Trachea and Oesophagus Compression by DAA

Case Report

Om Shankar Chaurasiya, Ravanagomagan MG, Sapna Gupta, *Prayas Vats
Department of Paediatrics, *Department of Radiodiagnosis, MLB Medical College, Jhansi

(Received: August, 2016) (Accepted: June, 2017)

ABSTRACT

A vascular ring is an anomaly of the great arteries (aortic arch and its branches) that compresses trachea or oesophagus. It presents as respiratory and GI symptoms. Double aortic arch is the most common vascular ring. It typically becomes symptomatic in infancy or early childhood. We present herein a rare case of an infant with double aortic arch who presented with respiratory distress and GI symptoms. The diagnosis was made by barium swallow, contrast-enhanced computed tomography chest, and MRI chest. Thereafter, the patient was referred to higher centre for surgical management.

KEY WORDS: barium swallow, computed tomography, double aortic arch (DAA), magnetic resonance imaging, respiratory distress

INTRODUCTION:

Vascular ring is a congenital anomaly in which the trachea and oesophagus (or its atretic remnant) is completely 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 (DAA) is the most frequently encountered vascular ring malformation characterized by a complete encirclement of trachea and oesophagus by the aortic arch. 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 may be sufficiently tight to cause clinical symptoms or loose enough to be asymptomatic. For children aged less than 2 years, symptoms are typically respiratory nature, while older children and adults frequently present with dysphagia and rarely show respiratory symptoms[4].

Diagnosing DAA is challenging because it has a wide clinical spectrum. Double aortic arch is usually diagnosed in infants with life-threatening respiratory symptoms due to severe compression of trachea. However, when it is minimal, DAA may remain undiagnosed until adulthood.

We report a case of complete double aortic arch with classical manifestations.

Corresponding Author:
Dr Om Shankar Chaurasiya
Department of Peadiatrics,
MLB Medical College, Jhansi-284001
Phone No.: +91 8934803482
E-mail: chaurasiyaom@gmail.com

CASE REPORT:

A 45 days old male child presented to our department with history of respiratory symptoms and recurrent regurgitation of feeds. The baby was delivered vaginally at term with birth weight of 3.1 kg and had cried immediately at birth. The patient started having symptoms of noisy breathing, increased respiratory rate and recurrent regurgitation of feeds from 5th day of life. There was no history of cyanosis. Patient was taken to several doctors but symptoms were not relieved in spite of medications. Several investigations were done, including 2D echo cardiography which did not reveal any abnormality. Patient reported in our department at 45 days of age.

On admission HR-132/min, RR-66/min with sub costal & intercostal retractions. On systemic examination bilateral rhonchi were heard. Initially patient was managed with antibiotics and inhalational steroids but no improvement occurred despite of use of proper antibiotic cover according to blood culture.

Then we suspected congenital anomalies and investigation were done accordingly. Barium swallow

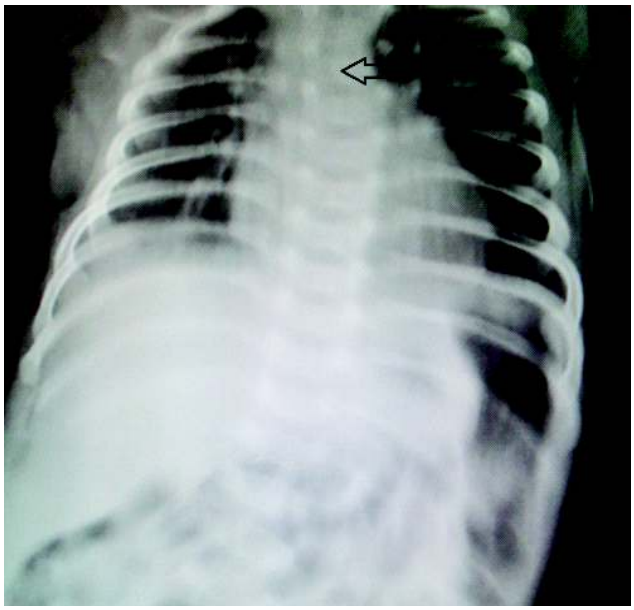

Figure 1: Chest x ray shows indentation of the oesophagus.

