Case Study

Clinical and Imaging Findings of Symptomatic Right Aortic Arch and Double Aortic Arch: Case Report

ABSTRACT

Aim: Anomalies of aortic arch are uncommon, accounting for only 1-3 % of all congenital cardiac diseases. A vascular ring results from the abnormal development of an aortic arch complex. Generally, it manifests with tracheoesophageal compression symptoms during infancy. We aimed to discuss clinical and imaging findings of two cases of vascular ring based on existing literature.

Cases: This article presents two cases of a 4-year-old child diagnosed with right sided aortic arch and a new-born child diagnosed with double aortic arch in neonatal unit. The first patient presented with complaints of coughing, wheezing and shortness of breath, the second patient suffered from respiratory distress. In the both cases, diagnosis were established using multi-detector computed tomography (MDCT). Both patients were diagnosed with vascular ring, considering their existing findings, and referred for surgery

Discussion: Apparent airway obstructions lead to critical complaints at early ages. Moderately severe symptoms and the non-existence of anatomic compression may cause resulting diseases to be diagnosed in a later period of life. A vascular ring can be diagnosed using chest radiography, barium oesophagus graphy, transthoracic echocardiography, MDCT, magnetic resonance imaging and angiography. However, pathologies of the studied cases (right aortic arch, double aortic arch, and tracheal compression) are effectively observed with MDCT and using the post-processing images of MDCT.

Conclusion: A vascular ring should be considered in the event of recurrent lung infections, unexplained coughs, wheezing and stridor during childhood as well as in the case of unexplained respiratory distress in new-born children. MDCT is an excellent modality for patients suspected of having a vascular ring.

Keywords: Right aortic arch, double aortic arch, multi-detector computed tomography, vascular ring, symptoms, congenital heart disease.

INTRODUCTION:

Complex embryological development may cause many different types of malformations in the aortic arch and its branches (1,2). Abnormal relationships can be seen between the tracheobronchial tree and vascular structures as a result of these malformations, leading vascular rings and secondary airway compression. A vascular ring is an important and unusual reason causing symptoms such as recurrent pulmonary infections, persistent wheezing and stridor during infancy and as well as respiratory distress in new-borns. These symptoms are associated with the degree of airway obstruction developing secondary to anatomic compression (3). In this study, we discuss the clinical and imaging findings of two cases of vascular ring, which is symptomatic and rarely appears.

CASES:

A four-year-old female patient with complaints of coughing, wheezing and shortness of breath was admitted to the chest diseases department of our hospital. The patient's history showed that she had persistent shortness of breath and wheezing and her complaints increased when playing games. she had no accompanying systemic complaints. Tests of the respiratory system and other systemic examinations of the patient returned normal. According to the patient's medical background, the patient and her family had no history of atopy. A complete blood count and biochemical tests of the patient showed that all results were within normal limits. A right sided aortic arch and related tracheal compression (Figure 1, 2) were observed during the examination of the thoracic MDCT taken in our radiology department (-kV: 100, mAs: 72, reconstruction section thickness 1.0 mm, approximate CT dose: 2,91 mSv-). In addition, it was observed that the left subclavian artery and the left common carotid artery (the left innominant artery) developed out of the aortic arch as the first branch with a shared root. Other branches included the right common carotid artery and the right subclavian artery (Figure 3). A surgical operation to fix the vascular ring identified based on the findings was planned, but the patient did not agree to the surgery.

