

CASE REPORT

Refractory Asthma Coexisting with a Double Aortic Arch: A Case Report and Literature Review

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Asthma that is poorly controlled despite adequate treatment should be investigated for medication compliance, treatment adherence, environmental control, asthma mimics, and comorbidities especially in infants and young children. Here, the case of a 2-year-old child with refractory asthma coexisting with a double aortic arch is reported.

Keywords asthma, children, comorbidity, double aortic arch

INTRODUCTION

Although the majority of patients with asthma can be well controlled with low-dose anti-inflammatory agents and occasional rescue medication, some patients with asthma will not be well controlled on standard medications. For those who remain uncontrolled despite adequate anti-inflammatory therapy, careful re-evaluation is required. This may include confirming the diagnosis of asthma, investigating compliance/adherence with medications, evaluation for the presence of comorbidities that may aggravate asthma, and excluding asthma mimics (1–3).

CASE REPORT

A 2-year-old female is presented with a history of recurrent wheezing and coughing that started at 1 year of age. She had continued episodes of wheezing triggered by viral respiratory infections and exercise; subsequently, the patient was referred to our center. There was no history of exposure to passive tobacco smoking and pets. She had suffered from mild atopic dermatitis. Her mother had a history of allergic rhinitis and younger brother had atopic dermatitis.

The patient was sensitized to *Dermatophagoides pteronyssinus* (17.4 KU/L) and *Dermatophagoides farinae* (29.5 KU/L), both measured by the UniCAP system (Pharmacia, Uppsala, Sweden). The response to a short-acting β_2 -agonist was good, and the PC20 was 3.70 mg/dL to the methacholine challenging test using trachea/chest auscultation with transcutaneous oximetry. The chest X-ray did not show any specific findings; the patient was diagnosed with asthma. Despite

adequate treatment with budesonide (1000 μ g/day) and montelukast, good compliance/adherence with the medications, and adequate environmental control measures, the asthma was poorly controlled and often severely exacerbated when combined with a viral infection.

Recently, the patient was hospitalized because of a severe pneumonia caused by adenovirus type 3. Tachypnea and dyspnea developed. Stridor and wheezing were auscultated in both lung fields. The chest radiographs demonstrated a narrow distal trachea (Figure 1) and a three-dimensional computed tomography scanning showed a double aortic arch (Figure 2). There was no associated cardiovascular anomaly.

The patient was successfully treated with a short-acting β_2 -agonist, budesonide, and intravenous aminophylline during the admission. Surgery is planned to repair the anomaly.

DISCUSSION

Diagnosis of asthma in children 5 years of age and younger is particularly difficult because episodic wheezing and coughing are common in this age group (3, 4). Therefore, the diagnosis of asthma has to be based largely on clinical judgment and an assessment of the symptoms and physical findings. A useful method for supporting the diagnosis of asthma in this age group is the response to a trial of short-acting β_2 -agonist and inhaled corticosteroids. Marked clinical improvement during the treatment and deterioration when it is discontinued supports the diagnosis of asthma (4).

Bronchial hyperresponsiveness (BHR) is a characteristic feature of asthma (3). BHR is usually measured by bronchial challenging test using direct or indirect stimuli such as methacholine, histamine, cold air, or exercise. The measurement of bronchial responsiveness is a potentially useful diagnostic tool in both children and adults. However, lung function measurement in children less than 5 years of age is technically difficult.

