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Double Aortic Arch: About One Case

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Abstract **Case Report**

A double aortic arch is a rare vascular anomaly. However, this is the most common cause of a symptomatic vascular ring malformation due to the absence of involution of the caudal dorsal aorta. It involves the complete encirclement and compression of the trachea and esophagus. The disease usually begins to show itself in very early clinical signs, already detectable in the neonatal period. It may lead to significant morbidity for the patient, a wide range of clinical symptoms ranging from life-threatening symptoms to no symptoms can be resulting; regardless, their detection is extremely important before undertaking procedures or making surgical decisions. If associated symptoms are present, surgical correction of the vascular ring should be performed. For accurate diagnosis and evaluation of aortic arch anomalies, cross-sectional imaging modalities such as CT or MRI play an important role in providing three-dimensional reconstructed images. We here report one case of double aortic arch to highlight the contribution of imaging in the difficult diagnosis of this anomaly.

Keywords: Double Aortic Arch, Imaging, CT-Angiography.

INTRODUCTION

The double aortic arch is a one of the most common forms of vascular ring malformation due to the absence of involution of the caudal dorsal aorta [1].

Most of the double aortic arch cases are diagnosed within the first year of life. In the absence of tracheal compression, this anomaly may go unnoticed. Classically, the double arch aortic is classified into three types depending on the relative size of the two arches and partial atresia of one arch. For accurate diagnosis and evaluation of aortic arch anomalies, cross-sectional imaging modalities such as CT-Angiography or MRI play an important role in providing three-dimensional reconstructed images.

CASE REPORT

Female infant, 7 months old, vaginal delivery at term, immediate crying, no history of consanguinity, who was presented since the age of 2 months with stridor, wheezing and bronchial congestion. The patient was treated with corticosteroids for 2 months with no improvement. Given the persistence of wheezing, she was admitted to our hospital for management with chest radiography (figure 1), The paraclinical work-up was completed by a thoracic CT- angiography, showing a double aortic arch forming a vascular ring around the trachea and reducing its lumen, morphologically, our patient has right arch dominance, left arch was hypoplastic (figure 2). With a normal cardiac ultrasound, the patient was referred to surgery for further management.

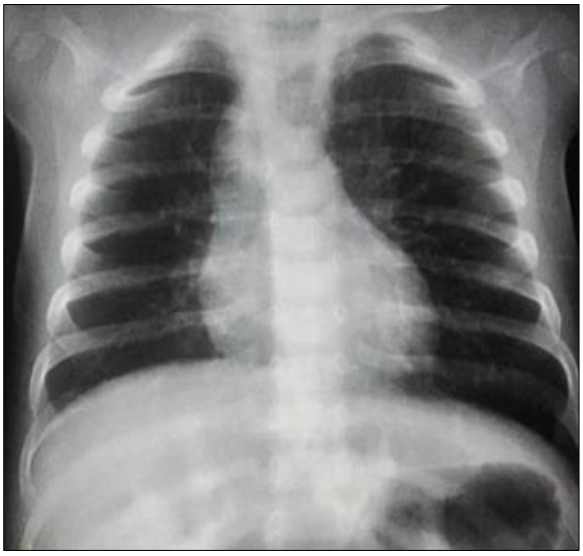

Figure 1: Front thoracic X-ray showing effacement of the aortic button and thoracic distension.

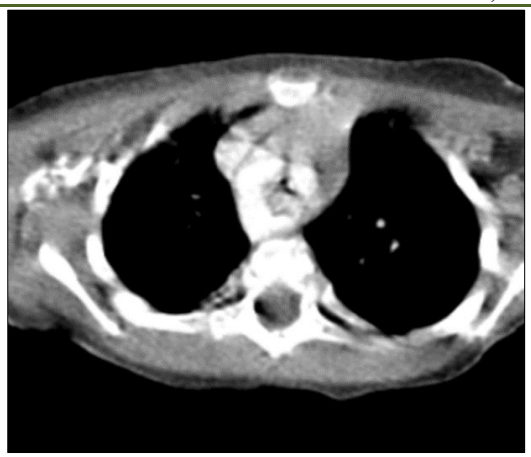

Figure 2: Thoracic CT-angiography in axial section showing a double aortic arch predominantly on the right, laminating the tracheoesophageal tract.

DISCUSSION

There is a wide range of congenital anomalies or variations of the aortic arch, from non-symptomatic variations that are most often detected accidentally, to clinically symptomatic variations that cause severe respiratory distress or esophageal compression. Some of these may be accompanied by other congenital heart defects or chromosomal abnormalities.

A double aortic arch is a rare anomaly found in only 0.05–0.3% of the population. However, this is the most common cause of a symptomatic vascular ring (symptoms occurs in 40–50% of cases) [2, 3].

Understanding the embryological development of the aortic arch helps to classify various subtypes of aortic arch anomalies and variants [4]. The development of the aorta occurs in the third week of gestation. In embryonic vascular development, six pairs of aortic arches connect the two primitive ventral and dorsal aortae [5]. A portion of the right fourth arch regresses, leaving the left aortic arch. If the right and left fourth arches both persist, it results in a double aortic arch.

Classically, the double arch aortic is classified into three types depending on the relative size of the two arches and partial atresia of one arch: right dominant (75%), left dominant (20%), and balanced (5%) [4]. The descending thoracic aorta in a double aortic arch is usually found on the contralateral side to the dominant arch. [6].