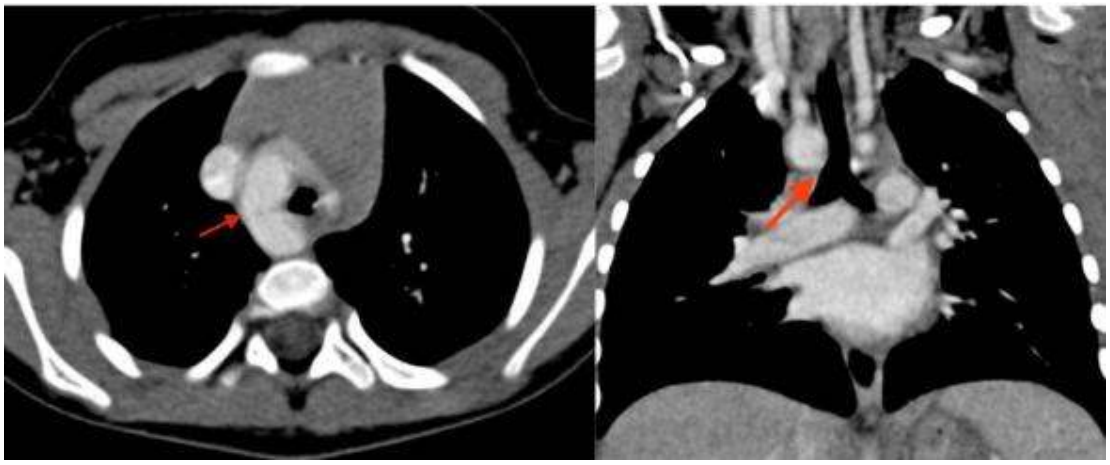

Figure 1: Axial Computed Tomography scan shows a right aortic arch (image on the left) and tracheal compression (image on the right)

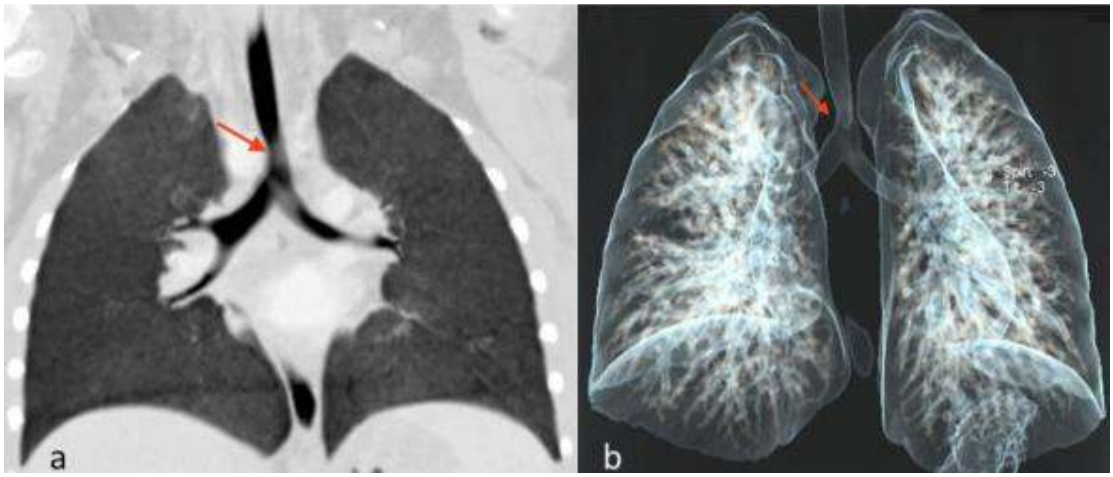

Figure 2: a) Tracheal compression (arrow) is seen on the MinIP image b) The Coronal volume-rendered (3D) image of the airway shows a narrowing of the airway (arrow) by the right aortic arch

