

American Thoracic Society International Conference

GREAT CASES

Clinical, Radiologic & Pathologic
Correlations by Master Physicians



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ATS 2018
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meets tomorrow's care™*

May 18 - May 23
San Diego, CA
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Great Cases:

Clinical, Radiologic & Pathologic Correlations

By Master Physicians

Organized by the Council of Chapter Representatives

Sunday, May 20, 2018
2:15 p.m. – 4:15 p.m.

San Diego Convention Center
Ballroom 20 D (Upper Level)

Chairing	Angela C. Wang, MD, La Jolla, CA Stephen P. Kantrow, MD, New Orleans, LA
Master Clinicians	Sharon I. Rounds, MD, Providence, RI Marvin I. Schwarz, MD, Aurora, CO Paul C. Stillwell, MD, Aurora, CO
Master Radiologist	Alison G. Wilcox, MD, Los Angeles, CA
Master Pathologist	Jeffrey L. Myers, MD, Ann Arbor, MI

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New York, NY

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Boston, MA

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Boston, MA

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Roanoke, VA

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Pittsburgh, PA

Case 1

A Persistent Narrowing

Presented by Leonard Riley, MD

A 54-year-old woman presented with a 12-year-history of intermittent dyspnea from tracheal stenosis attributed to an intubation for a bilateral oophorectomy. Beginning in 2005, she underwent multiple dilations. Biopsy of the tracheal mucosa revealed stromal fibrosis and chronic inflammation.

Despite a T-tube placement which provided some relief, she presented with another episode of dyspnea. Examination revealed decreased breath sounds of the left upper lung and no cutaneous lesions. Laboratory studies were unremarkable including normal urinalysis and serum creatinine, and negative ANCA, MPO antibody and PR3 antibody. C-reactive protein was 8.8 mg/L (0.0 – 5.0 mg/L). Chest computed tomography is shown in Figure A. She underwent a CT guided biopsy of the left upper lobe consolidation and was referred to our center. Flexible bronchoscopy of the vocal cords is shown in Figure B and the left main stem bronchus in Figure C.

Questions:

- 1) What is your diagnosis?
 - a. Tracheobronchial amyloidosis
 - b. Limited inflammatory bowel disease with organizing pneumonia
 - c. Relapsing polychondritis
 - d. Tracheobronchial tuberculosis
 - e. Granulomatosis with polyangiitis

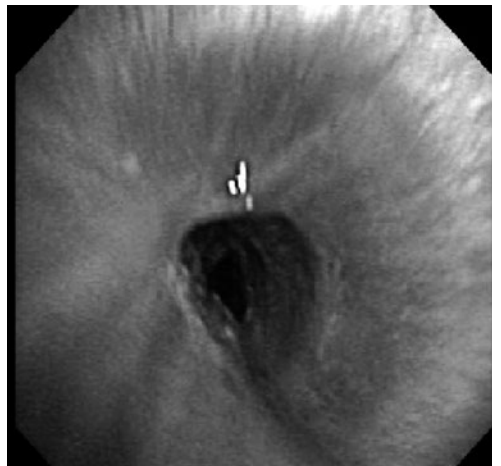
Figure A



Figure B



Figure C



Case 2

Homeless in LA

Presented by Stephanie Guo, MD

A 41-year-old undomiciled man with a history of heavy tobacco use presented with progressive dyspnea on exertion over 6 months without associated infectious, allergic or constitutional symptoms. He endorsed significant dust exposure as he was living outdoors under a bridge as well as remote silica exposure while working in construction. On exam, he was afebrile, with an oxygen saturation of 95% on room air and faint bilateral dry crackles on exam. The initial computed tomography (Fig.A) revealed diffuse, bilateral centrilobular ground glass opacities with subpleural sparing. Results of pulmonary function testing revealed a restrictive process with a TLC of 75% predicted and a DLCO of 55% predicted. There was no clinical or serologic evidence of infection or rheumatologic disease. Bronchoalveolar lavage and transbronchial biopsies also were nondiagnostic. Two months later, repeat imaging (Fig. B) showed diffuse peribronchiolar micronodular opacities, some of which were cavitory and cystic in nature, as well as irregular bilateral upper lobe calcified masses.

