Pulmonary Case Presentation

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Patient History

- 38 year-old Hispanic male
- 2 to 3 month history of progressive dyspnea
- 2 hospitalizations for “pneumonia”
- Treated with antibiotics, oxygen, steroids
- Readmitted with hypoxia, abnormal CXR
- Transferred to Wichita

Past Medical History

- Type 2 diabetes
- Dyslipidemia
- Nephrolithiasis

Social History

- Previous smoker – stopped 3 months ago
- Occasional cocaine use
- No alcohol use
- Works in the oil fields
- No pets or bird exposure
- No influenza or pneumonia vaccination

Review of Systems

- No fever or chills
- CV – no chest pain
- RESP – cough and dyspnea
- GI – no nausea, vomiting, diarrhea
- GU – no dysuria or hematuria
- MS – no joint pain or swelling

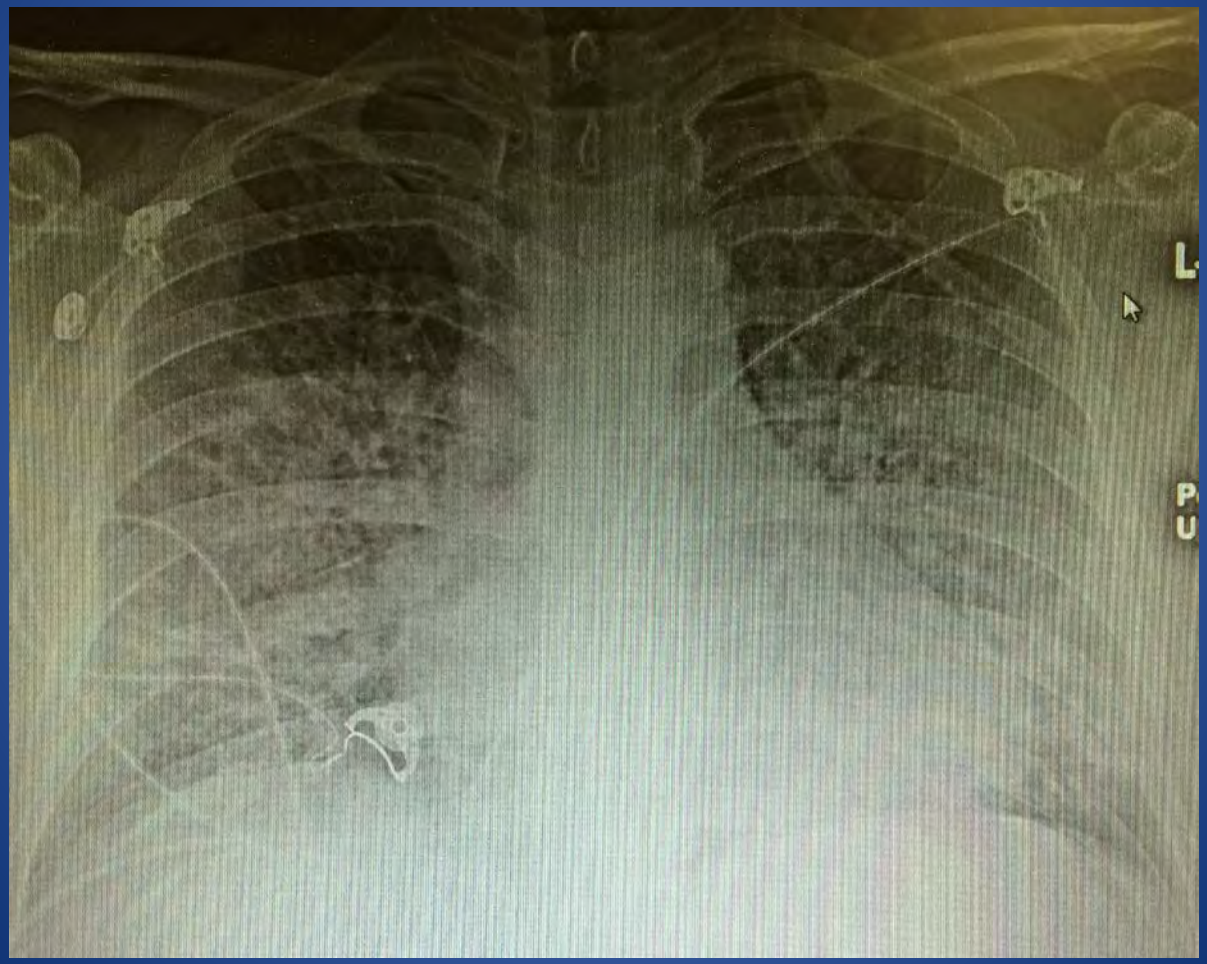
Physical Exam

- VS – BP 120/78, HR 62, RR 26, afebrile
- Awake and alert
- HEENT – on BiPAP
- Chest – decreased BS but clear
- CV – RRR, no murmur
- ABD – soft, non-tender, no masses
- EXT – no edema, cyanosis, or clubbing