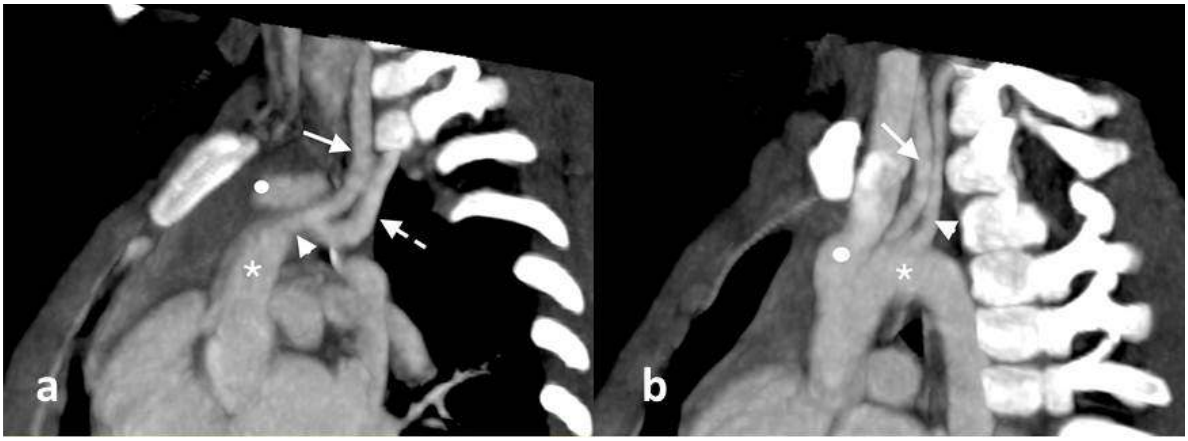

Figure 3: a) The ascending aorta (asterisks), left innominate artery (arrowhead), left main carotid artery (arrow), and left subclavian artery (dashed arrow) are seen in the MIP sagittal image. b) The aortic arch (asterisks), right main carotid artery (arrow), and right subclavian artery (arrowhead) are seen on the other MIP sagittal image. The white circle is veins in both images.

The second case was a new-born male infant who had been intubated and monitored from birth in the paediatric intensive care unit of our hospital. The infant, the first pregnancy of a 26-year-old mother, was born in the 27th week of the pregnancy. The mother did not have any specific conditions.

The case was delivered transvaginally in 1300 gr. The patient was intubated due to respiratory distress, an antibiotic therapy was initiated, based on the preliminary diagnosis of pneumonia. An attempt was made to extubate the patient, who recovered clinically and radiologically a week later. However, the patient was re-intubated due to a decrease in saturations and respiratory distress. The results of the chest X-ray was normal (Figure 4). A MDCT angiography was taken of the aorta as a vascular ring was suspected (-kV: 80, mAs: 28, reconstruction section thickness 1.0 mm, approximate CT dose: 0,36 mSv-). A double aortic arch, four artery signs (figure 5) and tracheal compression were discovered on the cross-sections. A 3D volume-rendered image displayed the ring and sizes of the arches effectively (Figure 6). The patient was referred to surgery. The patient had a surgical division of the right arcus aorta, thereby eliminating the compression on the trachea and oesophagus.

Approval of the relevant ethics committee was obtained for this study, and the patients provided their written informed consent.

Figure 4: There were no pathologic findings in the chest radiography

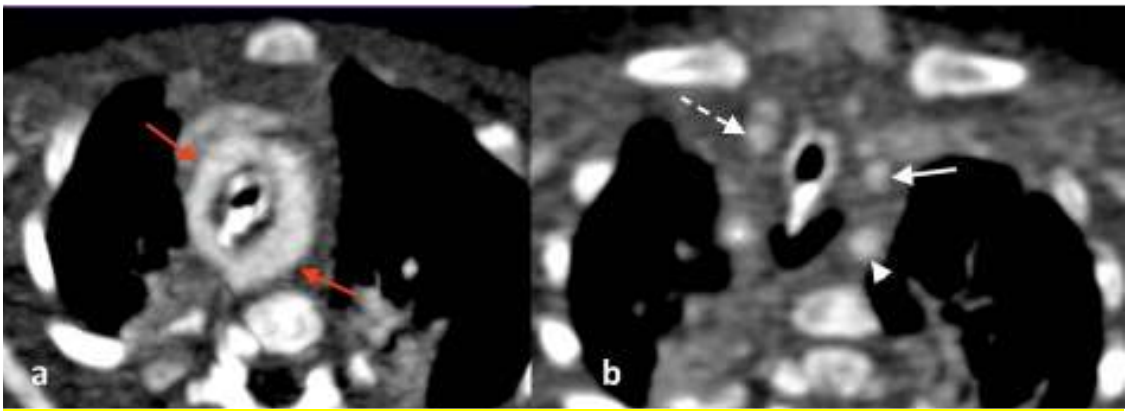

Figure 5: a) The axial MIP image shows a double aortic arch encircling the trachea and oesophagus. b) A 'Four-artery sign' , which is a characteristic finding of the double aortic arch, is observed on the axial image. The asterisk indicates the right subclavian artery, the dashed arrow points to the right main carotid artery, the arrow points to the left main carotid artery, and the arrowhead highlights the left subclavian artery.

