Bronchiolar disorders: Current perspective on diagnosis & management

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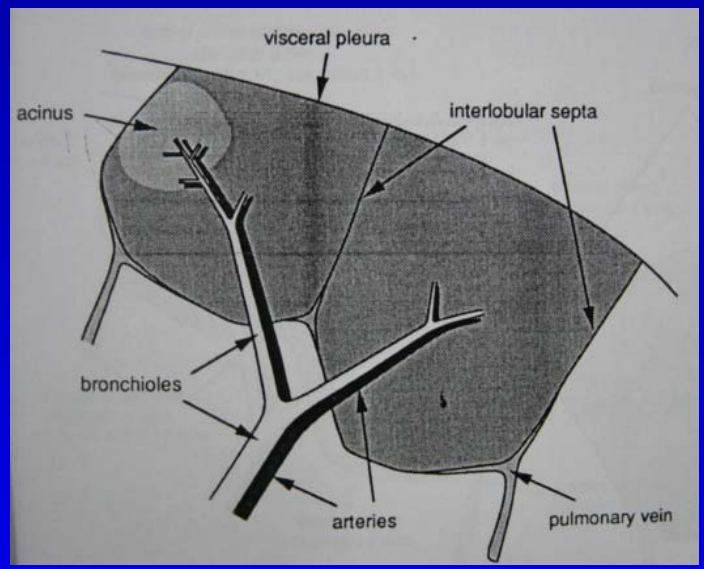
- Anatomic considerations
- Classification
- Diagnostic approach
- Specific disorders
- Quiz

Anatomic considerations

- Bronchioles are airways distal to those containing cartilage
- Terminal: last purely conducting airways
- Respiratory:
  in relation to alveoli
  lie within center of 2^0
  pulmonary lobule



- Normal bronchioles (0.6mm) not visible on CT

Classification of Bronchiolar disorders

- Aetiologic
- Pathologic
- Clinicopathological
- HRCT

Aetiologic classification

- Inhalational injury

  toxic gases, cigarette smoke, mineral dusts, organic dusts, fire smoke
- Postinfectious
- Drug induced

  ampho B, amiodarone, bleomycin, carbamazepine, cephalosporins, IFN-α, mtx, penicillamine, Paraquat
- Idiopathic

  No assoc. disease (BO, BOOP, DPB)

  Assoc. with other disease (organ Tx, CTD, others)

Pathologic classification

Colby, AJCP 1998

- Cellular bronchiolitis
  - Cellular bronchiolitis (Infectious, HP, FB, DPB)
  - Respiratory bronchiolitis (RB, RB-ILD,DIP)
- Constrictive bronchiolitis
  - Constrictive bronchiolitis/ BO
  - Constrictive bronchiolitis with polyps/ BOOP

HRCT classification

Muller, Radiology 1995

- Tree-in-bud pattern

  Asthma, ABPA

  Infections (bacterial, Mycoplasma,Chlamydia,TB, CMV, PCP)

  Diffuse panbronchiolitis

- Centrilobular nodules

  HP, RB-ILD, FB, sarcoidosis, LIP, CTD

- Decreased lung attenuation

  BO

- Ground-glass opacity and/or consolidation

  BOOP

Clinico-pathological classification

Ryu et al, AJRCCM, 2003

- Primary bronchiolar disorders

  BO, DPB, RB, FB, Mineral dust airway disease, others

- ILDs with prominent bronchiolar component

  BOOP, HP, RB-ILD, DIP, others (LCH, sarcoidosis)

- Large airway diseases with bronchiolar involvement

  Bronchiectasis, COPD, CF

Diagnostic approach to bronchiolar disorders

History & physical exam

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CXR, PFT

↓

HRCT

Is there bronchiolar disease?

What is the pattern?