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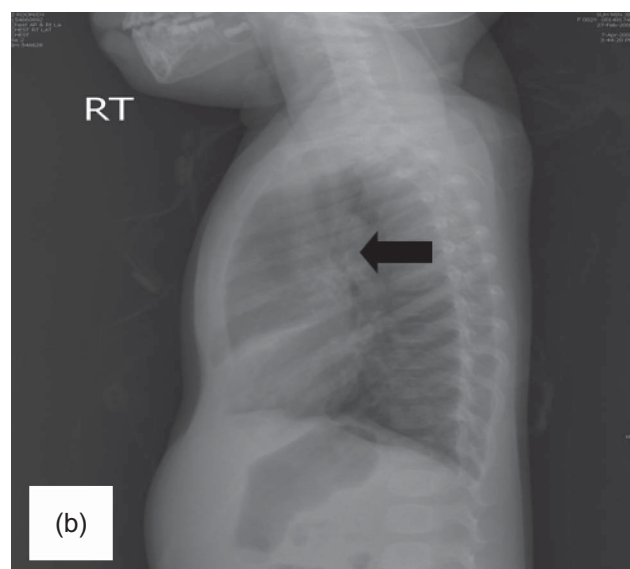
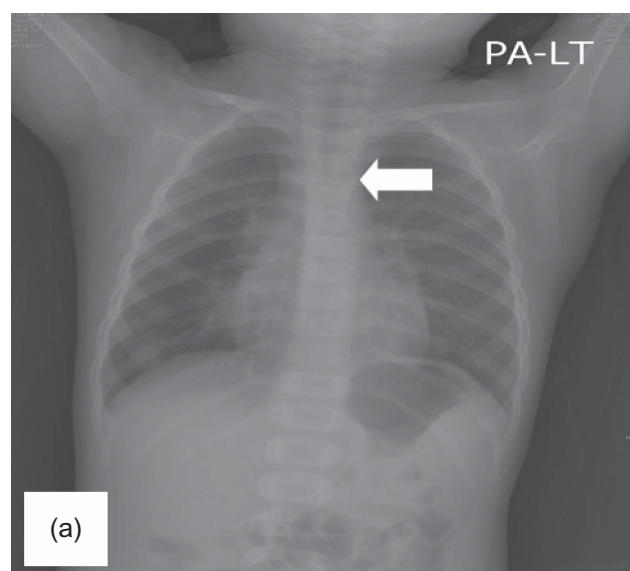


FIGURE 1.—(a) Chest X-ray (AP view) showing narrowed distal trachea (white arrow). (b) Chest X-ray (lateral view) showing narrowed distal trachea (black arrow).

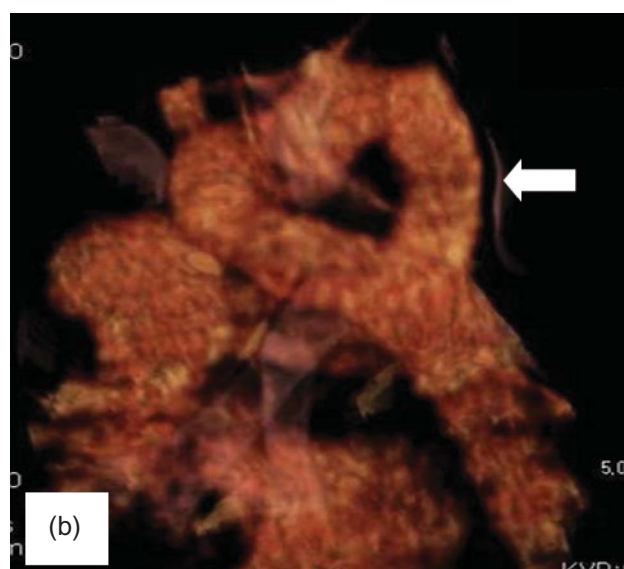
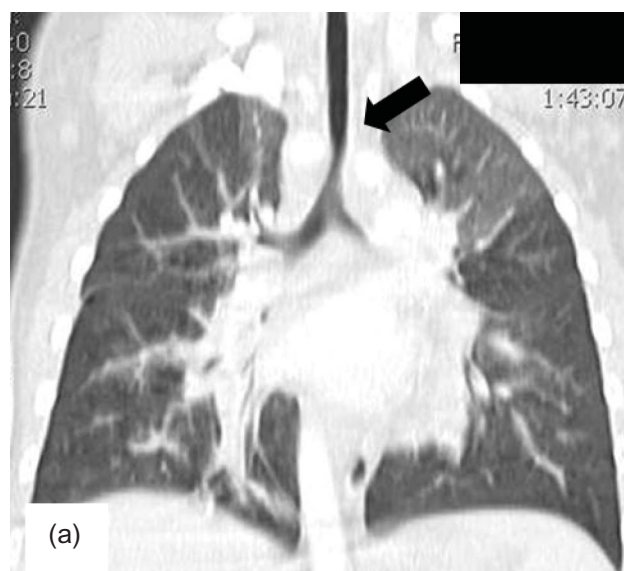


