

Case Study**Clinical and MDCT Findings of Symptomatic Right
Aortic Arch and Double Aortic Arch:Two Cases****ABSTRACT**

Aim: Anomalies of aortic arch are rarely seen. They are 1-3% of all congenital heart diseases. Vascular ring occurs in consequence of abnormal development of aortic arch complex. Usually it manifest itself with compression findings of tracheo-esophageal in infants. In this paper, we discussed clinical findings and characteristic imaging findings of the two cases with vascular ring.

Cases: We report two cases of a child in four years old age diagnosed with right aortic arch and a neonatal diagnosed with double aortic arch. In both diagnosis was made by MDCT.

Discussion: If airway obstruction is distinct complaints emerge critically in early age. If symptoms are mild and there is no anatomic compression disease may be diagnosed in later years of life. The first case is four years old age who suffered cough and shortness of breath. Another was a neonate who suffered from respiratory distress . Vacular ring diagnosis can be made by chest radiography ,barium esophagus graphy, transthoracic echocardiography, MDCT, magnatic resonance imaging and angiography. But pathologies of our patients (right aortic arch ,double aortic arch, and tracheal compression) are seen effectively by MDCT and the post-processing images of MDCT

Conclusion: Vascular rings must be taken into account in pulmonary infections repeated, unexplained cough, wheezing, stridor in childhood period and unexplained respiratory distress in newborn childs. And MDCT is an excellent modality in patients suspected to have a vascular ring

Keywords: Right aortic arch, double aortic arch, MDCT, vascular ring

INTRODUCTION:

Complex embryological development of aortic arch and its branches leads to anomalies in different types in this system (1,2). By means of those anomalies anormal relations between tracheobronchial tree and vascular structures can be seen. In other words, vascular ring and secondary airway pressure can be showed. Vascular ring is an important and rare reason of the symptoms such as pulmonary infection repeated in infancy, stertorous respiration, stridor and respiratory distress in neonates. These

33 symptoms are associated with the degree of airway obstruction that is developed as secondary to
34 anatomical compression (3).

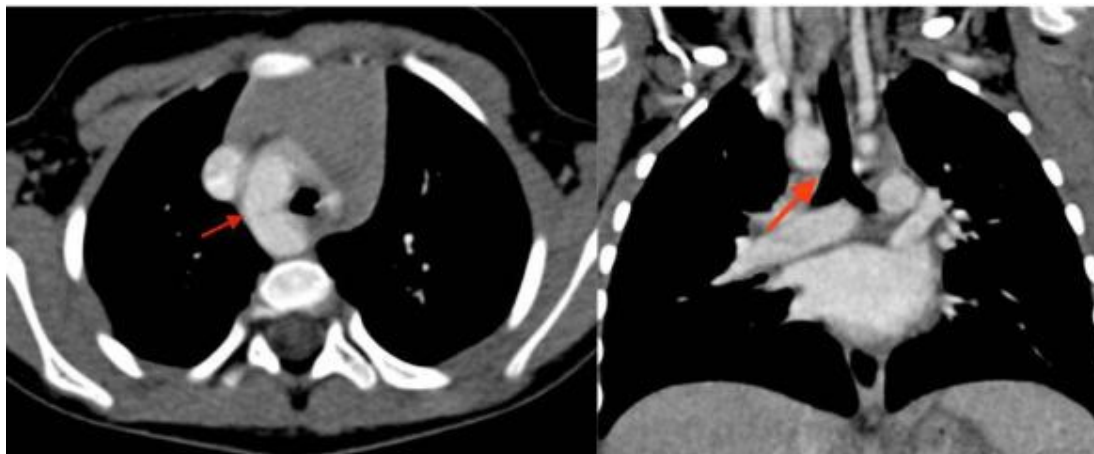
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36 **CASES:**

37 The female patient in four years old age applied to our hospital for complaints of cough, wheezing and
38 shortness of breath. It was understood from her history that her shortness of breath and cough
39 continued, her complaints increased while she was playing and running and she had no any systemic
40 complaint. Her respiratory system and all other systemic examination have been evaluated as normal.
41 There was no any atopic story both in her family and her. Complete Blood Count and biochemical test
42 was in normal limits. Right aortic arch and compression on trachea from the right (Figure 1) was seen
43 in MDCT thorax examination made in our Radiology Department. On the other hand, it was observed
44 that left subclavian artery and left common carotid artery were the first branch of aortic arch by a
45 common root (left innominate artery). Other branches were right main carotid artery and right
46 subclavian artery respectively. A surgical operation was planned for vascular ring that have been
47 defined by these findings.