↓

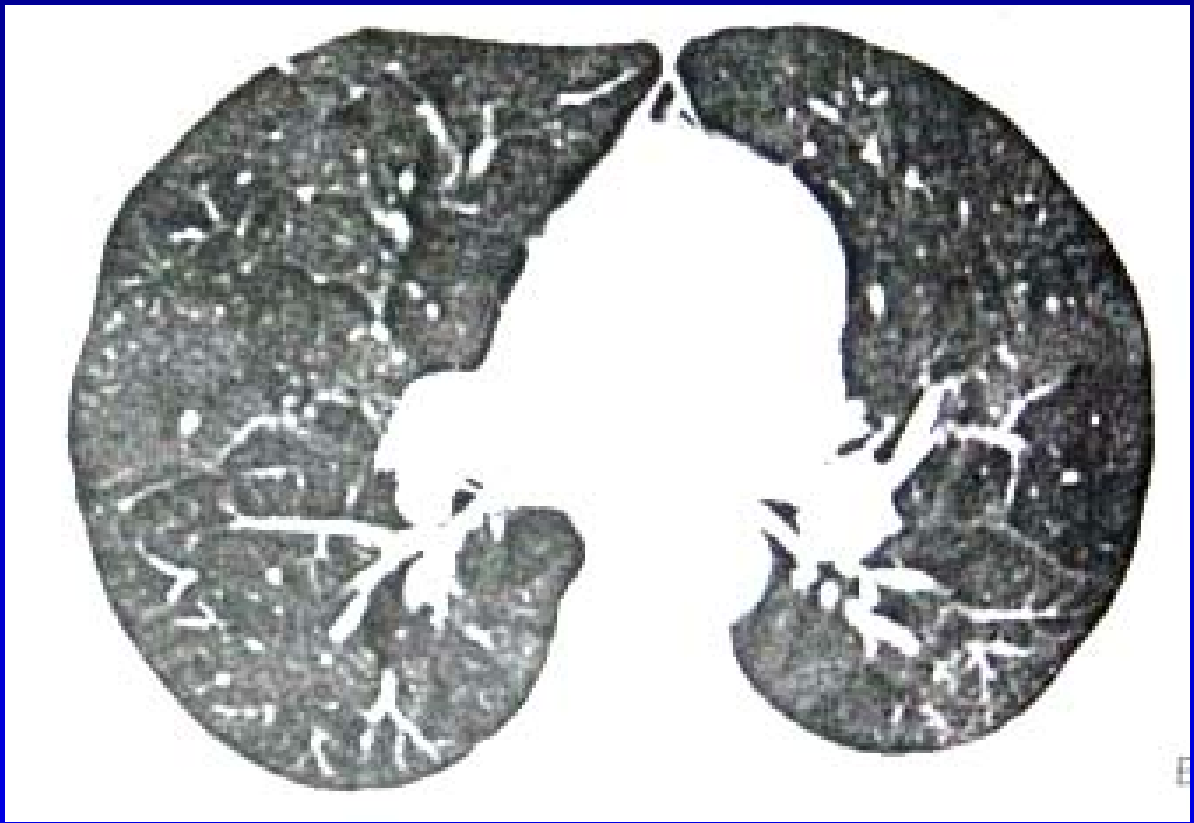
BAL,TBLB/OLB

HRCT

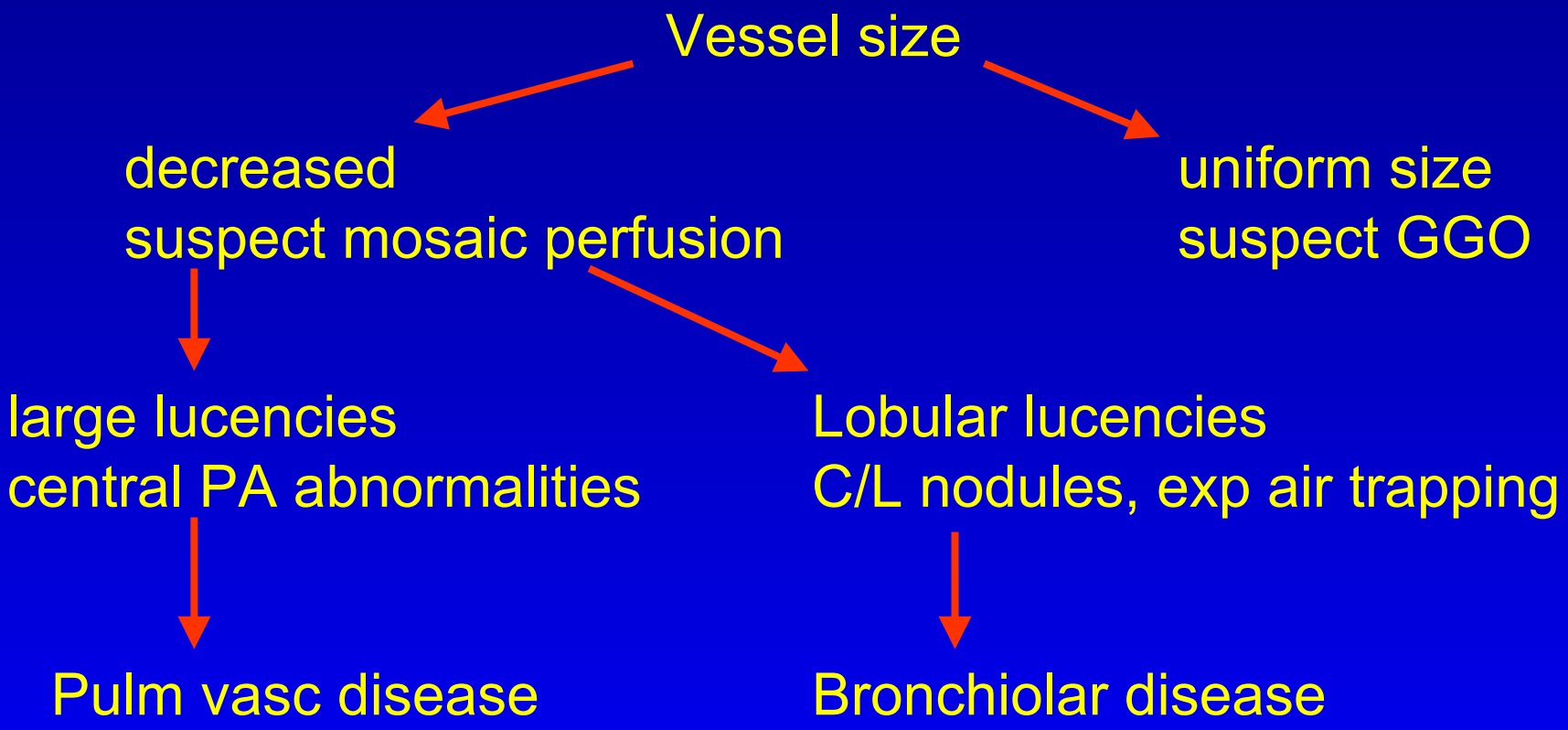
Is there bronchiolar disease?

