

**Name:** Mr. D.

**Age:** 25

**Gender:** male

**Race:** White

**Source/Reliability:** Patient/Very Good

### Subjective

**CC:** "coughing up blood" x 1 week

#### HPI

Mr. D. is a 25-year old male with cystic fibrosis presenting with a cough and hemoptysis for 1 week. The patient reports chronic strong coughs with up to 5 ml of clear/yellow sputum associated with his cystic fibrosis but noticed an increasing red tinge about 1 week ago with the same amount of sputum. He states that he has not tried anything in particular to alleviate the cough, but reported aggravation by dust and pet dander. He ranks the severity of the cough and hemoptysis a 4 out of 10 compared to other exacerbations of his cystic fibrosis he has had in the past. Patient admits to night sweats, fatigue, dyspnea (on exertion), nasal blockage, nasal drainage (yellow/green mucus), pharyngitis, chest pain (associated with cough), heartburn/reflux and diarrhea all of which are chronic conditions associated with his cystic fibrosis, but states that none of these conditions has worsened during this recent exacerbation. Patient denies weight changes, fevers/chills, abnormal bleeding or bruising, anemia, oral lesions and epistaxis. Patient denies wheezing, asthma, bronchitis, leg edema, palpitations, syncope, HTN, heart disease or family history of DVT. Patient denies positive PPD, but was last checked about 10 years ago.

#### Review of Systems

Constitutional: Negative except for HPI.

Eye: Denies visual disturbances.

Ear/Nose/Mouth/Throat: Denies decreased hearing and ear pain.

Respiratory: Negative except for HPI.

Cardiovascular: Negative except for HPI.

Gastrointestinal: Denies nausea, vomiting, constipation, belching, bloating and abdominal pain. Denies change in bowel habits and stool caliber.

Genitourinary: Denies dysuria, hematuria, change in urine stream and urethral discharge.

Musculoskeletal: No back pain, neck pain, joint pain and muscle pain.

Integumentary: No rash, pruritus and skin lesions.

Neurologic: No abnormal balance, confusion, memory loss, numbness, tingling and headache.

Psychiatric: No anxiety, depression, hallucinations, delusions and suicidal ideation.

#### PMH

- **Allergies:** Vancomycin (red-man syndrome) and Pediazole (reaction unknown).
- **Hospitalizations:** Approximately 2 hospitalizations per year since birth for CF related infections. Never intubated for respiratory distress.
- **Illnesses:** CF (DX at birth) without treatment until age 10, chronic sinusitis GERD, mild psoriasis (untreated). Patient reports history of depression (untreated).
- **Immunizations:** Up to date per patient.
- **Surgeries:** Sinus surgeries in 2005 and 2010 for nasal polyp removal.
- **Trauma:** none
- **Medications/supplements:** Pulmozyme (inhaled), azithromycin (oral), tobramycin (inhaled), albuterol (inhaled), fluticasone (inhaled), hypertonic saline (inhaled mist), pancrelipase (oral) and pantoprazole (oral). Patient has an

oscillation vest for airway clearance but admits to very infrequent use (once a week or less) and admits to poor medication compliance due to "life getting in the way".

- **Youth illnesses:** chickenpox

#### FH and SH

Patient enjoys working independently as a sound recording engineer. He was recently married and purchased a home in which he built a recording studio. He has no children. Patient is a moderate drinker (1-2 per week), does not smoke cigarettes, but admits to smoking marijuana several times a week. He categorizes his diet as fair to good, but exercises little due to fatigue and dyspnea with exertion (e.g. climbing 1 flight of stairs). His parents are both living and healthy. He has one sister (29) who also has cystic fibrosis and is doing well after a double lung transplant last year. He has a strong support system of friends and family. Patient denies recent travel.

#### **Objective**

Vitals:                    T 35.6 C (oral)                    RR 18 BPM, regular                    HR 84 BPM, regular  
                                   SPO2 94% on room air            BP-Rt arm sitting: 104/58 mmHg

General Inspection: 25 year old male, reclining comfortably in bed, in no apparent distress. Patient is cooperative, alert and oriented x 4. Speech is fluid and appropriate. Skin is warm and moist with adequate skin turgor. No pallor, jaundice, cyanosis. Mild erythematous plaque noted on both elbows. Capillary refill < 2 seconds. Nail clubbing noted bilaterally.

HEENT: Normocephalic/atraumatic. Eyes: PERRLA. Conjunctiva pink with no scleral jaundice. Nose: Nares patent, no sinus tenderness. Mouth: No lesions, inflammation or exudate to oral mucosa, tongue or gum line. No bleeding or tenderness on palpation of gums. No tonsillar enlargement or exudate. Neck: Supple and trachea midline. No thyromegally. No cervical or axillary lymphadenopathy.

Lungs: Equal and bilateral chest rise, breathing unlabored with no accessory muscle use. No pain or tenderness on palpation of sternum, anterior or posterior thorax. Mild increased tactile fremitus and E-to-A changes bilaterally in lower lobes. Lungs clear to percussion and auscultation in all lobes. No rales, rhonchi, stridor or wheezing. Diaphragmatic excursion 5cm bilaterally.