FIGURE 2.—(a) Contrast-enhanced computed tomography showing compression of the trachea by the double aortic arch (black arrow). (b) Three-dimensional computed tomography reconstruction showing the double aortic arch (white arrow).

Tracheal/chest auscultation with transcutaneous oximetry is suggested to measure the provocative concentration of methacholine which induces audible wheezing and is a sensitive and relatively safe bronchial challenge test for this age group (5, 6).

Recurrent episodes of cough, wheezing, chest tightness, and dyspnea are most often due to asthma in both children and adults. However, any conditions that narrow the intrathoracic airways or increase airway resistance can cause similar symptoms and must be considered in the differential diagnosis of asthma. Because conditions that mimic asthma could be misdiagnosed as asthma as well as coexist with asthma in refractory asthma, children with asthma poorly controlled by adequate treatment that have good adherence and compliance to medication protocols and adequate environmental control require a careful re-evaluation of the cause of their symptoms (2–4).

Comorbidities or asthma mimics include upper airway diseases (e.g., chronic rhino-sinusitis), gastroesophageal reflux, cystic fibrosis, bronchopulmonary dysplasia, congenital malformation causing narrowing of the intrathoracic airways (e.g., vascular ring and sling, enlarged cardiac chamber, lymph nodes enlarged by tuberculosis or lymphoma, tracheomalacia, bronchomalacia, and complete cartilage rings, cysts, webs), extrathoracic obstruction (e.g., tracheomalacia, adenotonsillar hypertrophy, and posterior deviated huge uvula), a foreign body in trachea or bronchus, primary ciliary dyskinesia syndrome, immune deficiency, congenital heart disease, vocal cord dysfunction (VCD), and bronchiectasis (4).

VCD is a functional disorder of the vocal cords, characterized by the inappropriate adduction of the vocal cords during inspiration. Dyspnea, inspiratory stridor, and wheezy respiration occur spontaneously or with exercise.

Direct visualization of vocal cords while the patient is symptomatic is the gold standard for the diagnosis (7).

Tracheomalacia is an anatomical and functional disorder characterized by flaccidity of the supporting tracheal cartilage, widening of the posterior membranous wall, and reduced anterior–posterior airway caliber. These factors cause tracheal collapse against the posterior wall during times of increased airflow such as coughing, crying, or feeding. The collapse occurs during inspiration and causes stridor in intrathoracic type. However, in extrathoracic type, the collapse occurs during expiration and causes wheezing. Gold standard for diagnosis is flexible bronchoscopy with conscious sedation and spontaneous respiration (8). Bronchomalacia is a similar disorder in main-stem bronchus.

A vascular ring is a rare cause of unexplained respiratory symptoms in infants and young children. The resulting compression of the trachea and esophagus may cause symptomatic obstruction of these structures, or patients may be completely asymptomatic. Patients with vascular rings may present with severe symptoms of respiratory distress soon after birth or may be asymptomatic for life (9). *Vascular rings* are more commonly associated with stridor in infancy; however, symptoms may develop in older children. Although inspiratory stridor is a classic sign of intrathoracic obstruction, it is not always clearly present. Some studies report that wheezing and stridor are the most common symptoms associated with vascular rings (10, 11).

Delayed diagnosis of a vascular ring can result in unnecessary investigations and prolonged periods of ineffective treatment. Prolonged airway compression may also be harmful. Long-standing compression of the trachea increases the risk of complications such as secondary tracheomalacia (12).

In conclusion, consideration of alternative diagnoses is essential in the management of children with refractory asthma.

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