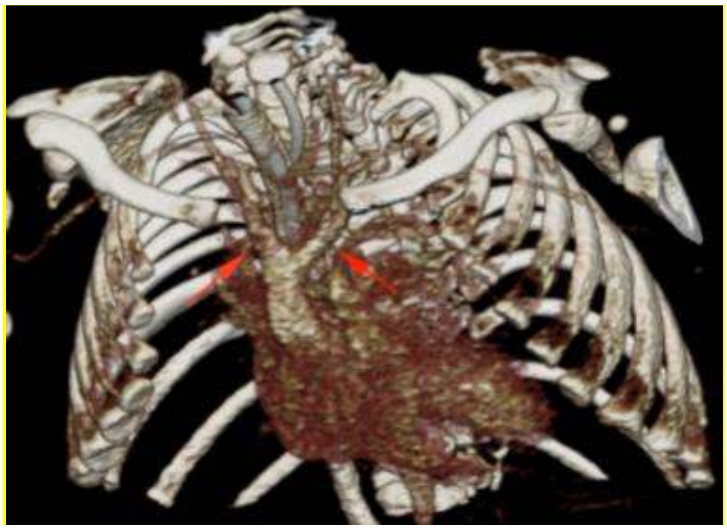

Figure 6: The 3D volume-rendered image demonstrates the double aortic arch (arrows).

DISCUSSION

A vascular ring, a congenital anomaly, is a defect in which thetrachea and oesophagus are surrounded by blood vessels; the ligamentous structures developed from this arch system remain

under pressure as a result of the abnormal development of the branchial arch system (4). A vascular ring results from a lack of normal involution of vascular structures forming an aortic arch during embryonic life or from the occurrence of the involution of these vascular structures in different sites (5). This defect, which accounts for 1-2% of congenital cardiac diseases, is an important factor causing airway obstruction during the neonatal and childhood period.

The severity of clinical symptoms varies by the degree of compression (6,7). Dyspnea, stridor, wheezing and cough are the most common symptoms (6). A vascular ring can cause severe respiratory distress developing immediately after birth as well as be asymptomatic for life (6). The first case examined in this paper was a four-year-old patient who presented with complaints of coughing and wheezing. The other case was a new-born with respiratory distress and related clinical symptoms that developed immediately after birth.

Although there are different classifications for vascular rings, more than 95% of these classifications are grouped into four main categories (Table 1) (8). The most common classification is double aortic arch anomaly (9). Typically, both arches are patent and the right arch is mostly dominant in this anomaly that almost always forms a tight ring (2). This anomaly usually shows a clinical picture that is more severe and emerge earlier than other vascular ring types. Two arches originate from the ascending aorta, cross on either side of the trachea-oesophagus, and join the descending thoracic aorta. (9). The right (posterior) arch is retrooesophageal. Usually one arch is dominant while the other arch is smaller or might be atretic (9,10). In our case, the left-side lumen was somewhat wider than the right-side one. We did not observe stenosis or occlusion at any level. Cardiovascular anomalies, such as tetralogy of fallot (the most common), the coarctation of aorta and patent ductus arteriosus, may accompany this anomaly (9,10). No additional cardiac abnormality was identified in our case.

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

I-Double arch aorta
II-Right arch aorta + left lig. arteriosus
III-Innominate artery compression
IV-Pulmonary artery sling

*These 4 groups comprise of more than 95% of all vascular rings.

The second most common type of vascular ring anomaly is the right - sided aortic arch. This anomaly is classified into three groups (11). The most common branching patterns of the right aortic arch are the mirror - image branching pattern (type II) (9). In this type, the great vessels originate from the arch in the following order: left innominate artery, right common carotid artery. and right subclavian artery (11). This type also has an aberrant left subclavian artery. A type II right aortic arch anomaly was seen in our case.The right aortic arch can be associated with congenital cardiac malformations, such as

persistent truncus arteriosus, tetralogy of fallot and pulmonary atresia with ventricular septal defect. Tetralogy of fallot is seen in 30 % of individuals in this patient group. However, no additional cardiac abnormality was identified in our case. An intact retro-oesophageal left ligamentum arteriosum, ductus arteriosus and Kommerell's diverticulum (an embryological diverticulum of the proximal descending aorta) may also accompany a right aortic arch anomaly.