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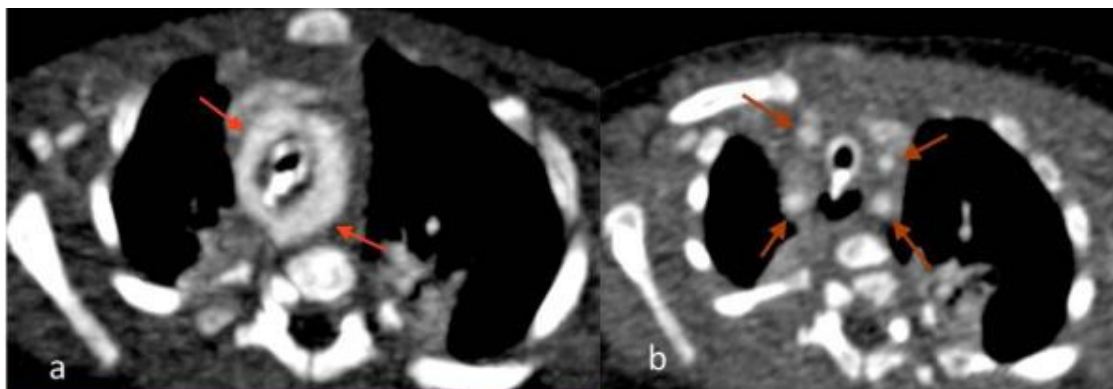
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51 **Figure 1: Axial CT scan shows a right aortic arch (small arrow) and tracheal**
52 **compression (big arrow)**

53 Second case was a male newborn that was followed as intubated since his birth at intensive care
54 unit of our hospital. The case that was the first pregnancy of a mother in 26 years old age born as 36
55 weeks. Mother had no any specific medical history. The case was born in transvaginal way in 3010 gr.
56 The patient was intubated due to respiratory distress and treated by antibiotherapy on diagnosis of
57 pneumonia. One week later the patient got well in radiologically and clinically and extubated. However,
58 the patient was intubated again due to falling saturation and respiratory distress. It was taken MDCT
59 angiography of aortic arch due to suspected vascular ring. It was detected double aortic arch, four-
60 artery sign (Figure 2) and compression to trachea findings in images. The patient was referred to
61 surgery.

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65 **Figure 2: a) Axial MIP image shows double aortic arch encircling the trachea and oesophagus. b)**
 66 **'four-artery sign' which is characterics findings of double aortic arch is seen on axial image.**

67 **DISCUSSION**

68 Vascular rings are congenital abnormalities of the aortic arch-derived vascular and ligamentous
 69 structures, which encircle the trachea and oesophagus to varying degrees, resulting in respiratory or
 70 feeding difficulties in children (4). Vascular ring occurs as vascular structures have no normal
 71 involutions or wrong involutions in embryonal life (5). This pathology including 1-2% of congenital heart
 72 disease, is one of the important factors of airway obstruction in neonatal and childhood

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74 Severity of clinical findings is changed according to compression degree (6,7). Dispne, stridor,
 75 wheezing and cough are the most frequent symptoms (6). Not only it makes way to severe respiratory
 76 distress but also be asymptomatic lifelong (6). The first case is 3 years old age who suffered cough
 77 and shortness of breath. Another was a neonate who suffered from respiratory distress .

78

79 Although vascular ring has different classifications more than 95% of it is classified 4 major categories
 80 (Table 1)(8). The most frequently seen is double aortic anomaly (9). Both aortic arch are patent in this
 81 anomaly. Right side arch is typically dominant(2). And right arch is retroesophageal. It has generally
 82 more severe and early symptoms than other ring types. Two arches originate from the ascending
 83 aorta, cross on either side of the trachea-oesophagus and join the descending thoracic aorta. (9).
 84 Usually one arch is dominant and the other arch is smaller or may be atretic (9,10). In our case left
 85 side lumen was a little wider than the other one. We did not observe any occlusion or stenosis in aortic
 86 arch. Cardiovascular anomalies such as fallot tetralogy, coarctation of aorta and patent ductus
 87 arteriosus may accompany that anomaly (9,10). In our case additional cardiac anomaly has not been
 88 seen.