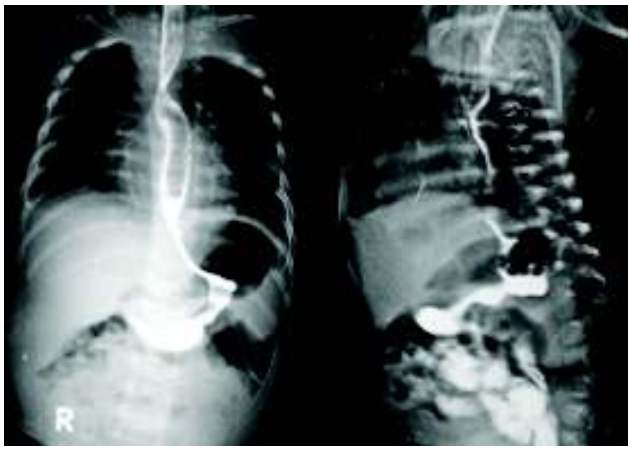

Figure 2: Barium swallow shows indentation of the oesophagus.

study revealed severe stenosis at upper oesophagus. CECT and MRI thorax revealed double aortic arch with dominant right side and right descending thoracic aorta with approximately 50% tracheal lumen narrowing.

MRI chest showed branches arising from the right arch, which were left common carotid, right common carotid and right subclavian artery, anterior to posterior. Left subclavian artery was arising from left aortic arch posteriorly.

Double aortic arch was encircling both trachea and oesophagus with indentation on trachea and oesophagus. Then the patient was advised for surgery and was referred to the higher centre for further surgical management. The vascular anomalies

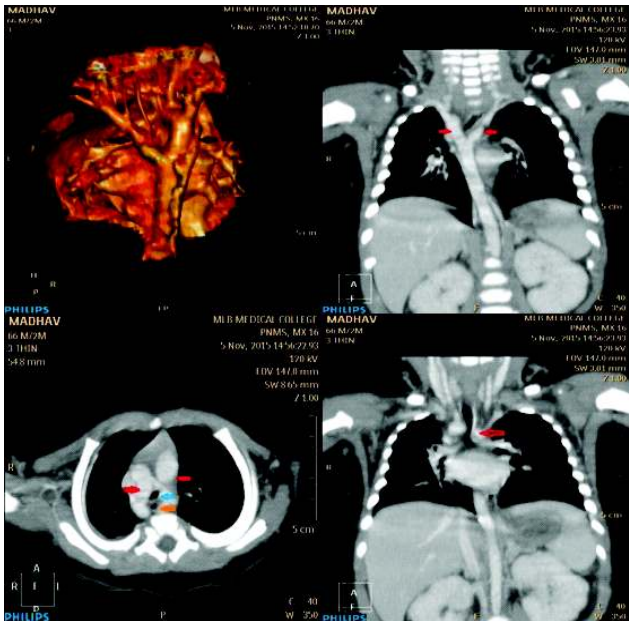

Figure 3: Plates 1, 2, 3 shows double aortic arch, plate 4 shows indentation of the trachea.

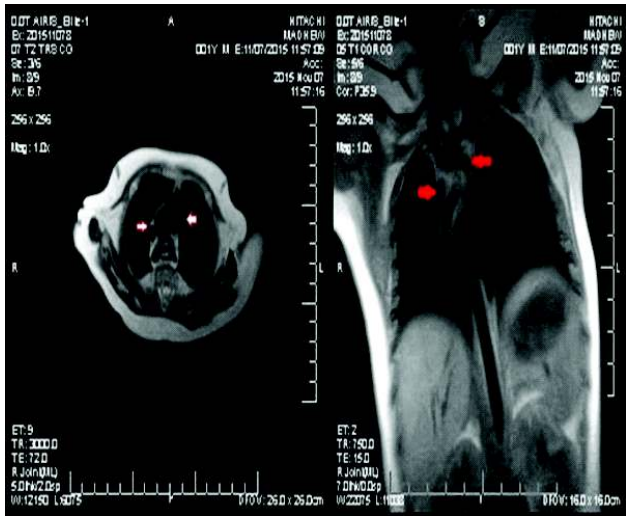

Figure 4: CT images, plate 1 & 2 shows double aortic arch.

were surgically corrected and patient was relieved of symtoms.

DISCUSSION:

Double aortic arch is due to failure of regression of the right aortic arch. Normally, absorption of the right (posterior) arch takes place between the right subclavian artery and its junction with the descending aorta. The remnant of the right arch becomes the right innominate artery and leaves a left (anterior) arch in normal development, freeing the trachea and esophagus. Failure of this process of

absorption in the right arch produces a vascular ring causing complete encircling and compression of oesophagus and trachea leading to severe feeding and respiratory difficulties.

Double aortic arch usually exists as an isolated problem, although the presence of associated congenital heart disease such as tetralogy of Fallot, ventricular septal defect, and Transpositions of great arteries have been reported.

In our patient there was no evidence of any other cardiac anomaly.