The diagnosis of a vascular ring is established utilizing chest radiography (posterior-anterior and lateral), a barium-contrasted oesophageal graph, transthoracic echocardiography (ECO), contrast-enhanced MDCT, magnetic resonance imaging (MRI), and angiography. Today,the ECO, MRI, and contrast-enhanced MDCT are the most preferred diagnostic methods for vascular rings (6,9,10). The PA chest radiography shows that there is a single-or double-sided emphysema due to compression and a right-or left-sided tracheal compression, while the lateral chest radiography shows a compression on the anterior of the trachea. Alsenaidi et al. (12) in their study of 81 patients found that chest radiographies were normal in 20% of the cases. We also did not observe any significant characteristic in the chest radiographies of the cases in our study. The barium-enhanced oesophagus radiographies show that the oesophagus is exposed to anterior, posterior, and lateral compression. Indentations caused by compression can occur at different levels and in different sizes (6,10). No barium-enhanced examination was performed for either case in this study. PA chest radiography and barium-enhanced radiographies can used to confirm the diagnosis of a vascular ring. However, cross-sectional imaging is usually required to define the exact morphology of the aortic arch malformation and for planning an optimal surgical approach (10).

The degree, level, and length of the tracheal compression of the aortic arch can be clearly monitored using MDCT and MRI. The ability to show surrounding structures in addition to vascular structures and to provide images in three planes is the most important advantage of cross-sectional imaging (6,9). The mirror-image branching of the right-sided aortic arch as well as the 'four-artery sign' and marked tracheal compression, the characteristic symptom of a double aortic arch, was clearly demonstrated in our case.

MDCT is a fast imaging method. Unlike MRI, it does not require sedation in most cases. Sedation can also cause serious problems in patients with airway obstruction (6). Whereas intubation instruments for patients requiring airway assistance must be MRI-compatible, such instruments are not required for CT imaging. MDCT provides more precise information for evaluating airways and lung parenchyma (10). Furthermore , multiplanar reformat (MPR) and volume rendering (VR) reconstruction allows for examining images in each of the three planes and three dimensions. A 3D volume-rendered display effectively indicated the ring and sizes of the arches in our cases. In addition, minimal compressions on the trachea can be observed using the minimum intension projection (MinIP). Using 3D methods to examine these compressions can only reveal airways and parenchyma. We use these post-processing images to distinguish the tracheal stenosis. The most significant drawback is the use of X-ray, whose use has a particular importance especially during the paediatric period (6,9,10).

The following parameters can be preferred for good MDCT images in paediatric patients (9). Tube current: 0–3 kg–60 mAs/slice, 3–6 kg–80 mAs/slice, 6–10 kg–100 mAs/slice, 10–15 kg–120 mAs/slice. Tube voltage: 80-100 kV. Reconstructed slice thickness: 0.9 mm. 80 kV value and iterative reconstruction algorithm can be used to reduce CT doses. We used 80 kV for the new-born patient and 100 kV for the four-year old patient.

Conventional angiography is used to specifically determine topographic vascular anatomy. However, this method has disadvantages such as inability to determine atretic vascular structures, tracheal and/or oesophageal compression (6, 10). It is important to perform ECO in order to exclude accompanying anomalies in suspected vascular ring (9).

Surgical intervention for vascular ring is recommended for symptomatic patients with airway compression identified radiologically (13). The surgeon must make a decision of approach method according to the vascular ring anatomy and the associated tracheal or cardiac anomalies (11).

CONCLUSION

A vascular ring should be considered in the event of recurrent lung infections, unexplained persistent wheezing, and stridor during childhood as well as in the case of persistent respiratory distress in new-born children. And MDBT is an excellent modality for patients suspected of having a vascular ring. It allows evaluation of the aortic arch and its branches and provides valuable information for planning surgical management with multiplanar and three-dimensional (3-D) imaging.

CONSENT

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

ETHICAL APPROVAL

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

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