- Direct evidence
  - CL nodules, TIB, bronchiolectasis
- Indirect evidence
  - subsegmental atelectasis
  - air-trapping
    - mosaic perfusion on inspiratory scans
    - expiratory CT (postexp, dynamic exp, spirometrically triggered exp CT)

Differential diagnosis?



d/d of patchy inhomogenous lung opacity



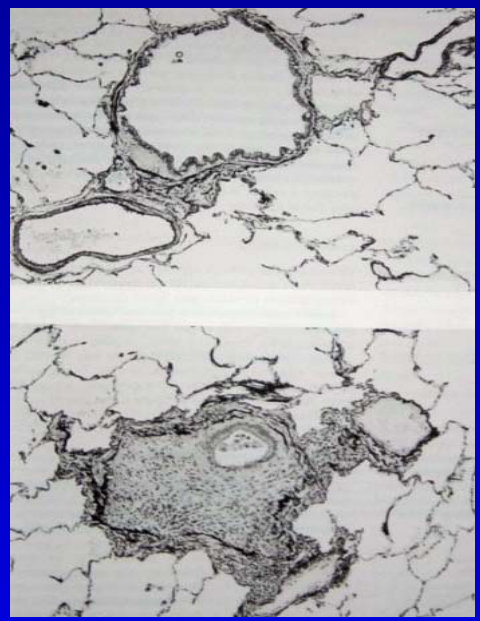
What is the pattern?

- Tree-in-bud pattern
- Centrilobular nodules
- Decreased lung attenuation
- Ground-glass opacity and/or consolidation

Primary bronchiolar disorders

Bronchiolitis obliterans/ Constrictive bronchiolitis

- Aetiology
  - Idiopathic
  - Postinfectious
  - CTDs
  - Inhalational injury, ingested toxins
  - Organ Tx
  - Drugs
  - Others (IBD, paraneoplastic pemphigus, NE cell hyperplasia)



Idiopathic Bronchiolitis obliterans/ Constrictive bronchiolitis

- Diagnosis: rare, middle aged women

  HRCT: category 3

- BAL: neutrophilia
- Treatment: poorly steroid responsive (early Rx imp, BAL neutrophilia↓s in responders)

  prog airflow limitation, resp failure

Post BMT Bronchiolitis obliterans

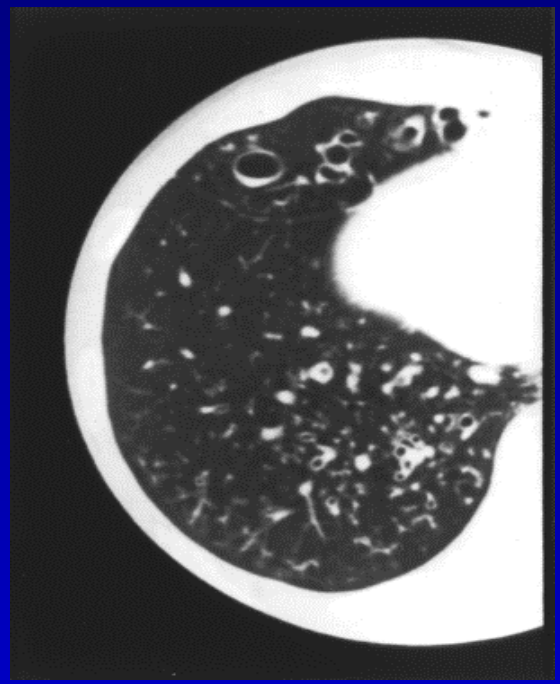
- 10% of patients, 1-10 months post Tx
- More common with allogenic (GVHD)
- Risk factors: older age, sinusitis, GVHD (esp with mtx prophylaxis)
- Hallmark: FEV1/FVC< 70%
- Rx: steroid, aza, cyclosporine, brochodilators

  50% mortality

Diffuse panbronchiolitis

- Asia
- Aetiology ? Genetic HLABw54 (63% vs 11% gen population)
  ? Environmental
- C/F: M:F 2:1, 50 years
  chronic sinusitis (75-100%)
  purulent sputum, BD reversibility
  raised ESR, CRP, cold agglutinins

HRCT: category 1

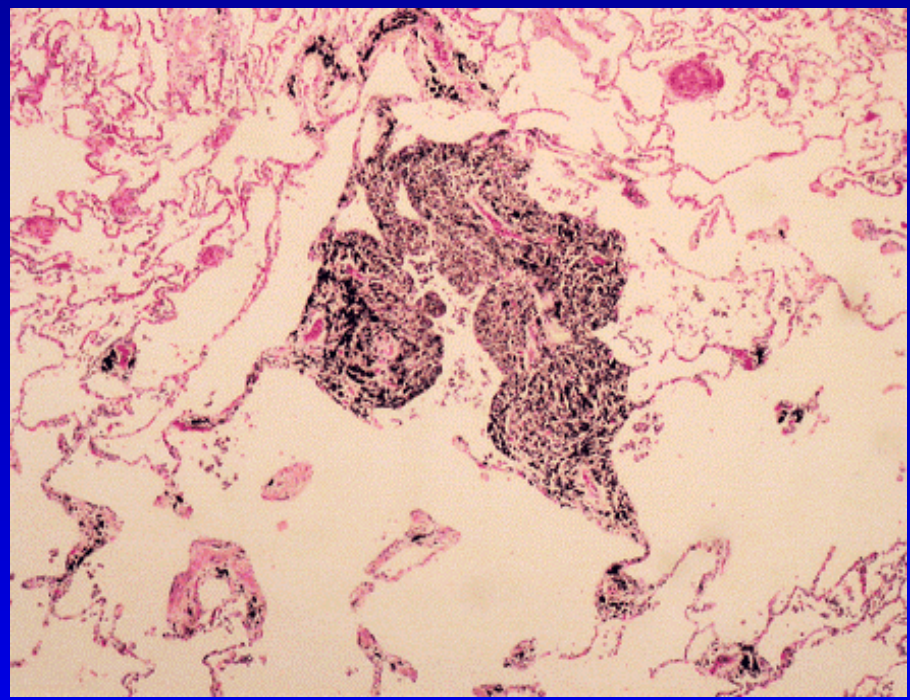


Rx

- Erythromycin 200-600 mg/day,
  Azithromycin 250mg 3/wk
  MOA: 2 fold
- NSAIDs: control bronchorrhea
- Bronchodilators
- Rx of sinusitis

