Two children referred for cardiac imaging were found to have an unusual form of a vascular ring. The first patient was an asymptomatic 10-year-old girl with a membranous ventricular septal defect, prolapse of the right coronary aortic cusp, and mild aortic regurgitation. Her past history was notable for gastroesophageal reflux disease as an infant, which had prompted a barium swallow at a few months of age demonstrating a posterior compression defect on the esophagus. Detailed imaging of the thoracic vasculature was not pursued at the time given the lack of respiratory symptoms and resolution of her reflux. She was now referred for cardiac magnetic resonance imaging to quantify the degree of left-to-right shunt, aortic regurgitation, and left ventricular dilation. Cardiac magnetic resonance demonstrated a small membranous ventricular septal defect with pulmonary-to-systemic flow ratio of 1.2, mild aortic regurgitation, and a mildly dilated left ventricle with normal systolic function. A right aortic arch was incidentally detected with branches arising in the following order: proximal to distal: right common carotid artery, right subclavian artery, and left innominate artery. The left innominate artery originated from a prominent diverticulum of Kommerell off the proximal descending aorta and bifurcated into the left subclavian and left common carotid arteries (Figure 1). The trachea was not compressed. Given lack of symptoms and a normal pulmonary function test, no further intervention was recommended.

The second patient was a 4-year-old boy with developmental delay, single kidney, and history of recurrent respiratory infections, asthma, and exercise intolerance. He had undergone a tonsillectomy. A barium swallow study obtained to evaluate noisy breathing and cough revealed posterior compression of the esophagus. Computed tomographic angiography of the chest demonstrated a right aortic arch with aberrant origin of the left innominate artery from a large diverticulum of Kommerell off the proximal descending aorta (Figure 2). The distal trachea and proximal right mainstem bronchus were compressed, and a left-sided ligamentum arteriosum completing a vascular ring was suspected. Given his respiratory symptoms and airway compression by the vascular ring, the patient underwent division of the vascular ring by video-assisted thoracoscopic surgery. Intraoperative findings confirmed the preoperative anatomic diagnoses, and the patient was discharged from the hospital the following day.

Figure 1. Graphic illustration of this anomaly with corresponding cardiac magnetic resonance turbo spin-echo images in the axial plane at various levels of the arch obtained from patient 1. AAo indicates ascending aorta; DAo, descending aorta; DK, diverticulum of Kommerell; Esoph, esophagus; LCCA, left common carotid artery; LIA, left innominate artery; LPA, left pulmonary artery; LSCA, left subclavian artery; Lig, ligamentum arteriosum; RCCA, right common carotid artery; RPA, right pulmonary artery; SVC, superior vena cava; *, trachea; and arrow, left innominate vein.
A vascular ring is an aortic arch anomaly in which the trachea and esophagus are completely surrounded by vascular structures. The most common forms are double aortic arch and a right aortic arch with an aberrant left subclavian artery from a diverticulum of Kommerell. The arch anomaly described here—right aortic arch with an aberrant left innominate artery from a diverticulum of Kommerell—is rare, and, to our knowledge, only 10 such cases have been reported in the literature. The earliest such report was in 1968, and, since then, the anomaly has been reported both in isolation and in conjunction with other congenital cardiac anomalies. This type of vascular ring is thought to arise from interruption of the embryonic left aortic arch between the ascending aorta and the left common carotid artery (Figure 3). A complete vascular ring is formed by the pulmonary arteries anteriorly, the aortic arch on the right, the diverticulum of Kommerell posteriorly, and either a left-sided ductal ligament or an atretic aortic arch segment between the ascending aorta and the left common carotid artery on the left.

Vascular rings can be diagnosed by several imaging modalities, including chest radiography, bronchoscopy, barium swallow, echocardiography, cardiac magnetic resonance, and computed tomographic angiography. Although each diagnostic technique is capable of establishing the diagnosis of a vascular ring, surgical division of the ring, especially by video-assisted thoracoscopic technique, requires detailed knowledge of the anatomy. As illustrated by the patients described in this report, cardiac magnetic resonance or computed tomographic angiography is usually required for complete anatomic assessment of the vascular ring and the airways. Exceptions to this are infants with a double aortic arch in whom echocardiography fully demonstrates the vascular structures. As with other vascular rings, the decision for surgical intervention is generally determined by symptoms of proximal airway obstruction or esophageal compression. The patients described in this report illustrate the spectrum of clinical manifestations of vascular rings; the first patient had no tracheal compression and was asymptomatic, requiring no intervention, whereas the second patient had tracheal compression with corresponding symptoms and underwent surgical division of the vascular ring.

Disclosures

None.

References

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