### Approach to Interstitial Lung Disease

#### History
- Allergic rhinitis and asthma – eosinophilic pneumonia
- Nasal polyps – EGPA (churg-strauss)
- Medications – amiodarone, anti-TNFα, nitrofurantoin
- Smoking – RBILD, DIP, EG
- Exposures – asbestos, silica, birds, etc

#### Physical Exam
- Fibrosis sounds like coarse, velcro-like crackles
- High pitched inspiratory squawk – bronchiolitis (HSP)
- Clubbing is nonspecific, usually in advanced fibrosis
- Extrapulmonary – rashes, Raynauds, arthritis, muscle weakness/pain, nailfold capillary

#### ILD that may have obstructive PFTs
- Sarcoidosis
- Hypersensitivity pneumonitis
- Combined pulmonary fibrosis emphysema
- LAM/tuberous sclerosis
- Eosinophilic granuloma

#### Clinical Pearl: Look at old imaging and PFTs to determine the time course and progression of disease!

#### Radiology

**Classic UIP pattern that indicates IPF and requires no further workup:**
1. Bilateral, subpleural reticular opacities in a basilar predominance
2. Traction bronchiectasis
3. Honeycombing

**Other typical radiographic patterns of ILD**
- Balanced hilar adenopathy - sarcoidosis
- Pleural plaques and septal lines – asbestosis
- Centrilobular nodules – hypersensitivity pneumonitis, eosinophilic granuloma, sarcoïdosis
- “Atoll sign” – airspace disease at the end of an airway with central clearing – COP
- Cysts – EG, LAM, Sjogren

**Clinical Pearl: If you have a classic UIP pattern and no inkling of autoimmune disease, you can call it IPF. We still recommend some screening serologies to help with prognosis and rule out myositis.**

### Serologies

**Screening serologies that I get on almost everyone**
- ANA – get excited with titer >1:80; pattern matters, i.e., nucleolar important, homogenous not so much
- RF- pretty nonspecific but can clue you in that something is afoot
- CK, aldolase, myositis panel, Jo-1: 70% of anti-synthetase syndrome presents with ILD and negative ANA
- ESR and CRP- these are not helpful and I do not get them

**Disease specific serologies that I only get if pretest probability is high**
- CCP- specific marker for RA, get hand XR as well if suspicion is high
- ssA/ssB, immunoglobulins: Sjogren, IgG f seronegative Sjogrens, IgG subclasses for IgG4 disease
- Scl 70, centromere, PM-1: scleroderma and the scleroderma/myositis overlap syndrome
- ANCA- order with suspicion of vasculitis: GPA, EGPA, MPA; interpretation can be difficult
- dsDNA, smith, RNP- SLE and its overlap syndromes, RNP is helpful if dsDNA and smith are negative

**Clinical Pearl: If the CT is not classic UIP fibrosis and/or you suspect autoimmune disease based on history, exam, serologies- try to biopsy something to get an actual diagnosis.**

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