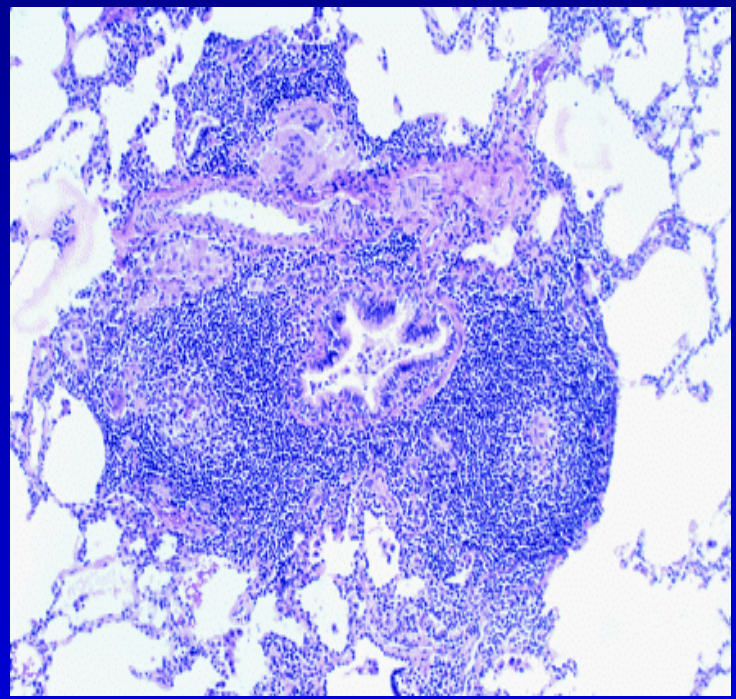
Mineral dust airway disease

- Asbestos, iron oxide, aluminium oxide, talc, mica, silica, coal
- C/F & Δ =
- Rx: remove from exposure steroids +/-



Follicular bronchiolitis

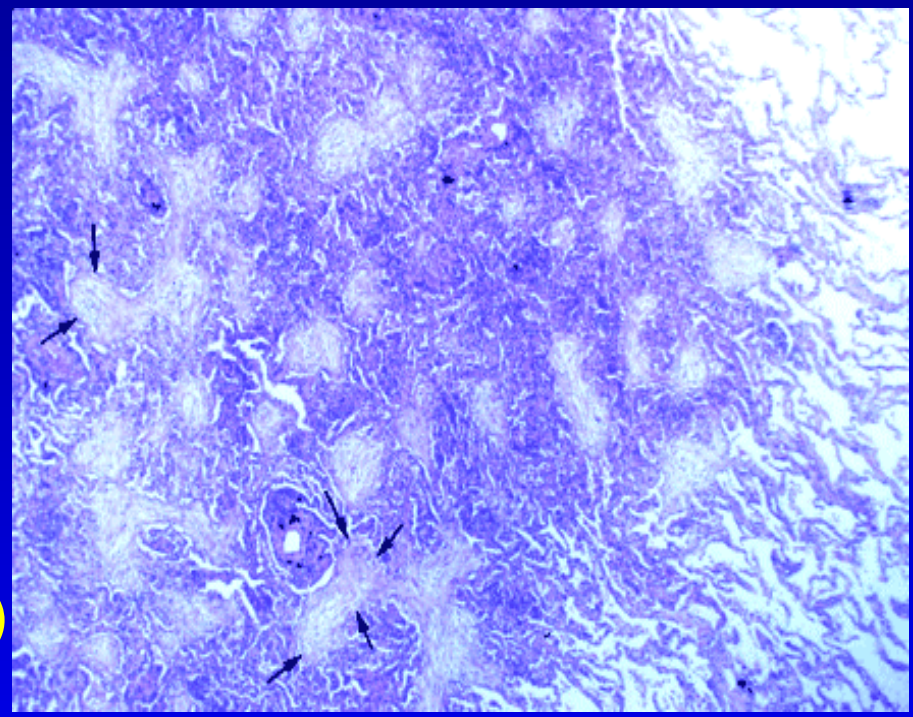
- Aetiology: Idiopathic
  CTD (RA)
  AIDS
  Infections
- C/F & Δ =
- HRCT: category 2
  (CL nodules)
  + peribronchial nodules, groundglass
- Rx: of underlying disease
  Idiopathic: bronchodilators, steroids, erythromycin



ILDs with prominent bronchiolar component

BOOP/COP

1. Patchy, peribronchial
2. Predom within alveoli
3. Uniform temporal app
4. Granul tissue ext
   through pores of Kohn
   (2 types of Masson bodies)
5. Honeycombimg unusual
6. Granulomas, giant cells, vasculitis -sent



BOOP/COP (contd.)

Although most commonly idiopathic, BOOP can be found in 3 categories of patients

i) BOOP is 1^0 cause of the resp illness

   Cryptogenic, postinfectious, CTDs, inhalation injury, organ Tx, drugs, IBD, radiation

