

Case Reports

Double Aortic Arch – A Case Study

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Abstract:

Double aortic arch (DAA) is one of the 2 most common forms of vascular ring, a class of congenital anomalies of the aortic arch system in which the trachea and esophagus are completely encircled by connected segments of the aortic arch and its branches. The aim of this study is to describe a case of DAA in a middle aged person. A 40 year old male came to outdoor patient department with cough and breathlessness since childhood which was diagnosed earlier as bronchial asthma, cold allergy, and dust allergy. The final diagnosis DAA was made after CT angiogram. Establishing a diagnosis of DAA in a middle aged person requires thorough understanding and clinical skills in performing steps.

Key Words :
Arch of the aorta, Vascular ring, CT angiogram

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Introduction:

Vascular ring is a congenital anomaly in which the trachea and esophagus (or its atretic remnant) are surrounded by vessels. Vascular rings, which constitute less than 1% of congenital heart diseases, were first identified by Gross in 1945.¹ Double aortic arch is the most frequently encountered vascular ring malformation characterized by a complete encirclement of trachea and esophagus by the aortic arch. Although the double aortic arch has various forms, the common defining feature is that both the left and right aortic arches are present.^{2,3}

Diagnosing DAA is challenging because it has a wide clinical spectrum. Extrinsic compression of the airway by vascular rings causes respiratory symptoms and/or feeding abnormalities that usually result in prompt diagnosis during early infancy. However, there are occasions when DAA remain undiagnosed.⁴

We report a case of a middle aged man with DAA which is diagnosed after doing a CT angiogram for chronic cough and dyspnea.

Case presentation:

Mr. J, a 40 years old male, came to OPD with the complaints of cough and breathlessness since childhood. His mother also had dust allergy, so he was given a diagnosis of bronchial asthma. Over the time treatment with salbutamol, fluticasone,

montelukast failed to benefit. When he grew older he was treated for gastro-oesophageal reflux disease resulting in no benefit. He continued to have episodes of coughing triggered by cold weather and viral respiratory infections, and was diagnosed with asthma again. His physical examination was insignificant except for tachycardia. Chest auscultation revealed no abnormality. Laboratory findings were all within the normal range. Chest radiograph showed slight tracheal compression and ECG was normal. Then Echocardiography was done showing normal features. Later Chest CT scan was advised and demonstrated compression of trachea by vascular structure consistent with anomalous arch of aorta as well as some nodular shadows in both lower lobes. To confirm the diagnosis CT angiogram was performed revealing double arch of aorta with right and left arch encircling the trachea and oesophagus. Right subclavian and right common carotid arteries arise from the right arch and left subclavian and left common carotid arteries arise from the left arch. He was referred to a cardiac surgeon.

Discussion:

Double aortic arch is the most common symptomatic vascular ring, accounting for 50–60 % of vascular rings.¹¹ Developmentally it is caused by persistence of both the right and left fourth aortic arches and the right and left dorsal aortae.¹² Classically, DAA

has three types; right dominant aortic arch, left dominant aortic arch, and balanced-type aortic arch. In 75% of the cases, the right arch is dominant, whereas the left arch is dominant in approximately 20% of the cases, and the remaining 5% of cases, both arches are equally dominant.^{2,3}

Double aortic arch usually occurs without associated cardiovascular anomalies. It is associated with a chromosome band 22q11 deletion in approximately 20% of patients. Occasionally, patients with double aortic arch may have anomalies consistent with either vertebral, anal, cardiac, tracheal, esophageal, renal, and limb (VACTERL) or posterior coloboma, heart defect, choanal atresia, retardation, genital, and ear (CHARGE) associations.

Presentation of symptoms in patients with double aortic arch depends on several factors, including the severity of tracheal compression, esophageal compression, or both and whether associated anomalies are present.

Breathing symptoms include:

- High-pitched sound during breathing (stridor).
- Noisy breathing.
- Repeated pneumonias.
- Wheezing.

Digestive symptoms may include:

- Choking.
- Difficulty eating and swallowing.
- Vomiting.

Physical findings can vary, often in accordance with the patient's history. Newborns with associated anomalies may have no evidence of a vascular ring on physical examination, but this situation is the exception because most patients have readily recognizable physical signs. The classic sign of double aortic arch and of vascular rings in general is nonpositional stridor; however, many young infants with double aortic arch have adventitious expiratory breath sounds, as well as the characteristic inspiratory stridor. Respiratory findings typically do not improve with nebulized bronchodilator therapy and usually are more prominent with agitation or crying.

In differentiating double aortic arch from upper or lower respiratory infections, a WBC count and respiratory viral studies may be helpful. However, because patients with double aortic arch are

predisposed to respiratory infections, the diagnosis of an infection does not exclude the possibility of double aortic arch. Also no characteristic ECG findings are associated with double aortic arch, and ECG findings are usually normal, except in patients with associated cardiovascular anomalies. However, in most patients, the diagnosis of double aortic arch can be made reliably based on echocardiography; helpful information is obtained with suprasternal, high parasternal, and subcostal imaging. However, because patients with a vascular ring typically present with respiratory symptoms, the diagnosis is usually made based on other imaging modes.