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Table 1. International Nomenclature and Database Conferences for Pediatric Cardiac Surgery(1998-1999)

I-Double arch aort

II-Right arch aorta + left lig.arteriosus

III-Innominate artery compression

IV-Pulmonary artery sling

90 *This 4 groups comprises of more than 95% of all vascular rings.

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93 Right aortic arc is the second most common type of vascular ring anomalies. This anomaly is
94 classified in three form (11). The most common branching patterns of the right aortic arch are the
95 mirror image branching pattern(type II)(9). In this type, the great vessels originate from the arch in the
96 following order, left innominate artery, right common carotid artery and right subclavian artery(11). And
97 this type has aberrant left subclavian artery. In our case Type I right aortic arch anomaly was seen.

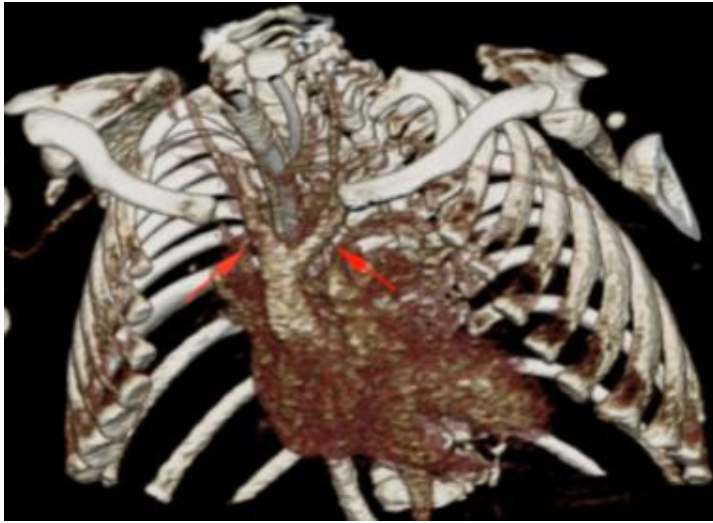
98 Vascular ring diagnosis is made by chest radiography (PA and Lateral), barium esophagus graphy,
99 transthoracic echocardiography (ECO), contrast MDCT, magnetic resonance imaging (MRI) and
100 angiography. Today, MDCT, MRI and ECO are the most preferred diagnosis methods (6,9,10). In PA
101 chest radiography single or bilateral emphysema depending on compression and compression on
102 trachea from right or left may be observed. In lateral chest radiography compression on the anterior of
103 trachea may be observed. In the study of 81 cases made by Alsenaidi et al (12) it was determined in
104 20% of cases that chest graphy was normal . We did not observe a distinct findings in the chest
105 radiographies of our cases. In barium esophagus graphies it can be seen that esophagus is subjected
106 to compression from anterior, posterior or lateral sides . Notchings related to compression may be
107 seen in different levels and sizes (6,10). In both cases barium esophagus examinations have not been
108 performed. PA chest radiography and barium graphies may varifies vascular ring diagnosis. But cross
109 sectional imaging is usually required to define the morphology of the aortic arch formation and for
110 planning an optimal surgical approach (10).

111 Degree, level and lenght of compression related aortic arch in trachea can be clearly seen by MDCT
112 and MRG. The most important advantages of cross-sectional imaging are the ability for showing
113 environmental structuring as well as vascular structures and supplying images in three plans(6,9). In
114 our case right aortic arch is seen by mirror image branching and 'four-artery sign', which is
115 characterics findings of double aortic arch, and significant tracheal compression are seen effectively
116 by MDCT(9).