ii) Minor component of another disease

   sarcoidosis, HP, LCH

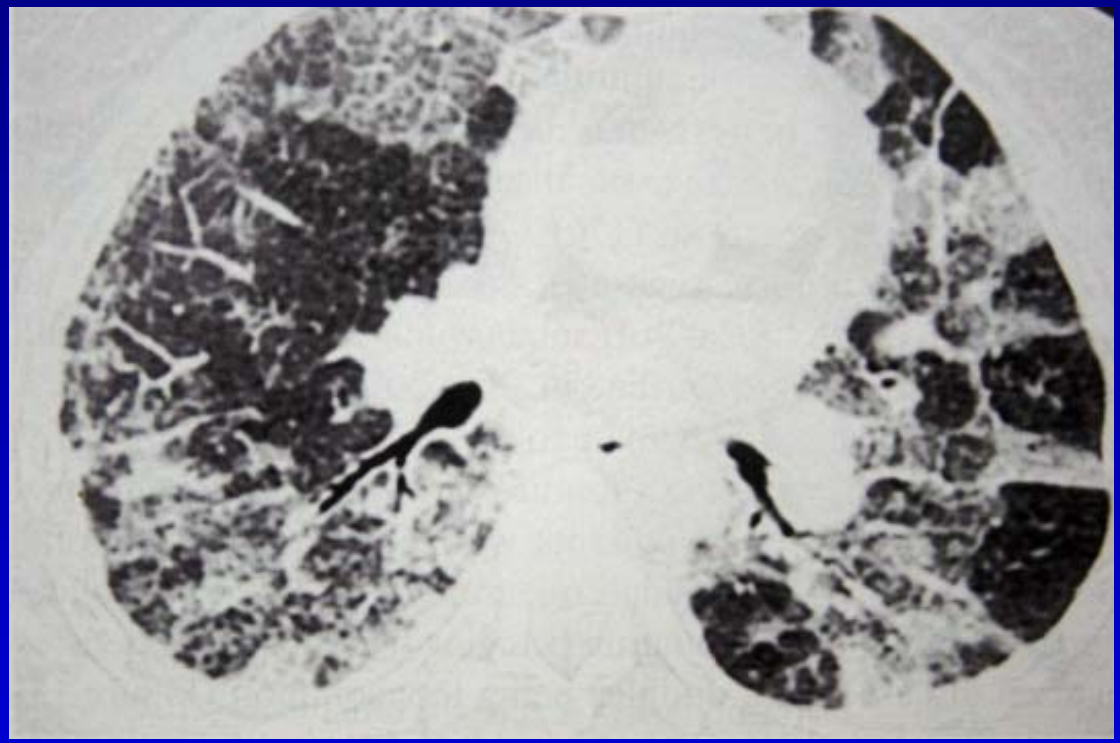
iii) Found as a nonspecific reaction at periphery of unrelated pathologic process

   bg ca, wegener's, pulm infarcts etc.

Idiopathic BOOP

- C/F: 50-70 yrs, subacute dyspnoea, cough
  fever, malaise LOA, LOW
  rarely: ARDS
  O/E: Velcro crackles, clubbing rare
  raised TLC, ESR (100s!), CRP
- CXR: B/L patchy, fleeting alveolar infiltrates
  If reticular pattern +: CTD, poorer prognosis
  Less common patterns: focal consoldation, masses
  Pleural effusions in 25%, no hyperinflation

- HRCT:



- BAL: increased lymphocytes, Th1-related cytokines, r/o other causes of OP
- PFT: restriction, decreased DL_{CO}

Role of TBLB

Sufficient for diagnosis if 3 conditions are +

i) clinical and CT findings appropriate

ii) biopsy specimen large enough to contain all elements of lesion

iii) close follow up possible

If not: OLB/VATS

Rx: steroid responsive

2/3 complete resolution, improvement in < 48 hrs

prednisolone 0.75-1.5mg/kg/day (max 100mg) x 4-8 wks, taper over 6-12 months (relapse in 1/3 if dur < 3mo)

2nd line: Cyclophos, Cy A, Aza (trial of > 3-6 mo)

Poor prognostic factors:

