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March 2023 Critical Care Case of the Month: A Bad Egg

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History of Present Illness

You are asked to see a 35-year-old man who was admitted to the ICU from the ER the previous night with an exacerbation of his chronic obstructive pulmonary disease (COPD). He has a long history of COPD and came to the ER for COVID-19 testing because he was at a party where a friend was later found to COVID-19. He denies any change in his chronic respiratory symptoms but his spirometry was significantly worse than his baseline in the ER and despite his protests he was admitted. He was treated with empiric antibiotics (amoxicillin and clavulanic acid), corticosteroids (methylprednisolone 125 mg every 6 hours), bronchodilators (albuterol/ipratropium every 4 hours) and oxygen. He says his breathing has not improved and he wants to go home. He has had gradually increasing shortness of breath for the past 8-10 years. He has minimal cough but denied any fevers, systemic symptoms, or wheezing.

PMH, FH, and SH

He had a history of multiple pneumothoraces which eventually led to bilateral pleurodesis. He has had not pneumothoraces since. He had a benign bone tumor removed about 25 years ago and a history of manic-depression. There is no FH of any similar type of problems. He does smoke about 3/4 pack of cigarettes per day and has more than occasional marijuana use.

Physical Exam

Physical examination was unremarkable expect for a well-healed scar on the left thigh.

Spirometry

Previous spirometry performed as an outpatient showed his FVC 2.54 L (53% of predicted) with an FEV1 1.25 L (31% of predicted). These improved to 2.99 L and 1.52 L after a bronchodilator. His spirometry last night in the ER was FVC 1.63 L (29 % predicted) and FEV1 0.80 L (18 % predicted).

Radiography

A chest radiograph was performed (Figure 1).

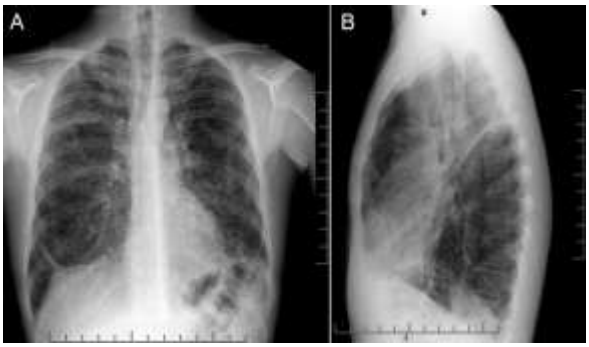

Figure 1. PA (panel A) and lateral (panel B) chest x-ray.

What should be ***done at this time***?

1. Continue his antibiotics, corticosteroids and bronchodilators
2. Order an alpha-1 antitrypsin level
3. Transfer to the floor
4. 1 and 3
5. All of the above

Correct!
5. All of the above

The patient's COVID-19 was negative and his chest x-ray does not show any focal areas on consolidation. He is stable and could be transferred to the floor. His present therapy could probably be stopped but that is not one of the options.

It is important to recognize that this patient does not likely have COPD from cigarette smoking. Some assume that all shortness of breath is either heart failure or COPD probably since those are the two most common diagnosis. COPD would be unusual in this age group unless the patient has alpha-1 antitrypsin deficiency (A1AT). For reasons that are not entirely clear, patients with A1AT often present with reversible airway obstruction and can be misdiagnosed with asthma.

He is transferred to the floor and discharged on a short course of antibiotics, a rapid taper of his corticosteroids, and bronchodilators by metered dose inhaler. He is scheduled for follow up in the clinic in 10-14 days.

Which of the following should be ***done at his follow up appointment***?

1. Repeat spirometry
2. HRCT of thorax
3. Review pathology
4. 1 and 3
5. All of the above

Correct!
5. All of the above

His repeat spirometry done in the office shows a FVC of 2.15 L and a FEV1 of 1.05 L. A HRCT of the thorax is interpreted as showing reticulonodular changes and small cysts bilaterally in the mid and upper lung zones which is stable compared to a CT scan done 2 years ago. His alpha-1 antitrypsin level had not returned. A request for his pathology report was submitted to another hospital.

Which of the following is the ***most likely diagnosis***?

1. Alpha-1 antitrypsin deficiency
2. Birt-Hogg-Dubé syndrome (BHD)
3. Langerhans cell histiocytosis
4. Lymphangioleiomyomatosis (LAM)
5. Lymphoid interstitial pneumonia (LIP)

Correct!
3. Langerhans cell histiocytosis

All the diagnosis has been associated with pneumothoracies (1). LAM is a rare disease that is associated with cystic lung disease, angiomyolipomas, and obstruction of the lymphatic system. Birt-Hogg-Dubé syndrome (BHD) is a hereditary condition associated with multiple non-cancerous (benign) skin tumors, lung cysts, and an increased risk of kidney lesions (cysts, benign tumors, and kidney cancer). Lymphoid interstitial pneumonia (LIP) is an uncommon form of interstitial lung disease in adults that is characterized histopathologically by infiltration of the interstitium and alveolar spaces of the lung by lymphocytes, plasma cells, and other lymphoreticular elements.

Langerhans cell histiocytosis (LCH) is the patient's diagnosis as you may have suspected from the title. The disease has gone by several other names, including eosinophilic granuloma (EG), Hand–Schüller–Christian disease, Abt-Letterer-Siwe disease, and histiocytosis X. LCH is a rare disease involving clonal proliferation of Langerhans cells, abnormal cells deriving from bone marrow and capable of migrating from skin

to lymph nodes (2). Clinically, its manifestations range from isolated bone lesions to multisystem disease. Chest radiographs in patients with LCH reveal diffuse reticulonodular infiltrates that are symmetric with sparing of the lung bases a frequent finding. Unlike other interstitial pneumonitides, the lung volumes may be normal or increased and often present with a mixed obstructive and restrictive picture on pulmonary function testing.

Our patient's diagnosis was confirmed when his pathology from his bone lesion returned. His alpha-1 antitrypsin level was normal.

Which of the following is the recommended initial ***treatment in patients with LCH***?

1. Glucocorticoids with or without vinblastine in multisystem LCH
2. Early lung transplantation
3. Smoking cessation
4. 1 and 3
5. All of the above

Correct!

3. Smoking cessation

Evaluation of treatment in LHC is difficult because spontaneous remissions can occur. Glucocorticoids are often empirically given which seem to help with systemic symptoms and radiographically (2). Vincristine is sometimes added. LHC is predominately a disease of smokers and smoking cessation seems to help. There are multiple reports of improvement with smoking cessation (3,4). Interestingly, patients with LHC seem to have difficulty quitting smoking.

Lung transplantation has been performed in both children and adults with LCH. Recurrence of disease does occur in the allograft at a high rate (20% in one series) (3). LCH has also been associated with malignancy in several reports. Given the small numbers of transplants and the reported outcomes to date, lung transplantation remains an appropriate therapeutic option for patients with advance disease due to LCH.

Our patient was followed and seems stable. He did quit cigarette smoking with the aid of Nicorette gum which he still uses. Patients with LCH often do not quit smoking, although when they do, their disease often stabilizes or improves. His marijuana smoking might contribute to his lack of improvement.

References

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