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118 Advantages of MDCT include the rapid acquisition time. In most cases sedation is not needed unlike
119 MRG. In the patients suffering from airway obstruction sedation may give rise to severe problems (6).
120 In the evaluation airway and lung parenchyma MDCT gives more precise information (10). Images can
121 be examined in three dimensional (3D) in each plan by Multiplanar reformation (MPR) and volume
122 rendering (VR) reconstruction. 3D volume-rendered displayed the ring and sizes of the arches in our
123 case effectively((Figure 3). Additionally minimal compression on trachea can be examined by
124 minimum intension projection(MinIP). On the other hand these compression can be also examined by
125 3D methods concretizing airway and parenchyma. In our cases, we use these post-processing images
126 and show tracheal narrowing distinctly (Figure 4). And The most important disadvantage, especially in
127 the pediatric age is the use of X-rays(6,9,10).

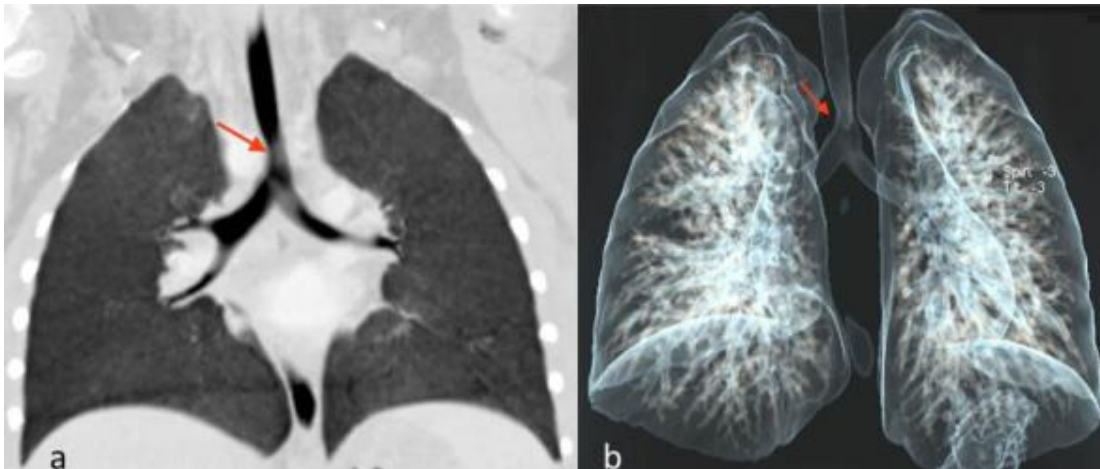
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130 **Figure 3: 3D volume-rendered image demonstrates the double aortic arch (arrows).**

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133 **Figure 4: a) Tracheal compression (arrow) is seen on MinIP image b) Coronal volume-rendered (3D)**
134 **image of the airway shows narrowing of the airway (arrow) by the right aortic arch**

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137 Conventional angiography determines topographic anatomy specifically. However it has
138 disadvantages in failing determination of atretic vascular structures and indication of tracheal and/or
139 esophagus compression (6,10). It is important to perform ECO in order to exclude accompanying
140 anomalies in doubt of vascular ring (9).

141 It is recommended surgical intervention directed to vascular ring in symptomatic patients in which
142 airway compression is evidenced radiologically (13). The surgeon must make a decision of
143 approachment according to the vascular ring anatomy and the associated tracheal or cardiac
144 anomalies(11).

145 **CONCLUSION**

146 Vascular rings must be taken into account in pulmonary infections repeated, unexplained cough,
147 wheezing, stridor in childhood period and unexplained respiratory distress in newborn children. And
148 MDCT is an excellent modality in patients suspected to have a vascular ring. It allows evaluation of the
149 aortic arch and its branches. It also provides valuable information for planning surgical management
150 with multiplanar and three-dimensional(3-D) imaging.

151

152 **CONSENT**

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154 All authors declare that 'written informed consent was obtained from the patient (or other approved
155 parties) for publication of this case report and accompanying images. A copy of the written consent is
156 available for review by the Editorial office/Chief Editor/Editorial Board members of this journal
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158 **ETHICAL APPROVAL**

159

160 All authors hereby declare that all experiments have been examined and approved by the appropriate
161 ethics committee and have therefore been performed in accordance with the ethical standards laid
162 down in the 1964 Declaration of Helsinki."

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