Case Study: Bronchiolitis Obliterans

Can an interstitial lung disease be disguised as poorly controlled Asthma?

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Case Study

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**Chief Complaint**

A 15-month old Hispanic male with a history of asthma and allergic rhinitis presented to the primary care office with his mother after having been recently discharged from his second hospitalization for viral pneumonia. The presenting symptoms for this visit were wheezing, cough, and fever for the past 24 hours.

**History of Present Illness**

The patient had been hospitalized for three days with viral pneumonia; he did not require intubation. He has had a persistent cough post hospitalization. Fevers began in the previous 24 hours along with wheezing. He has been receiving nebulizer treatments about every 4 to 6 hours with no improvement. He has been taking fluids, but his appetite is poor. He has been urinating as usual. There is no vomiting or diarrhea.

**Past Medical History**

His mother is 21 years old, a Gravida 2, Para 1, AB 0. She sought prenatal care at 2 ½ months of pregnancy and took prenatal vitamins throughout the pregnancy. There were no maternal complications, and no exposure to medications, alcohol, recreational drugs, or tobacco. The patient was delivered via Cesarean section due to post term dates. He weighed 6 pounds 9.9 ounces at birth and was discharged at 2 days of age.

The child’s mother reported that the patient did not have any respiratory problems at birth. At five months of age, the patient was seen in the emergency department (ED) with cough and fever and diagnosed with bronchiolitis. He returned to the ED at seven
months of age with fever, cough, and bronchiolitis. Thereafter, he presented to the primary care clinic approximately every 3 to 4 weeks for persistent cough and subjective fevers and was treated with antibiotics, albuterol, and oral steroid therapy. At 12 months of age, the patient was hospitalized with a fever of 104.6 degrees Fahrenheit, cough, and diarrhea; he was diagnosed with adenoviral pneumonia. After this hospitalization, he was referred to the pediatric pulmonologist.

He presented to the clinic again at 14 months of age for persistent cough, wheezing, fever with a maximum temperature reading of 102 degrees Fahrenheit, and diarrhea for one week. A chest x-ray revealed prominent findings of interstitial infiltrates bilaterally, and he was treated with antibiotics. He returned to the clinic for a follow up visit two days later with improvement of the cough. A chest x-ray was repeated which revealed bronchopneumonia with bilateral interstitial and hilar infiltrates without consolidation. The child was seen again in the ED at 15 months of age, and was then hospitalized with viral pneumonia. His mother reported that the patient has required multiple ED visits since his birth.

Family History

The child’s mother has a history of anemia which has resolved. She states that her grandmother had heart disease and hypertension. There is also a history of asthma and cancer, but was unable to identify which family members had these illnesses. The child’s father is healthy and reports no history of chronic illnesses in his family.

Personal/Social/Developmental History

The child lives at home with his parents and 5-year-old sister. He does not attend daycare. His primary caregiver is his mother; she does not work outside the home. She is
from Mexico and speaks only Spanish. His father is employed as a day worker in construction. He speaks mostly Spanish with some English. At one time, the family kept parakeets as pets, but the family no longer has these birds in the home. There is no smoking in the home. The child speaks three to five words in Spanish. He sat with support by six months of age, and sat alone by eight months of age. He started walking at 13 months of age and loves to play games such as peek-a-boo and patty cake.

**Review of Systems**

The mother states that the child has a poor appetite and has been slow to gain weight. There is no history of cardiac problems. He has not vomited recently. He has 4 soft stools per day. He has had several bouts of diarrhea during antibiotic therapy but does not currently have diarrhea. There has been no blood or mucus in his stools.

**Medications**

The patient takes fluticasone (Flovent), levalbuterol (Xopenex), montelukast (Singulair), and albuterol nebulizer treatments for asthma; however, despite this medication regimen his asthma remains poorly controlled. Levalbuterol (Xopenex) nebulizer treatments are administered every 6 hours as needed for wheezing and cough, but the treatments do not seem to help him much, according to his mother.

**Physical Exam**

The patient is a thin appearing 15-month-old Hispanic toddler in no acute distress. He weighs 18 pounds and 5 ounces (<3rd percentile), his height is 28 inches (<3rd percentile) and his head circumference is 45 cm (<15 percentile). His vital signs are as follows: T 96.5 degrees Fahrenheit; HR 125; RR 36; and the oxygen saturation is 94% on room air. He is alert, and his skin is pink, warm and dry with no cyanosis. His head is
normocephalic. There are dark circles under his eyes (allergic shiners). His tympanic membranes are grey with normal light reflexes bilaterally. He has clear nasal discharge. His trachea is midline and neck is supple without masses. There is symmetrical thoracic expansion on inspiration. His respiratory effort is unlabored, but he has mild expiratory wheezing bilaterally. The cardiovascular exam reveals a regular rate and rhythm without murmurs. Peripheral pulses are strong bilaterally; his capillary refill is less than 3 seconds. There is no clubbing noted or edema of his upper and lower extremities. The abdomen is soft with no distension and no liver or spleen enlargement.

