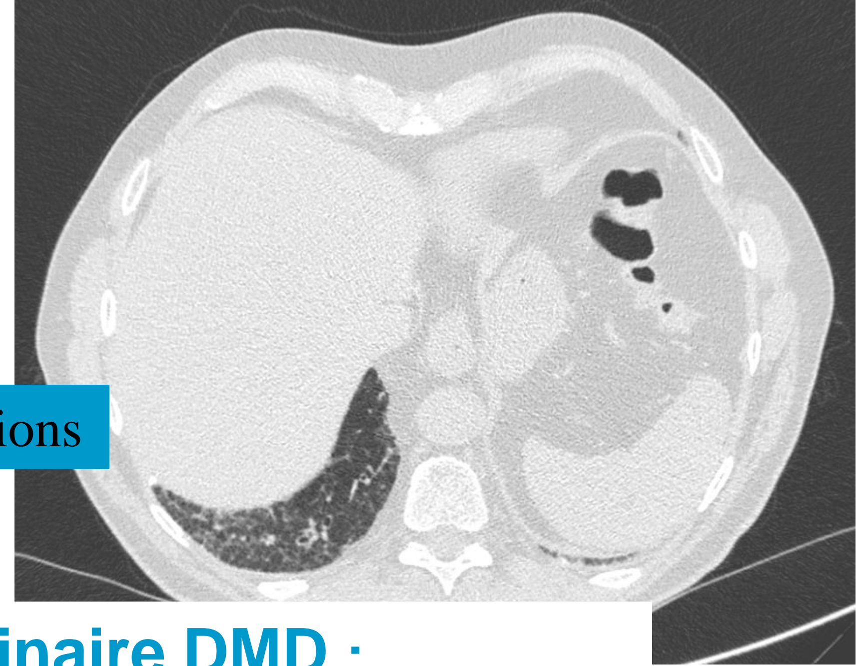
- Microbiologie négative





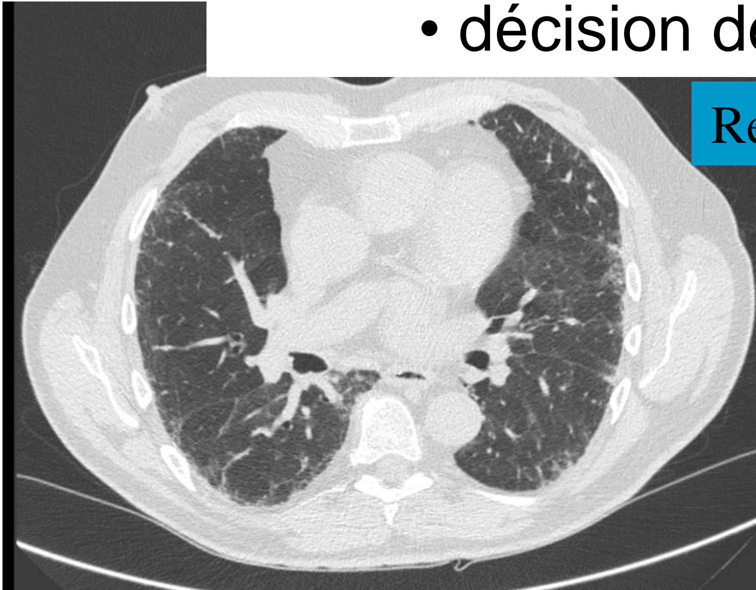
ATTENTION
Pourcentage de lymphocytes > 30 %
Profil cytologique

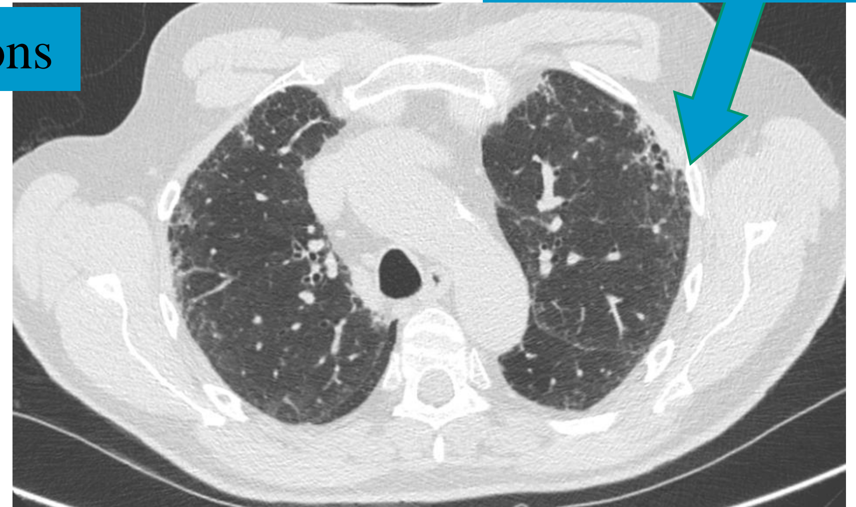


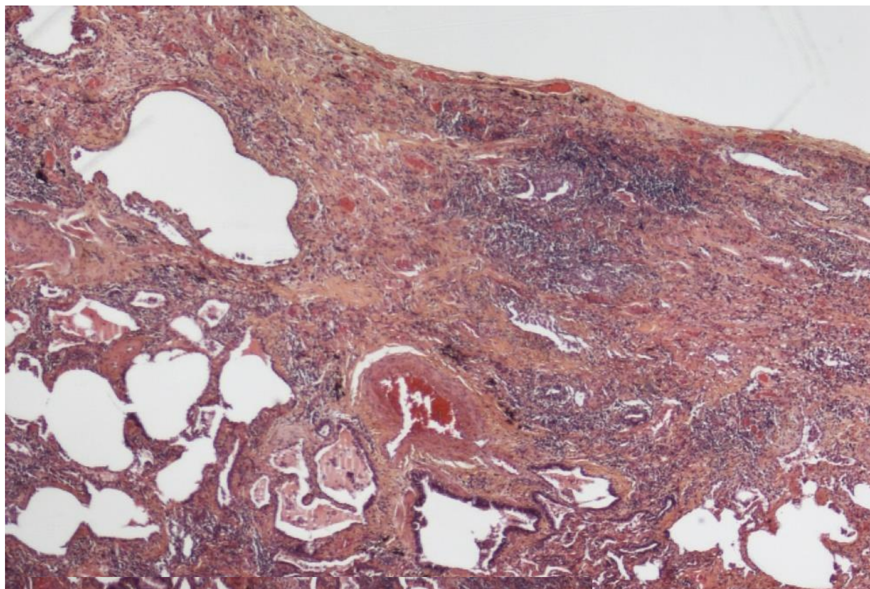


Discussion Multidisciplinaire DMD :

- TDM : **PIC possible**
- décision de biopsie chirurgicale



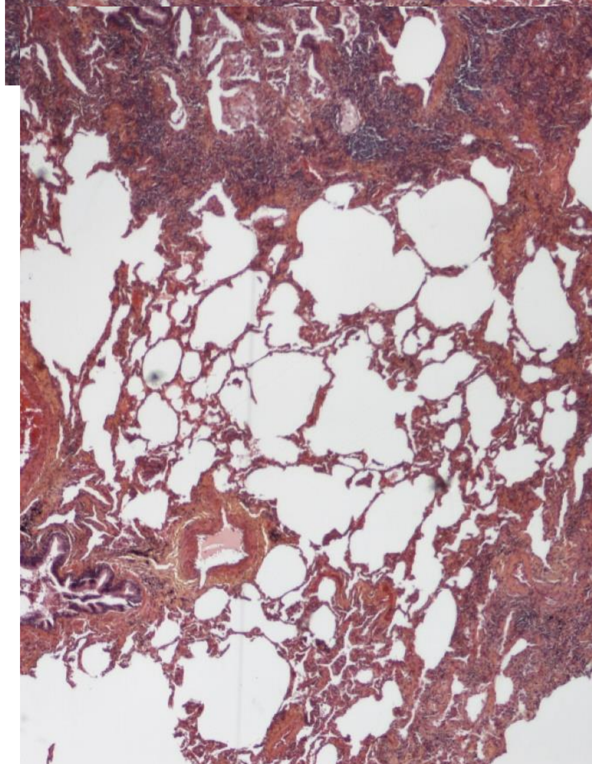


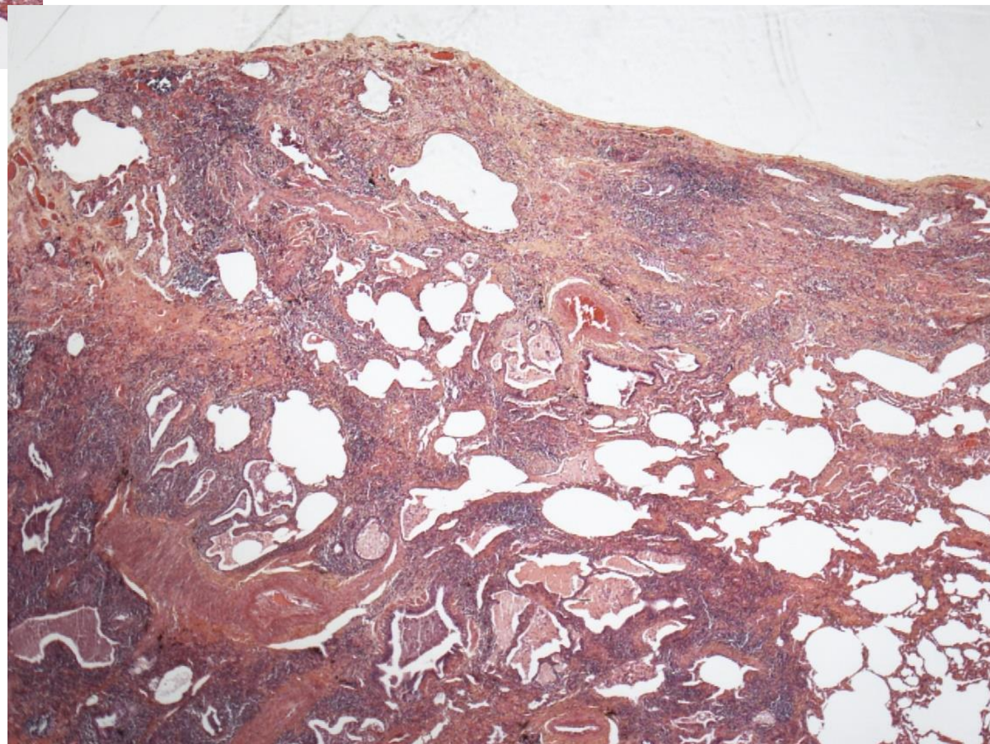


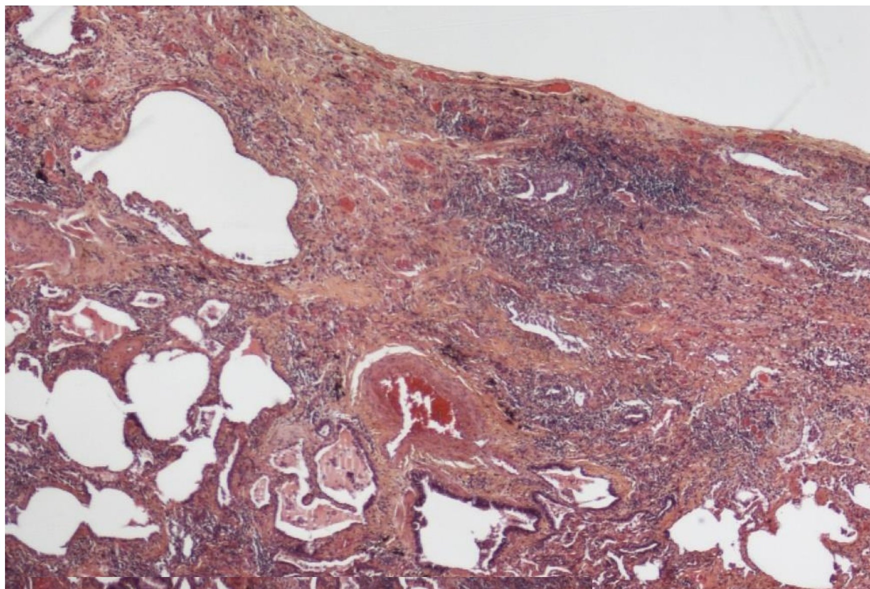
POUMON DROIT – Biopsie chirurgicale

Trois séries de prélèvements adressées, intitulées :

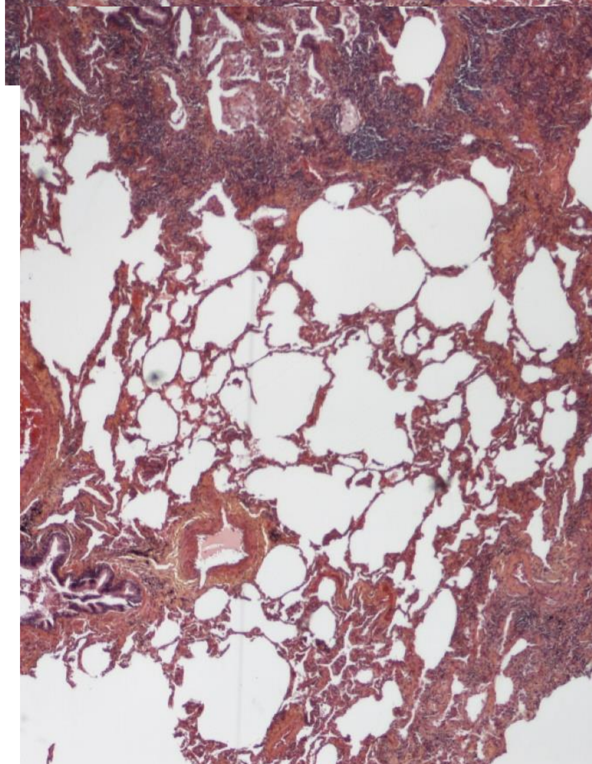
- ✓ **lobe moyen** de 4,5 x 1,5 x 0,8 cm ; surface de la plèvre mamelonnée.
  Inclusion en totalité selon des tranches sériées et numérotées [trois blocs/1RP].
- ✓ **lobe supérieur** de 5,2 x 2,2 x 1 cm ; aspect assez superposable.
  Inclusion en totalité selon des tranches sériées et numérotées [trois blocs/2RP].
- ✓ **lobe inférieur** de 4,5 x 2 x 1,5 cm ; même aspect.
  Inclusion en totalité [trois blocs/3RP].

