Care of the Hospitalized ILD patient

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Disclosures

No disclosures
Outline

Clinical Features
- Acute
- Acute on Chronic
- AEILD

Prognosis/Treatment
- Medical therapy
- Mechanical Ventilation
- Transplant

ILDs Presenting in Hospitalized Patient

Acute ILDs
- Acute interstitial pneumonia
- CTD-ILD
- Synthetase syndrome
- Eosinophilic pneumonia
- Organizing pneumonia
- Drug induced

Chronic ILD
- Chronic ILD presenting for first time.
- Chronic progression of underlying ILD.
- Acute exacerbation of chronic ILD
  - Acute exacerbation of IPF
  - Acute exacerbation of a CTD-ILD
  - Acute exacerbation of hypersensitivity pneumonitis.
Acute ILDs

Acute Interstitial Pneumonia (AIP)

- Diffuse uniform thickening of alveolar septa
- Cellular interstitial infiltrate
- Lymphocytes and plasma cells
- Often will see hyalin membranes

Katzenstein and Myers, Am J Resp Crit Care Med, '98
Acute Interstitial Pneumonia (AIP): Treatment

- Approach same as ARDS
- Supportive care
- Antibiotics
- Low tidal volume ventilation
- Mortality: > 60%

Ichikado, AJRCCM, 2002

AIP: Mimics

AIP is diagnosed after ruling out secondary causes

- Viral Infection
- Drugs
  - Amiodarone
  - Macrodantin
  - Chemotherapy
- Autoimmune disease
  - Dermatomyositis
  - Anti-synthetase syndromes
**Anti-synthetase syndromes**

**What are they?**

Diseases associated with autoantibodies against t-RNAs.

- Anti-Jo1 (histidine) (36%)
- Anti-PL-7 (Threonine) (18%)
- Anti-PL-12 (Alanine) (12%)
- Anti-OJ (Isoleucine) (5%)
- Anti-KS (Asparagine) (8%)
- Anti-EJ (Glycine)
- Anti-Zo
- Anti-Ha
- Anti-SRP

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**Anti-synthetase syndromes: Clinical Clues**

Hamaguchi et al, PLOS1 2013
**Anti-synthetase syndromes: Treatment**

- Cyclosporin
- Tacrolimus
- Rituximab

Sem et al, Rheum 2009

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**Dermatomyositis: PCP**

Patients with DM are at risk for PCP (8%)
Lymphopenia is a risk factor
60% occurred during first month of therapy!
PCP onset before therapy

Viguier, Medicine 2003
Chronic ILDs

Clinical Classification

Pulmonary Fibrosis

Exposure-related:
- Occupational
- Environmental
- Avocational
- Medication

Idiopathic interstitial pneumonia (IIP)

Idiopathic pulmonary fibrosis (IPF)

Connective tissue disease:
- Scleroderma
- Rheum. arthritis
- Sjogrens
- UCTD

Desquamative interstitial pneumonia (DIP)

Acute interstitial pneumonia (AIP)

Nonspecific interstitial pneumonia (NSIP)

Respiratory bronchiolitis interstitial lung dis. (RBILD)

Cryptogenic organizing pneumonia (COP)

Lymphocytic interstitial pneumonia (LIP)

Other:
- Sarcoidosis
- Vasculitis/Diffuse alveolar hemorrhage (DAH)
- Langherhans cell histiocytosis
- Lymphangioleiomyomatosis (LAM)
- Pulmonary alveolar proteinosis (PAP)
- Eosinophilic pneumonias
- Neurofibromatosis
- Inherited disorders
- Chronic aspiration
- Inflammatory bowel disease
Clinical Course: 
Individual IPF Patient

Acute Exacerbation of 
Underlying ILD
**Case**

A 75 yo man underwent elective right hip replacement surgery. 48 hours after surgery he developed shortness of breath and an increasing oxygen requirement to 6 liters. At the time he denied cough, sputum production, or recent symptoms c/w a viral infection. An echocardiogram was normal, and blood and sputum cultures negative for bacterial or viral pathogens, CT angiogram w/o PE.

**HRCT**
The HRCT was interpreted as c/w IPF, with acute exacerbation. He was treated with prednisone (60 mg/d). He was discharged after 3 weeks in the hospital on 2 liters of oxygen and was limited to walking 2 blocks.

CT 3 months later

Diagnosis of Acute Exacerbation of an ILD

Diagnosis of ILD
Unexplained worsening dyspnea for < 4 weeks
New opacities on HRCT
Absence of secondary cause: lung infection, CHF, PE.

Exacerbation: HRCT

Background of:
- Bilateral subpleural reticulation
- Traction bronchiectasis
- Honeycombing
- Increased parenchymal opacities

Exacerbations Are Not Limited to IPF

Exacerbations have been reported in other ILD’s:
- CTD-ILD
  - SSC, RA, SLE
  - Chronic Hypersensitivity pneumonitis

Clinical Features
- HRCT: New opacities on background of fibrosis
- Pathology: Fibrosis w/ DAD
Exacerbation: Pathology

UIP Pathologic changes
- FF
- Honeycomb
- Dense fibrosis

In Addition:
DAD
- Interstitial edema, Type II cell hyperplasia
- Hyalin membranes
- Intra-alveolar Organizing Pneumonia/fibrosis

Who Gets an Exacerbation?

Incidence: 2-20%/patient/yr

**Triggers of an Exacerbation?**

- Unknown
- Mechanical ventilation (*diagnostic VATS biopsy*)
- Pollution ([Johannson et al., ERJ 2013](#))
- Radiation therapy for lung cancer
- GERD, microaspiration ([Lee et al., ERJ 2012](#))

**Management of AEIPF**

- Supportive care
  - High flow oxygen, NIV, (MV)
  - Antibiotics
- No medical therapy is proven effective
- Steroids?......Maybe if the CT has regions consistent with organizing pneumonia
Management of Hypoxic Respiratory Failure in ILD Patient

23 Patients w/ IPF and acute respiratory failure (14 w/ AE)
Median survival: 3 d
Overall Survival: 4%
MV can be used in select pts with AE as bridge to transplant

Stern et al, Chest 2001
Ventilatory support: One size does not fit all

120 ILD patients admitted to ICU
80% with IPF
No pattern explaining successes

Gungor et al, Resp Care 2013

Lung Transplant in IPF

- Should be considered in patients less than age 70.
- 50-60% 5 year survival after transplant
- Patients that do well: younger, few co-morbidities, minimal steroids, BMI, pulmonary rehab


Getting Hospitalized Patients to Transplant: ECMO

ECMO can be used as a bridge to lung transplant.

31 lung transplants at UCSF off ECMO

42% were for pulmonary fibrosis

comparable survival
high acuity patients transplanted without ECMO

Hoopes et al, J Thorac Cardiovasc Surg 2013

Palliative Care

57% of IPF patients died in the hospital.
13.7% received PC referral
Timing:
1 mo prior 71%
6 mo prior 18%
12 mo prior 11%

Lindell et al, Chest 2014 (in press)
Conclusions

The majority of ILD patients will be hospitalized at some point during the course of their illness.

Present acutely:
- Think autoimmune
- Consider infection
- Present as an exacerbation of underlying ILD
  - Manage with supportive care

Requirement for mechanical ventilation is poor prognostic marker.
- Use MV as bridge to transplant.

Consider accelerated lung transplant workup for patients hospitalized with ILD.

Consider early palliative care/hospice consult when disease advances.