Questions:

- 1) What would you do next?
 - a. Repeat bronchoscopic exam with lavage and transbronchial biopsies
 - b. Proceed to thoracoscopic surgical biopsy
 - c. Prolonged course of oral steroids
 - d. Avoidance of dust exposure and repeat PFTs/CT chest in 3 months
 - e. PET scan
- 2) What is the diagnosis?
 - a. Sarcoidosis
 - b. Hypersensitivity pneumonitis
 - c. Chronic silicosis
 - d. Pulmonary Langerhan's cell histiocytosis
 - e. Pulmonary tuberculosis

Figure A

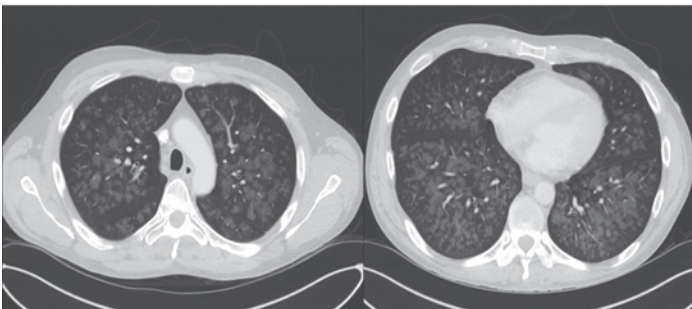
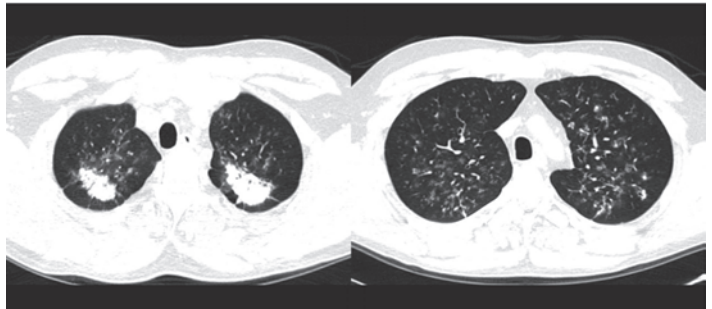


Figure B



Case 3

Looking Past the Heartache

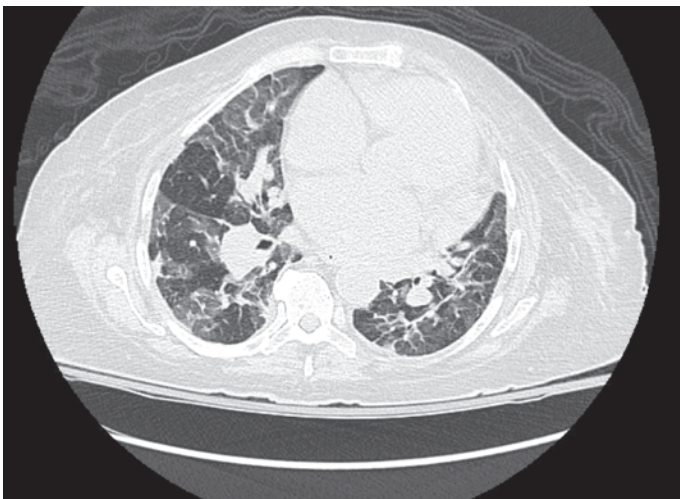
Presented by Kerri I. Aronson, MD

A 78-year-old woman with a 2 year history of diastolic heart failure, pulmonary hypertension, and restrictive lung disease on home oxygen was admitted for suspected acute on chronic heart failure exacerbation after several months of worsening dyspnea. Computed tomography (CT) scan of the chest revealed mosaic attenuation and interlobular septal thickening. Serologic testing revealed positive anti-centromere and SCL-70 antibodies. Her creatinine was elevated to 1.35 mg/dL. She lacked symptoms or examination findings of systemic connective tissue disease, and denied significant occupational or allergen exposure history. Pulmonary function testing revealed severe restriction. Right heart catheterization revealed elevation of pulmonary arterial pressure (60/16 mmHg), a pulmonary capillary wedge pressure of 17 mmHg, and no vasodilator response to nitric oxide. Cardiac MRI was negative for infiltrative disease. Her hypoxemia and dyspnea did not resolve after diuresis and repeat CT scan of the chest was unchanged (Figure A).

