Case of a 16 year Female with Cystic Fibrosis and Persistent Dyspnea

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Summary: 16 year old African American female with cystic fibrosis presents with persistent dyspnea that failed to improve despite therapy with intravenous antibiotics.

HPI: Two weeks prior to admission the patient developed cough, malaise, and fatigue due to influenza B. She was treated with 5 days of oseltamivir. Her symptoms initially improved but 5 days prior to admission she developed a worsening productive cough as well as shortness of breath and dyspnea on exertion. Upon presentation her PFTs demonstrated FVC 1.34L (47%) and FEV1 0.89L (35%), which were decreased from FVC 1.36L (54%) and FEV1 0.94L (41%). Therefore, she was admitted for IV antibiotics therapy.

PMH: The patient has CF, due to F508del and c438_440delTCA CFTR mutations, which was diagnosed in infancy due to failure to thrive. She has pancreatic insufficiency, allergic bronchopulmonary aspergillosis (ABPA) and poor weight gain despite a gastrostomy tube. The patient has undergone a left upper lobectomy for severe bronchiectasis in 2010 and subclavian Port -A- Cath placement in 2008. She is colonized with Pseudomonas aeruginosa and MRSA.

Hospital course: The patient was initially treated with aggressive pulmonary clearance, IV antibiotics (trimethoprim/sulfamethoxazole, cefepime, and tobramycin), and therapy for ABPA (prednisone and itraconazole). Her cough, sputum production and lung function improved after 1 week of therapy. However, she continued to have significant dyspnea that was out of proportion with her other respiratory symptoms. Given her persistent shortness of breath an echocardiogram was performed to evaluate for pulmonary hypertension. The echocardiogram demonstrated a non-occlusive but hypermobile thrombus (1.5cm x 1.2cm) associated with the tip of the catheter in the right atrium with a portion of the thrombus adherent to the junction of the right atrium and coronary sinus os. Estimated right ventricular systolic pressure = 43% systemic. A subsequent chest CT demonstrated chronic thromboembolism of the right lower lobe pulmonary artery as well as significant progression of cystic bronchiectasis within the left lung and nodular consolidation in the right upper lobe. Anticoagulation with enoxaparin was initiated. Repeat echocardiograms demonstrated stable clot size. Her Port-a-Cath was used during her anticoagulation without complication. Her dyspnea and PFTs improved significantly prior to discharge.

Discussion points:
1. What are the risk factors for venous thromboembolism (VTE) and pulmonary embolism in CF?
2. What is the relationship between central venous catheters, venous thromboembolism (VTE) and pulmonary embolism in CF?
3. Is there a role for prevention of VTE in individuals with CF and central venous catheters?
4. When should the diagnosis of pulmonary embolism be considered?
5. What is the best approach to diagnosis of pulmonary embolus in CF?
6. What is the appropriate therapy for VTE or pulmonary embolus?