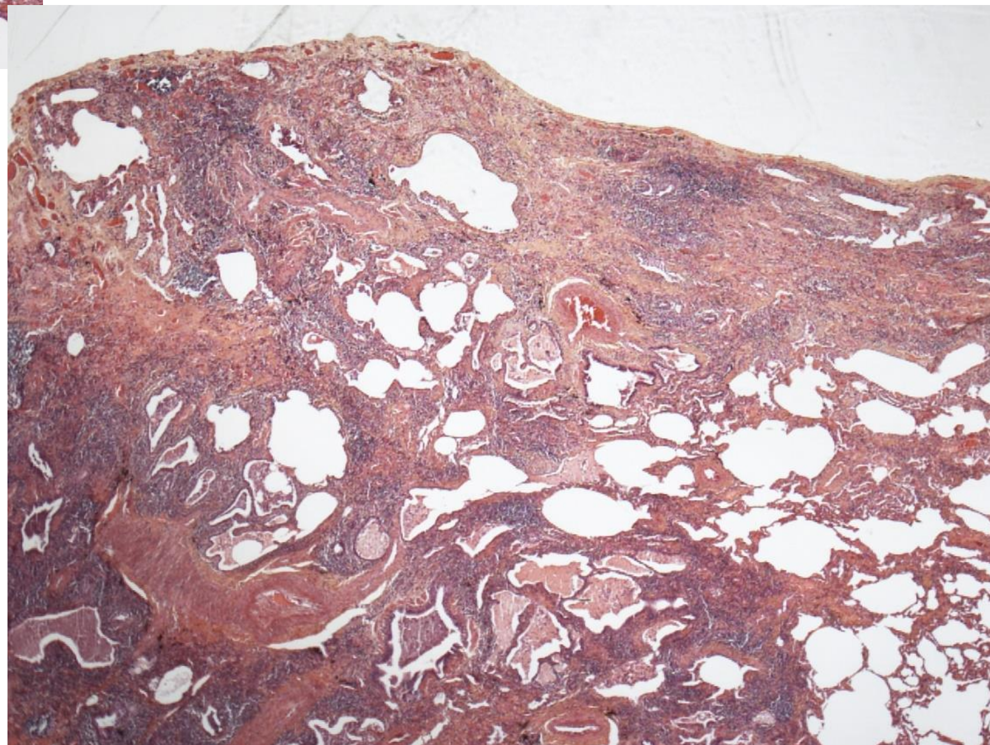




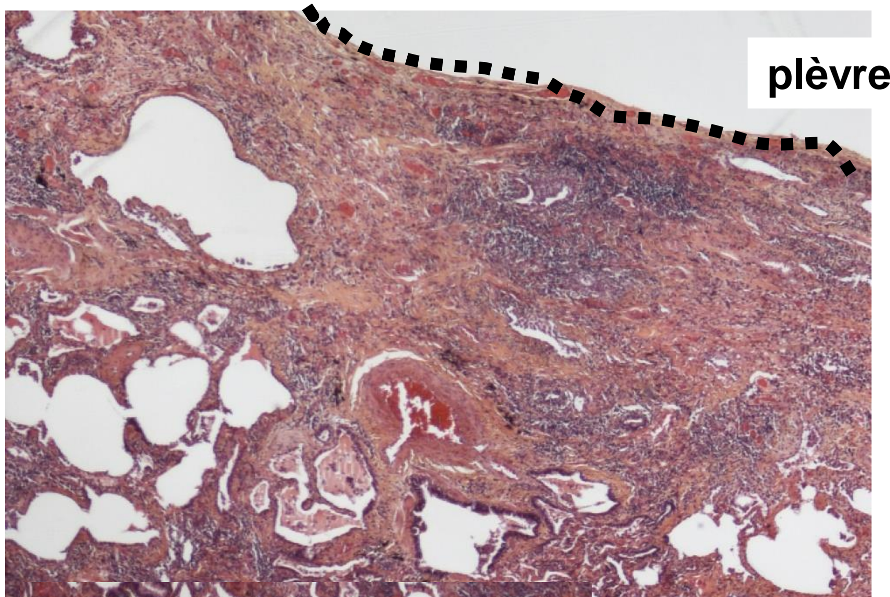


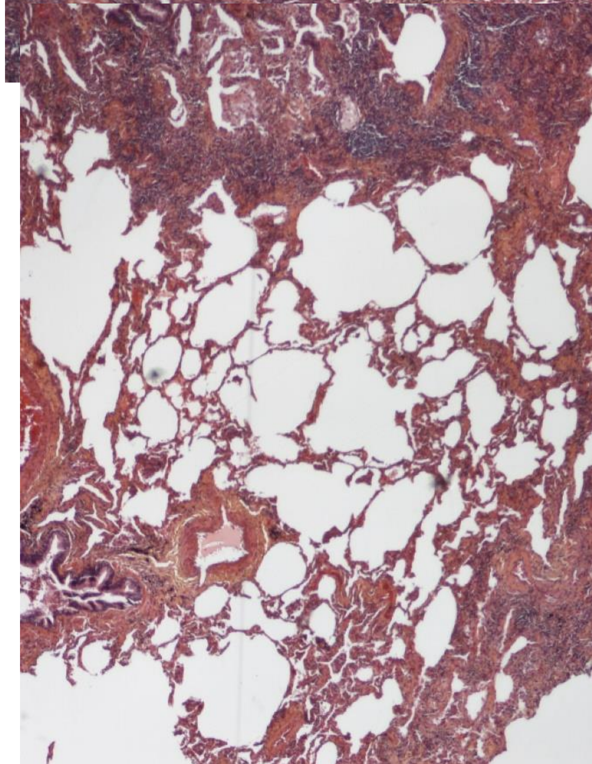
Lésions fibrosantes
Fibrose collagène

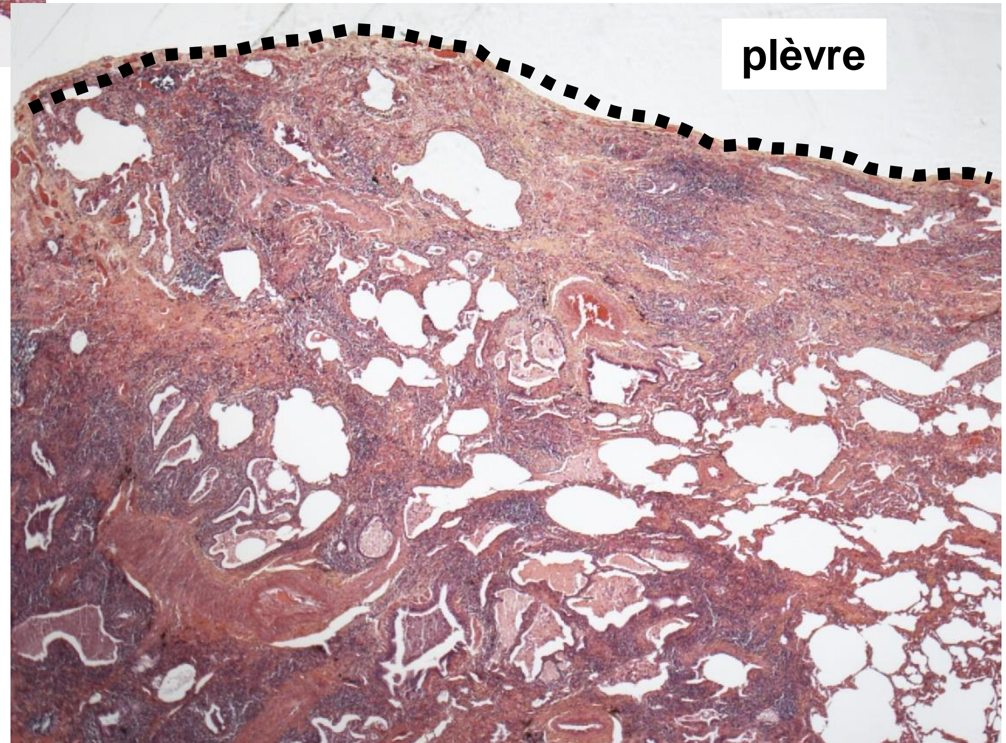




Lésions sous pleurales



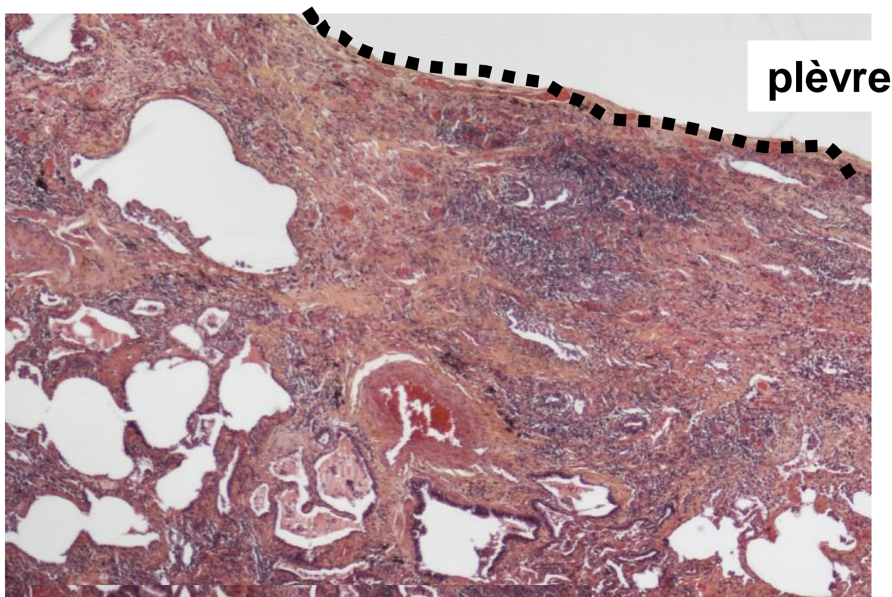


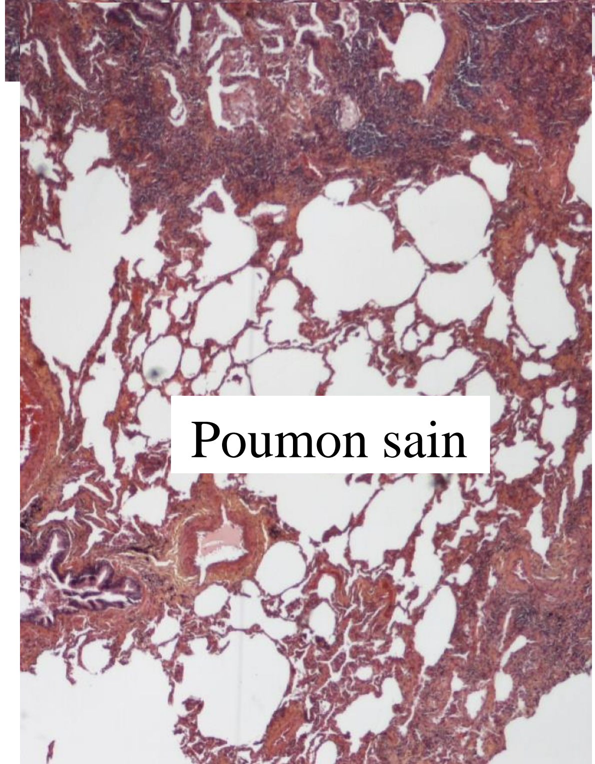


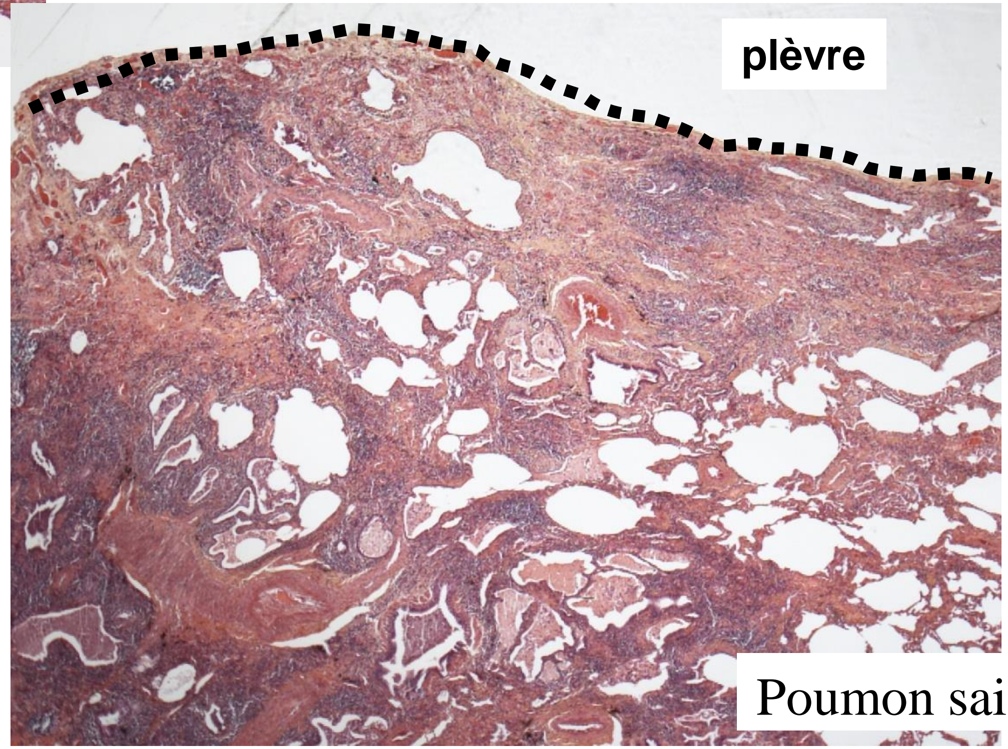
Lésions de distribution hétérogène

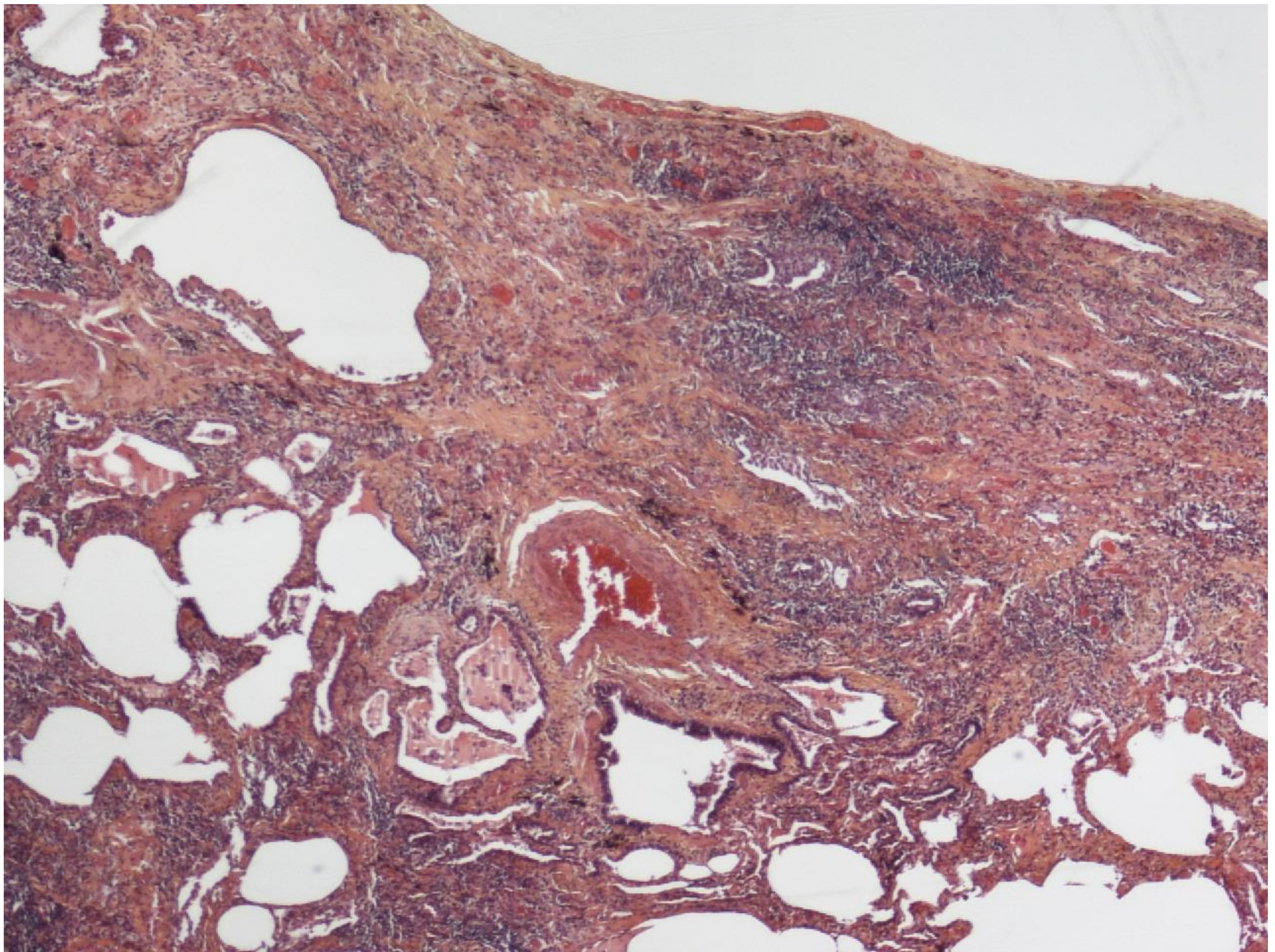
lésions disséminées

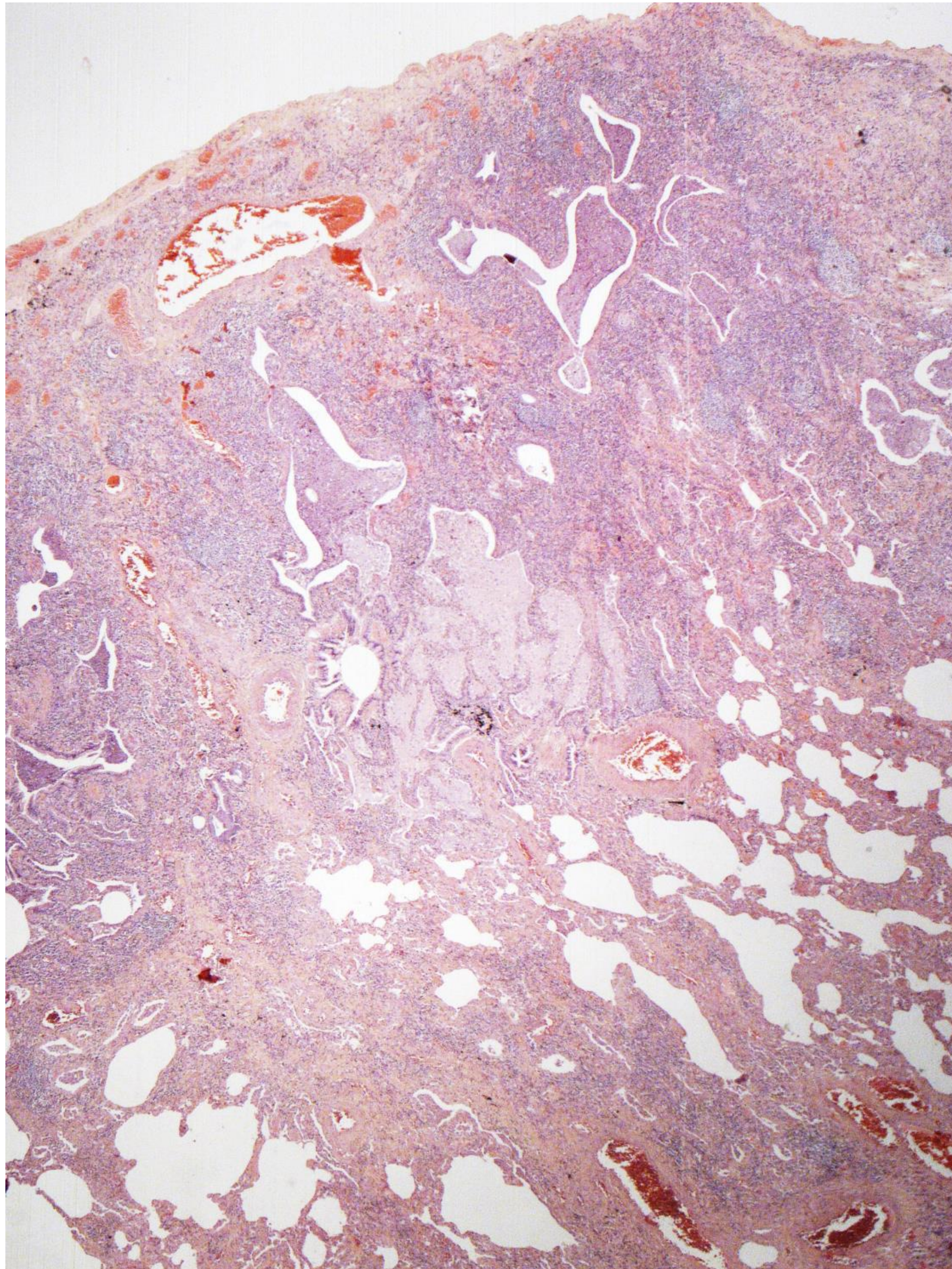
Hétérogénéité spatiale

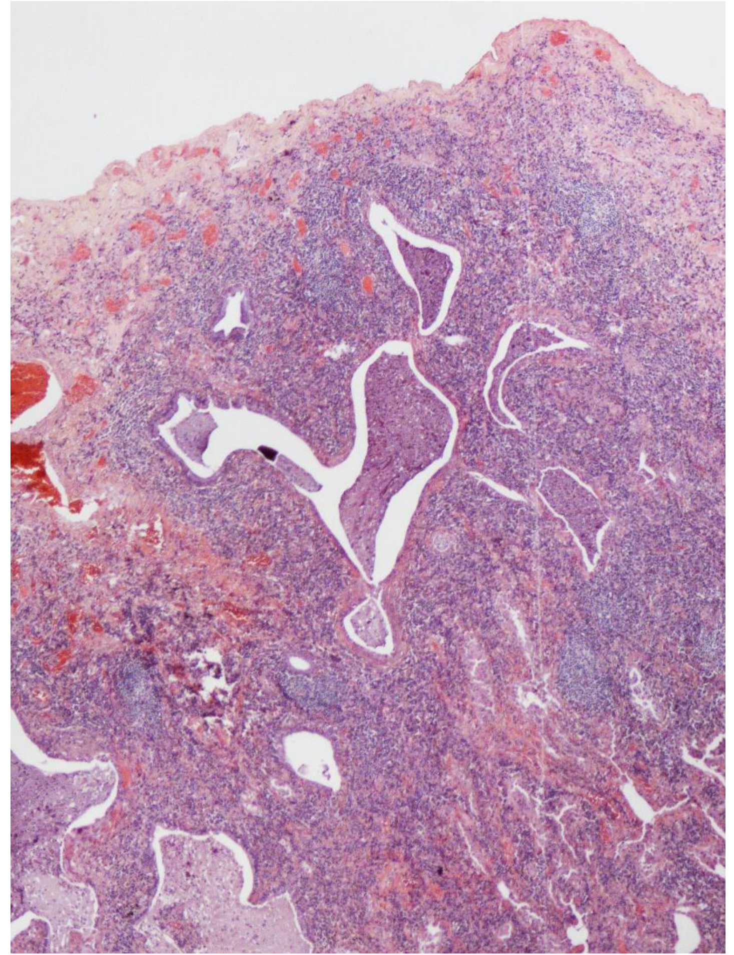


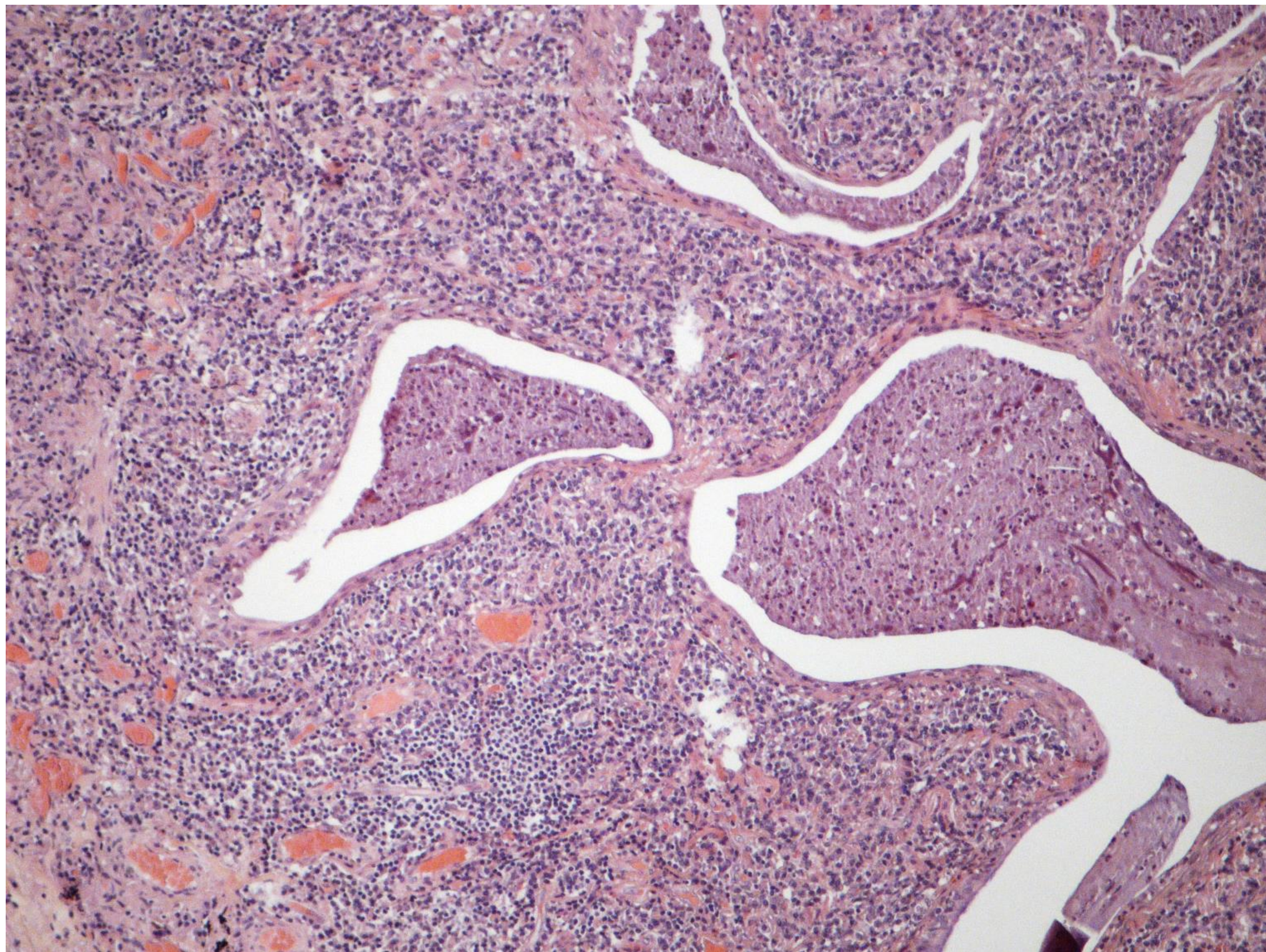












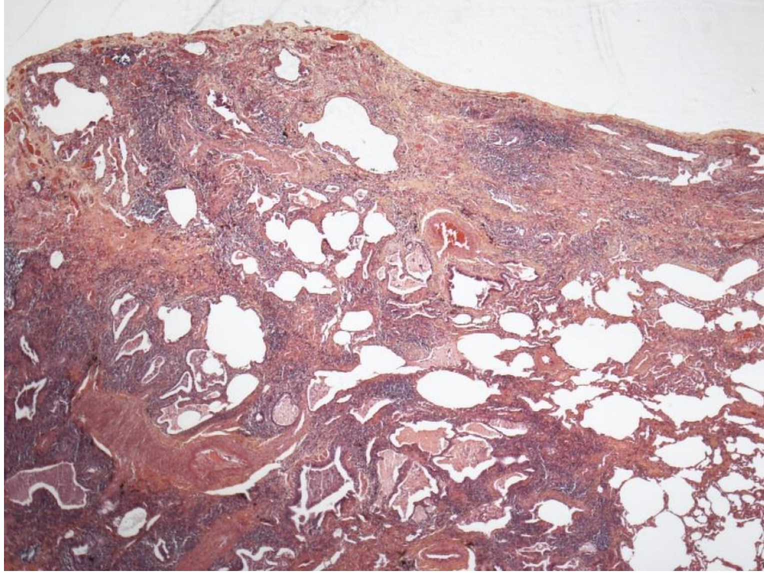
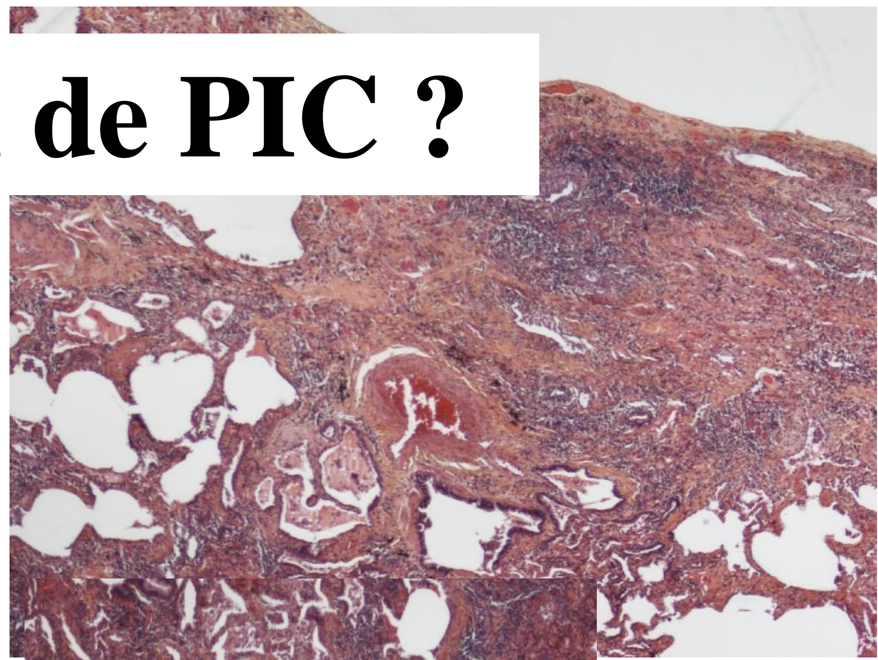
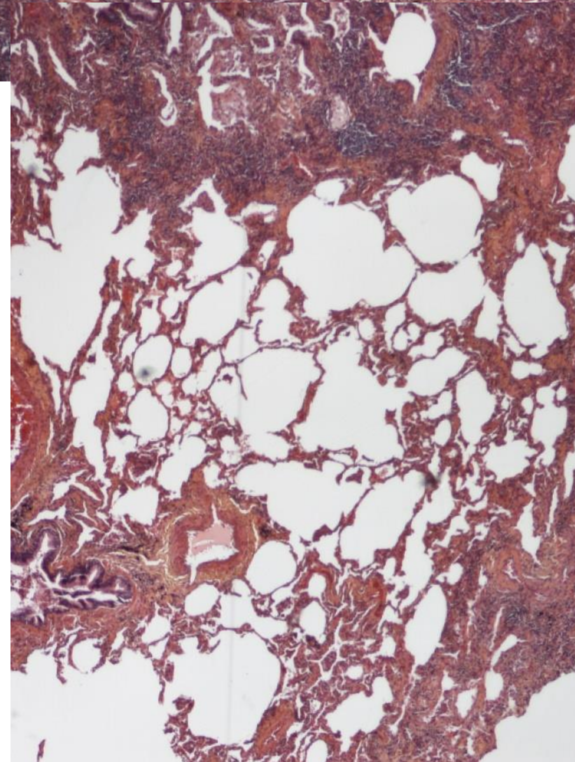
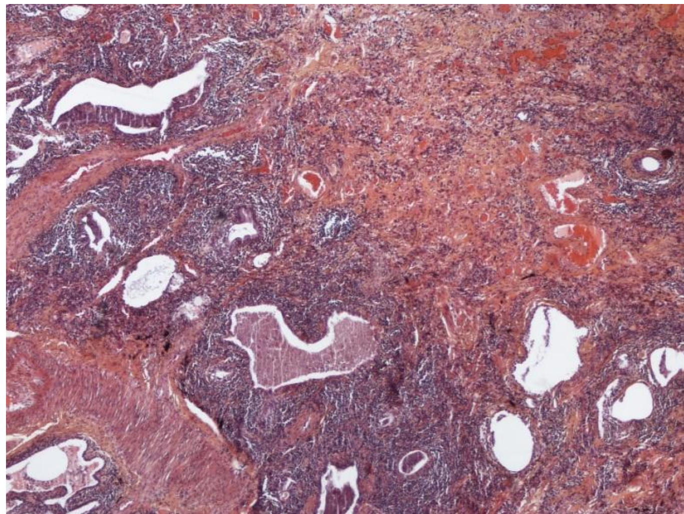
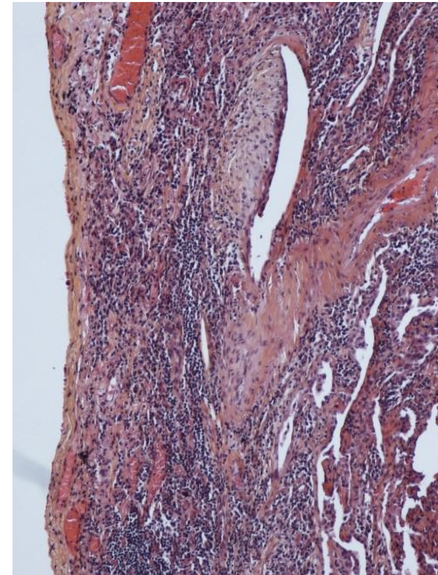
Fibrose collagène

Fibrose jeune
Foyers fibroblastiques

Fibrose d'âge variable

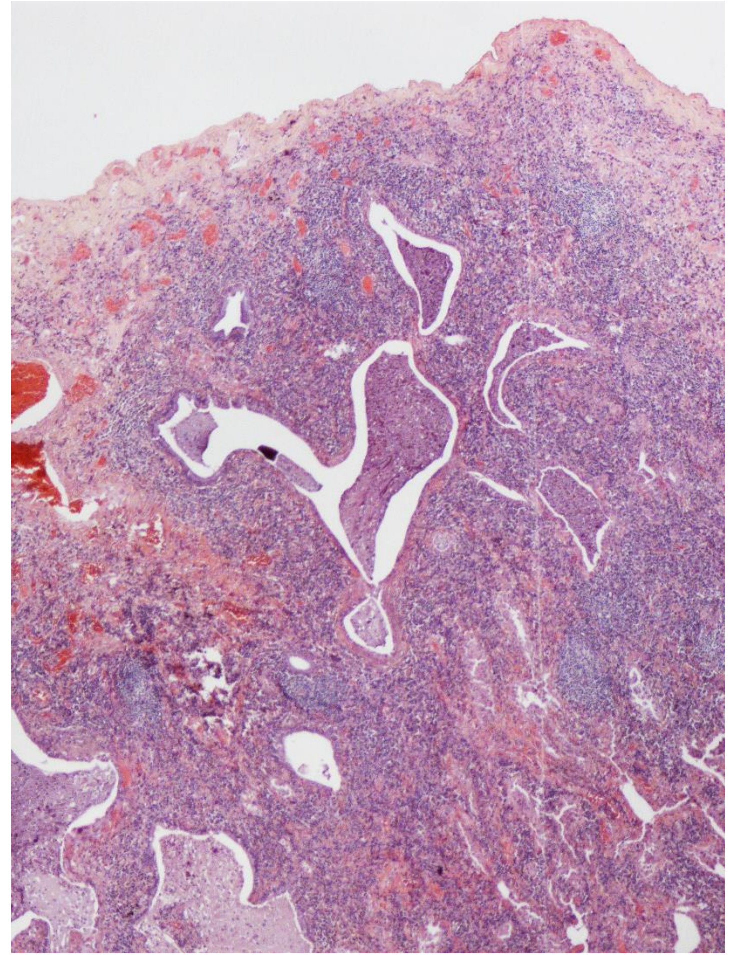
Hétérogénéité temporelle

profil lésionnel de PIC ?

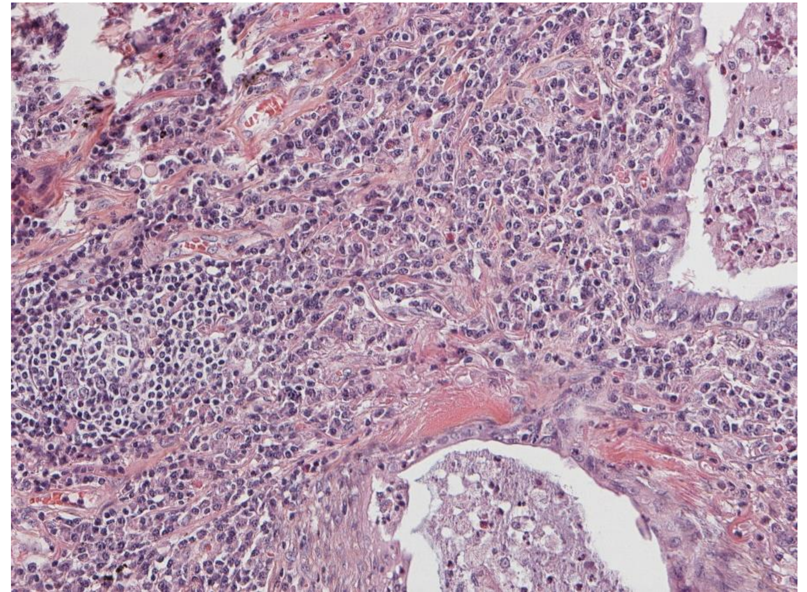
2) Connaitre les critères anapath définissant le profil lésionnel de PIC

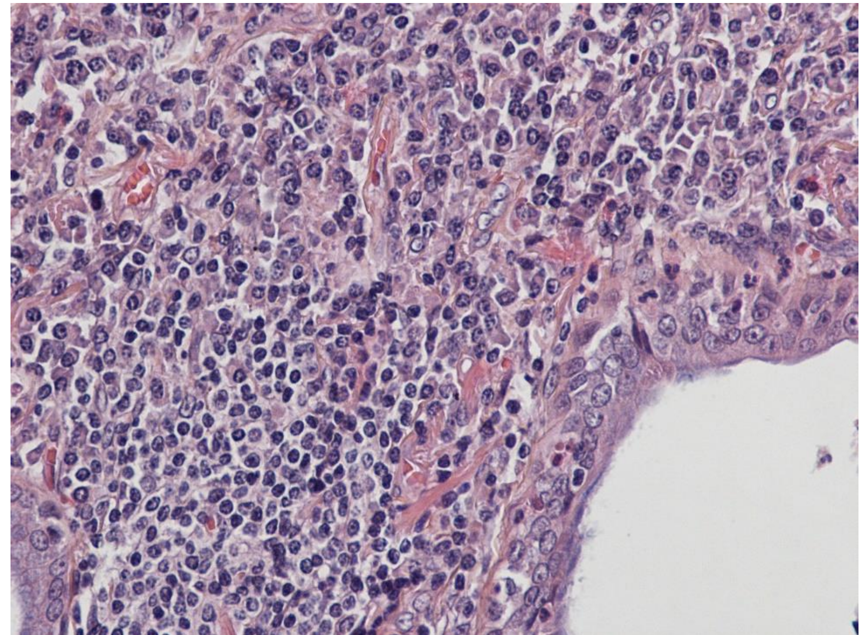
PIC certaine (présence des 4 critères)	PIC probable (présence des 3 critères)	PIC possible (présence des 3 critères)	Signes incompatibles avec un aspect de PIC (au moins un des 6 critères)
• Fibrose marquée/remodelage architectural, +/- rayon de miel de distribution sous-pleurale/paraseptale prédominante • Atteinte disséminée du parenchyme par la fibrose • Présence de foyers fibroblastiques • Absence de signes suggérant un autre diagnostic	• Fibrose marquée/remodelage architecturale, +/ rayon de miel • Absence soit d'atteinte disséminée du parenchyme par la fibrose, soit de foyers fibroblastiques (mais pas d'absence des 2 critères) • Absence de signe[illegible] suggérant un autr[illegible] diagnostic	• Fibrose parenchymateuse disséminée ou diffuse, avec ou sans inflamm[illegible]on [illegible] critères de P[illegible] • Absence de signes suggérant un autre diagnostic	• Membranes hyalines • Pneumopathie organisée (bourgeons fibro-inflammatoires alvéolaires) • Granulomes • Infiltration inflammatoire interstitielle marquée à distance du rayon de miel • Anomalies prédominantes centrées sur les voies aériennes • Autres signes suggérant un autre diagnostic



