CLINICAL VIGNETTE

Wheezing Beyond Asthma

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Presenting History

A 27-year-old male with mucopolysaccharidosis type II, also known as Hunter syndrome, and moderate-persistent asthma was admitted for wheezing and shortness of breath. His symptoms were refractory to his chronic inhalers and steroid bursts. His baseline asthma regimen consisted of fluticasone-salmeterol 500/50 BID, montelukast 10mg daily and as needed albuterol. In the last year, he reported two episodes of left-sided pneumonia requiring hospitalization and prolonged recovery, but no intubation. He reports his wheezing typically is more pronounced on his left side. He otherwise denied sick contacts, recent travel, fevers, or rhinorrhea.

On initial ED evaluation, he was afebrile and normotensive, but was tachycardic to 120/min, tachypneic to 28/min, with 90% O_2 saturation. His exam was notable for congenital short stature secondary to Hunter Syndrome. Breath sounds were notable for bilateral wheezing, more pronounced on the left, with left lower lobe rhonchi. Heart sounds were unremarkable, and there was no lower extremity edema, or jugular venous distension. He was placed on bilevel positive airway pressure (BPAP) with subjective improvement.

Laboratory data included normal CBC, negative respiratory viral panel, negative procalcitonin, and BNP < 20 ng/L. Initial venous blood gas while on BPAP was notable for pH 7.32 and pCO2 of 55 mmHg. Basic metabolic panel was remarkable for HCO3 26 mmol/L.

Initial portable chest XR (Figure 1) showed mild left perihilar and retrocardiac airspace disease with blunting of the left costophrenic angle. CT chest without contrast revealed severe main distal trachea collapse (Figure 2) secondary to tracheobronchomalacia (TBM) with extension of collapse into the left mainstem bronchus (Figure 3) resulting in distal bronchiectasis, mucus plugging, and post-obstructive pneumonia.

Clinical Course

After finding significant TBM correlating to left laterality of wheezing, steroids were stopped, and he was placed on high flow nasal cannula during the day and nighttime non-invasive positive pressure ventilation (NIPPV) using average volume-assured pressure support (AVAPS) to provide some degree of pressure airway stenting to reduce hypercapnia. Albuterol and ipratropium every 6 hours replaced his home inhalers. Systemic steroids were weaned in favor of inhaled corticosteroids which were subsequently weaned. A pulmonary hygiene regimen of N-acetylcysteine, 3% saline, and chest physiotherapy using IPV and bidaily oscillatory vest therapy were started to help mucus clearance. He improved clinically over the week. At discharge, he was set up with a home AVAPS machine and oscillatory vest therapy to aid with pulmonary hygiene. He has not required re-admission in 6 months, and is currently undergoing evaluation for more definitive airway therapy at a tertiary care center.

Discussion

Wheezing is a common exam finding characterized by high-pitched, musical respiratory sounds. It is often associated with asthma or chronic obstructive pulmonary disease (COPD). Wheezing occurs when air moving through a narrowed airway lumen creates resonant vibrations and sound waves. While small airways are most often affected, it is important for clinicians to recognize that this can occur anywhere along the respiratory tract, from terminal bronchioles to the oropharynx. When wheezing is refractory to usual therapy, a broader differential is necessary.

In this patient, tracheobronchomalacia (TBM) was initially overlooked. TBM refers to the weakening or collapse of the tracheal and bronchial cartilaginous structures, resulting in airway obstruction and respiratory symptoms including wheezing, dyspnea, cough, and recurrent respiratory infections.[1] It can be acutely exacerbated by respiratory infections, or present chronically with underlying conditions. Early identification of TBM is important as prolonged steroids can further weaken cartilaginous structures, causing progressive airway collapse, and further predisposition to infections.[2]

TBM can be difficult to diagnose as routine XRs are generally nondiagnostic. Pulmonary function testing will typically suggest obstruction but can be difficult to distinguish from other obstructive lung conditions. Flow volume loop morphology will also depend on the pattern and severity of airway collapse. Flexible bronchoscopy has traditionally been considered the gold standard for diagnosis as it allows for real-time assessment of airway properties.[3] Paired inspiratory and expiratory CT scans allow static visualization of airway collapse. Finally, ultrafast multidetector CT scanners with dynamic airway CT allow further asynchronous cine visualization.[4,5] These are currently less widely available.