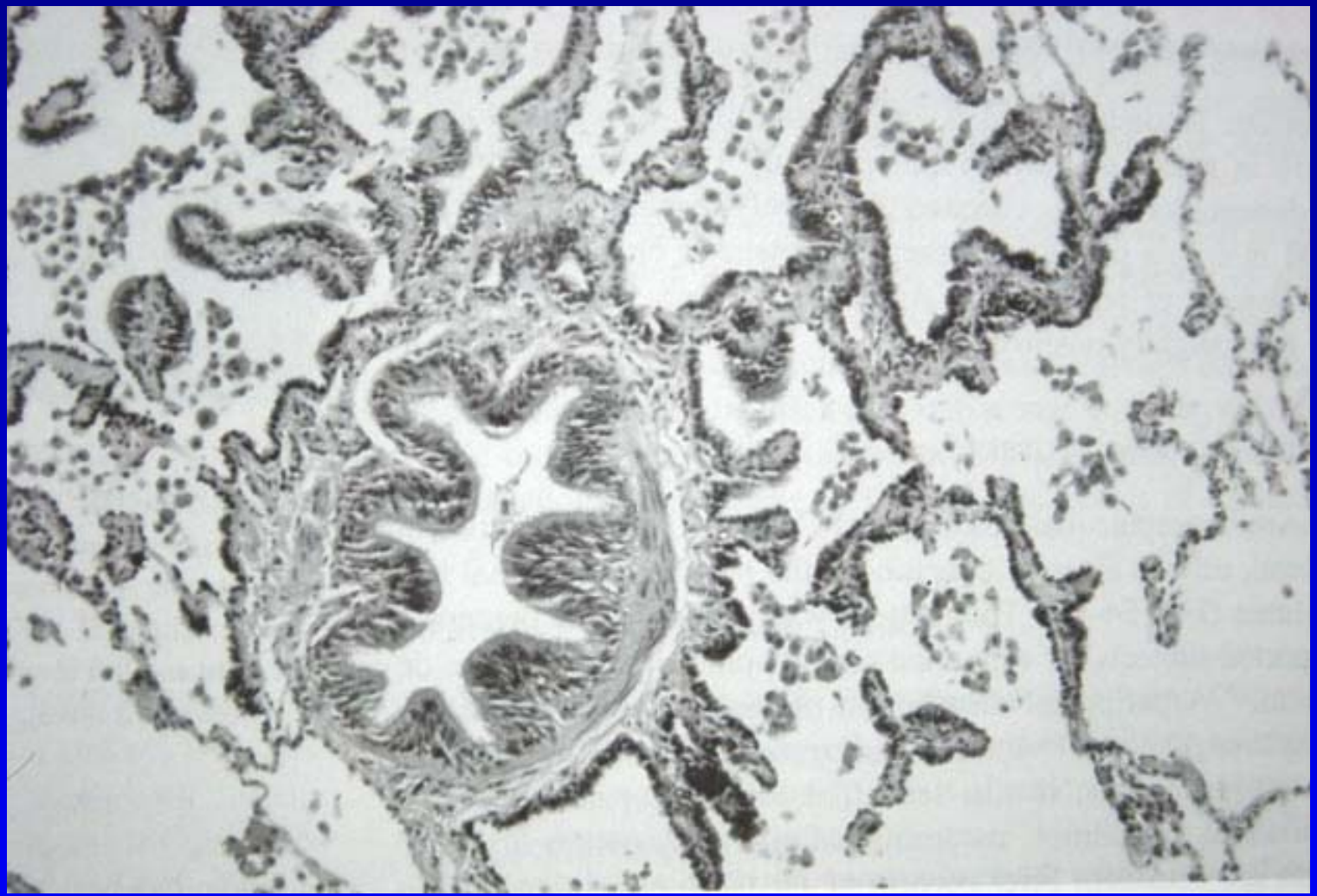
i) non-idiopathic BOOP

ii) predominantly interstitial pattern on CT

iii) Lack of BAL lymphocytosis

iv) Type 2 Masson bodies on biopsy

RB-ILD & DIP



RB-ILD vs DIP

30-50 yrs, > 30 pack years cig smoking

- mild dyspnoea
- clubbing occasional
- PFT: N/ mixed
- CXR: lung volumes N
- CT: predominant CLN
- Rx: smoking cessation

- more marked
- clubbing 50%
- PFT: restrictive
- CXR: lung volumes ↓
- CT: predominant GGO
- Rx: smoking cessation
- + steroids

Summary of diagnostic approach to bronchiolar disorders

History & physical exam

↓

CXR, PFT

↓

HRCT

Is there bronchiolar disease?

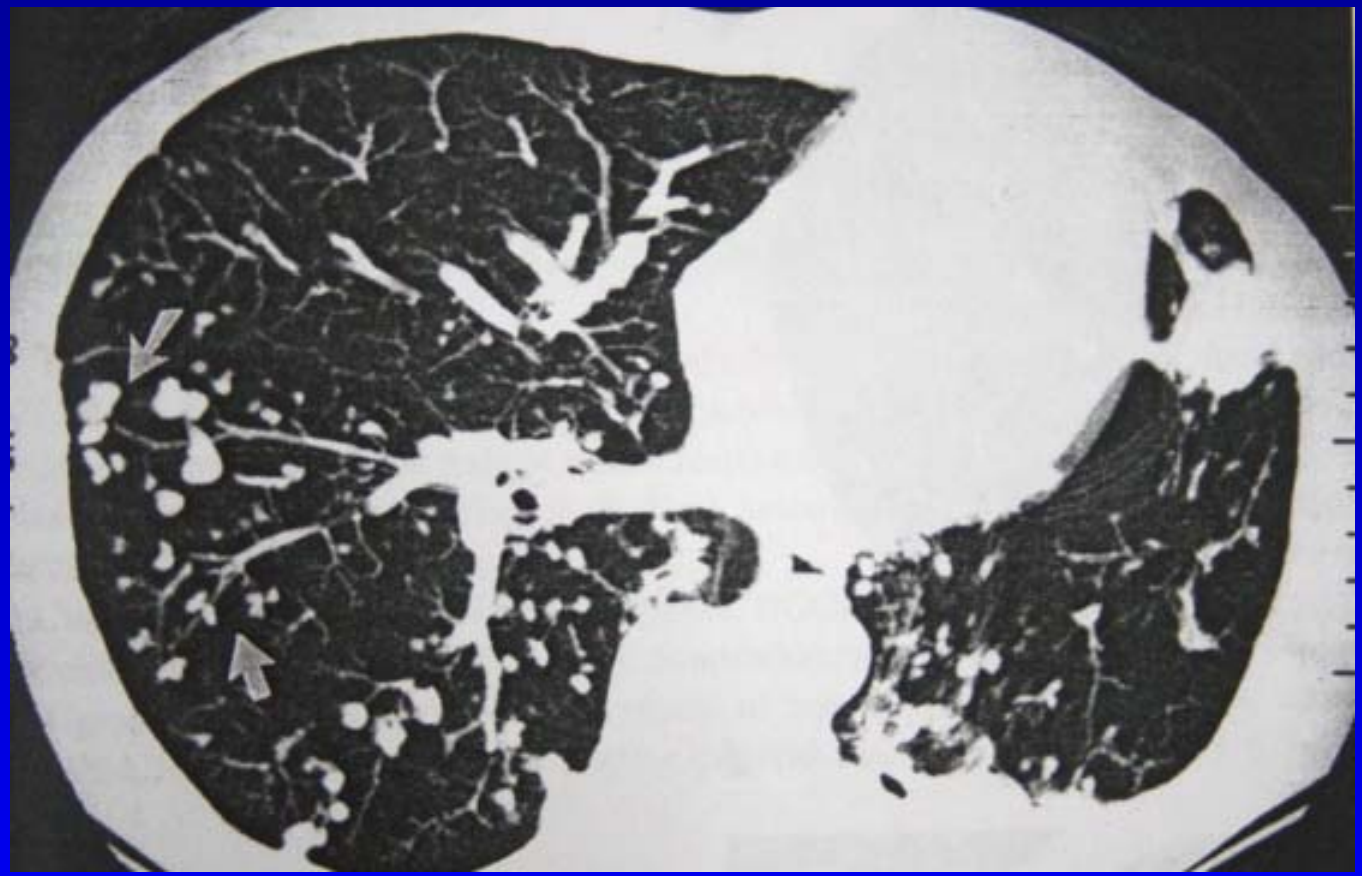
What is the pattern?