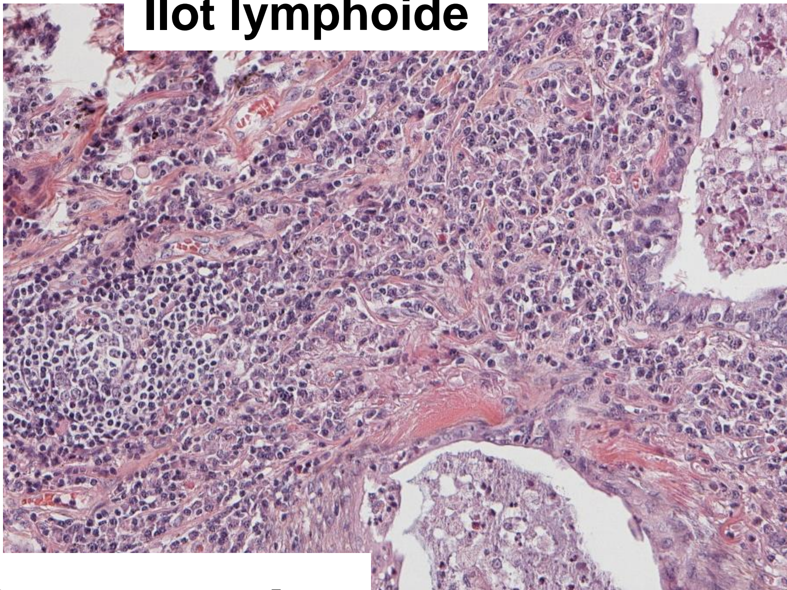
plèvre

Atteinte de la plèvre

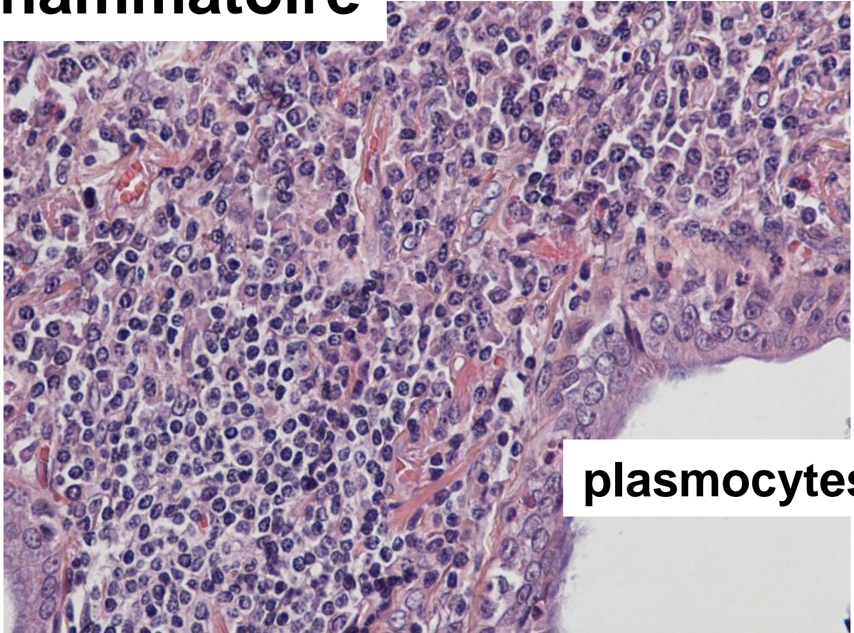




Atteinte de la plèvre



Infiltrat inflammatoire



Differential diagnosis of usual interstitial pneumonia: when is it truly idiopathic?

Wim A. Wuyts[1], Alberto Cavazza[2], Giulio Rossi[3], Francesco Bonella[4], Nicola Sverzellati[5] and Paolo Spagnolo[6]

Eur Respir Rev 2014

Usual interstitial pneumonia-*pattern* fibrosis in surgical lung biopsies. Clinical, radiological and histopathological clues to aetiology

Maxwell Smith,[1] Mercedes Dalurzo,[2] Prasad Panse,[3] James Parish,[4] Kevin Leslie[1]

ABSTRACT
Pulmonary fibrosis in surgical lung biopsies is said to have a 'usual interstitial pneumonia-*pattern*' (UIP-*pattern*) of disease when scarring of the parenchyma is present in a patchy, 'temporally heterogeneous' distribution. These biopsies are one of the more common non-neoplastic specimens surgical pathologists encounter and often pose a number of challenges. UIP is the expected histopathological pattern in patients with clinical idiopathic pulmonary fibrosis (IPF), but the UIP-*pattern* can be seen in other conditions on occasion. Most important among these are the rheumatic interstitial lung diseases (RILD) and chronic hypersensitivity pneumonitis (CHrHP). Because theses entities have different mechanisms of injury, approach to therapy, and expected clinical progression, it is imperative for the surgical pathologist to correctly classify them. Taken in isolation, the UIP-*pattern* seen in patients with IPF may appear to overlap with that of RILD and CHrHP, at least when using the broadest definition of this term (patchy fibrosis). However, important distinguishing features are nearly always present in our experience, and the addition of a multidisciplinary approach will often resolve the critical differences between these diseases. In this manuscript, we review the distinguishing clinical, radiologic and histopathological features of UIP of IPF, RILD and CHrHP, based, in part, on the existing literature, but also lessons learned from a busy lung biopsy consultation practice.

include a heterogeneous group of mainly non-neoplastic disorders. ILDs are mostly inflammatory in nature and the resulting injury to the lung parenchyma can lead to fibrosis and eventual honeycomb cystic remodelling. Some ILDs are characterised by the presence of fibrosis. Dominant among these is 'usual interstitial pneumonia' (UIP) which is the most common histopathological form of diffuse lung fibrosis occurring in older adults, first described by Liebow in 1969.[1] According to Liebow, UIP was idiopathic in about half the affected patients, and the idiopathic form of UIP was referred to clinically as 'idiopathic pulmonary fibrosis' (IPF). The other half of UIP patients had other causes of diffuse lung fibrosis. The common use of the term UIP by pathologists to describe diffuse fibrosis is often confusing to clinicians who tend to assume that a pathological designation of UIP denotes a diagnosis of IPF, not realising that some forms of the UIP-*pattern* of advanced fibrosis can have other aetiologies. Our goal in this review is to shed light on the multidisciplinary clues that are most helpful in distinguishing those forms of UIP that are manifestations of pulmonary fibrosis from other causes.

From a histopathological perspective, the UIP-*pattern* of fibrosis is characterised by two key features:

- Spatial or geographic heterogeneity which refers to a patchy distribution of dense parenchymal scar alternating with areas of less affected or normal parenchyma (figure 1A).

J Clin Pathol 2013

Chronic Hypersensitivity Pneumonitis With a Usual Interstitial Pneumonia-Like Pattern
Correlation Between Histopathologic and Clinical Findings

Sahoko Chiba, MD; Kimitake Tsuchiya, MD, PhD; Takumi Akashi, MD, PhD; Masahiro Ishizuka, MD, PhD;
Tsukasa Okamoto, MD, PhD; Haruhiko Furusawa, MD, PhD; Tomoya Tateishi, MD, PhD;
Mitsuhiro Kishino, MD, PhD;

Chest 2016

Differential diagnosis of usual interstitial pneumonia: when is it truly idiopathic?

Wim A. Wuyts[1], Alberto Cavazza[2], Giulio Rossi[3], Francesco Bonella[4], Nicola Sverzellati[5] and Paolo Spagnolo[6]

Eur Respir Rev 2014; 23: 308–319

Le **profil lésionnel de PIC /UIP**

Support histologique de la fibrose pulmonaire idiopathique

MAIS peut **se rencontrer dans d'autres pathologies** dans lesquelles il signe une évolution fibrosante.

- **PID associées aux Collagénoses**
- **Pneumopathie d'hypersensibilité PHS**
- **Asbestose**
- **PID d'origine médicamenteuse ….**

differences between these diseases. In this manuscript, we review the distinguishing clinical, radiologic and histopathological features of UIP of IPF, RILD and CHrHP, based, in part, on the existing literature, but also lessons learned from a busy lung biopsy consultation practice.

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J Clin Pathol 2013;66:896–903

Differential diagnosis of usual interstitial pneumonia: when is it truly idiopathic?

Wim A. Wuyts[1], Alberto Cavazza[2], Giulio Rossi[3], Francesco Bonella[4], Nicola Sverzellati[5] and Paolo Spagnolo[6]

Eur Respir Rev 2014; 23: 308–319

TABLE 1 Histological clues that, when present in the usual interstitial pneumonia pattern, suggest the possibility of an underlying connective tissue disease (CTD) or chronic hypersensitivity pneumonitis (HP)

Feature	CTD	Chronic HP
Cellular (lymphocytic and/or plasmacellular) interstitial infiltrate	+	+
Plasma cells	+	+
Cellular bronchiolitis	+	+
Centrilobular fibrosis, with or without bridging fibrosis between bronchioles and pleura	-	+
Pleuritis	+	-
Small interstitial/peribronchiolar granulomas	–#	+
Coexistence of more than one pattern in the same biopsy	+	-/+

positive; –: negative. #: except in Sjögren's syndrome, in which small granulomas can be found.

Differential diagnosis of usual interstitial pneumonia: when is it truly idiopathic?

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Eur Respir Rev 2014; 23: 308–319

Usual
in su
histo

Maxwell

ABSTRACT
Pulmonary f
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J Clin Pathol 2013;66:896–903

Le **profil lésionnel de PIC /UIP**

Support histologique de la fibrose pulmonaire idiopathique

MAIS ... es

dans l

-
- Pneumopathie d'hypersensibilité PHS
- **Asbestose**
- **PID d'origine médicamenteuse ….**

Le profil lésionnel de PIC n'est pas spécifique

Alerter le pneumologue
sur caractère **non idiopathique**
des lésions fibrosantes

antécédents

- Ancien ajusteur
- Asthme apparu en 1989, sévère - actuellement de palier 2 du GINA contrôlé par SERETIDE 250X2
- Rhinite inflammatoire et obstructive – 1993: septoplastie, ethmoidectomie antérieure, méatomie moyenne bilatérale
- Diabè
- Non fu

biologie

- Hb : 15,6 – leucocytes : 5 540 – éosinos 4,7%- plaquettes : 266 000 – CRP 1
- Clearance créatinine : 75 ml/mn, iono Normal
- SGOT 28 SGPT 28 Ph alcaline 105
- Pro BNP : 60
- Électrophorèse des protides normale :augmentation polyclonale sans pic
- Enzyme de conversion de l'angiotensine : 29 UI
- VIH 1 et 2 : négatif

Discussion

Bilan auto immun ? A refaire ?

Manifestations extra-pulmonaires ?

Hi

Dyspne

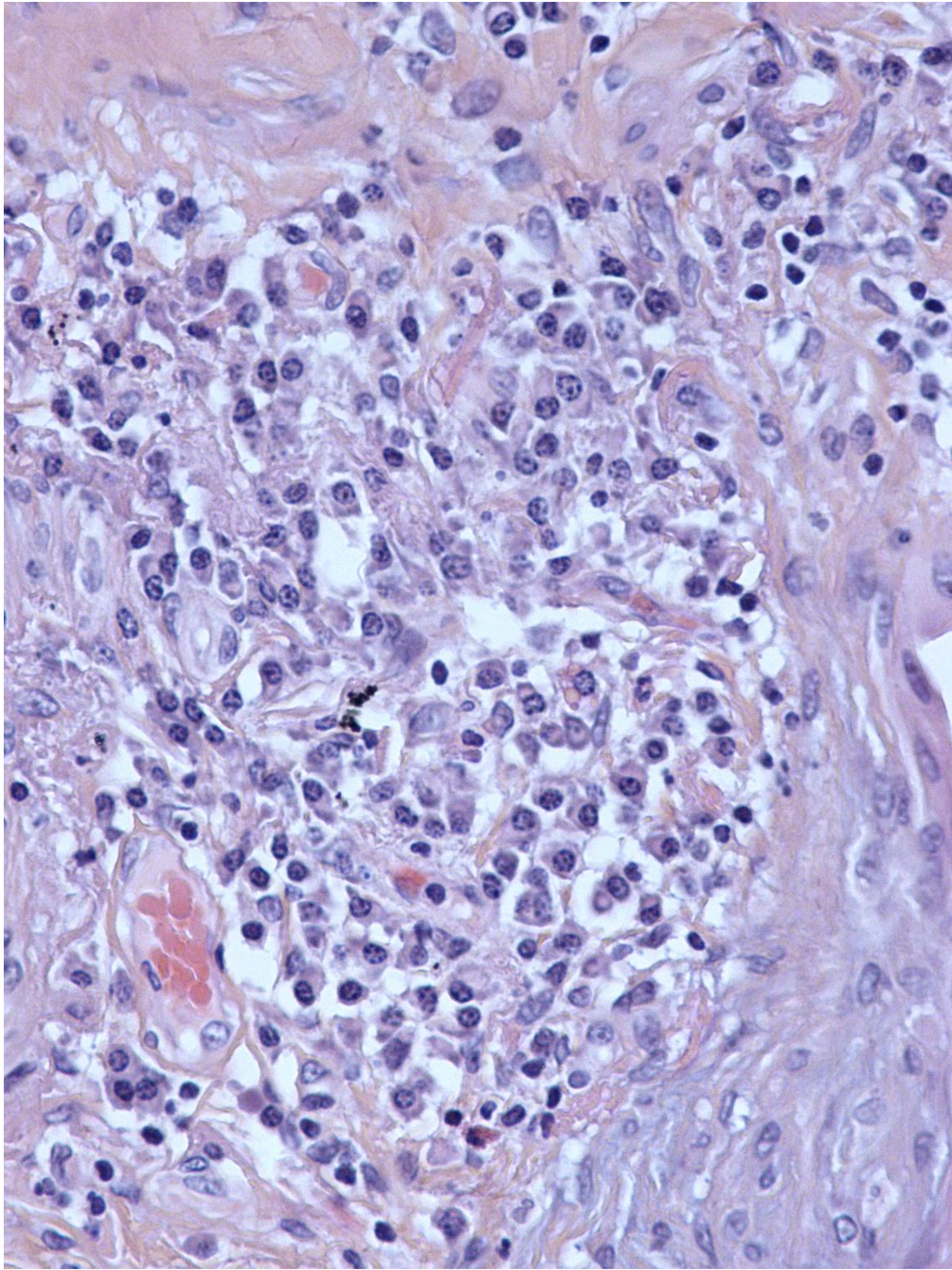
Découverte de crépitants velcro

Bon EG – 75 kg pour 1m72

Pas d'hippocratisme digital

- **EFR**
  CV 48 % (1L750) – VEMS 61,7% - CPT 43,7
- **GDS sous air**
  PaO2 77 – PaCO2 46 – SaO2 95,4%
- **TCO**
  73,8%

Dossier clinique Dr T.Pigeanne pneumologue Sables d'Olonnes



infiltrat plasmocytaire polytypique
nombreux IgG4 répartition variable
IgG4 par champ au grossissement 400 > 50
un rapport IgG4/IgGG > 40 %

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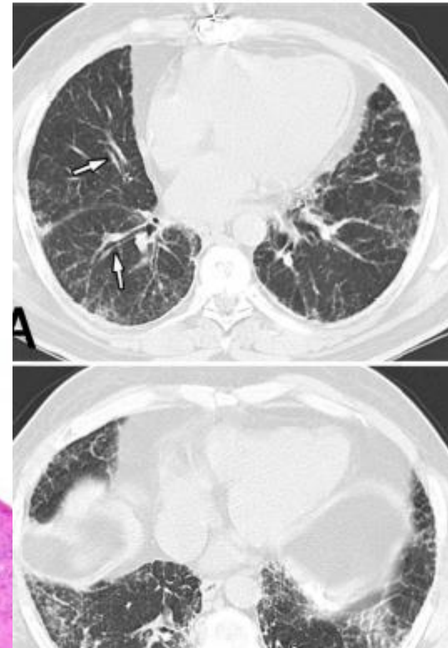
DOI: 10.2174/1874312901610010033

IgG4-Related Lung Disease Associated with Usual Interstitial Pneumonia

Frank Schneider[1,*], Kristen L. Veraldi[2], Marc C. Levesque[2], Thomas V. Colby[3] and Eunhee S. Yi[4]



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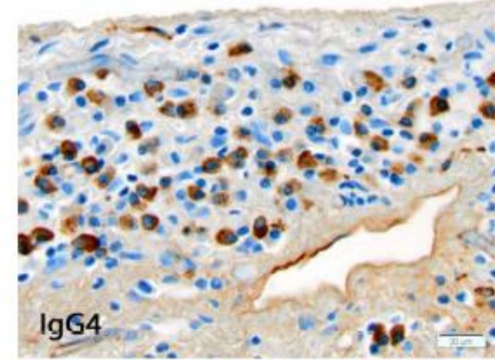
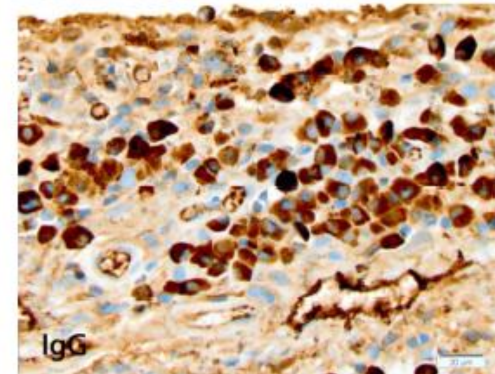
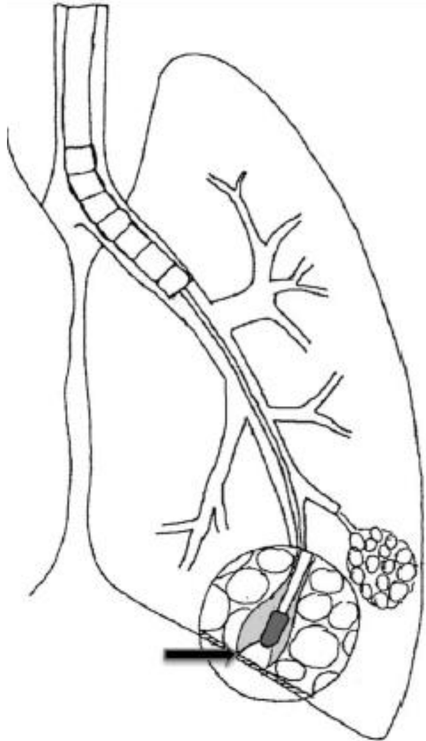




Observation - Cryobiopsies



Respirology

INVITED REVIEW SERIES:
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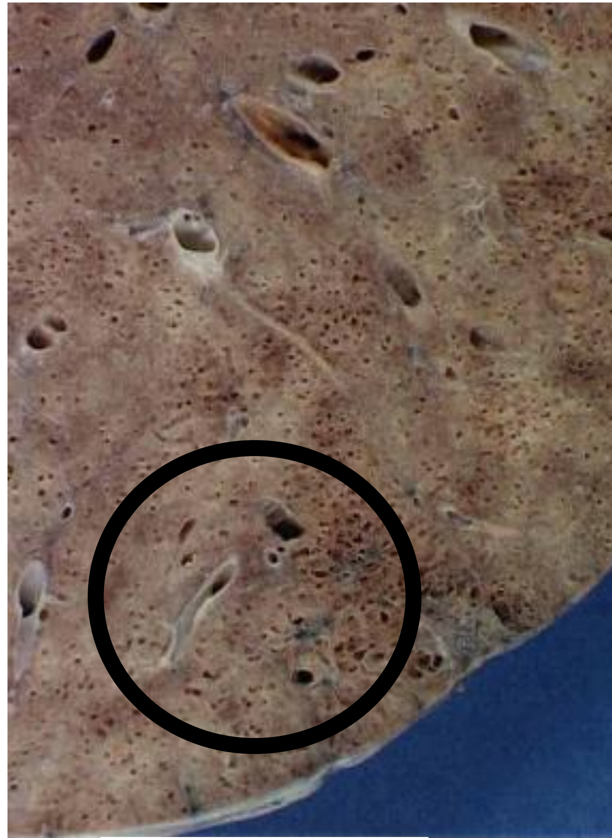
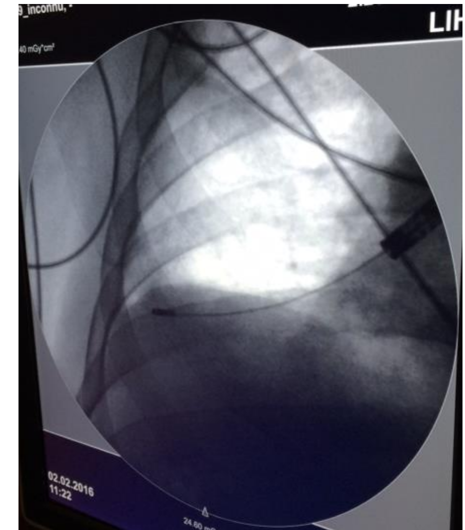
Lung cryobiopsies: A paradigm shift in diagnostic bronchoscopy?

