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

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 inhomogeneous lung opacity

Vessel size
- decreased
  - suspect mosaic perfusion
- uniform size
  - suspect GGO

large lucencies
- central PA abnormalities
  - Pulm vasc disease
Lobular lucencies
- C/L nodules, exp air trapping
  - Bronchiolar disease
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. Honeycombing unusual
6. Granulomas, giant cells, vasculitis -sent
Although most commonly idiopathic, BOOP can be found in 3 categories of patients

i) BOOP is the 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 consolidation, masses
  - Pleural effusions in 25%, no hyperinflation
• HRCT:

• BAL: increased lymphocytes, Th1-related cytokines, r/o other causes of OP
• PFT: restriction, decreased DL\textsubscript{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 d
- 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