Usually, both archs are permeable. In 75% of cases, according to the literature [4-7], as in our patient's case, the right arch was dominant.

The age of revelation of the disease was early in our case (7months), which seems in concordance with the literature when the age of revelation is generally before 3 years [1], although in 25% of cases, the diagnosis is made in an adult age [1].

Most of the double aortic arch cases are diagnosed within the first year of life, likely due to symptoms of tracheal compression leading to persistent cough, stridor, wheezing, recurrent pulmonary infections, and choking on feeds. In the absence of tracheal compression, this anomaly may go unnoticed [8].

The double aortic arch can be difficult to diagnose, as the symptoms are not typical of a cardiac disorder [8]. The clinical presentation is dominated by respiratory symptoms (stridor, respiratory distress, barky cough, severe cyanosis…) and digestive symptoms (dysphagia) [8-9]. The importance of the signs depends on the space between the two aortic arches. Sometimes the diagnosis can be confused with asthma, bronchiolitis or recurrent pneumonia [9]. It like was the case of our patient treated for asthma with high-dose inhaled corticosteroids but with no recovery.

Accompanying intracardiac defects are found in 20% of double aortic arch patients. This congenital vascular anomaly may be isolated or associated with cardiac malformations such as tetralogy of Fallot, ventricular septal defect, patent ductus arteriosus, pulmonary atresia, coarctation of the aorta or extracardiac malformations such as microdeletion of chromosome 22q11 in Di George syndrome [4]. Ultrasound of our patient came back normal.

Imaging techniques available to diagnose arch anomalies include echocardiography, CT angiography, MRI, catheter angiography and barium esophagram [10]. The conventional techniques such as barium esophagram and catheter angiography are not frequently used as primary because they are difficult to use in paediatric patients and provide only two-dimensional (2D) information [11].

Currently, Computed tomography angiography, MRI and echocardiography are the main modalities used to detect and evaluate the double aortic arch [4]. Chest

radiography may be required to suggest vascular compression.

CT provides high spatial resolution of the anatomy of the vessels and surrounding structures. Additionally, reformation in multiple planes or three-dimensional (3D) imaging can provide detailed anatomical information, including relative positional relationships, which is advantageous, especially when assessing tracheal and esophageal compression [12].

CT can be performed without anesthesia or breath holding in pediatric patients [13]. Despite improvements in CT techniques, however, there are still concerns regarding radiation exposure and administration of iodine contrast media.

Diagnostic aortic arch imaging by CT angiography has several minimal requirements for optimal quality [14]. First, the minimal scan range must include the ascending thoracic aorta, descending thoracic aorta, and large aortic arch branches (brachiocephalic artery [BCA], common carotid artery [CCA], and SA). Second, at least 200-250 HU should be used for adequate aortic enhancement [15]. Computed tomography angiography (CTA) contrast enhancement depends on various factors such as contrast medium volume, injection rate, scan duration, scan timing, and patient-related factors. By adjusting these variables, appropriate contrast enhancement can be achieved [16]. Third, thin slice thickness is an important factor affecting CTA spatial resolution. For adequate diagnostic aortic arch imaging, slice thickness should not be more than 2 mm. Further multi-planar reconstruction using thin slices can provide additional anatomic information for diagnosis.

When scanning pediatric patients, more variables should be considered including small body volume and sensitivity to radiation exposure [17]. Pediatric CT-Angiography optimization aims to minimize radiation exposure while obtaining images with adequate diagnostic quality.

MRI is an alternative cross-sectional imaging modality that allows precise assessment of double aortic arch without ionizing radiation. Contrast-enhanced MR angiography and non-contrast enhanced black blood and bright blood sequences can be performed to evaluate anatomical relationships between the aortic arch, esophagus, and airway [18]. Like CT, 3D volumetric acquisitions provide multi-planar reformatting for detailed assessment of tracheal compression. However, the biggest drawback of MRI is the long acquisition time, patient cooperation is a factor to consider due to the prolonged time requirements.

Echocardiography is often used as the initial imaging modality in pediatric patients as it is non-invasive, there is no need for intravenous contrast medium, and there is no radiation exposure [19]. Echocardiography not only provides anatomic information on the vascular and surrounding structures, but functional changes can also be assessed by Doppler ultrasonography. However, limited access to the entire thoracic aorta and dependency on operator skills are limitations of echocardiography as a major imaging modality.

Any symptomatic infant whose TOGD reveals an abnormal esophageal impression should be referred for cross-sectional imaging.

If associated symptoms are present, surgical correction of the vascular ring should be performed [20]. The surgical procedure is based on left posterolateral thoracotomy involving cooperation of an experienced surgical team and good anesthetic preparation. During surgery, the smaller segment or atretic portion of the aortic arch is usually resected to disrupt the complete vascular ring and resolve tracheoesophageal compression. The long-term prognosis of patients with a double aortic arch who have undergone surgical treatment is excellent [21].

CONCLUSION

The widespread use of multidetector computed tomography (CT) in clinical practice has resulted in detection of the double aortic arch like a several variation of the aortic arch. Thus, radiologists and clinicians should carefully look for imaging features associated with a high risk of clinical symptoms.

Nowadays, with new technology, an angio-CT with reconstruction gives excellent images in two or three dimensions. MRI, like CT, gives a precise anatomical precise anatomical analysis, it is not irradiating and is considered no invasive but it necessitates heavy sedation.

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