**Diagnostic Testing**

A complete blood count at this visit was normal; the chest x-ray revealed hyperinflation of the lungs. Multiple chest x-rays during his recent hospitalization revealed the presence of pneumonia. All chest x-rays were reviewed by the pulmonologist; extensive peribronchial thickening with more confluent bibasilar atelectasis/infiltrate were noted in both the left lower lobe and right middle lobes. No pleural fluid was present. The heart and pulmonary vasculature appear normal.

Cultures were obtained; chlamydia, fungal, mycoplasma, legionella, and psittacosis were subsequently found to be negative. A sweat test was done which was negative.

CT scan of chest showed mosaic perfusion with slight vascular attenuation and marked peribronchial thickening and some bronchiectasis in both lower lobes. In addition, there was prominent tree-in-bud pattern especially in the left lower lobe. These findings are consistent with bronchiolitis obliterans.

**Case Study Questions**
1. What are the differential diagnoses for this patient?

Differential diagnoses for a child who presents with chronic cough and wheezing include asthma with exacerbation, pneumonia, pertussis, bronchiolitis, cystic fibrosis, foreign body obstruction and laryngotracheobronchitis (Benich and Carek, 2011; Boynton et al., 2010). A comprehensive lab panel, viral culture panel and chest x-ray can rule out pneumonia, pertussis, and foreign body obstruction.

Cystic fibrosis was on the list of differential diagnoses for this child. His weight and length were in the 50th percentile at birth; however, at the time of this visit, his weight and length were less than the 3rd percentile. His disease process affected his appetite and he developed failure to thrive. He had a negative sweat test, which ruled out cystic fibrosis.

Laryngotracheobronchitis is usually seen in children under the age of three years old and characterized as a harsh barking cough with severe inspiratory stridor and elevated temperature (Boynton et al., 2010). This toddler presented with fever, but did not have stridor or a barking cough.
Bronchiolitis is a respiratory infection that affects children most commonly under the age of six months old but can occur in children up to the age of two years (Moonnumakal & Fan, 2008). Respiratory syncytial virus (RSV) is the most common agent that causes bronchiolitis (Dawson-Caswell & Muncie, 2011). RSV has an incubation time of two to eight days and viral shedding is present for three to eight days, although it may last up to four weeks: initial symptoms include fever and cough followed by dyspnea and wheezing (Dawson-Caswell & Muncie, 2011).

A severe case of acute bronchiolitis increases a child’s risk of developing post-infectious bronchiolitis obliterans. Bronchiolitis obliterans is manifested by tachypnea, increased anterior-posterior chest diameter, crackles, wheezing and hypoxemia that persist for at least 60 days after the initial infection that causes lung injury (Moonnumakal & Fan, 2008, p. 274). Children with acute adenovirus pneumonia can develop bronchiolitis obliterans resulting in severe respiratory compromise and may require intensive care unit admission, the use of oxygen, mechanical ventilation, corticosteroids and B-2 agonist administration (Castro-Rodriquez, Daszenies, Garcia, Meyer, & Gonzales, 2006; Colom, Tepper, Vollmer, & Diette, 2006). The adenovirus is the leading infectious cause of bronchiolitis obliterans worldwide, although other causative agents include influenza, parainfluenza, measles, RSV, varicella, and Mycoplasma pneumonia (Moonnumakal & Fan, 2008).

2. **What are the diagnostic criteria for bronchiolitis obliterans?**

The diagnosis of bronchiolitis obliterans is made through a careful investigation of the patient’s history, a thorough physical exam, an infectious disease evaluation, imaging studies, lung function tests, and a lung biopsy (Moonnumakal & Fan, 2008). An
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Infectious-disease evaluation should include appropriate viral cultures, rapid tests by immunofluorescence or radioimmunoassay, paired serology and DNA probes for adenovirus and other viruses during the acute illness (Moonnumakal & Fan, 2008, p. 274).

During the acute phase, a chest x-ray will show bilateral interstitial prominence and marked hyperinflation with consolidation or atelectasis (Moonnumakal & Fan, 2008). One-third of the children will also show Swyer-James syndrome, which is a unilateral, small hyperlucent lung (Moonnumakal & Fan, 2008).

As with all progressive chronic lung disorders, lung biopsy is the gold standard (Smith et al., 2010). In today’s advanced healthcare, however, a high resolution computed tomography (CT) scan is taking the lead for diagnosis of bronchiolitis obliterans because it is less invasive and a lung biopsy is not always able to identify bronchiolitis obliterans lesions due to their heterogeneous nature. Moonnumakal and Fan (2008) noted that lung biopsy in children may be either normal or non-diagnostic in up to one-third of the patients who already have a diagnosis of bronchiolitis obliterans.

The CT should be performed when lung volumes can be controlled with breath holding and complete exhalation (Smith et al., 2010). Clinical findings on a CT that are indicative of bronchiolitis obliterans include parenchymal hypoattenuation, vascular attenuation, and bronchial lumen narrowing (specificity >95.5%) (Smith et al., 2010, p. 258). When these findings are found in combination, specificity for bronchiolitis obliterans improves (Smith et al., 2010). If the results of the CT are highly specific for bronchiolitis obliterans, a lung biopsy is not necessary for confirmation of diagnosis (Mattiello et al, 2010). If the results of the CT are not indicative of bronchiolitis...
obliterans, a lung biopsy from multiple sample sites should be performed for confirmation (Smith et al., 2010).

