Kommerell’s Diverticulum With a Twist: A Case of Recurrent Wheeze in an 8-Year-Old Boy

Tiffany Amber Robles, BS1, Aditya Srinivasan, BS1, Lynnette Mazur, MD, MPH1, and Anand Gourishankar, MD1

Abstract
Kommerell’s diverticulum is a rare, congenital aortic arch anomaly, usually associated with other vascular abnormalities. When present with a concurrent right-sided aortic arch and an aberrant subclavian artery, this triad can form a vascular ring that encompasses the trachea and esophagus. This anatomical variant is usually asymptomatic but can present with respiratory symptoms due to compression of the trachea. In this report, we discuss a case of a Kommerell’s diverticulum, which presented as frequent and recurring asthma exacerbations in a pediatric patient.

Keywords
Kommerell’s diverticulum, right-sided aortic arch, wheezing

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Introduction
Kommerell’s diverticulum (KD) is an uncommon congenital vascular anomaly described as a dilation occurring at the origin of an aberrant subclavian artery. It is usually associated with a right aortic arch and is present in 0.05% to 0.1% of the population.1,2 Symptoms include dysphagia, wheezing, and shortness of breath. Most often, KD is an incidental finding; however, patients with symptoms can be misdiagnosed with asthma or asthma exacerbations.2 Rarely, patients present with rupture of the diverticulum or aortic dissection. We present an uncommon clinical case of congenital vascular anomaly in a pediatric patient with persistent right tracheal deviation and a history of asthma. We will discuss the pathophysiology, imaging, and management of this condition as a differential diagnosis that physicians, residents, and students should consider in patients with chronic dyspnea.

Case Report
An 8-year-old male presented to the emergency room with a 2-day history of shortness of breath and wheezing. He had a history of moderate-persistent asthma and had several emergency department visits over the past 2 years for similar complaints attributed to asthma exacerbation. He was on albuterol and inhaled corticosteroids for his persistent asthma, but he reported that it was not effective in relieving his exacerbation. He was admitted for acute asthma management due to increasing work of breathing and wheeze. He denied any dysphagia or odynophagia.

The patient’s growth was appropriate, respiratory rate at 40 breaths per minute, heart rate at 130 beats per minute, blood pressure of 90/60 mm Hg, and pulse oximetry of 95% in room air. The rest of the physical examination was unremarkable.

On admission, his frontal and lateral chest X-rays showed only an incidental finding of right tracheal deviation. His previous chest X-rays showed persistent right tracheal deviation, which was not of prior concern due to its mild nature. A transthoracic echocardiogram revealed mild tracheal compression of the left atrium just above the bifurcation, at the level of descending aorta, either by the aorta or the surrounding structures. A contrast-enhanced computed tomography (CT) scan...
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of the chest demonstrated a right-sided aortic arch with aberrant left subclavian artery and KD, mild tracheal deviation, and moderate esophageal deviation and compression (Figures 1 and 2). Pulmonary function tests read mixed an obstructive/restrictive pattern, and barium swallow demonstrated posterior narrowing of the proximal thoracic esophagus. Esophageal compression was likely secondary to right-sided aortic arch with aberrant left subclavian artery sling. With these findings on imaging, foreign body aspiration, tracheomalacia, retropharyngeal abscess, cysts, neoplasms, and bony dysplasia were subsequently ruled out.

Our patient’s respiratory distress improved with albuterol and systemic steroids. We discharged him with an increase in his albuterol and inhaled corticosteroid doses and a course of prednisolone. He had an outpatient follow-up for surgical intervention (left thoracotomy division of the vascular ring) of his KD.

Discussion

Kommerell’s diverticulum is a scarce anatomical variation, referring to the bulbous configuration of the origin of an aberrant left subclavian artery in the setting of a right-sided aortic arch. The aberrant subclavian artery courses posterior to the esophagus in 80% of the cases and may be asymptomatic or may cause symptoms due to mass effect on the trachea and esophagus. Left tracheal deviation is commonly noted due to the tension of the left subclavian artery pulling the right-sided aortic arch leftward. An embryologic remnant of the left fourth aortic arch, which did not undergo complete involution, presents posteriorly and creates a vascular ring that compresses the trachea causing shortness of breath and wheezing. Interestingly, our patient presented with right-sided tracheal deviation, hypothesized to be due to mass effect secondary to the presence of the expanding KD.