Questions:

- 1) What is your diagnosis?
 - a. Erdheim-Chester disease
 - b. Lymphangitic spread of malignancy
 - c. Amyloidosis
 - d. Pulmonary veno-occlusive disease
 - e. Lymphocytic interstitial pneumonia
- 2) What would you do next?
 - a. Bone marrow biopsy
 - b. Fiberoptic bronchoscopy with transbronchial biopsy
 - c. Video-assisted thoracoscopic lung biopsy
 - d. Fat pad biopsy
 - e. Low dose pulmonary vasodilator therapy

Figure A



Case 4

Casting a Wide Diagnostic Net in Pediatric Respiratory Failure

Presented by Jonathan Levin, MD

A 13-year-old male with high-functioning autism was admitted to the intensive care unit (ICU) with acute hypoxemic and hypercapnic respiratory failure of unclear etiology. He had presented two weeks earlier with chest pain and tachycardia; a CT chest revealed scattered peripheral pulmonary nodules but no pulmonary embolism. Four days prior to onset of respiratory failure, he developed cough and diffuse expiratory wheezes; chest X-ray showed perihilar infiltrates. At that time, he was started on prednisolone 2mg/kg/day, albuterol, and azithromycin. After transient improvement, his respiratory symptoms acutely worsened, and examination was notable for severe respiratory distress and diffuse expiratory wheezes. Laboratory evaluation found an elevated WBC 15,000 cells/ μ L (61% neutrophils, 24% lymphocytes, 11% monocytes, 4% eosinophils), with hematocrit 43% and platelet count 467,000 cells/ μ L. CRP was 1.1 mg/dl. Systemic corticosteroids were stopped and broad-spectrum antibiotics were started. Despite aggressive bronchodilator support, he required intubation and eventual cannulation to VV-ECMO after about 12 hours of ventilator support due to hypercapnia. He had been hospitalized for RSV as an infant and had intermittently used albuterol as a toddler. Family history was notable for multiple first-degree relatives with asthma.

At time of admission to the ICU, his chest X-ray demonstrated progressive opacification of the right upper lobe (Figure A). Bronchoscopy, performed while on VV-ECMO, revealed erythema and inflammation of the mucosa throughout the tracheobronchial tree, and numerous bronchial casts exclusively in the right lung, which were sent for pathology (Figure B). Infectious studies for bacterial, fungal, and viral etiologies were sent as well.

Questions:

- 1) What is your diagnosis?
 - a. Foreign body aspiration
 - b. Influenza infection complicated by respiratory failure
 - c. Idiopathic acute eosinophilic pneumonia
 - d. Acute severe asthma exacerbation
- 2) Which therapy would most likely lead to clinical improvement in the patient?
 - a. Methylprednisolone 4mg/kg/day
 - b. Broad-spectrum antibiotics including MRSA, antiviral, and antifungal coverage
 - c. Dornase alfa inhalation
 - d. N-acetylcysteine bronchoscopic instillation

Figure A

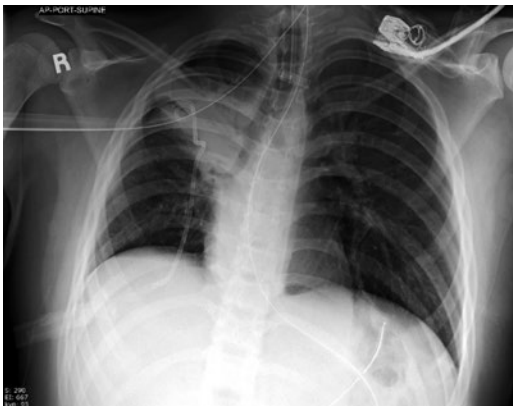
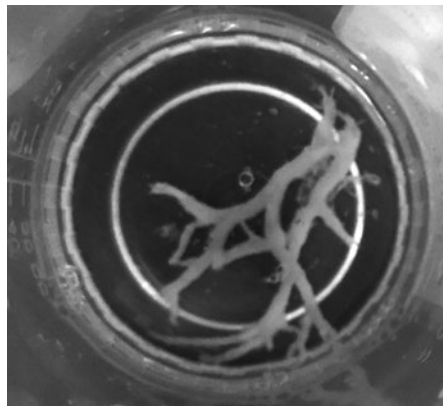


Figure B



Case 5

Things That Go Bump in the Airway

Presented by Julie Ng, MD

A 67-year-old non-Hispanic white male underwent bilateral lung transplantation (CMV D+/R+, EBV D+/R+) in 2016 for idiopathic pulmonary fibrosis. He was maintained on mycophenolate mofetil, tacrolimus and prednisone. Surveillance bronchoscopies and biopsies done at 1, 3 and 6 months post transplantation did not show any evidence of acute rejection or infection. His post-transplantation course was complicated by endstage renal disease and he was started on dialysis. He was lost to follow-up until 15 months after transplantation, when he presented with 1 week of dyspnea, cough and fever. He was started on broad spectrum antibiotics and intubated for hypoxemic respiratory failure.