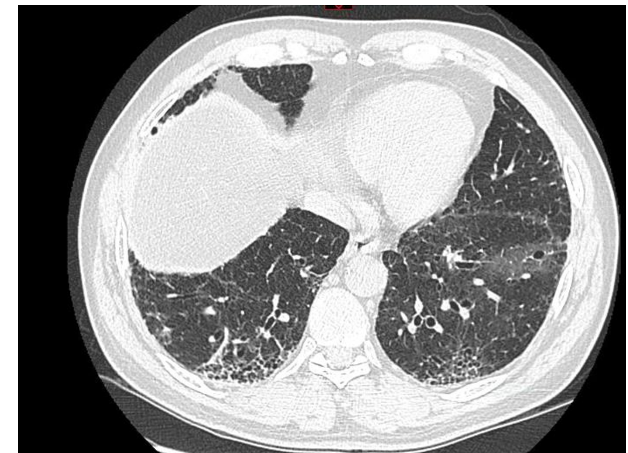
VENERINO POLETTI,[1] GIAN LUCA CASONI,[1] CARLO GURIOLI,[1] JAY H. RYU[2] AND SARA TOMASSETTI[1]

[1]Department of Diseases of the Thorax/Pulmonology Unit, Ospedale GB Morgagni, Forlì, Italy, and [2]Respiratory and Critical Care Medicine, Mayo Clinic, Rochester, Minnesota, USA



Poumon cortical





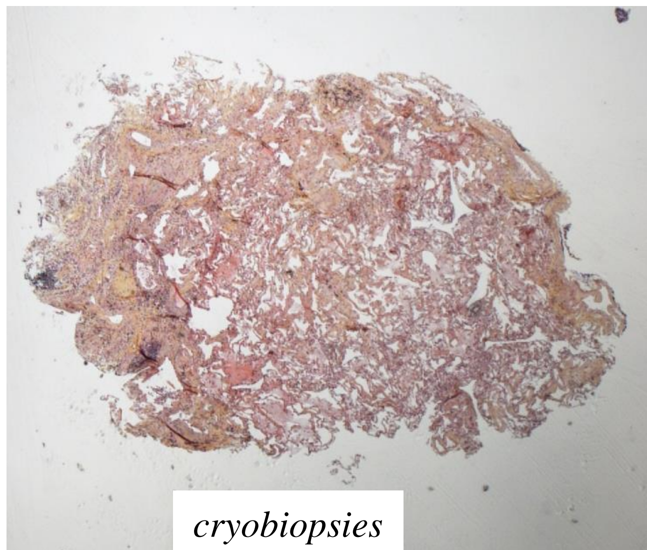
Observation - Cryobiopsies

Official Journal of the Asian Pacific Society of Respirology
Respirology

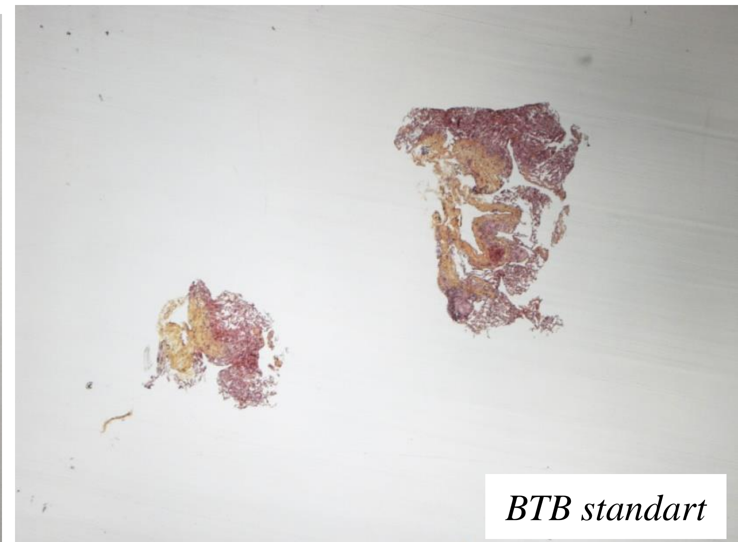
EDITORIAL

Transbronchial cryobiopsy in the diagnosis of interstitial lung disease: A cool new approach

Respirology (2014) **19**, 623–624

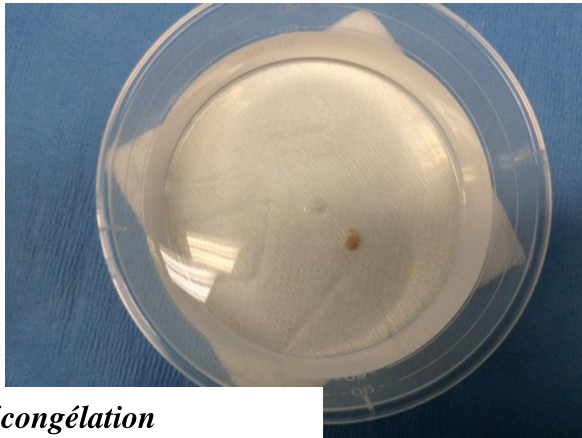


cryobiopsies



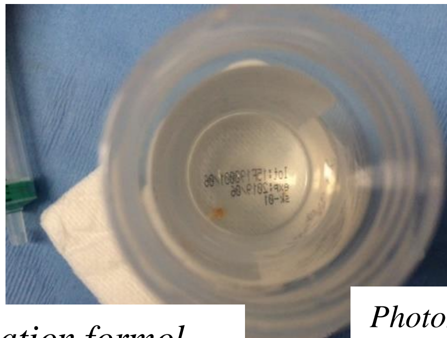
BTB standart

Prélèvement de qualité



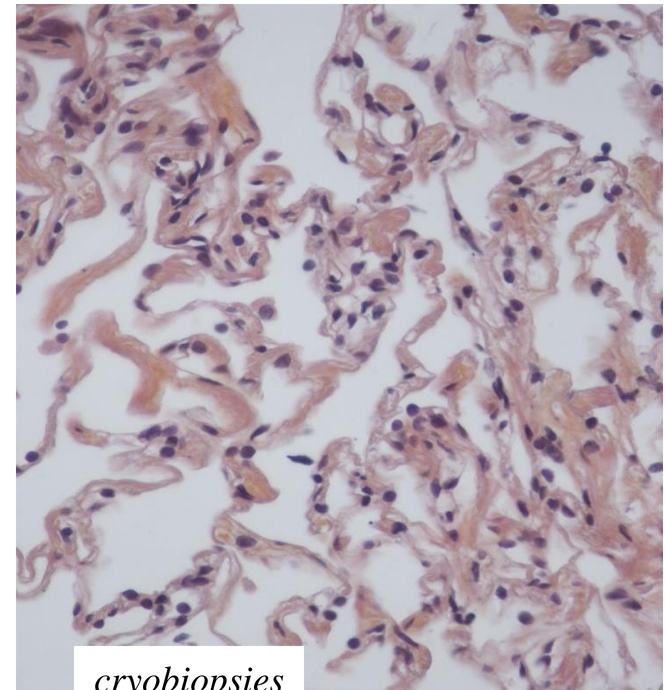
Décongélation
Sérum physiologique…

Décongélation ***hors*** *de la bronche*
dans sérum physiologique
au moins 2 secondes
avant fixation dans formol

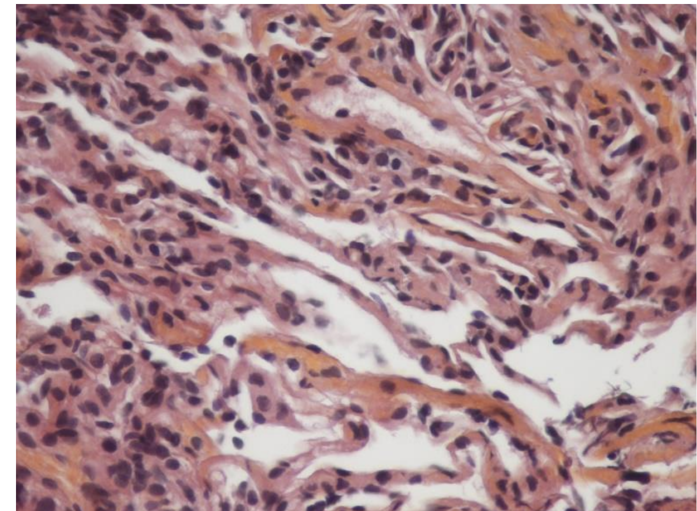


Puis fixation formol

Photos A cavailles

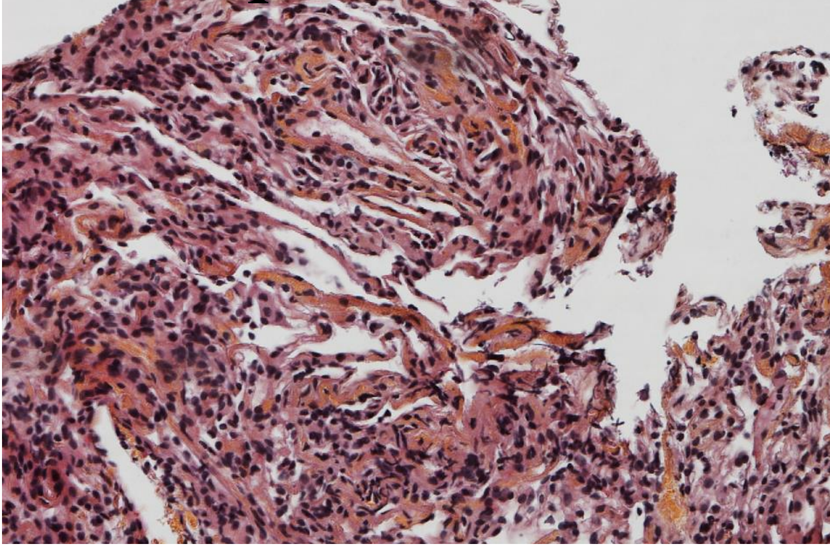


cryobiopsies

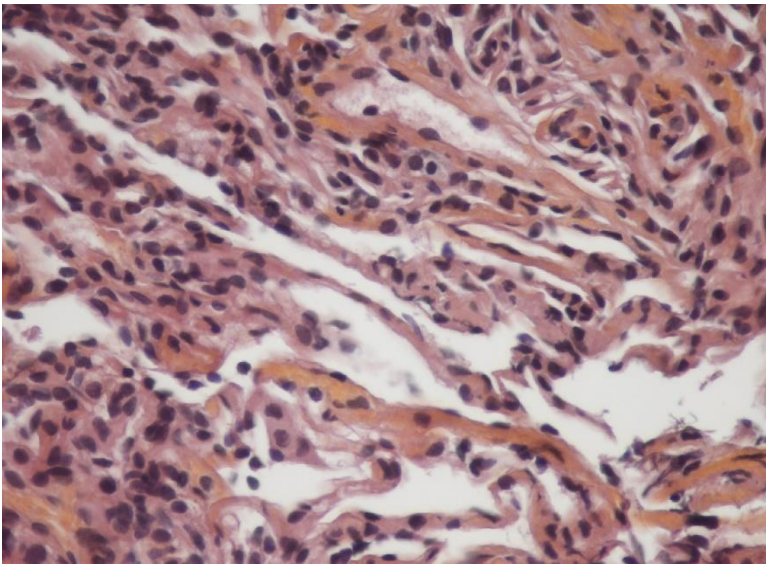


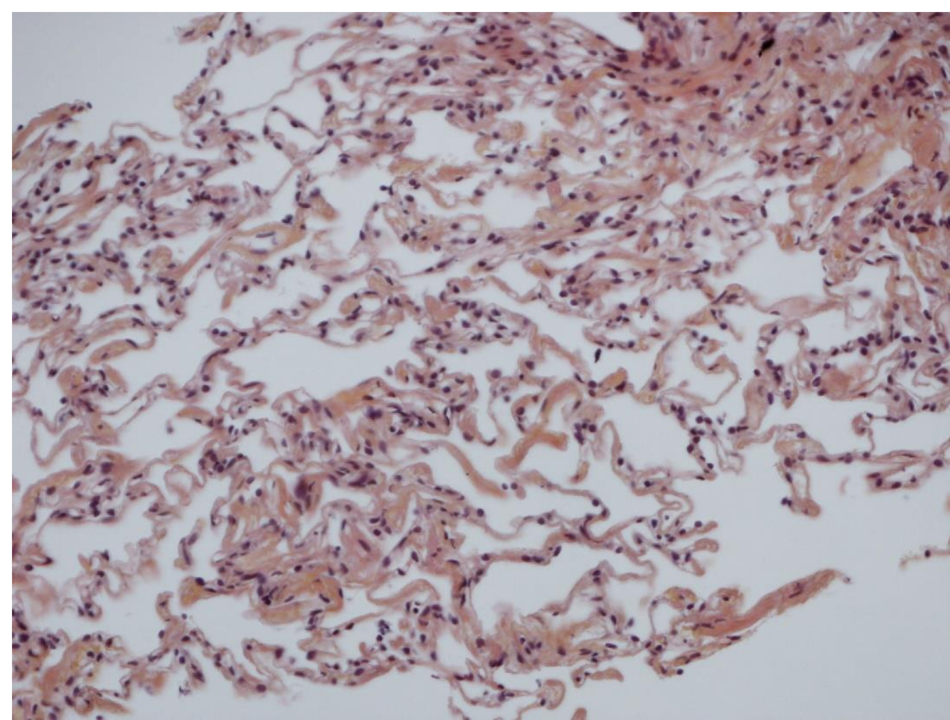
BTB standart

Prélèvement de qualité ?