Table 1. Edwards Classification of Right-Sided Aortic Arch.

| Type | Definition                  | Prevalence (%) |
|------|-----------------------------|----------------|
| I    | Mirror image aortic arch    | 59             |
| II   | Aberrant left subclavian artery | 39.5          |
| III  | Isolated subclavian artery  | 1.5            |

Edwards classification of a right-sided aortic arch are categorized into 3 groups based on anatomy and associated anomalies, seen in Table 1. Our patient presented with a type II variant, which involves a right-sided aortic arch with aberrant left subclavian artery and associated KD. These classifications play an important role in identifying anatomic variability to guide future surgical intervention.

Seldom, Kommerell lesions reach large dimensions and cause symptoms of severe compression. In the pediatric population, respiratory symptoms predominate due to the pliability of the immature tracheal rings. This cartilage portion of the airway is soft and flexible until calcification of the larynx and trachea occurs in
the teenage years. In adults, however, symptoms most commonly present after age of 40 years when the aortic wall is atherosclerotic and rigid, with dysphagia being most prevalent.

A thorough history and physical examination differentiates vascular rings from the other possible diagnoses when evaluating a patient with dyspnea and can help identify any comorbidities. In our case, our patient’s history of moderate persistent asthma and recurrent respiratory distress undeterred by ideal medical management led to additional imaging and the discovery of a comorbid condition. Other comorbidities such as eczema, skin allergies, and gastroesophageal reflux disease have been reported more frequently in patients with asthma and, on treatment, can improve respiratory symptoms.

The optimal choice for imaging has not been studied or established. Radiographs are often done initially, due to their accessibility and utility in identifying vascular abnormalities. In a study done to evaluate radiographic abnormalities in patients with a symptomatic vascular ring, Pickhardt et al found that the combination of both frontal and lateral X-rays displayed at least one abnormality in each patient, attesting to the importance of obtaining a 2-view X-ray on suspicion of a vascular ring.

Barium esophagography is performed in patients with dysphagia, while bronchoscopy is used to evaluate children with respiratory symptoms to rule out trauma or foreign body aspiration. Cardiac magnetic resonance imaging is a radiation-free technique that may provide a safe alternative in the evaluation of structural abnormalities of the heart, vascular dilations, or compressions. The most commonly used imaging modality is CT, which allows physicians to visualize the patient’s unique vascular anatomy and provide the details necessary for the planning of future surgical intervention. These imaging tools altogether provide a sound way to diagnose KD before deleterious complications arise.

If left undiagnosed, complications of an unmanaged diverticulum may include aneurysm rupture, which could be fatal, aortic dissection or recurrent pneumonia. Surgical treatment is recommended for patients with a large asymptomatic diverticulum or symptomatic patients, as in the case of our patient. For pediatric patients, Backer et al proposed that any diverticulum more than 1.5 times the diameter of the subclavian artery is worthy of surgical intervention due to the potential for rupture.

**Conclusion**

In conclusion, this rare case demonstrates that anatomical abnormalities consistent with vascular rings can compress the airway and present as increased frequency of asthma despite optimal medical management. Differential diagnosis of recurrent wheeze in children is warranted. While no imaging modality is the gold standard, CT scans are reliable and allow proper preoperative planning. KD is one such lesion that can present in young children and if symptomatic, must be managed operatively.

**Author Contributions**

TAR: Contributed to acquisition, analysis, and interpretation; drafted manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.

AS: Contributed to acquisition, analysis, and interpretation; drafted manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.

LM: Contributed to conception and design; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.

AG: Contributed to conception and design; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.

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**Ethics Approval**

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**Informed Consent**

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**ORCID iDs**

Aditya Srinivasan https://orcid.org/0000-0003-1753-006X

Anand Gourishankar https://orcid.org/0000-0003-4344-7032

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