

# Adult Incidental Asymptomatic Symmetrical Double Aortic Arch Case as a Variation of Congenital Vascular Ring Malformation

## Konjenital Vasküler Ringin Bir Varyasyonu Olarak Yetişkin Bir Olguda Asemptomatik Simetrik Doble Aortik Ark

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Geliş Tarihi/Received: 06.06.2011  
Kabul Tarihi/Accepted: 04.07.2011

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**ABSTRACT** We report a case of a rare and asymptomatic vascular ring in an adult patient. A vascular ring presents a complex of anomalous anatomy of aortic arch and related vascular structures which encircle trachea and esophagus as a 'ring' which presents only less than 1% of all congenital cardiac abnormalities. Vascular ring may be with or without concomitant congenital cardiac defects. This defect of vascular ring is free of any geographical predominance and the male to female ratio is not distinguishing. Most of cases among vascular ring in a percentage of 85-95% are double aortic arch as in our case.

**Key Words:** Aortic arch syndromes; aorta, thoracic

**ÖZET** Biz bu yazıda nadir ve asemptomatik olarak gözlenen erişkin bir hastadaki vasküler ring olgusunu sunduk. Bir vasküler ring trakea ve özofagusu çevreleyen aortik ve vasküler yapılarla ilişkili olan bir kompleks anomalidir. Bu anomali konjenital kardiyak patolojilerin %1'inden daha azını oluşturmaktadır. Vasküler ring eşlik eden konjenital defektlerle birlikte olabilir. Vasküler ringin bu defektlerinde herhangi bir cinsiyet farkı ve coğrafi farklılık mevcut değildir. Vasküler ringli hastaların çoğu bizim vakamızda da olduğu gibi %85-95 oranında double aortik ark olgularından oluşmaktadır.

**Anahtar Kelimeler:** Aort kavsi sendromları; aort, torasik

**Türkiye Klinikleri J Cardiovasc Sci 2013;25(1):49-52**

Vascular rings are formed by double aortic arches with equal right and left components or with a smaller or atretic right or left component; the descending thoracic aorta may be left or right sided, and there may be a left, right, or bilateral patent ductus arteriosus, or a ligamentum arteriosum.<sup>1,2</sup>

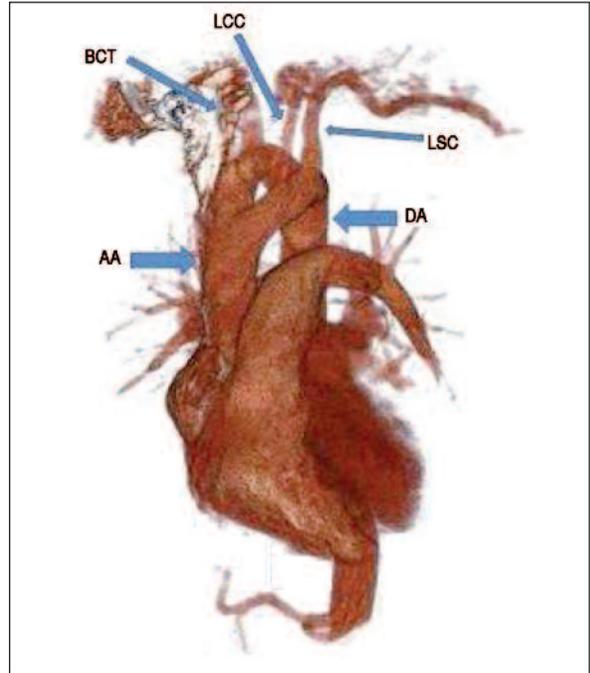
For vascular ring; presence of symptoms are indications for surgery at every age. Especially symptom-free adults may not necessitate surgery. Nevertheless we advocate that even cases like ours may require close follow-up with regular laboratory studies and appropriate physical examination. Detention of surgery is with irreversible complications of respiratory system which may lead to a severe morbidity and death.

## CASE REPORT

A 47 years old female patient administered to our department with a sporadic/incidental computerized tomography scans revealing a double aortic arch. Physical examination was within normal limits. The electrocardiogram showed a normal sinus rhythm for 80/min. Arterial blood pressure was 140/90 mmHg on the right upper extremity and 150/90 on the left upper extremity. There was no significant pressure difference between two upper extremities. Lungs and heart were clear to auscultation. There was no evidence of cardiac murmur and/or respiratory stridor.

Laboratory blood tests showed no abnormalities with the cardiac panel. Neurologic evaluation was also detected free of any findings. All arterial areas were palpable and without any murmur to auscultation including carotid system. The chest radiograph and M-mode echocardiograms (TTE) showed no concomitant intra-cardiac malformations. A computed tomography scan (CT) was presented by the patient who was performed by pulmonologists department as a part of a routine preoperative evaluation for elective cholecystectomy. Patient was referred to our department due to CT angiography findings of double aortic arch. Figure 1 presents the anterior view of double aortic arch with a split after ascending aorta. Posterior branch of double aortic arch gives origin to the truncus brachiocephalicus and arteria carotid communes sinister with arteria subclavian sinister arise from the anterior branch, respectively from proximally to distally. From the posterior aspect in Figure 2, descending aortas reunites right after arteria subclavia sinister distinction. No abnormalities are detectable during the ascending aorta and continued existence of descending aorta in CT scans.