Laboratory studies

- WBC 8.3, HB 14.9, platelets 216 K
- ABG – pH 7.46, pCO2 31, pO2 53
- CMP – normal
- HIV – negative
- QuantiFERON Gold - indeterminate

CXR



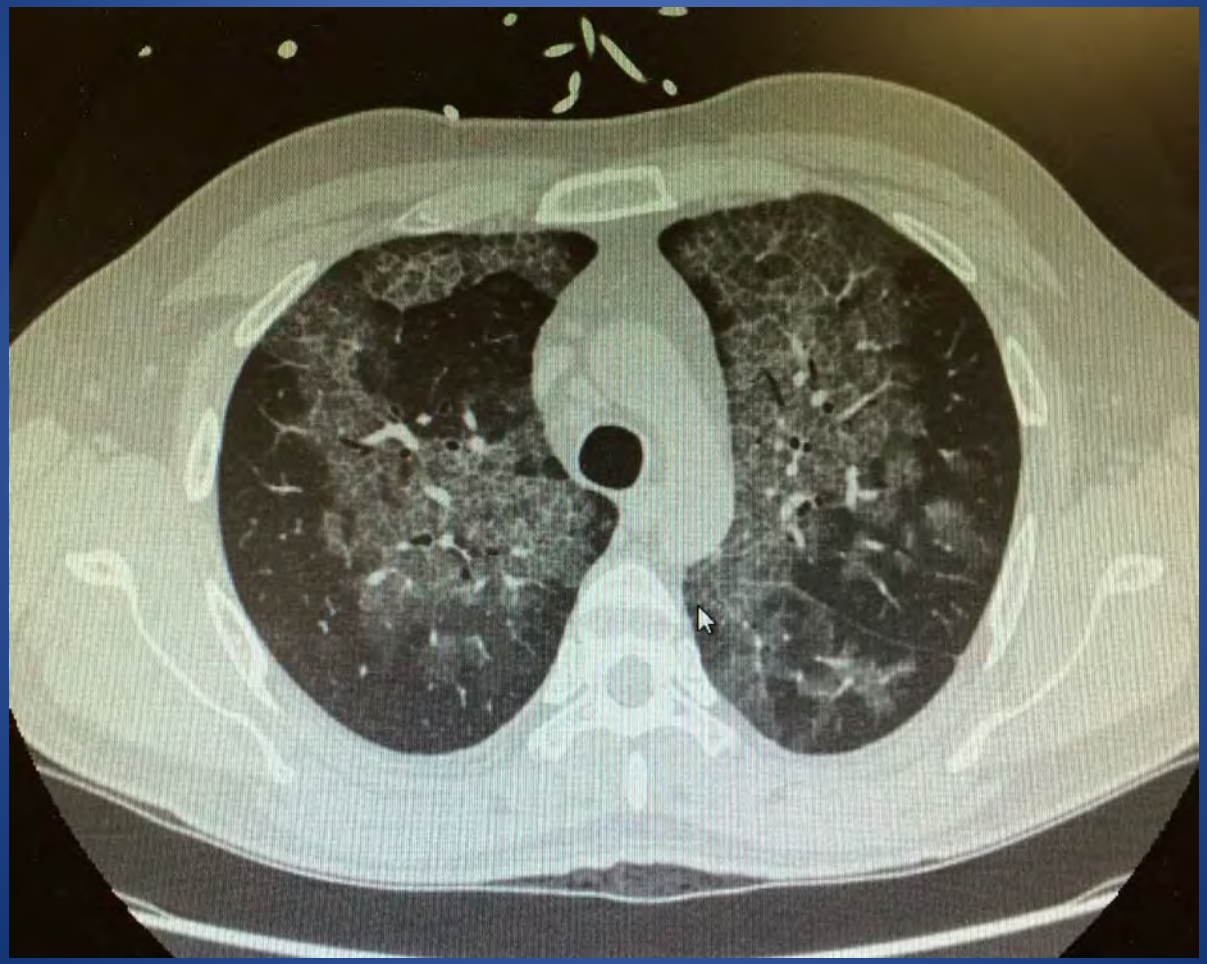
Hypoxic respiratory failure with pulmonary infiltrates

- Infection ?
- Inflammation ?
- Neoplastic ?
- A combination of the above ?