A high-resolution computed tomography chest scan demonstrated new consolidative and ground-glass opacities in all lobes of the right lung (Figure A). Bronchoscopy revealed endobronchial nodules extending from the native airways at the right anastomosis to the segmental airways of the transplanted lung (Figure B). Bacterial and fungal cultures were negative.

Questions:

1. What is your diagnosis?
 - a. Post-transplant lymphoproliferative disorder
 - b. Large T cell lymphoma
 - c. Kaposi sarcoma
 - d. Pulmonary papillomatosis
 - e. Viral pneumonia
- 2 What viral infection is associated with this disease?
 - a. HHV-8
 - b. EBV
 - c. HTLV-1
 - d. CMV

Figure A

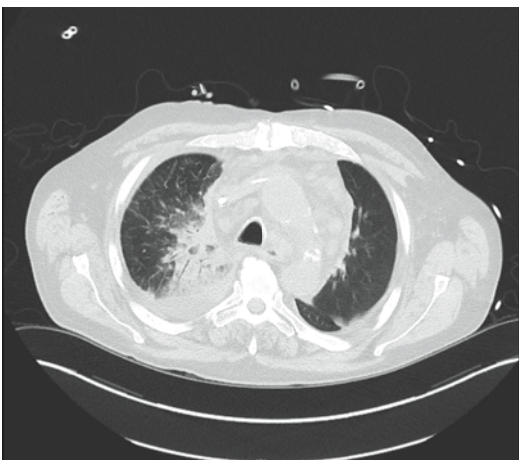
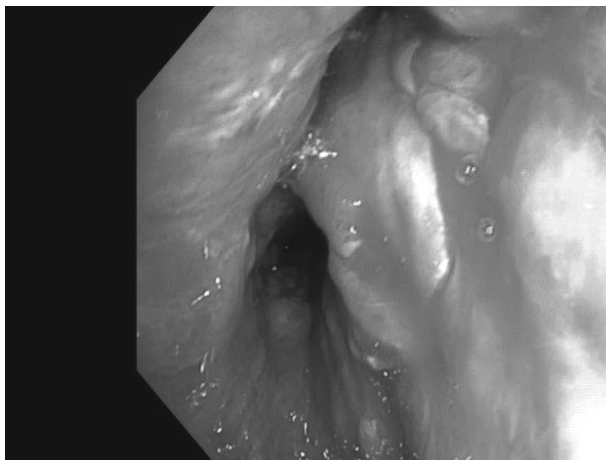


Figure B



Case 6

A Mosaic Mystery

Presented by Bradley L. Icard, DO

A 71-year-old woman with asthma treated with inhaled steroid and long acting beta agonist, diabetes mellitus type 2 and melanoma resected 9 years earlier presented with worsening dyspnea on exertion over 6 months unresponsive to albuterol. She is a lifelong non-smoker and has no occupational, hobby or mold exposures. She reported occasional wheezing and outdoor seasonal allergies. She was afebrile with a respiratory rate of 16/min, oxygen saturation of 95% on room air and a body mass index of 41 kg/m². Her lungs were clear to auscultation without wheeze. Complete blood count was normal with 1.5% eosinophils. Spirometry revealed a FEV₁/FVC 0.76, FEV₁ 2.4L (59% predicted), FVC 3.1L (57% predicted) and DLCO of 61%. Echocardiogram demonstrated diastolic dysfunction and an estimated right ventricular systolic pressure of 43 mmHg. An autoimmune evaluation included negative anti-nuclear antibodies, rheumatoid factor and anti-nuclear cytoplasmic antibodies. Computed tomography images are shown below (Figures A and B). Bronchoscopic examination and bronchoalveolar lavage were normal; all culture results were negative.