In this case, the vascular ring encircles trachea and esophagus but without any symptoms of laryngeal stridor, frequent pulmonary infection, dysphasia, cough or respiratory obstructions at any level. Thus, symptom free nature of patient's clinical features, normal physical examination and the success in arriving to adult age took us away from surgical interventions. Patient was discharged at



**FIGURE 1:** Multislice computed tomography presents a vascular ring, anterior view. Posterior branch of double aortic arch gives origin to the truncus brachiocephalicus (arrow indicating BCT). Arteria carotid communes sinistra (arrow indicating LCC) with arteria subclavian sinister (arrow indicating LSC) arise from the anterior branch, respectively from proximally to distally. No abnormalities are detectable in the ascending aorta (AA) and descending aorta (DA).

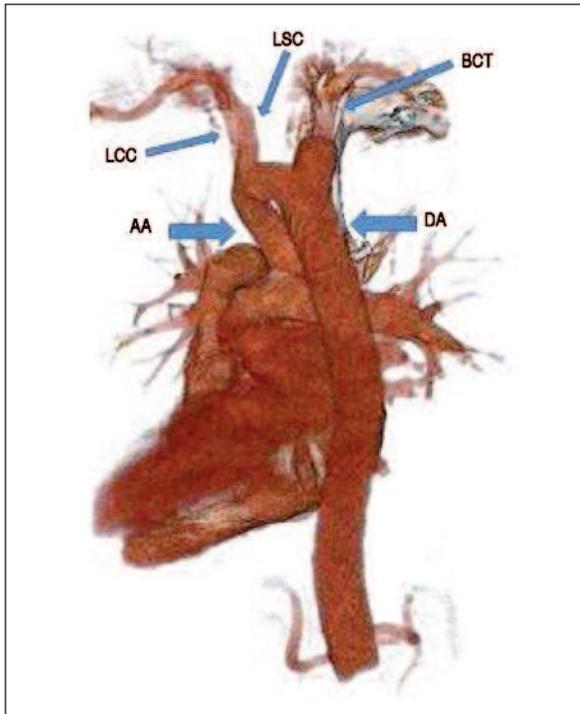
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the sixth hospitalization day with decisions of regular clinical follow-ups in every year.

## DISCUSSION

Vascular rings are unusual major congenital vascular malformations. First diagnose was presented by Hommel in 1737. Double aortic arch was circumstantially described by Abbot Maude in 1932. The term of vascular ring was used by Gross in New England Journal of Medicine. A vascular ring presents a complex of anomalous anatomy of aortic arch and related vascular structures which encircle trachea and esophagus as a 'ring'. This disease presents only less than 1% of all congenital cardiac abnormalities.

Vascular ring may be with or without concomitant congenital defects free of any geographical predominance and the male to female ratio is not distinguishing. Most of cases among vascular



**FIGURE 2:** Multislice computed tomography presents a vascular ring, posterior view. No malformations are presented with continued existence of descending aorta after reunited anterior and posterior branches. AA= Ascending aorta, DA=Descending aorta, BCT= Truncus brachiocephalicus, LCC=Arteria carotis communis, LSC=Arteria subclavia sinistra.

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ring in a percentage of 85-95% are double aortic arch as in our case. Besides, tetralogy of Fallot cases present a double aortic arch in 30% cases. Other types include right aortic arch in mirror image arising vessels with left ligamentum arteriosum and left aortic arch with right subclavian artery abnormality in less frequency than 1%. Associated vascular abnormalities may include at the localizations of pulmonary artery with pulmonary sling, arterial branches of arcus aorta and other intra-cardiac chambers and structures. These anatomical findings are results of abnormal aortic arch developments of embryonic phases in terms of incomplete involution phases.<sup>2-4</sup>

Laboratory studies and radiologic investigations include; chest x-ray, barium esophageal graphy, TTE, magnetic resonance imaging, CT, digital subtraction angiography, aortic angiography with/without cardiac catheterization and for selected cases bronchoscope. There are several med-

ical literatures which compare the efficiency of these radiologic tests. In our opinion CT with TTE may present valuable finding on these cases. On the other hand, a barium esophageal graph is an inexpensive and easily performable technique with valuable results.<sup>5,6</sup>

As in our case, a double aortic arch is quite more frequent in this topic. Co-morbidities of intra-cardiac defects are rare with this type of vascular ring which was also the feature of this patient. Double aortic arch forms a ring around trachea and esophagus resulting a trachea-esophageal compression.

Symptoms mostly present in infancy and/or early ages of childhood. Adult age diagnosis, as in our case at the age of 47, is extremely rare. Most common clinical features include inspiratory stridor and wheezing with respiratory difficulties, cyanosis for severe compressive rings, and apnea with coughing caused by tracheal component of compression. Furthermore, on the side of esophageal compression, patient suffers from dysphasia, lower weights and feeding distress causing cyanosis. More severe cases may presents intercostals retractions and discontinuance of breath resulting apnea periods with level changes in consciousness.

Presence of symptoms is indications for surgery at every age. Medical therapy is an additive choice to surgical treatment. Surgery for division of aortic arch so to reach a relief of trachea-esophageal structures can be achieved via left or right thoracotomy. A maximum effort to avoid of vascular and neighboring nervous structure is essential for acceptable postoperative results. Postoperative complication is pneumonia, atelectasis, clinical reflections of trauma in vagal nerve and continuance of preoperative symptoms such as dysphasia and respiratory distress. Nevertheless, long-term survival after surgery is about 90-95% of all cases.<sup>6,7</sup>

Especially symptom-free adults may not necessitate surgery. Nevertheless we advocate that even cases like ours may require close follow-up with regular laboratory studies and appropriate physical examination. Detention of surgery is with irreversible complications of respiratory system which may lead to a severe morbidity and death.

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