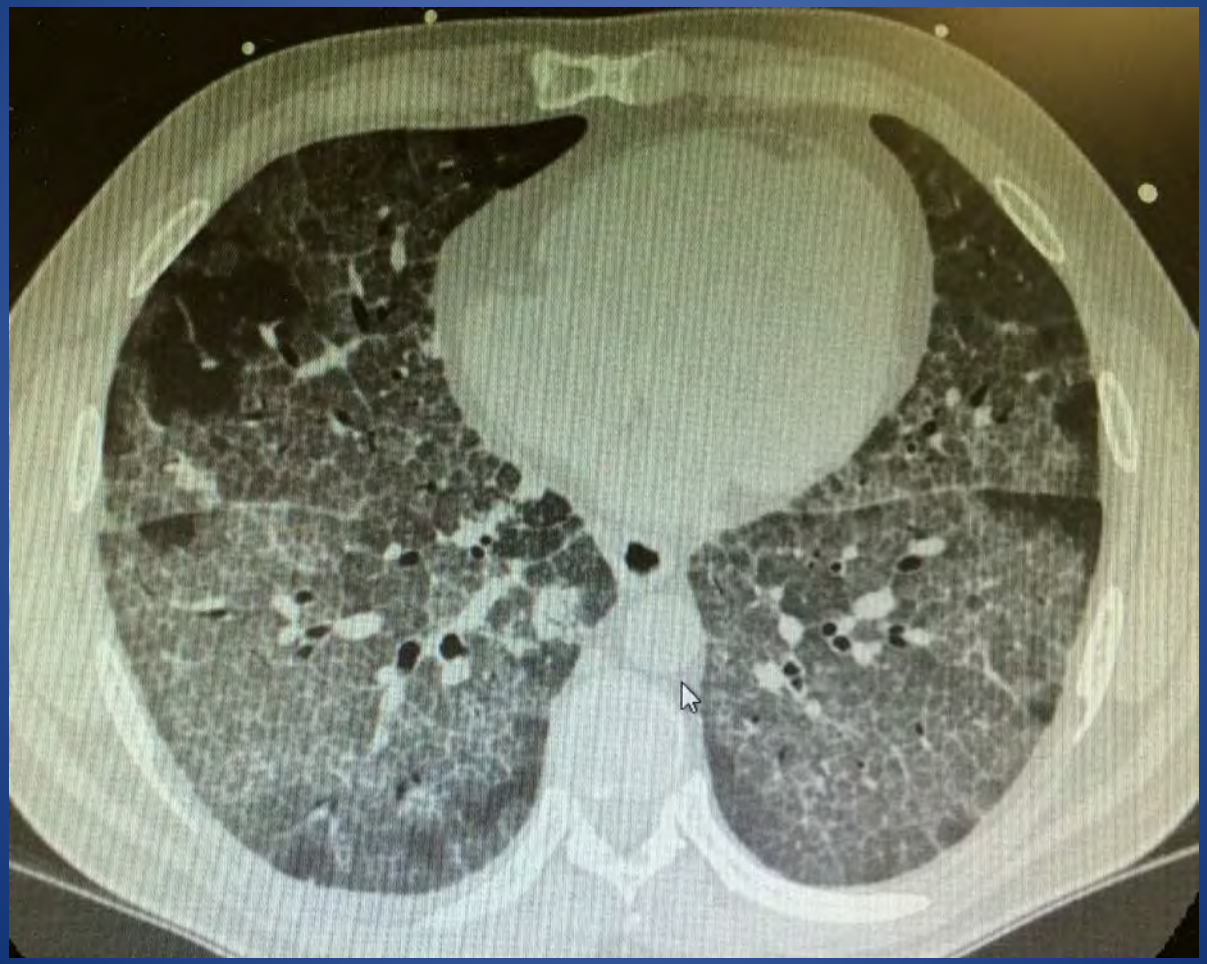
Clinical Course

- Progressive dyspnea and hypoxia
- No improvement despite antibiotics and steroids
- Viral and bacterial cultures negative