Questions:

1. What is your diagnosis?
 - a. Hypersensitivity pneumonitis
 - b. Follicular bronchiolitis
 - c. Diffuse idiopathic pulmonary neuroendocrine cell hyperplasia
 - d. Respiratory bronchiolitis with interstitial lung disease
 - e. Constrictive bronchiolitis
2. This diagnosis is associated with which one of the following?
 - a. Immunoglobulin deficiency
 - b. Collagen vascular disease
 - c. Male gender
 - d. Carcinoid tumor
 - e. Cigarette smoking

Figure A

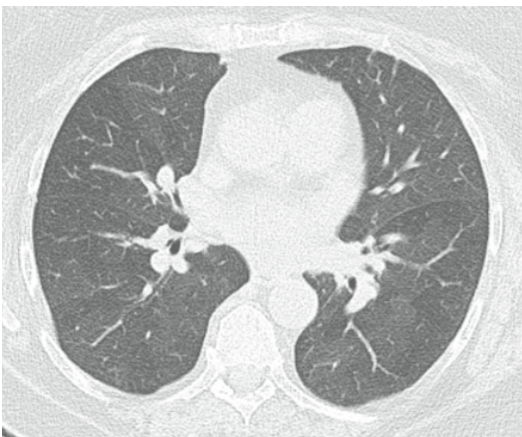
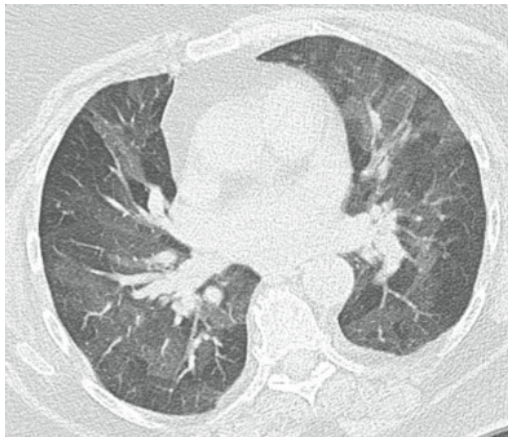


Figure B



Case 7

THE LUNGS AS “THE PUNISHER”

Presented by Maritsa Scoulos-Hanson, DO

A 52-year-old Caucasian woman with a 30 pack-year smoking history presented to the hospital with progressive fatigue and dry cough following a flu-like illness one year prior. She also endorsed a 50 pound unintentional weight loss and progressive shortness of breath with exertion that acutely worsened the week prior to her presentation. She denied travel outside the state of Pennsylvania with the exception of one trip to Niagara Falls.

On physical exam, the patient was hypoxic and had decreased breath sounds bilaterally with expiratory wheezes. Chest radiograph (Figure A) and chest CT (Figure B) demonstrated diffuse bilateral cavitary lung disease, thick walled cavities with air-fluid levels, interlobular septal thickening, bilateral mild mediastinal and hilar adenopathy. Hemoglobin 9.5 g/dL, WBC 12,000/uL, procalcitonin 0.08 ng/dL. Immunoglobulins (mg/dl) IgG 2172, IgM 143, IgA 1076, IgE 548. Sputum samples were negative for respiratory pathogens. The patient underwent bronchoscopy with bronchoalveolar lavage and biopsy. Cytology and bacterial, mycobacterial and fungal studies were negative. Transbronchial biopsy demonstrated normal lung parenchyma.

Questions:

1. What is your diagnosis?
 - a. Sarcoidosis
 - b. Tuberculosis
 - c. Granulomatosis with polyangiitis
 - d. Squamous cell carcinoma
 - e. IgG4-related lung disease
2. Which would be considered first line therapy for this diagnosis?
 - a. Rituximab
 - b. Steroids
 - c. Pembrolizumab
 - d. Plasmapheresis
 - e. Isoniazid, rifampin, pyrazinamide and ethambutol

Figure A

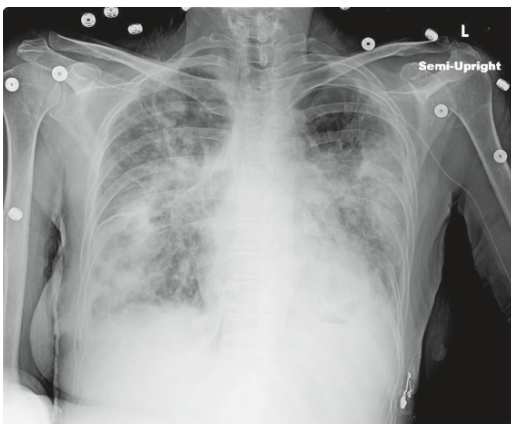


Figure B



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