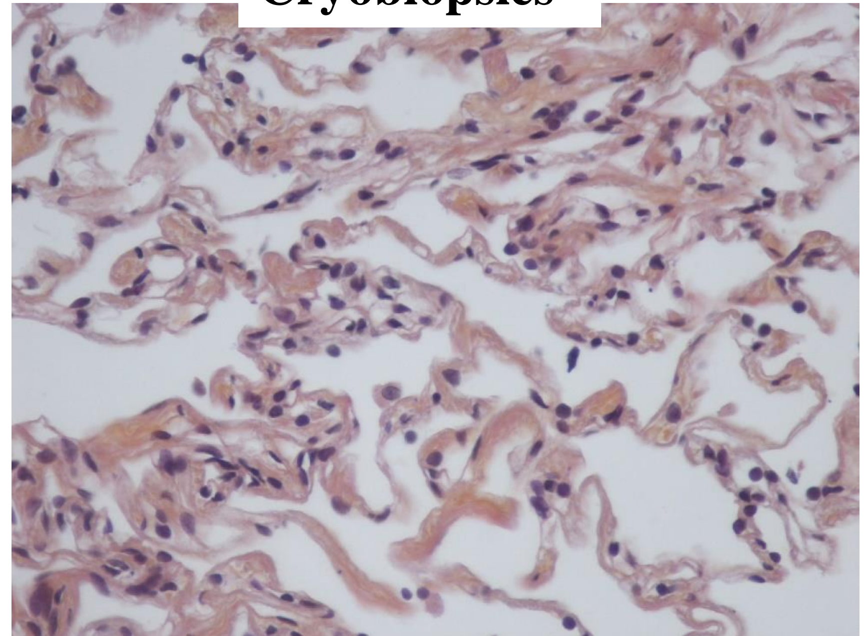


Biopsies transbronchiques standarts

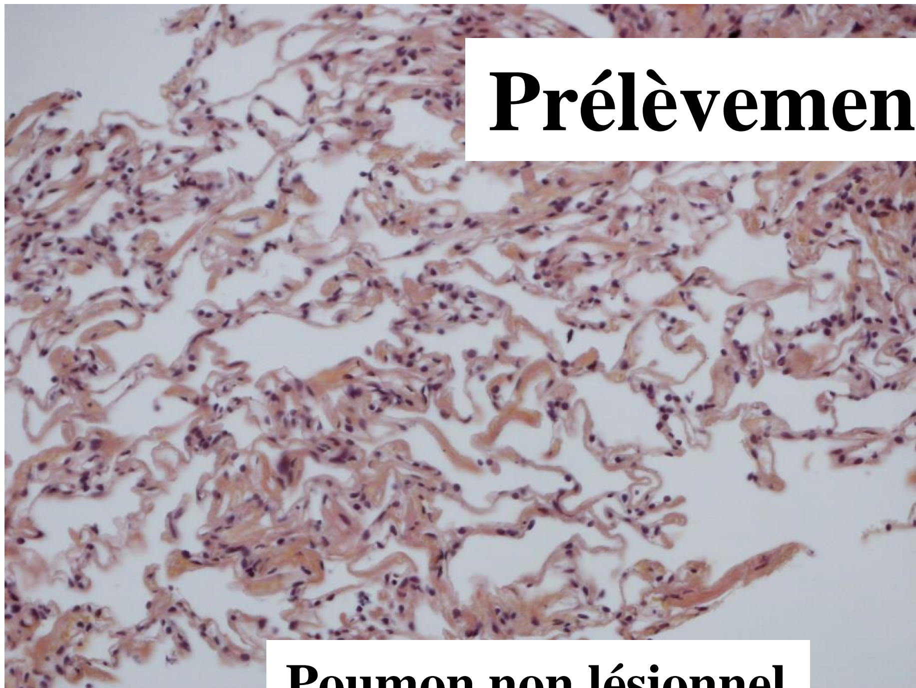




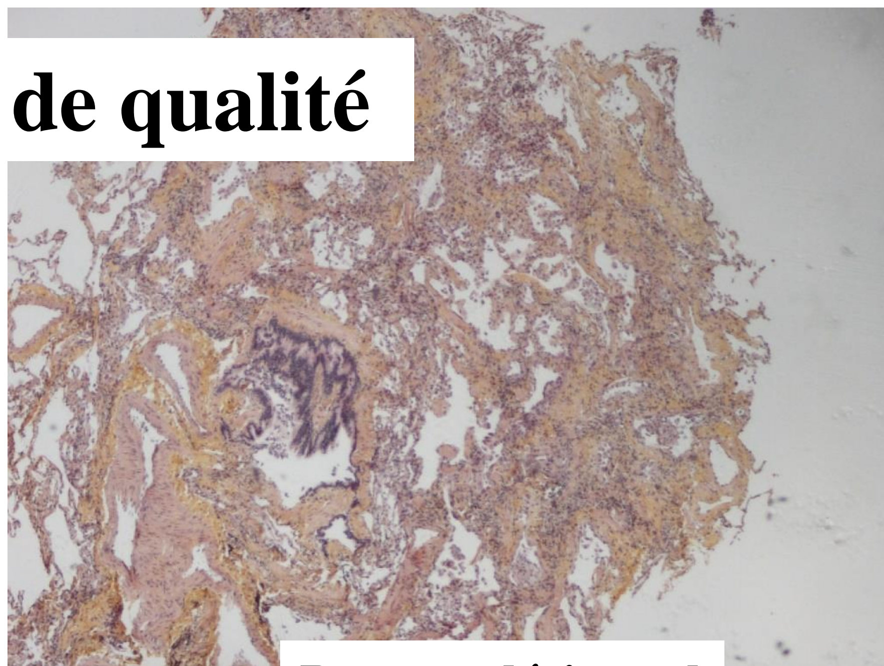
Cryobiopsies



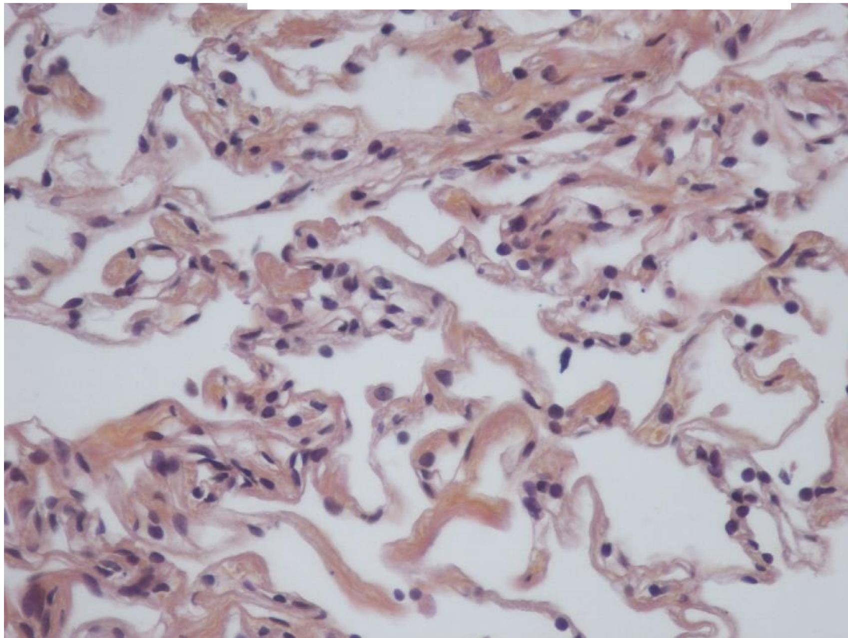
Prélèvement de qualité

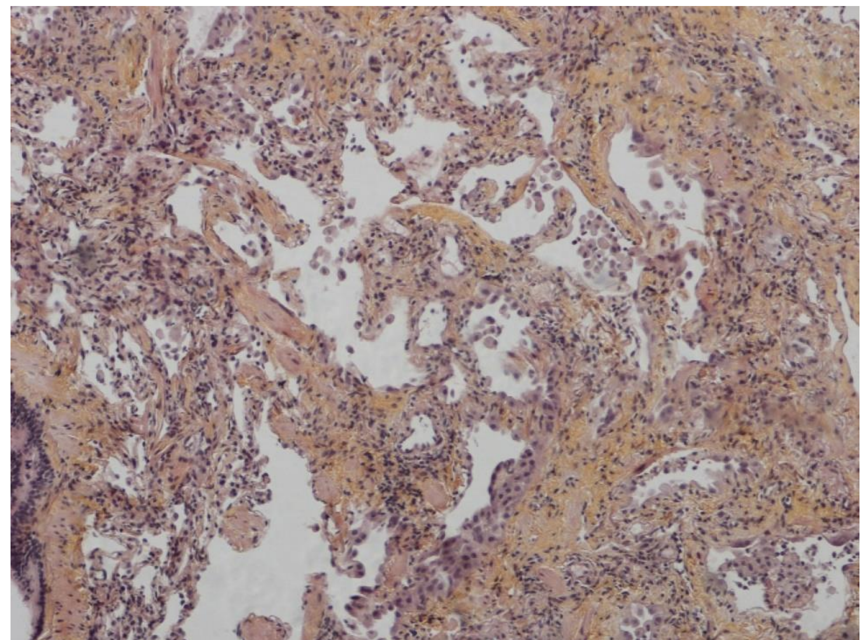


Poumon non lésionnel



Poumon lésionnel





Observation - Cryobiopsies

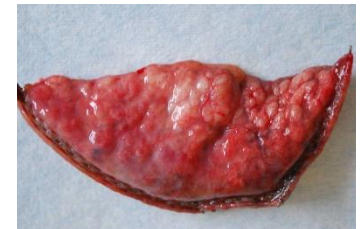
EDITORIAL

Transbronchial cryobiopsy in the diagnosis of interstitial lung disease: A cool new approach

Respirology (2014) **19**, 623–624

American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias

THIS JOINT STATEMENT OF THE AMERICAN THORACIC SOCIETY (ATS), AND THE EUROPEAN RESPIRATORY SOCIETY (ERS) WAS ADOPTED BY THE ATS BOARD OF DIRECTORS, JUNE 2001 AND BY THE ERS EXECUTIVE COMMITTEE, JUNE 2001



Transbronchial Cryobiopsy in Diffuse Parenchymal Lung Disease: Need for Procedural Standardization

V enerino Poletti[a, b] Jürgen Hetzel[c]

[a]D epartment of Respiratory Diseases and Allergology, Aarhus University Hospital, A arhus, Denmark; [b] D epartment of Diseases of the Thorax, Ospedale GB Morgagni, F orlì, Italy; [c]D ivision of Pulmonary Medicine, Department of Internal Medicine, University of Tüb ingen, T üb ingen, Germany

Transbronchial Cryobiopsy in Diffuse Lung Disease

Update for the Pathologist

Thomas V. Colby, MD; Sara Tomassetti, MD; Alberto Cavazza, MD; Alessandra Dubini, MD; Venerino Poletti, MD

Arch Pathol Lab Med 2016

10.5858/arpa.2016-0233-RA

- **Bonne qualité**
    - Taille au moins 0,5cm – 4 fois plus large / BTB
    - multiples
- **Diagnostic possible** *Critères consensus ATS /ERS 2001*
    - 297 patients - 17,2% prélèvements inadéquats (*Ravaglia 2016)*
    - 15series – 780 patients 82% diagnostic cryobiopsie /98% chirurgie
- Complication : pneumothorax 10 30%
- **Impact clinique – place des cryobiopsies dans les DMD**

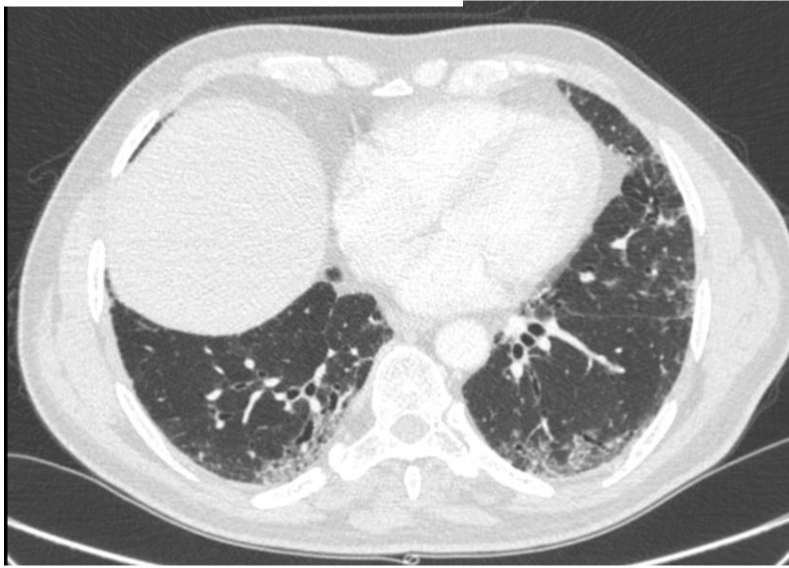
Bronchoscopic Lung Cryobiopsy Increases Diagnostic Confidence in the Multidisciplinary Diagnosis of Idiopathic Pulmonary Fibrosis

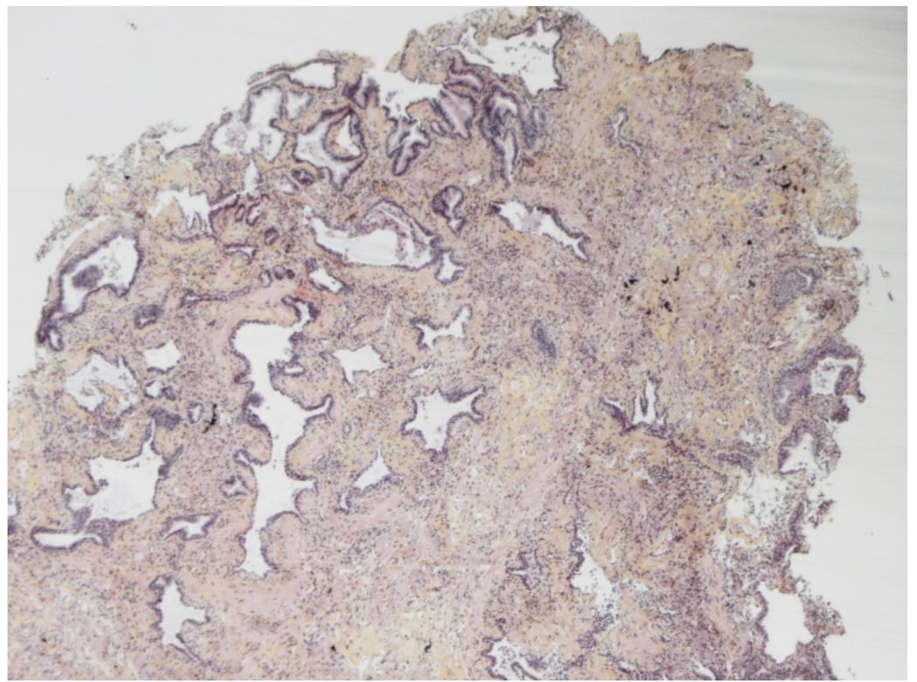
Sara Tomassetti[1], Athol U. Wells[2], Ulrich Costabel[3], Alberto Cavazza[4], Thomas V. Colby[5], Giulio Rossi[6], Nicola Sverzellati[7], Angelo Carloni[8], Elisa Carretta[9], Matteo Buccioli[1], Paola Tantalocco[1], Claudia Ravaglia[1], Christian Gurioli[1], Alessandra Dubini[10], Sara Piciucchi[11], Jay H. Ryu[12], and Venerino Poletti[1,13]

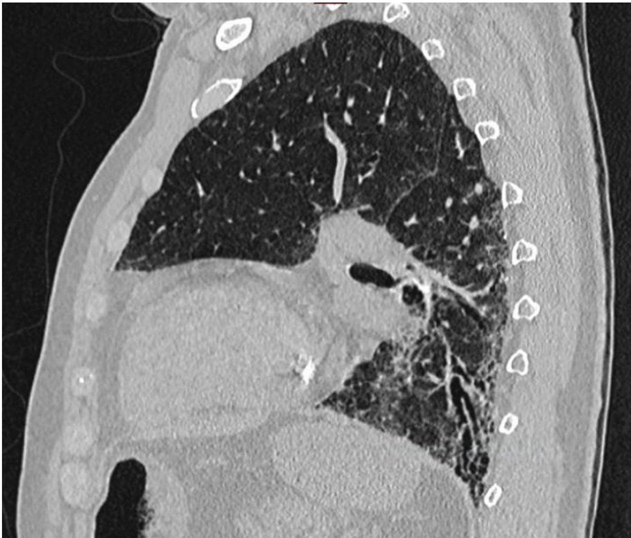
58 cryobiopsies
59 biopsies chirurgicales