Treatment strategies for TBM aim to maintain airway patency to allow pulmonary toilet and improve respiratory function. In mild cases, conservative measures such as positional maneuvers, humidified air, and bronchodilator therapy may provide symptomatic relief. More severe symptomatic cases may necessitate interventions tailored to the degree of airway collapse. High-flow nasal cannula (HFNC), continuous positive airway pressure (CPAP), NIPPV, or air pressure stenting may help non-invasively maintain airway patency, improve ventilation, and facilitate pulmonary toilet. At this time, there are no large-scale trials evaluating the long-term efficacy of these modalities, and much is left to clinical judgment. In our experience, other modalities including cough assist devices, intrapulmonary percussive ventilation (IPV),[6] and oscillatory vest therapy have also had varying degrees of success when combined with routine measures. Surgical options including tracheal stenting or tracheobronchoplasty are a last resort and generally limited to severe TBM that affects a limited tract of trachea.

This patient with Hunter syndrome developed severe TBM with atypical wheezing and was evaluated for less common etiologies. In TBM, steroids may need to be stopped or tapered to prevent further exacerbation of dynamic airway collapse. Paired inspiratory and expiratory or dynamic imaging may provide a reliable non-invasive diagnostic alternative to bronchoscopy. Various modalities of non-invasive positive pressure ventilation and pulmonary hygiene may help maintain airway patency and improve ventilatory function.

Figures

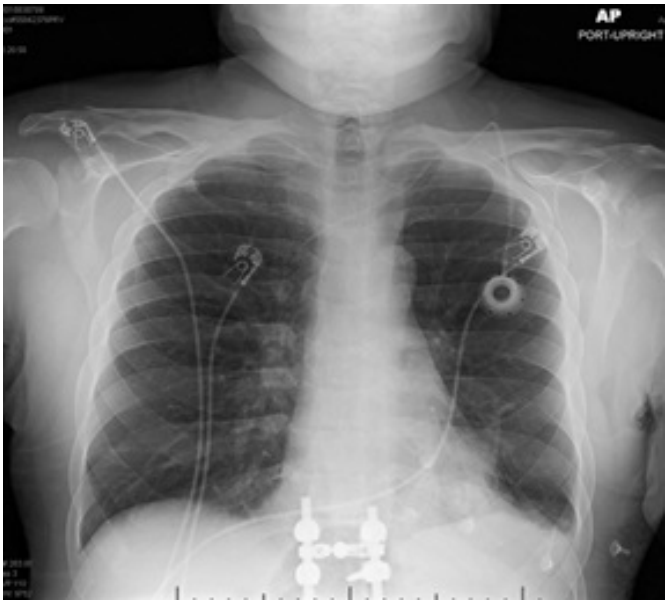

Figure 1.

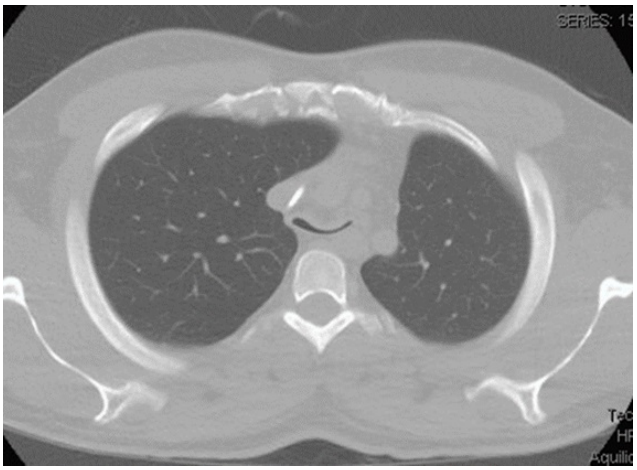
Figure 2.

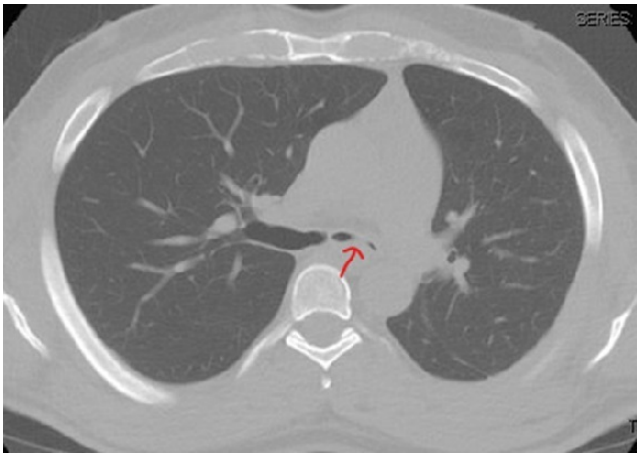
Figure 3.

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