Vascular rings can be identified by several imaging examinations and occasionally multiple imaging tests may be required to make a diagnosis.⁸ A radiograph is often the initial imaging test and some abnormalities are found in almost all patients with vascular rings.⁹ Arch laterality may be inferred from the anteroposterior (AP) radiograph by the pattern of indentation of the tracheal air column, which is from the right in a right arch, left in a left arch and bilateral in a double arch. On the lateral view, tracheal narrowing may be apparent. Pulmonary hyperinflation may occur with a pulmonary sling. Presence of a right arch with tracheal compression is highly suggestive of a ring. The location of the descending thoracic aorta can be inferred from the paraspinal line and azygo-oesophageal recess. Barium oesophagography is often performed in children with feeding difficulties. The specific type of vascular ring can often be diagnosed based on the pattern of oesophageal indentation on the oesophagram in combination with the pattern of tracheal indentation on the radiograph.¹⁰

MRI is one of the two commonly used imaging modalities diagnosing and characterizing vascular rings. Advantages of MRI include a wide field of view, multiplanar imaging capabilities and adequate spatial resolution to detect vascular ring and associated airway anomalies, without the use of ionizing radiation or iodinated contrast material. Disadvantages of MRI include limited availability, the long acquisition times and the need for deep sedation or general anaesthesia in young children, which may involve high risks in patients with airway compromise.

CT has emerged as the preferred imaging examination for the diagnosis and characterization

of vascular rings. It is performed in symptomatic patients with a suspected vascular ring in other imaging modalities to delineate the anatomy and help surgical planning. Advantages of CT include the rapid acquisition time without the need for sedation or general anaesthesia; high spatial and temporal resolution; large field of view; isotropic voxels with multiplanar reconstruction capabilities; and simultaneous evaluation of the vasculature, airways and, to a lesser degree, the oesophagus. The 3D volume-rendered and shaded surface display images can be helpful for surgical planning and depicting the anomalous anatomy. Ionizing radiation and the use of potentially nephrotoxic iodinated contrast material are the primary disadvantages. Moreover, CT angiography (CTA) for the evaluation of vascular rings can be performed without any sedation and with quiet breathing since the latest CT scanners have fast gantry rotation times and high z axis coverage, as a result of which artifacts are minimal.

Medical care prior to surgical repair of double aortic arch depends on the clinical presentation. In most patients, only supportive care is required. Catheter interventions are not used in the management of double aortic arch. Surgical division of the vascular ring is indicated in any patient with symptoms of airway or esophageal compression and in patients undergoing surgery for repair of associated cardiovascular or thoracic anomalies. The fundamental principle of surgical management of double aortic arch is division of the ring to relieve compression of the trachea and esophagus. In general, this is achieved by dividing the minor arch through an ipsilateral thoracotomy.

Conclusion:

Vascular rings are complex and diagnosis is most often daunting because of variable and non-specific clinical presentations. CT angiography plays an important role in the identification and definition of the anatomy of these complex anomalies, thus providing a roadmap to surgeons. Careful analysis

of the arch laterality, branching pattern and position of the ductus or ligamentum is essential for accurate characterization.

Conflict of Interest - None.

References:

1. Gross, R.E. (1955) Arterial Malformations Which Cause Compression of the Trachea or Esophagus. *Circulation*, 11, 124-134. <http://dx.doi.org/10.1161/01.CIR.11.1.124>
2. Kau T, Lesnik G, Eicher W, Sinzig M, Gassez J, Rabitsch E, *et al.* Aortic Development and Anomalies. *Semin Intervent Radiol* 2007; 24: 141-152. <http://dx.doi.org/10.1055/s-2007-980040>
3. Baraldi, R., Address, S.S., Bigli, S. and Mannella, P. Vascular Ring Due to Double Aortic Arch: A Rare Cause of Dysphagia. *Eur J Radiol* 2004; 52: 21-24. <http://dx.doi.org/10.1016/j.ejrex.2004.06.001>
4. Umegaki T, Sumi C, Nishi K, Ikeda S, Shingu K. Airway Management in an Infant with Double Aortic Arch. *Journal of Anesthesia* 2010; 24: 117-120. <http://dx.doi.org/10.1007/s00540-009-0850-4>
5. Weinberg PM. Aortic Arch Anomalies. *Journal of Cardiovascular Magnetic Resonance* 2006; 8: 633-643.
6. Jonas RA. Comprehensive Surgical Management of Congenital Heart Disease. London: Arnold, 2004: 497.
7. Kirklin JW, Barratt-Boyes BG. Eds. Cardiac Surgery. 3rd ed. New York: Churchill Livingstone, 2003: 1415.
8. Browne LP. What is the optimal imaging for vascular rings and slings? *Pediatr Radiol* 2009; 39(Suppl 2):S191-S195. doi: 10.1007/s00247-008-1118-0.
9. Pickhardt P, Siegel M, Gutierrez F. Vascular rings in symptomatic children; frequency of chest radiographic findings. *Radiology* 1997; 203: 423-426. doi: 10.1148/radiology.203.2.9114098.
10. Neuhauser E. The roentgen diagnosis of double aortic arch and other anomalies of the great vessels. *Am J Roentgenol Radium Ther* 1946; 56: 1-12.
11. Siegal MJ. Multiplanar and three-dimensional multi-detector row CT of thoracic vessels and airways in the pediatric population. *Radiology* 2003; 229: 641-650. doi: 10.1148/radiol.2293020999.
12. Hernanz-Schulman M. Vascular rings: a practical approach to imaging diagnosis. *Pediatr Radiol* 2005; 35:961-979. doi: 10.1007/s00247-005-1529-0.