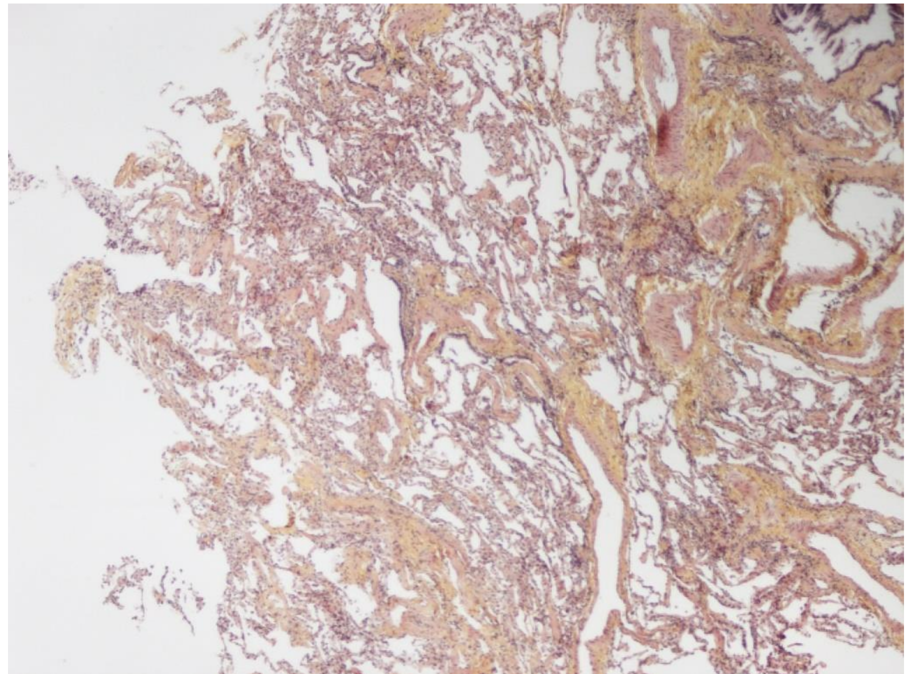
Am J Respir Crit Care Med 2016

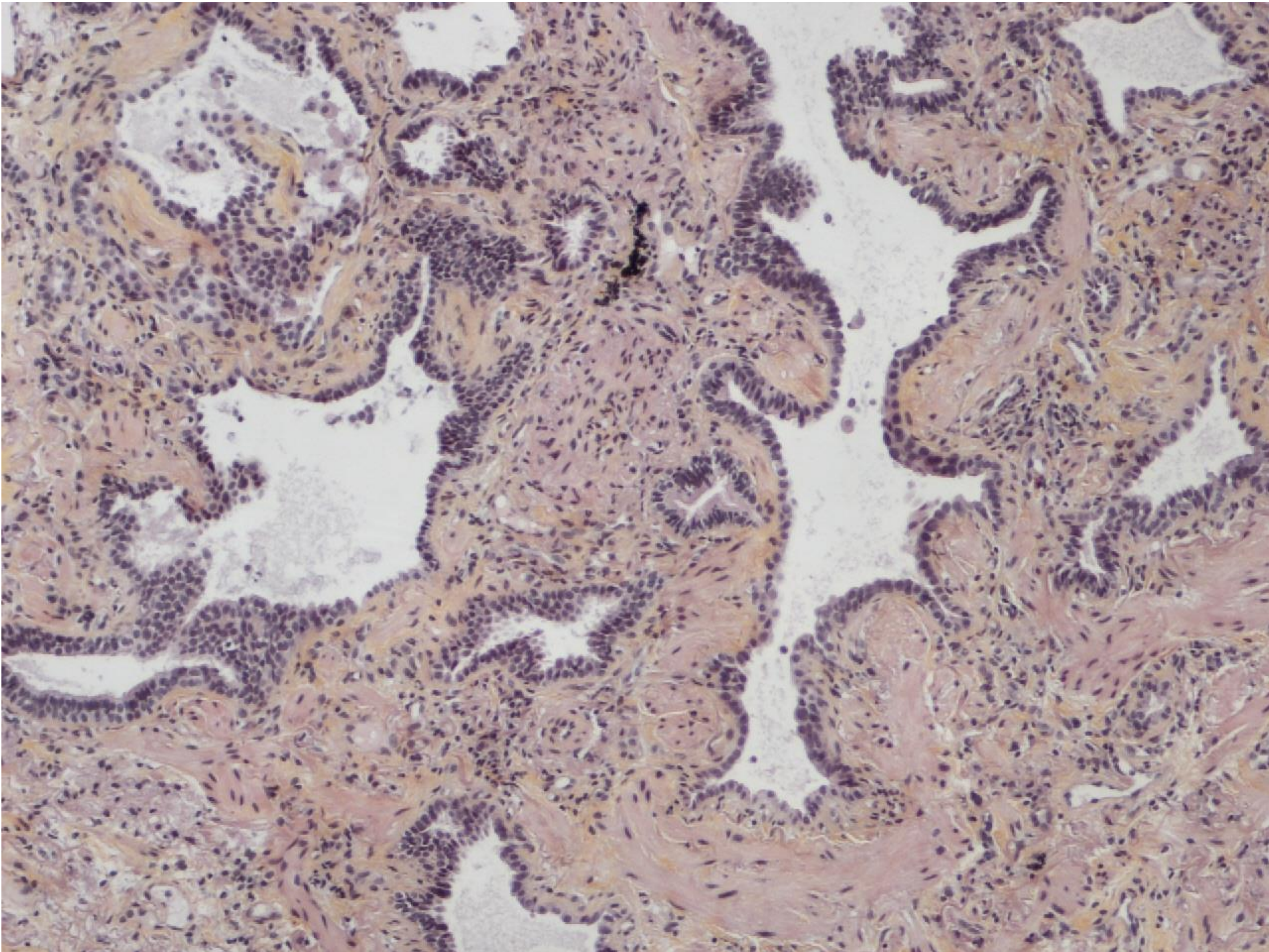
Homme né en 1942
PIC possible

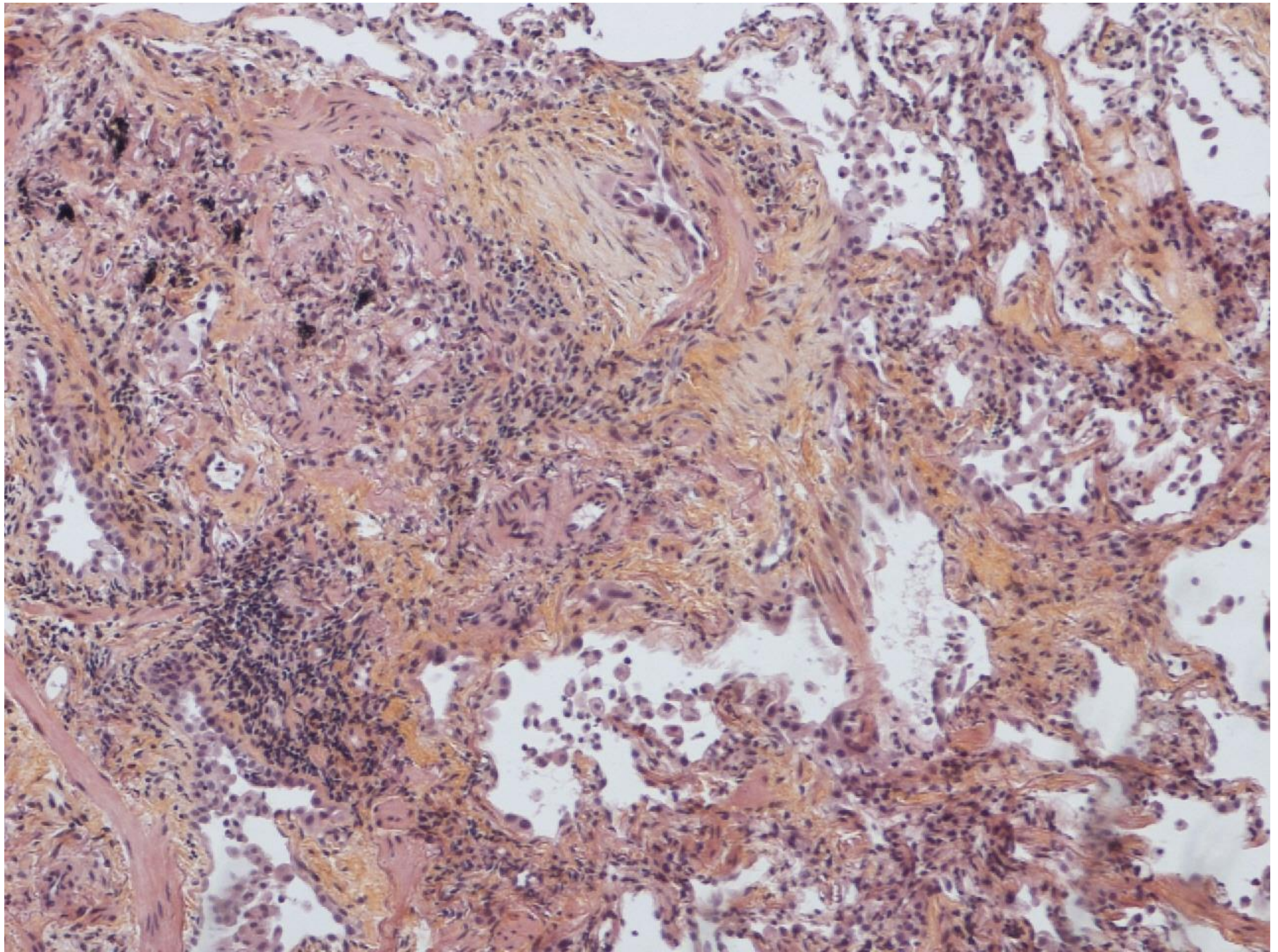


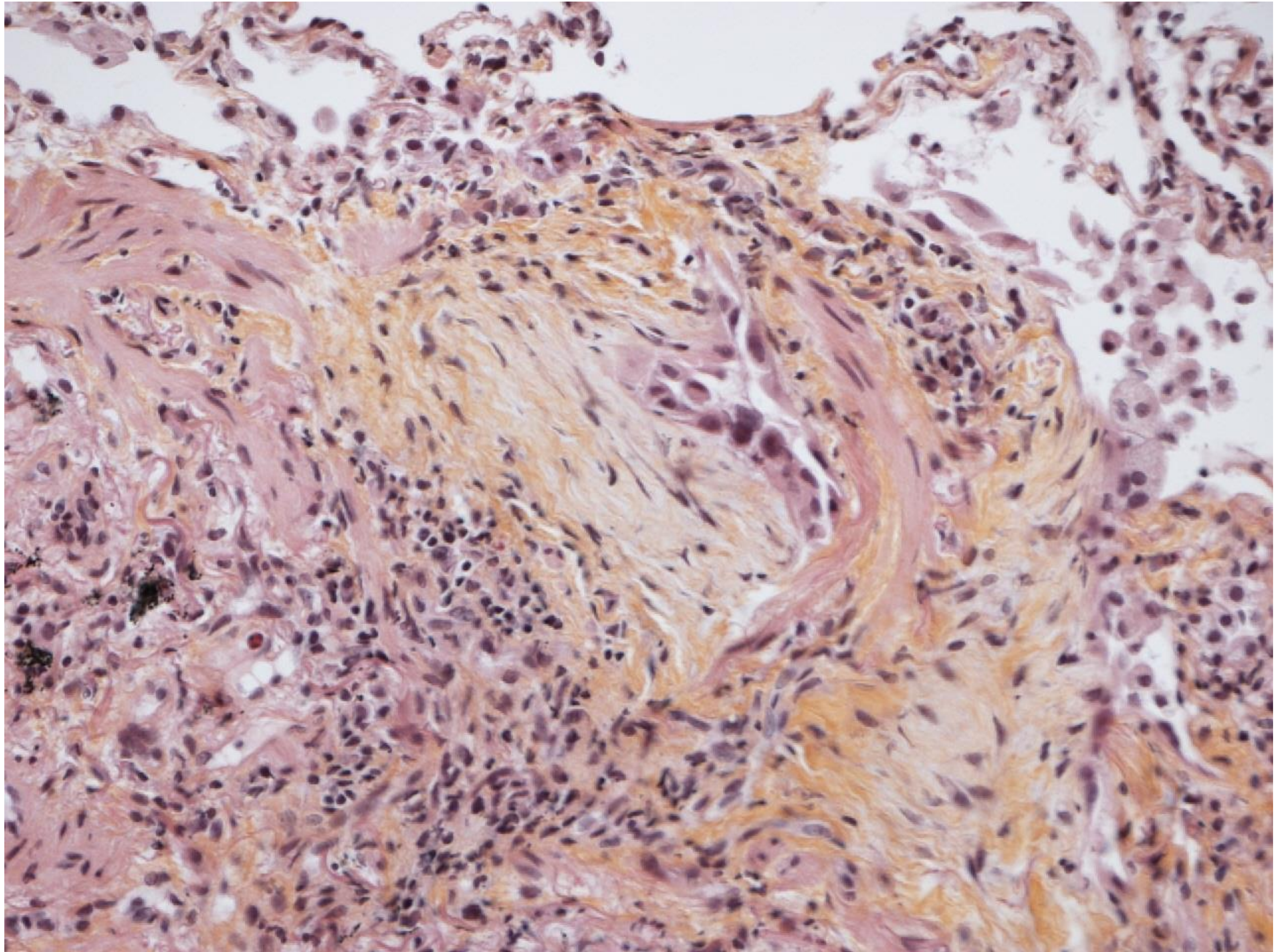


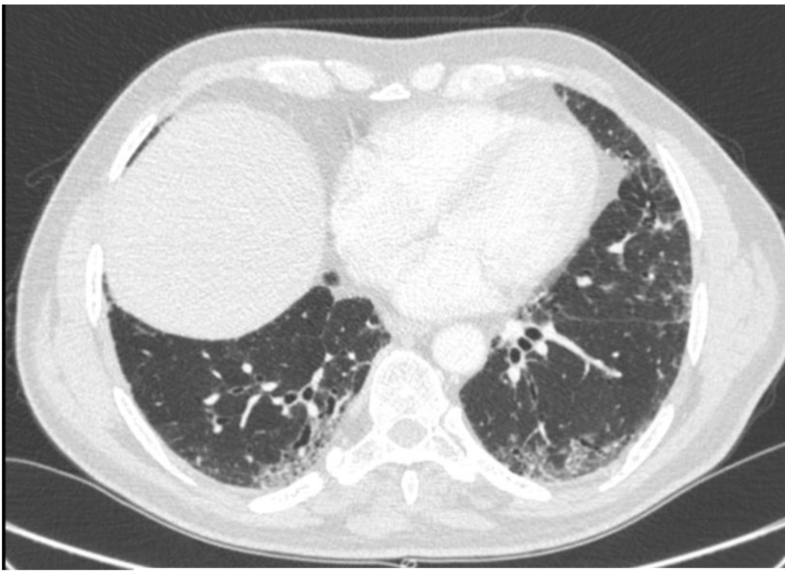


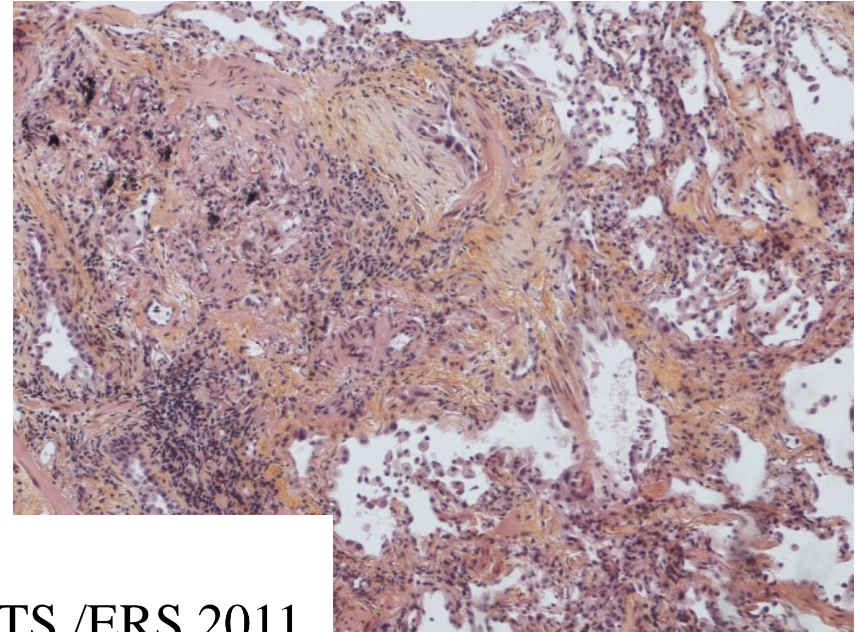






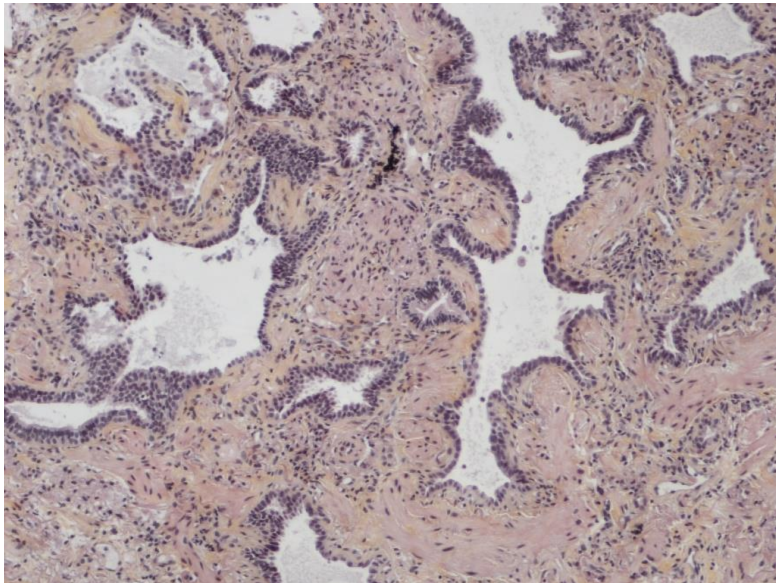


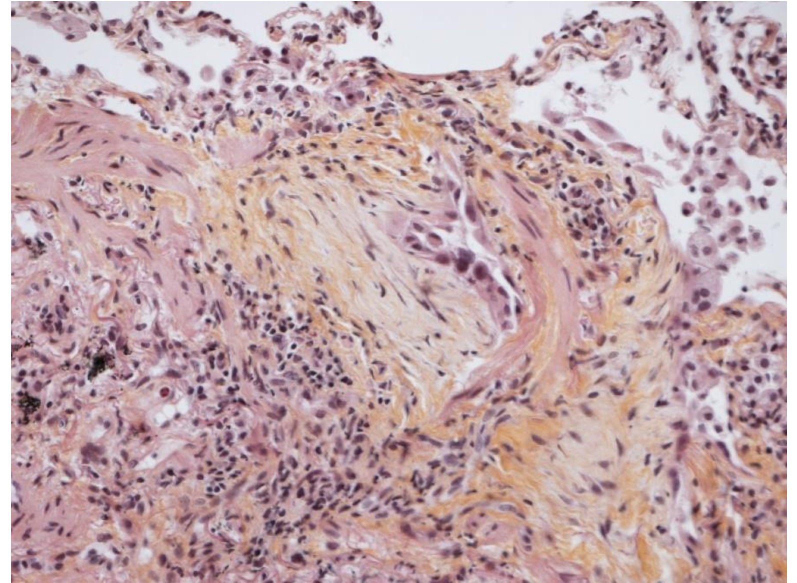




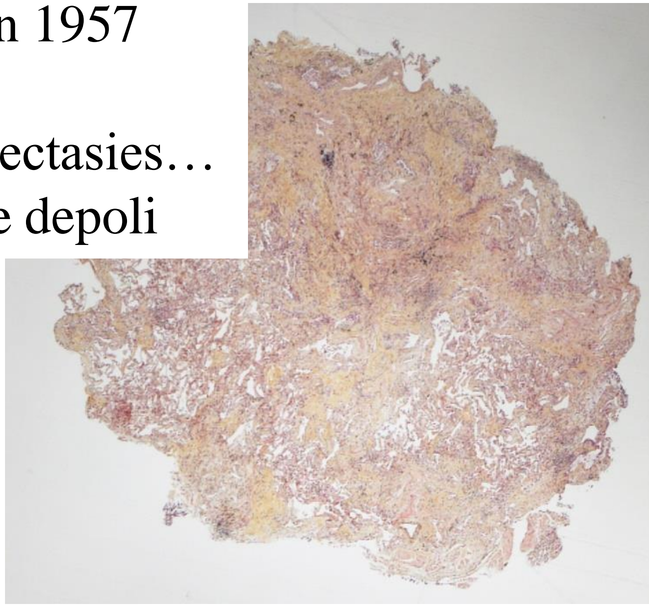
Profil lésionnel de PIC

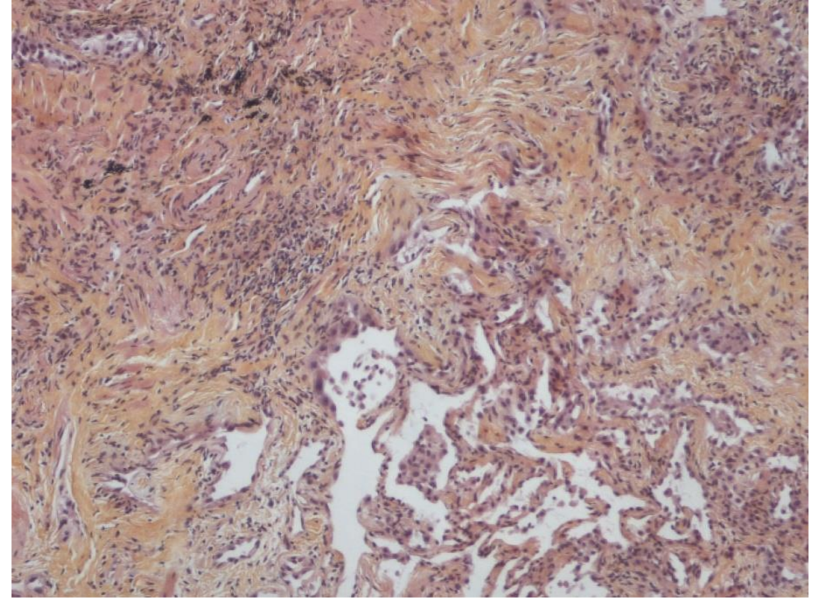
PIC « **probable** » selon le consensus ATS /ERS 2011

