CASE REPORT

Dyspnea on Exertion... An Asthma Masquerader!

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ABSTRACT

Children with asthma should not be exercise-limited and consulting with an expert is recommended if symptoms remain uncontrolled. We present a case of severe pulmonary hypertension (PH) that was misdiagnosed with asthma for several years based on dyspnea on exertion. We emphasize the role of the pediatrician in providing asthma education to the families – as well as to the patients with asthma to improve the quality of life of patients and avoid missing fatal diagnoses with detrimental outcomes that masquerades asthma.

CASE PRESENTATION

Our patient is a 13-year-old African American girl who was diagnosed with asthma for several years (almost five years) prior to presentation to our institution. Her diagnosis was based on dyspnea on exertion, and she was treated with inhaled β-agonist as needed for her symptoms. Following involvement in a minor motor vehicle accident as a passenger in the back seat, she presented to the emergency department with left-sided chest pain. Physical exam showed weight: 32 kg (< 3rd), temperature: 97.7F, heart rate: 89 bpm, respiratory rate: 23 breaths per minute, oxygen saturation 85% on room air and blood pressure: 100/75 mmHg. Chest examination was positive for decreased air entry bilaterally and signs of respiratory distress with mild suprasternal retractions. There was no evidence of trauma on the chest wall. Cardiac exam with single loud S2 and normal pulses. Musculoskeletal exam was positive for clubbing of the fingers and toes and kyphoscoliosis. The rest of the exam including the abdomen, skin, and genitourinary systems was unremarkable. Due to the above respiratory findings, a chest radiograph was performed and revealed severely enlarged pulmonary arteries suggestive of elevated pulmonary artery pressure. A Computed tomography (CT) [Figure-1] of the chest confirmed this finding with moderate-severe airway compression at the level of mid-distal trachea and ruled out interstitial lung disease and abnormal pulmonary venous system. An echocardiogram showed severely decreased right ventricular (RV) systolic function, severely dilated pulmonary artery, and tricuspid valve regurgitation with jet peak gradient consistent with elevated RV pressures, all suggestive of chronic severe PH. The patient was admitted to the pediatric cardiac intensive care unit and immediate therapy was started

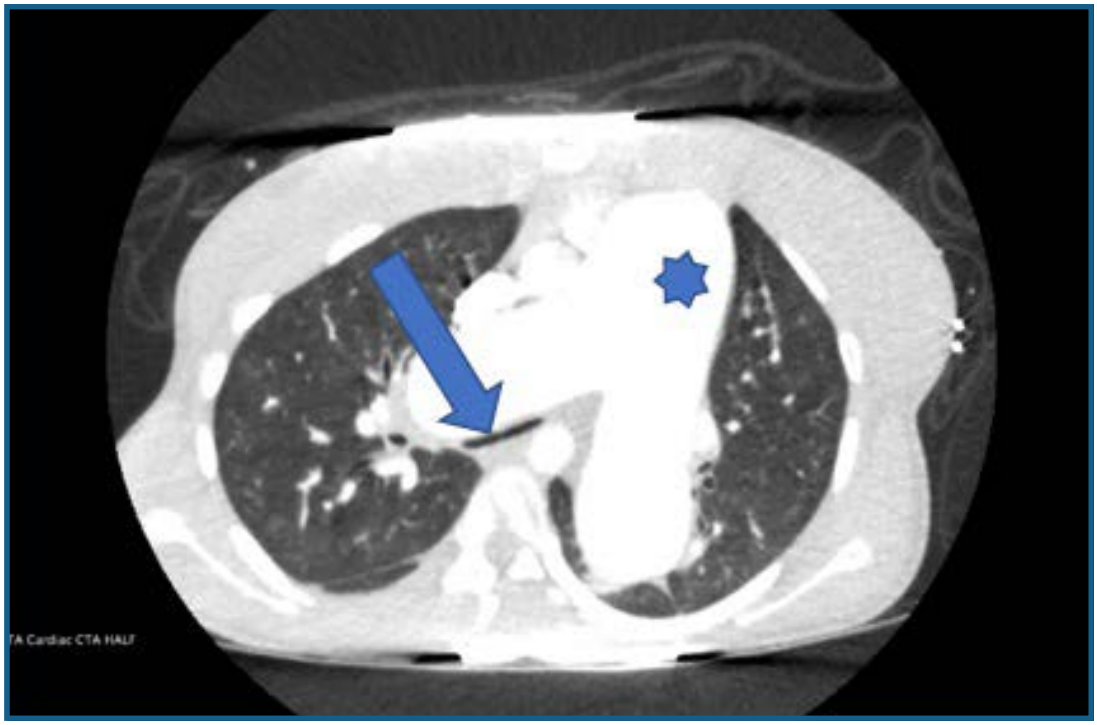

Figure 1: CT scan of the chest with IV contrast showing severely dilated pulmonary artery (star) with evidence of airway compression (arrow).