↓

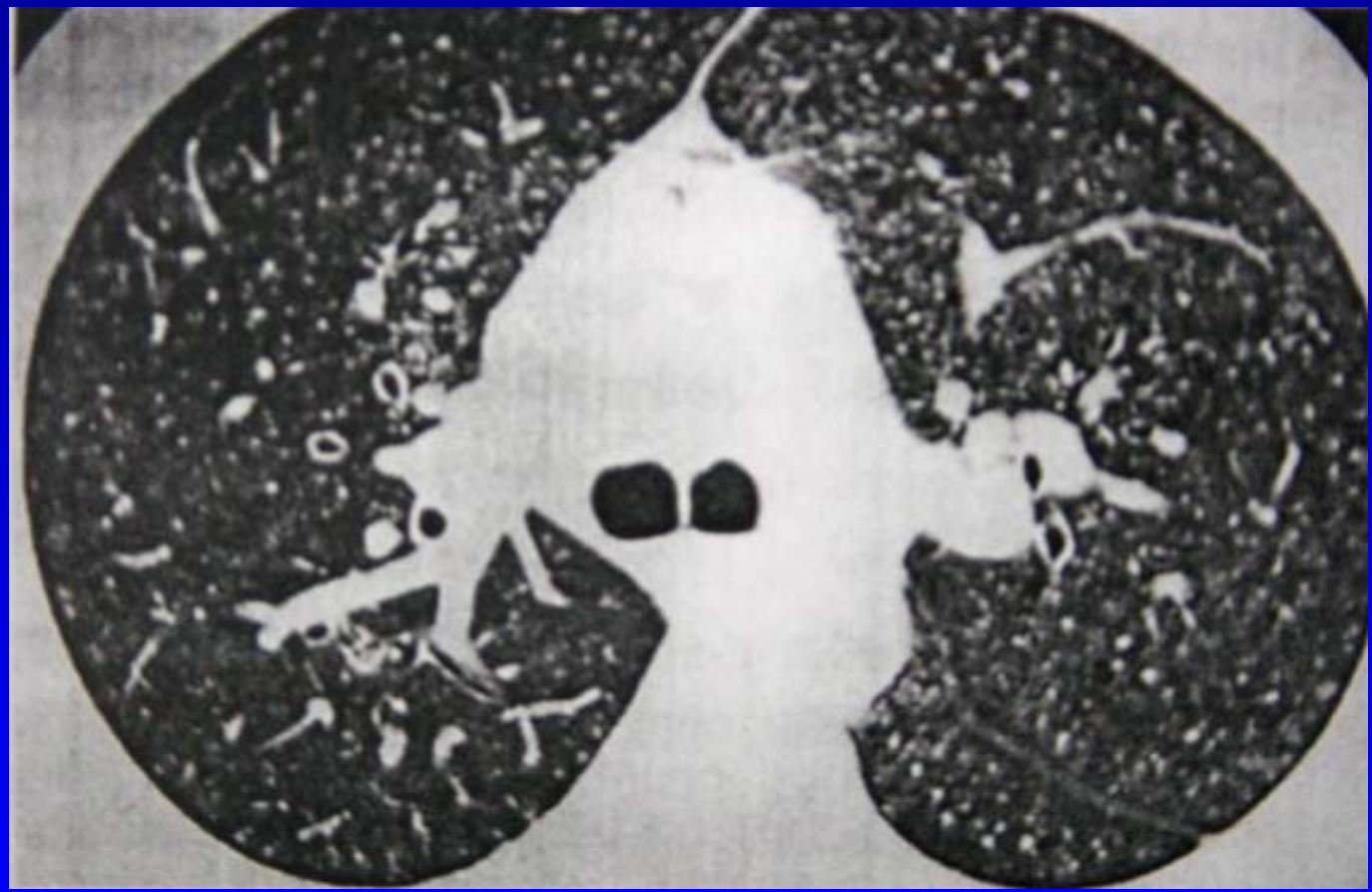
BAL,TBLB/OLB

Quiz

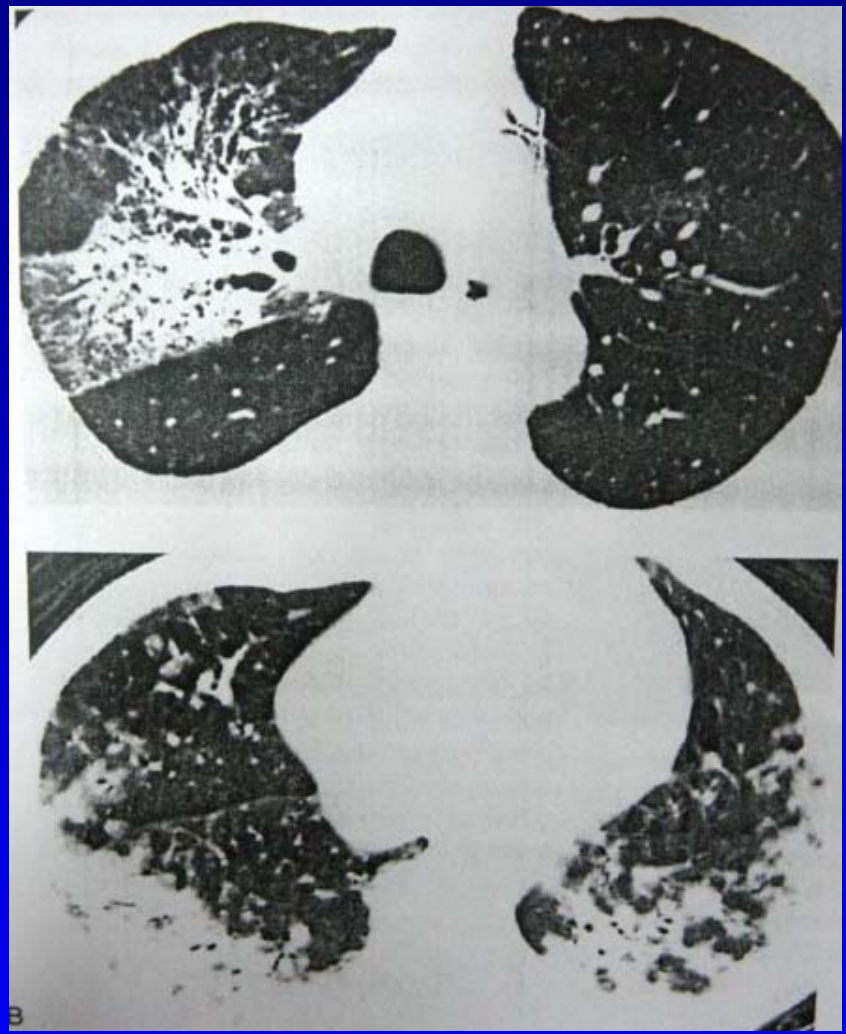
A 37 year old alcoholic with fever and haemoptysis



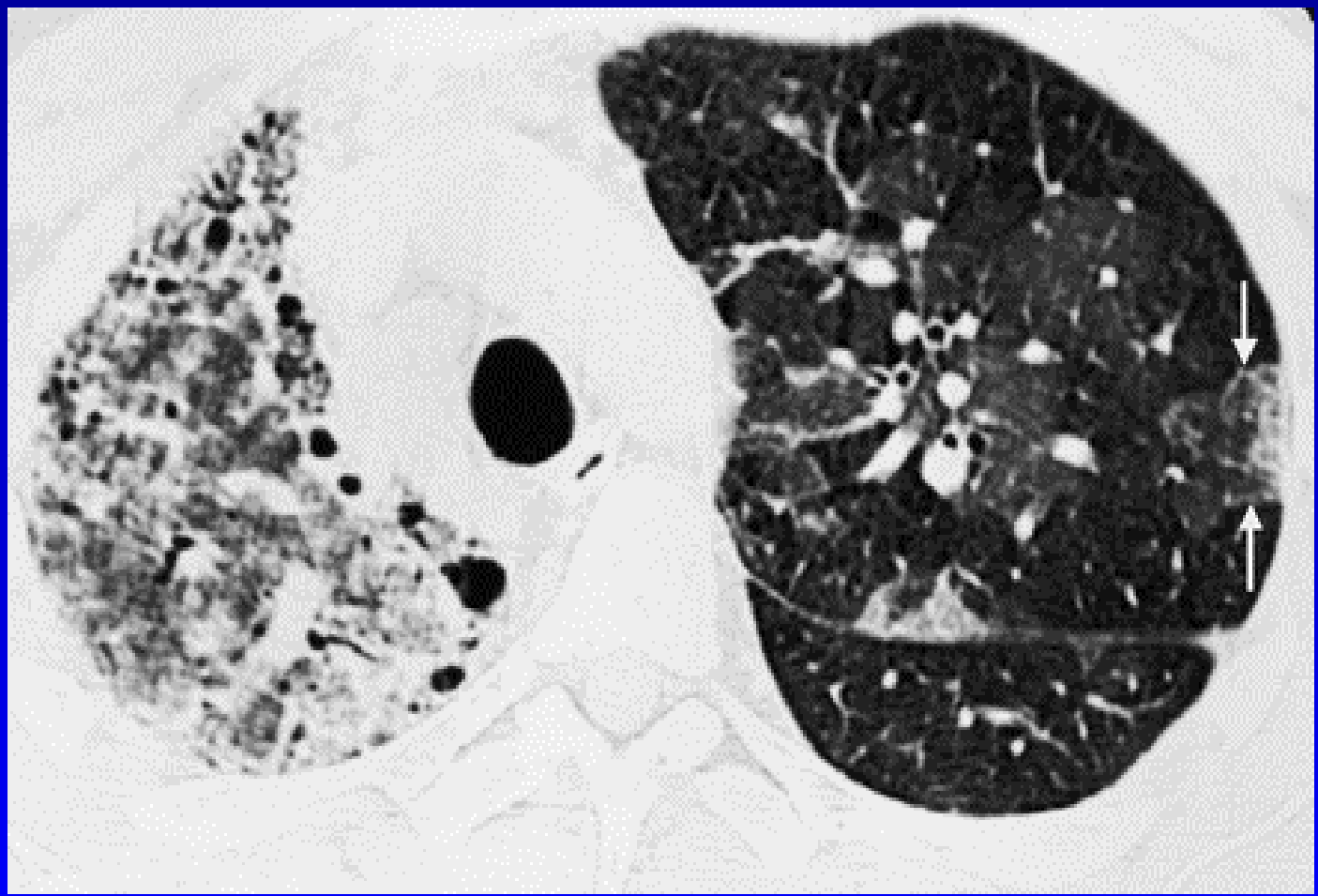
A 40 year old lady with h/o blood transfusion 7 years back presenting with oral thrush and dyspnoea



A 35 year old female with RA



A 58 year old smoker with end-stage lung disease, develops progressive dyspnoea after a major surgery



Thank you