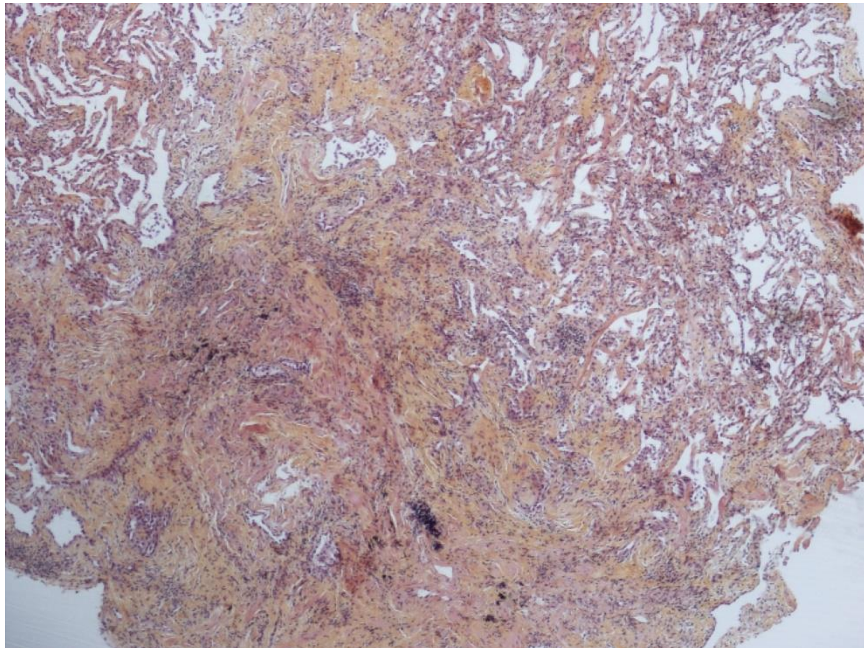


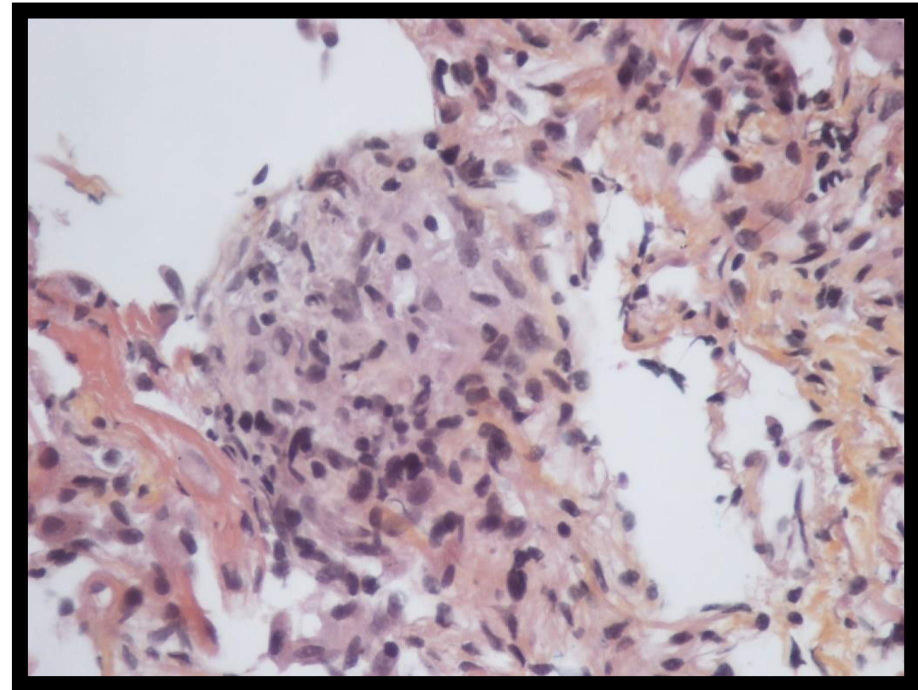


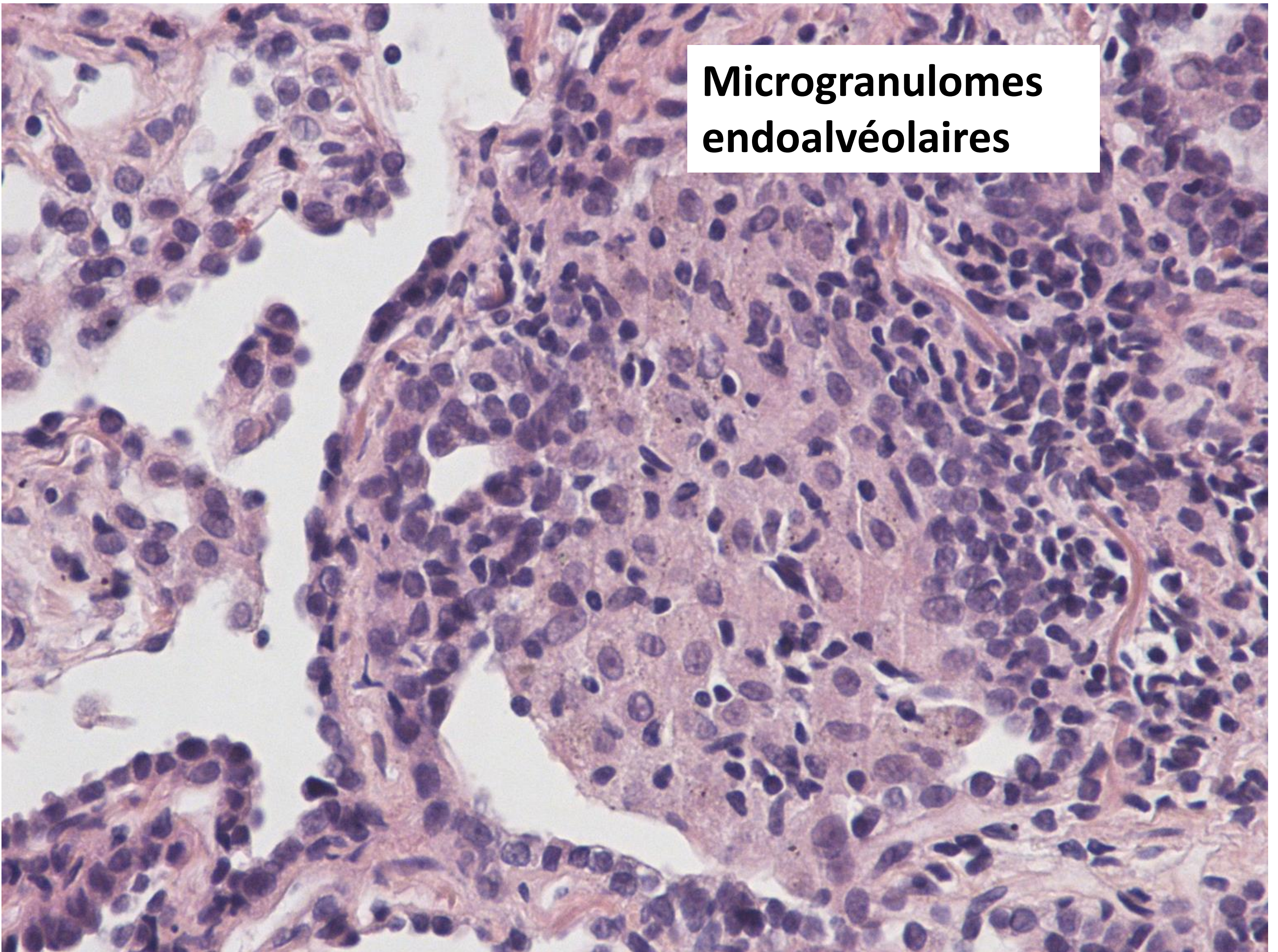
Homme né en 1957
PIC possible
Apex bronchectasies…
Minime verre depoli










Microgranulomes
endoalvéolaires

Homme né en 1957
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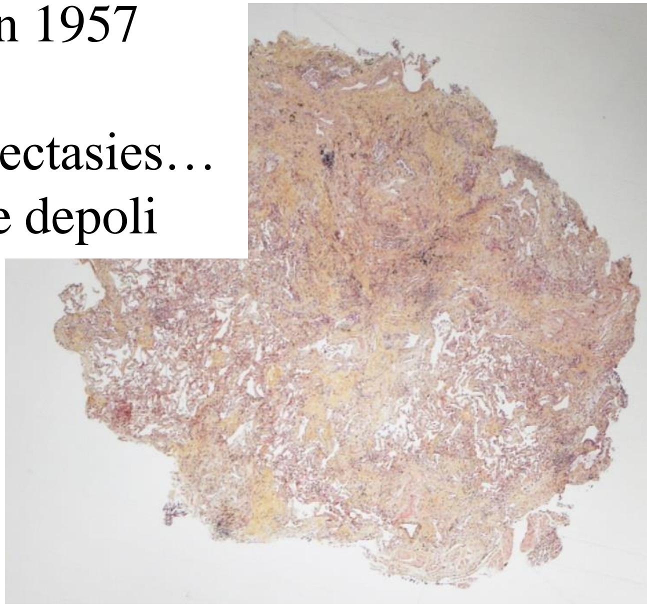
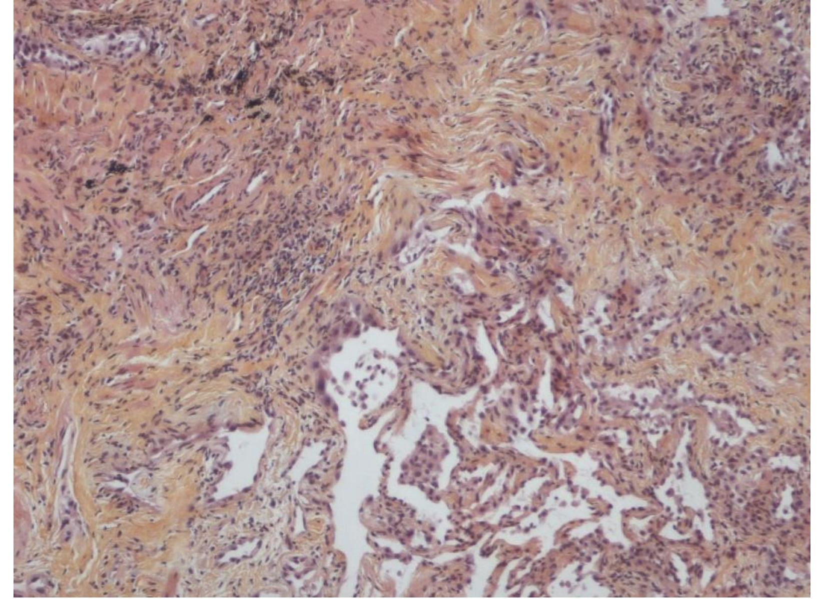
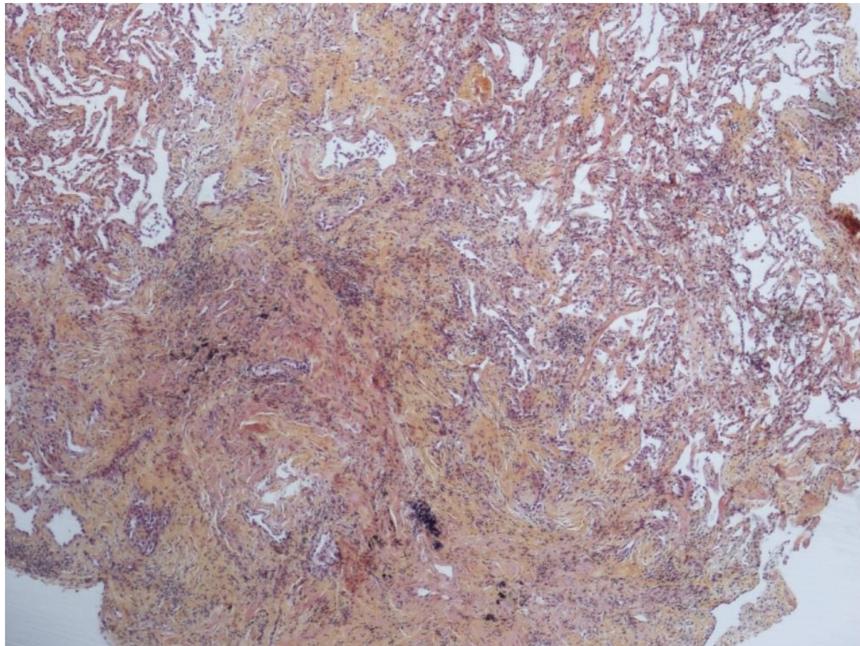
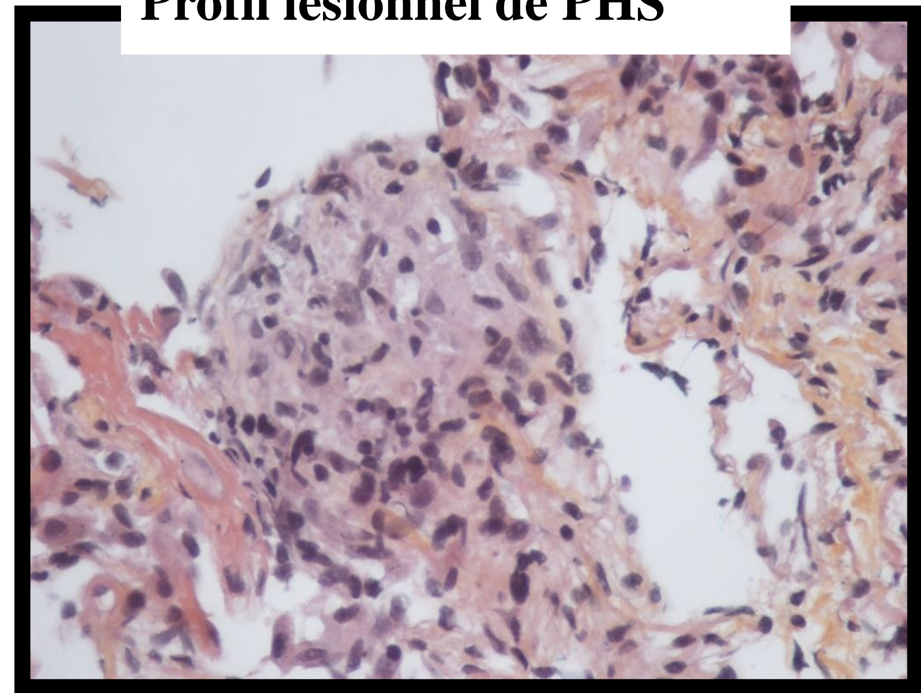
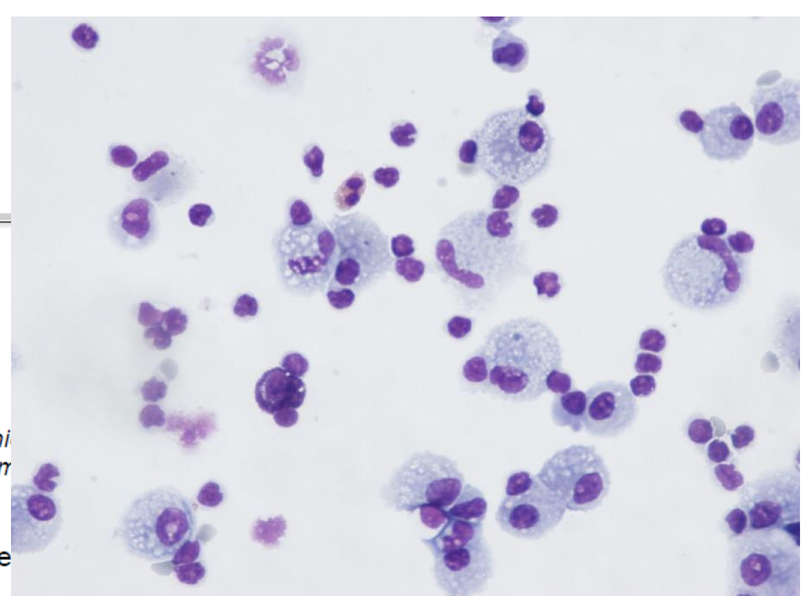






Profil lésionnel de PHS

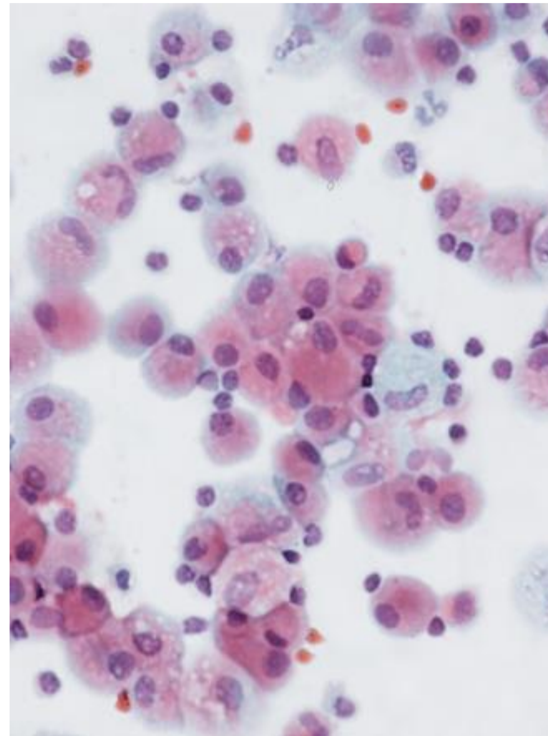


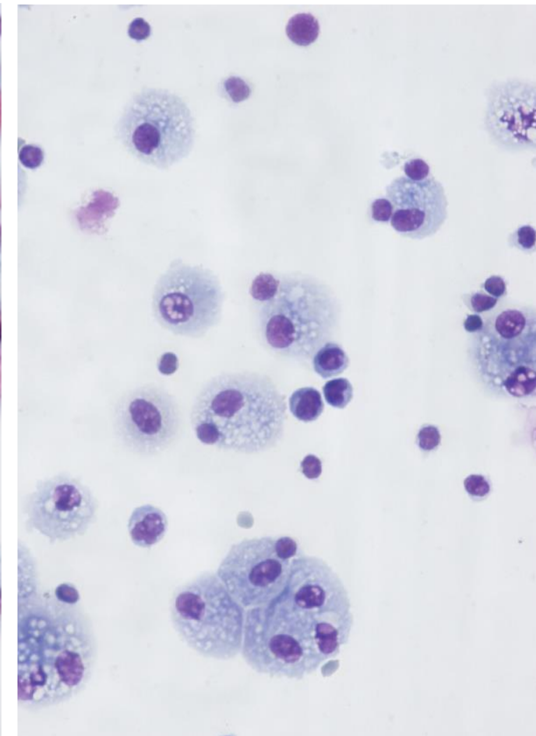


POUMON DROIT – Lavage broncho-alvéolaire du lobe moyen
(données endoscopiques : 90 ml instillés ; 46 ml recueillis)

Aspect : trouble
Volume : 16 ml
Cellularité : 410.000 cellules/ml
Technique : examen après cytocentrifugation sur quatre lames (colorations de Papani Grünwald Giemsa, Perls et Grocott) et sur culot de centrifugation (coloration d'hém safran). .

- Lavage bien préservé, peu contaminé (quelques traînées mucus, rare cellules bronchiques) – Fond avec exceptionnelles hématies.
- Formule :
  - 57 % de macrophages
  - 42 % de lymphocytes
  - 1 % de polynucléaires éosinophiles
- Aspect cytologique : lymphocytes principalement de pet activées et éléments de plus grande taille ; présence de parfois en transformation épithélioïde ; quelques macrophage vacuoles ; desquamation de quelques macrophages en paque
- Perls : pas de sidérophage
- Grocott : pas d'agent infectieux
- Culot de centrifugation : pas de constatation supplémentai





Conclusion :

- **lavage broncho-alvéolaire avec lymphocytose (42 % de petite taille ; rares formes activées)**
- **minimes modifications des macrophages pouvant d'alvéolite immune**

PID prise en charge par le pathologiste

- **Respect des prérequis indispensable**
  - **qualité des prélèvements**
  - **connaissance des profils lésionnels de PII**
  - **prise en charge avec Discussion mltidisciplinaire DMD**
- **Démarche sémiologique**
- **Etre attentif aux atypies dans les profils proposés**

PID
prise en charge par le pathologiste

Merci de votre attention

Multidisciplinary discussions and interstitial lung disease diagnosis: how useful is a meeting of the minds?

K C Meyer
Lancet Respi Med 2016