High Resolution CT Chest



High Resolution CT Chest



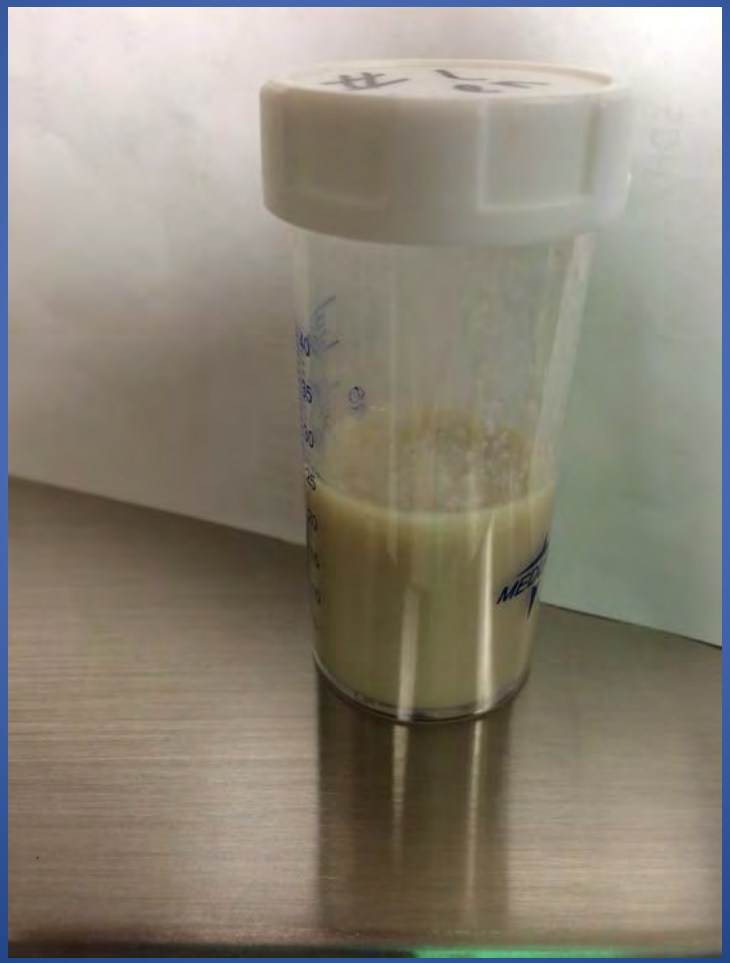
CT finding

- Extensive groundglass opacities
- Interlobular septal thickening
- “crazy paving pattern”
- No fibrosis

Bronchoscopy

- No airway lesions
- Increased secretions bilaterally
- BAL revealed thick tan secretions
- PAS stain positive consistent with the diagnosis of PAP (Pulmonary Alveolar Proteinosis)