SIX MINUTE WALK TEST	
Gender	Female
Age	13
Indication/Diagnosis	PAH
Supplemental Oxygen	Y
O2 Flow (L/M)	4
O2 Device	Nasal Cannula
Baseline SPO2 %	92
Baseline Dyspnea	0
Baseline Fatique	0
Baseline Heart Rate	112
Baseline BP	92/54
End Of Test SPO2 %	83
End Of Test Dyspnea	3
End Of Test Fatique	3
End Of Test Heart Rate	145
End Of Test BP	104/70
Stopped Or Paused Before 6 Minutes	Y
Reason For Stopping	Other
Total Distance Walked In 6 Minutes (Ft)	259
Total Distance Walked In 6 Minutes (M)	78.94

Figure 2: 6-Minute-Walk test showing extreme limitation of physical activity

with Oxygen (FiO2 100%), inhaled nitric oxide and milrinone infusion. Following stabilization, right heart catheterization (RHC) confirmed severely increased pulmonary artery pressure at 90 mmHg and supra-systemic RV pressure at 120 mmHg with negative acute vasoreactivity test. PH-targeted therapy was started including treprostinil infusion, ambrisentan, and tadalafil. Comprehensive PH workup including autoimmune and thromboembolic diseases was negative. PH genetics revealed a mutation of unknown significance in the KCNK3 gene. Functional classification with 6-minute walk test was severely impaired [Figure-2]. Severe (Class IV) Primary PH was diagnosed, and the patient is currently being evaluated for lung transplantation.

DISCUSSION

Asthma clinical practice guidelines recommend medical history and physical examination to establish the diagnosis of asthma and determine the presence of episodic airflow limitation.[1] Spirometry is recommended to confirm the diagnosis of asthma in children > 5 years of age, and more advanced pulmonary function tests are currently available at specialized pulmonary centers for children younger than 5 years, including impulse oscillometry and measurement of airway resistance.[2] Imaging is not recommended in the initial diagnostic process or in severe exacerbations unless complications are expected or in the exclusion of other comorbid conditions such as the use of chest X-rays to rule out pneumothorax. However, evidence is available to highlight the utilization of lung imaging in chronic management of the disease.[3] An example is the use of computed tomography (/CT) scans of the chest to aid in the diagnosis of conditions that might be associated with chronic severe asthma, such as bronchopulmonary aspergillosis, eosinophilic pneumonia and eosinophilic granulomatosis with polyangiitis.[4] In addition, growing research is now focusing on identifying specific radiological measures (including airways diameter and geographic hyperinflation on CT scans) to assess severe asthma and correlate that with pulmonary function test findgins, hoping to identify a unique group of patients who may benefit from certain therapies in the future.[3] The lack of evidence of airflow limitation or failure to respond to initial therapy mandates considering alternative diagnoses and early consultation with a pediatric pulmonary specialist.

Our patient was misdiagnosed for several years, and her exercise limitation was incorrectly attributed to asthma. Her physical exam findings including the severely decreased weight and the presence of clubbing as well as her echocardiogram findings of impaired RV systolic function affirm the long-standing history of her condition.

Exercise is a well-recognized major trigger for asthma, and many patients seek medical advice due to experiencing dyspnea on exertion. However; activity should not be limited in asthma patients, and goals of treatment should include symptom-control and improvement of the quality of life for asthma patients as highlighted by the National Asthma Education and Prevention Program (NAEEP expert panel report-3)[1] and the Global Initiative of Asthma Guidelines (GINA).[5] In our case, the limited understanding of the pathophysiology of the disease, and the lack of understanding of the goals of asthma therapy, lead to the serious misconception by the patient as well as her family that patients with asthma "cannot exercise" which consequently resulted in acceptance of her limited endurance and reduced physical capacity.

There is general agreement that asthma education has positive outcomes on symptom control as well as on other outcomes, including the rate of hospitalization and ER visits for asthma, unscheduled doctors' visits, costs and improvement in quality of life[6], with better outcomes for programs that provided self-management skills. As a result, repeated assessment of families' and patients' perceptions of the disease is an integral component of asthma care visits. The pediatrician is encouraged to utilize educational tools and strategies to deliver the information at different levels of health literacy to caregivers; an example is asthma action plans that became an essential part of a pediatric asthma visit. Other tools of education include more interactive learning that incorporates either individual or group learning.

PH is rare in pediatrics and is associated with considerable morbidity and mortality that can affect all age groups from the newborn period to infancy and childhood. Symptoms of PH are commonly nonspecific[7], and the diagnosis may be missed in the early disease stages. Exertional dyspnea and progressive fatigue are the most frequent complaints in the older child. Special considerations should apply to the child with exertional dyspnea that is not responding to asthma management and a lower threshold for investigating the child with uncontrolled asthma is recommended with early referral to a specialist.

CONCLUSION

Pediatricians are encouraged to educate patients and caregivers about asthma and utilize the widely available variable tools and strategies to provide an understanding of the expected symptoms, disease progression, and goals of care.

Timely referral to a specialist is mandatory when symptoms remain uncontrolled. PH is rare in pediatrics but with considerable mortality and morbidity and should be considered in pediatric cases of dyspnea on exertion.

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