Cardiovascular: Regular rhythm of S1 and S2 heart sounds audible at the aortic, pulmonic, tricuspid, and mitral areas. No pulsations, thrills, or heaves. PMI located in 5<sup>th</sup> intercostal space, midclavicular line. No murmurs, gallops, ectopic beats or pericardial friction rub. No JVD at 30.

Extremities: Bilateral radial and dorsalis pedis peripheral pulses present and 2+. No ankle edema, calf swelling or tenderness. Negative Homan's sign.

#### Admission labs and imaging

Viral studies:            Respiratory culture TB/AFT:            pending  
                                   Cystic Fibrosis Respiratory Panel:    Pseudomonas aeruginosa

139	108	11	109
4.0	26	.81	

Absolute Neutrophils: 6.28

10.1	14.5	165
	39.6	

ALT: 36

Alk Phos: 166

AST: 26

XR chest PA + lateral: Findings of cystic fibrosis characterized by widespread bronchial impaction and bronchial thickening essentially stable without acute superimposed process.

### Assessment

Mr. R is a 25-year-old man with cystic fibrosis presenting with hemoptysis for 1 week. Patient is not experiencing any additional exacerbation of chronic symptoms associated with cystic fibrosis, but admits to only infrequent airway clearance therapy at home. Patient is not in acute respiratory distress. Physical exam shows bilateral dullness to percussion in lower lobes and sputum culture is positive for pseudomonas aeruginosa.

1. Pulmonary exacerbation of cystic fibrosis bronchiectasis: Most likely diagnosis due to history of cystic fibrosis and hemoptysis as well as patient's admitted inconsistency with medications and airway clearance. Elevated WBC/neutrophils indicates bacterial origin and sputum culture confirms increased colonization with pseudomonas aeruginosa. Chest x-ray shows confirms bronchial impaction and bronchial thickening.
2. Pulmonary TB: Patient has low environmental risk factors for TB (travel to endemic areas, exposure to TB, unstable living), however, the symptom complex for TB and cystic fibrosis bronchiectasis are similar (cough, dyspnea, night sweats, hemoptysis). Patient has not experienced recent weight loss or fever and chest x-ray shows no hilar lymphadenopathy, consolidation or infiltrates. TB AFB is pending.
3. Pulmonary hypertension: Possible due to underlying chronic lung disease, dyspnea, fatigue, chest pain and noted noncompliance with treatment. On exam patient is not cyanotic and without peripheral edema, increased JVD or findings on cardiac auscultation.

### Plan

#### Diagnostic

- Admission labs, imaging and viral cultures confirmed infection with pseudomonas aeruginosa.
- Monitor AFB culture reports, consider Quantiferon Gold testing to R/O TB in conjunction with infection.
- Continue monitoring patient vitals including SPO2. Increased work of breathing may indicate need for intubation and mechanical ventilation.
- ECG monitoring.
- ABG for respiratory acidosis if signs of hypo/hyperventilation, cyanosis, tachypnea, tachycardia and/or altered mental status.
- Transthoracic Doppler echocardiography may be indicated to screen for pulmonary hypertension if antibiotic treatment does not resolve symptoms.
- After resolution of exacerbation measure peak expiratory flow or FEV1/FCV assess current stable lung function.

## Therapeutic

- Recommended treatment for pseudomonas aeruginosa infections in cystic fibrosis patients is IV anti-pseudomonal beta-lactam (e.g. piperacillin/tazobactam, ceftazidime sodium) and aminoglycoside (e.g. gentamicin or tobramycin). Hospital susceptibility testing showed intermediate susceptibility to aminoglycosides (both gentamicin and tobramycin) so alternative therapy of with colistimethate sodium is indicated.
- Respiratory Therapy: Provide supervised daily airway clearance sessions during hospitalization to facilitate restoration of lung function and to assess proper technique and treatment duration. Administer bronchodilators (albuterol) prior to airway clearance and inhaled antibiotics (tobramycin) after.
- Continue pancrelipase (oral) and pantoprazole (oral) during hospitalization.

## Patient Education

Inquire about the patient's understanding of cystic fibrosis and the medications prescribed to manage the symptoms and reduce hospitalizations. Stress the importance of airway clearance and discuss barriers to performing daily clearance and work with patient to come up with plans to increase their use. Introduce alternative airway clearance therapies that may be more amenable to the patient's lifestyle. Offer advice about the detrimental effects of his marijuana use on his declining lung function and assess patient's willingness to try to stop. Discuss patient's depression and offer resources for therapy during hospitalization and/or after discharge.

Problem list

1. Cystic fibrosis
  - a. Cough
  - b. Dyspnea
  - c. Diarrhea
2. GERD
3. Psoriasis
4. Depression
5. Chronic Sinusitis s/p 2 surgeries
6. Marijuana use

## References:

*Epocrates*, iPhone app.

*Skyscape*, iPhone app.

Zobell JT1, Young DC, Waters CD, Ampofo K, Stockmann C, Sherwin CM, Spigarelli MG., Optimization of anti-pseudomonal antibiotics for cystic fibrosis pulmonary exacerbations. *Pediatr Pulmonol*. 2013 Jun;48(6):525-37. doi: 10.1002/ppul.22757.