Development of vascular rings can be explained by Edwards' double ring hypothetical model[5]. The development of the arch and branch vessels is more complex, and a thorough understanding of this process is imperative for recognising and accurately characterising the variations of vascular rings, which is critical for surgical planning [6]. Development of the great vessels begins at 20–22 days by vasculogenesis, in which networks of endothelial channels are formed by aggregation of angioblasts. These networks fuse to form the dorsal aortae and aortic arches. The lumen is established within these vessels when the small endothelial channels merge into larger channels[7].

Smooth muscle cells of the media are formed from neural crest cells in the arch and mesenchymal cells in the dorsal aorta[8-10]. Six aortic arches are formed in the fourth and fifth weeks of development and run in the centre of the pharyngeal arches connecting the paired ventral and dorsal aortae[11]. The paired ventral aortae fuse to form a single ventral aorta, the aortic sac. Fusion of the dorsal aortae into a single dorsal aorta begins distally and progresses retrograde to the seventh somite. Proximally the aortic sac connects to the heart through the truncus arteriosus. Eventually, the truncus is divided into the ventral aorta and pulmonary trunk by the spiral septum.

The six arches develop in a craniocaudal fashion and also regress in the same fashion; hence all six arches are not seen at the same time . Also the fifth arch is rarely seen in humans, due to either nonexistence or very early regression[6]. The normal pattern of modelling and regression depends on neural crest cells, although the exact mechanism remains unknown [12,13,6,14].

CONCLUSION:

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. Factors responsible for the aberrant persistence of certain aortic arch segments have not been clearly identified, and the pathogenesis of this anomaly remains a mystery. Double aortic arch typically occurs without associated cardiovascular defects, although other lesions may be present, and accordingly, it is not usually found as part of a syndromic complex. In most centers, the mortality risk for surgery is between zero and 2%. A specific risk of open surgical repair of double aortic arch is injury to the recurrent laryngeal nerve, which can cause vocal cord paralysis. Injury to the lymphatic system can lead to postoperative chylothorax. Additional risks include lung injury, bleeding with the need for blood transfusions and wound infection.

REFERENCES:

1. Gross RE. Arterial Malformations Which Cause Compression of the Trachea or Esophagus. Circulation. 1955:11:124-134.
2. Kau T, Lesnik G, Eicher W, Sinzig M, Gasser J, et al. Aortic Development and Anomalies. Seminars in Interventional Radiology. 2007:24:141-152.
3. Baraldi R, Address SS, Bighi S, Mannella P. Vascular Ring Due to Double Aortic Arch. A Rare Cause of Dysphagia. Eur J Radiol Open. 2004:52:21-24.
4. Umegaki T, Sumi C, Nishi K, Ikeda S, Shingu K. Airway Management in an Infant with Double Aortic Arch. J Anesthesia. 2010:24:117-120.
5. Hernanz-Schulman M. Vascular rings: a practical approach to imaging diagnosis. Pediatr Radiol. 2005: 35:961-979.
6. Effmann EL, Whitman SA, Smith BR. Aortic arch development. Radiographics. 1986: 6(6):1065-1089.
7. Waldo K, Kirby M. Development of the great arteries. In: de la Cruz MV, Markwald RR (Eds); Living Morphogenesis of the Heart. Birkauser, Boston, MA. pp187-217.
8. Hungerford JE, Little CD Developmental biology of the vascular smooth muscle cell: building a multilayered vessel wall. Vasc Res. 1999: 36: 2–27.
9. Suri C, Yancopolous GD Growth factors in vascular morphogenesis: insights from gene knockout studies in mice. In: Little CD, Mironov V, Sage H (eds) Vascular Morphogenesis: In vivo, in Vitro, In Menter.

Birkhauser, Boston. 1999: pp 65–72.
10. Kuo CT, Veselitis ML, Barton KP et al.,The LKLF transcription factor is required for normal tunica media formation and blood vessel stabilization duringmurine embryogenesis.Genes Dev. 1997: 11(15): 2996–3006.
11. Congdon ED. Transformation of the aortic-arch system during the development of the human embryo. Conf Embry. 1922: 68: 47-110.
12. Edwards FR Vascular compression of the trachea and esophagus. Thorax; 1959: 14: 187–200.
13. Kussman BD, Geva T, McGowan FX Cardiovascular causes of airway compression. Paediatric Anaesth. 2004: 14: 60-74.
14. Gruber PJ,Wessels A, Kubalak SW Development of the heart and great vessels. In Pediatric cardiac Surgery. Eds Mavroudis C, Backer CL. Blackwell Publishing. 2013:1-26.

Cite this article as: Chaurasiya OS, Ravanagomagan MG, Gupta S, Vats P: Trachea and Oesophagus Compression by DAA**.** PJSR. 2017:10(2):78-81.
Source of Support: Nil, **Conflict of Interest**: None declared.