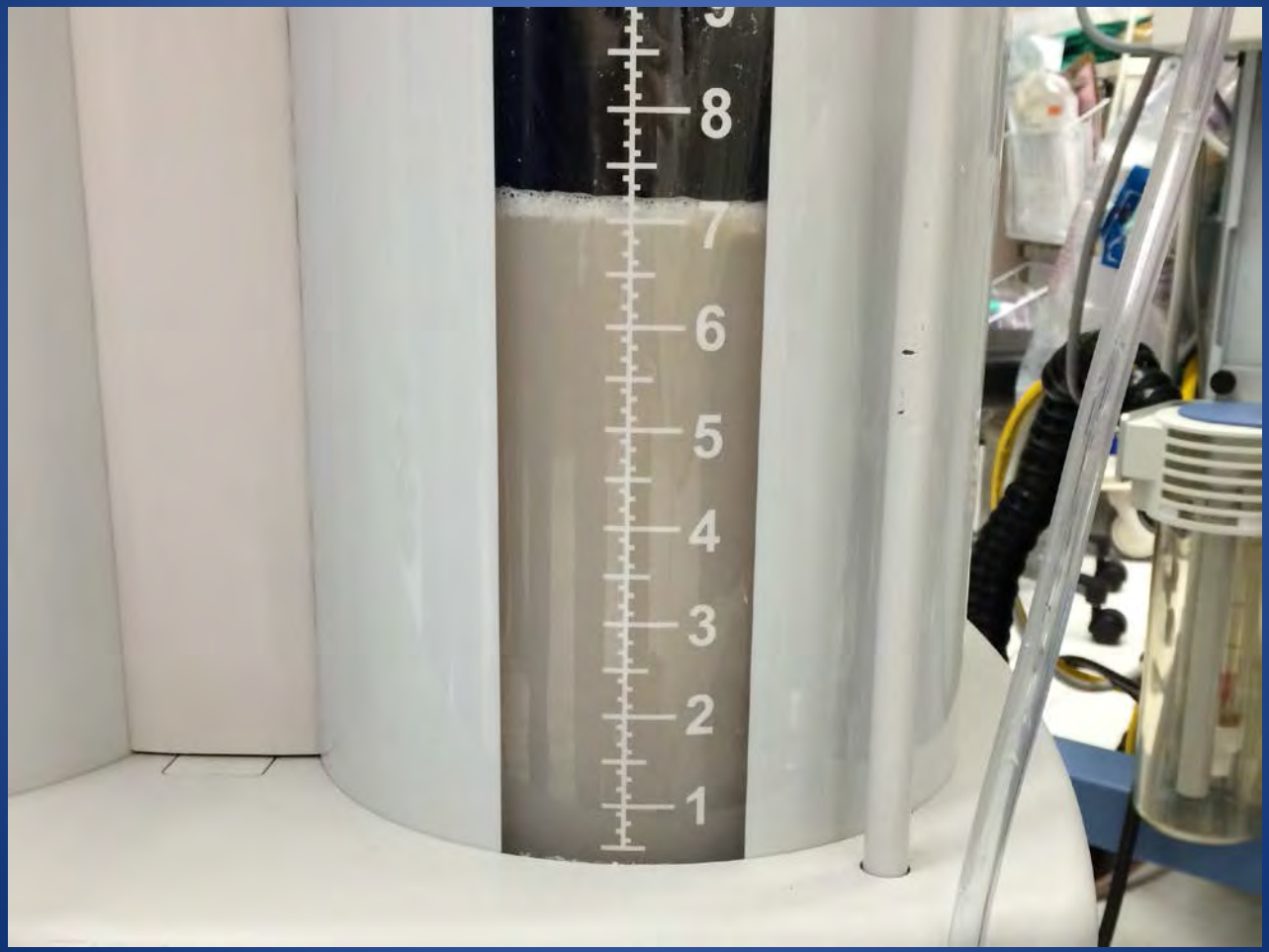
BAL return



Treatment

- Whole lung lavage under general anesthesia
- Day 1 – right lung
- Day 3 – left lung

Whole Lung Lavage - Right



Pulmonary Alveolar Proteinosis

- Discovered in 1958
- Rare 0.37 per 100,000
- Acquired disorder
- Median age diagnosis 39
- Most are men
- 72% have a smoking history

Clinical Findings

- Progressive dyspnea and cough
- Inspiratory crackles 50%
- Cyanosis 25%
- Clubbing < 10%

Clinical Findings

- CXR ill-defined nodular pattern
  "bat wing" appearance
  confused with pulmonary edema

- LAB usually normal

- PFT restriction
  reduced DLCO

Diagnosis by BAL

- Opaque, milky appearance
- Few inflammatory cells
- PAS positive staining
- Establishes the diagnosis 75%

Natural History

- Stable with persistent symptoms
- Progressive worsening
- Spontaneous improvement
- Five-year survival 75%

What causes PAP?

- Surfactant in the lungs is recycled or catabolized
- GM-CSF is required for catabolism
- High levels of antibodies to GM-CSF are present in PAP (1994)
- This leads to surfactant protein accumulation in the lung

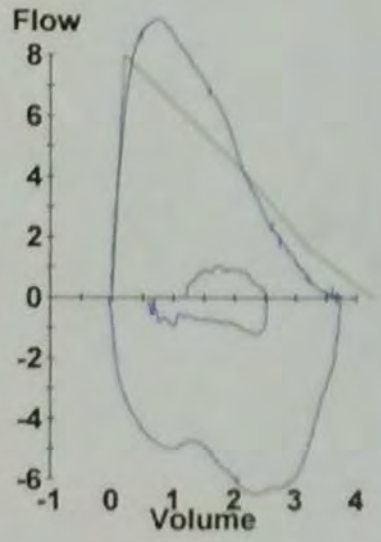
Treatment

- Whole lung lavage
- Immunotherapy with GM-CSF

Patient Follow Up

- 8 weeks post hospitalization
- Off oxygen
- Back to work with minimal dyspnea
- Normal spirometry with a mild reduction in DLCO

Follow Up Spirometry



Spirometry

		Ref	Pre Meas	Pre % Ref
FVC	Liters	4.29	3.74	87
FEV1	Liters	3.41	3.24	95
FEV1/FVC	%	78	87	
FEV3	Liters		3.61	
FEV1/FEV6	%		87	
FEF25-75%	L/sec	3.79	4.19	111
FEF25%	L/sec		9.09	
FEF75%	L/sec	1.68	1.87	111
PEF	L/sec	7.97	10.31	129
FET100%	Sec		6.26	
FEF/FIF50		<1.00	1.09	
FVL ECode			000000	
FVL Time			15:23	
MVV	L/min	147		