Other studies, including ventilation-perfusion lung scans, have been used to assist with the diagnosis of bronchiolitis obliterans, but have been largely replaced by CT scans due to better specificity (Chan & Dell, 2005; Moonnumakal & Fan, 2008). Bronchoscopy can be used during acute illness to assist in identifying the infectious agent (Moonnumakal & Fan, 2008).

3. What is the pathology of bronchiolitis obliterans?

Bronchiolitis obliterans is a rare chronic obstructive lung disease disorder characterized by fibrosis and inflammation of the respiratory bronchioles resulting in narrowing, which causes complete or partial obliteration of the small airway (Smith et al., 2010). The histological picture of bronchiolitis obliterans reveals scarring in the epitheliums of the small airways (Moonnumakal & Fan, 2008). The epithelial cell function is changed or becomes necrotic and this leads to an accumulation of fibrinopurulent exudate that causes collagen to be deposited (Moonnumakal & Fan, 2008). These lesions can progress and cause scarring. The lumen size then decreases which leads to obstruction (Moonnumakal & Fan, 2008).

4. What is the management plan for a child with post infectious bronchiolitis obliterans?

While there is not a definitive treatment therapy for patients with bronchiolitis obliterans due to the lack of research available to date, the treatment plan is largely supportive and ranges from preventive to symptomatic treatment. Preventative treatment consists of avoiding tobacco smoke and other inhaled irritants, annual influenza vaccine,
pneumococcal vaccine, a tailored exercise and fitness regime, pulmonary physiotherapy, and adequate nutritional intake (Moonnumakal & Fan, 2008). Nutritional support is vital as developmental needs are increased due to respiratory compromise (Bosa et al., 2008; Fischer, Sarria, Mattiello, Mocelin, & Castro- Rodriguez, 2010). As with any chronic lung disease, rehabilitation programs should be instituted to promote proper use of respiratory muscles through the incorporation of aerobic exercises (Fischer et al., 2010).

Other supportive measures for the treatment of bronchiolitis obliterans consist of supplemental oxygenation if hypoxemia is present, bronchodilators for exacerbation of wheezing, antibiotics for secondary bacterial infections, and inhaled corticosteroids which treat the inflammatory component of bronchiolitis obliterans (Dosanjh, 2007; Fischer et al., 2010). There is some controversy related to the use of corticosteroids due to a lack of research; however, based on current evidence, treatment with steroids should be implemented if the patient responds by clinical, radiographic, and pulmonary function (Dosanjh, 2007). The efficacy of corticosteroid therapy should be assessed on an individual basis until further larger scale clinical trials are performed (Dosanjh, 2007).

Surgical excision of bronchiectasis may be indicated in the first year of the disease once conservative measures have failed. Other treatment measures include treatment for gastro-esophageal reflux, which commonly occurs secondary to post infectious bronchiolitis obliterans (Fischer et al., 2010). In severe, progressive forms of the disease, lung transplant remains the ultimate option; however, lung transplant is not usually performed in cases of post infectious bronchiolitis obliterans. It is more commonly utilized in patients with bronchiolitis obliterans secondary to bone marrow transplant and Steven Johnson syndrome (Moonnumakal & Fan, 2008).
5. *What is the recommended follow up care for children with bronchiolitis obliterans?*

Current literature shows that close monitoring and preserving respiratory activity may be the most prudent intervention in order to prevent subsequent mortality and morbidity, especially in the first year of follow up (Chiu et al., 2008). Along with treating exacerbations, it is vital to monitor nutritional status, as this can have an impact on the efficiency of the respiratory muscles (Fischer et al., 2010).

More studies are needed to better understand the course of bronchiolitis obliterans in order to provide more effective treatment and long term follow up care. The BOLAT initiative (Bronchiolitis Obliterans in Latin America) intends to utilize research findings to further enhance the knowledge of the pathophysiology, implications, complications and long term follow up with the participation of multiple centers (Fischer et al. 2010).

**Summary**

Although this patient’s primary diagnosis had been asthma, he was hospitalized with adenoviral pneumonia and post-hospitalization he returned to the clinic multiple times with continued wheezing, cough and persistent fever. This challenging case involved collaboration between the primary health care provider and the pulmonary specialist. Eventually, the diagnosis of bronchiolitis obliterans was made. This child received intravenous pulse steroid therapy and his symptoms resolved over the next few weeks. He continues on daily fluticasone (Flovent), montelukast (Singulair) and takes levalbuterol (Xopenex) and albuterol as needed.

Nurse practitioners care for many children with asthma and may encounter a similar situation in which standard treatment is not successful and should consider
collaboration with a pulmonary specialist. A case of “poorly controlled” asthma needs further investigation, as there may be other diagnoses to consider, such as bronchiolitis obliterans.
References


