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The role of CT angiography in preoperative evaluation of aortic coarctation

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ABSTRACT

Aortic coarctation (CoA) constitutes 5-8% of all congenital heart diseases. Physical examination findings and imaging methods are helpful in diagnosis. Computed tomographic (CT) angiography and transthoracic echocardiography are the common diagnostic tools for aortic coarctation. In this study, we aimed to evaluate the intracardiac and extracardiac anomalies that we detected in our cases. We also evaluated the contribution of preoperative diagnosis of extracardiac anomalies in preventing surgical complications. From January 2016 and May 2018, we enrolled 37 infants with clinically suspected CoA who underwent CT angiography and transthoracic echocardiography and operated in our hospital. The extracardiac and intracardiac anomalies associated with CoA were evaluated. Extracardiac anomalies that were not seen in transthoracic echocardiography but diagnosed by CT angiography were evaluated. The contribution of CT angiography in surgical planning was determined. The patients (24 males and 13 females) were aged from one day to 240 months. Of this sample, 54% of thirty-seven patients were in the neonatal period. When we examined the accompanying intracardiac pathologies, ventricular septal defect was present in three cases, atrial septal defect in seven cases, subaortic membrane and Shone complex in two cases. Extracardiac anomalies such as, tracheal duplication, Scimitar syndrome, pulmonary vein course anomaly and left bronchial pressure were detected by CT angiography in 2.7% of the patients. Abnormal right subclavian artery was present in 10.8% of the cases and the surgical team was more sensitive to paraplegia measures. In 19% of the longsegment coarctation; if the surgical team consider it too far for end-to-end anastomosis, they chose synthetic tube graft for the patient prior to the operation. The results indicated that the CT angiography is a beneficial non-invasive method for confirming CoA and detecting the accompanying extracardiac anomalies in children with CoA. Preoperative morphological features and extracardiac anomalies which were detected by CT angiography were found to be reliable for surgical planning.

1. Introduction

Coarctation of the aorta (CoA) is a common congenital heart disease that appears approximately three in every 10000 births (Ringel et al., 2012; Beekman, 2013). Generally, the diagnosis is made during childhood, but there are frequent cases that are not diagnosed

until adulthood (Brickner et al., 2000). It often occurs in a discrete or tubular form in the distal of the left subclavian artery. CoA could be an isolated disorder as well as it could be seen with aortic arch hypoplasia, left heart lesions (aortic stenosis, mitral stenosis, hypoplastic left heart syndrome), atrial septal defect (ASD), ventricular septal defect (VSD), bicuspid aorta, Turner syndrome and aneurysm of circle of Willis (Attenhofer et al., 2002).

In addition to the physical examination, imaging methods are helpful in the diagnosis (Mellander et al., 2006; Gómez-Montes et al., 2013). Today transthoracic echocardiography, CT and MR angiography are the most preferred diagnostic methods. Computerized tomography angiography is one of the most frequently used methods due to its short duration of examination, sedation time, and its applicability in patients with poor general condition (Lee et al., 2004; Tsai et al., 2008). The location of coarctation, collateral circulation, relation with the left subclavian artery, arcus hypoplasia, and other extravascular anomalies that may accompany can be detected by CT angiography (Frush, 2005; Taylor, 2008). However, the role of detecting these extracardiac anomalies for the success of surgery has not been studied enough yet. Catheter angiography is used today for treatment rather than diagnosis. Compared to catheter angiography, CT angiography is exposed to a lower dose of radiation.

In the current study, we investigated the role of CT angiography in the preoperative evaluation of patients and its effectiveness on mortality during surgery.

2. Materials and methods

Patient population

We reviewed the charts of 37 consecutive infants who were diagnosed as CoA clinically and admitted to our hospital between January 2016 and May 2018. The research procedure was approved by the local ethical committee (No:2018/7-13). Demographics of the patients were also analyzed: Symptoms, age, gender distribution, weight, height, imaging data and operation procedure. At the same time, we evaluated the frequency of congenital heart defects associated with aortic coarctation. We included patients with mild associated cardiac anomalies that were not addressed at the time of the coarctation repair (ie, atrial septal defect or ventricular septal defect bicuspid aorta, aortic stenosis and Shone complex) but excluded complex congenital heart defects (ie, single ventricular physiology double outlet right ventricle -DORV and hypoplastic left heart syndrome- HLHS etc.). We evaluated the surgical benefits of knowledge of extravascular anomalies not detected in echocardiography and detected in CT angiograhy. We investigated the role of CT angiography in the evaluation of preoperative patients and its effectiveness on mortality.

Transthoracic echocardiography

All transthoracic echocardiography was performed by the pediatric cardiologist using the Philips Ultrasound System and the S8-3, S3-1 probe. The transthoracic echocardiography protocol recommended by the European Guidelines was applied to all patients (Evangelista et al., 2008). Intracardiac defects were evaluated by the apical four-chamber view, left ventricular long axis, suprasternal examination, parasternal short-long axis and subcostal examinations. During the suprasternal examination, the location and length of the CoA, diameter of the coarctation zone and the transverse aorta were evaluated. Color Doppler imaging was used to evaluate the blood flow and measure the differential pressure and maximum speed in the location of the CoA.

CT image acquisition

Common indications for CT angiography included transverse arch hypoplasia, poor echo windows, complex geometry, or uncertain arch branching pattern; surgical preference for three-dimensional (3-D) imaging was also a considerable factor. Computerized tomography angiography scan was performed with a Siemens Definition AS 128 section device and required cases were treated with chloral hydrate in a sedative manner with a dose of 50 mg/kg rectally or orally. Patients were examined in supine position, and we took images extending from the base of the neck to the diaphragm. A low radiation and contrast agent protocol was applied in the procedure according to age and weight. The non-ionic contrast agent was administered with a dosage of 0.8 cc/kg using antecubital vein. The lowest radiation dosage was applied to the tissue using 'care dose' technology according to the body area (collimation 16x1.25 mm, slice thickness 0.75 mm, slice interval 0.625 mm, pitch set automatically, reconstruction interval 0.625 mm).

The following analyses were performed in each patient for diagnostic evaluation.

i) Axial single-phase low dose multidetector CT scans and two-dimensional sagittal and coronal reconstructions.

ii) Maximum intensity projection.

iii) 3D image reconstruction included multiplanar reformatting (MPR), curved planar reformatting, maximum intensity projection (MIP), and volume rendering (VR).

All data were evaluated by a pediatric cardiologist and cardiac surgeon on the basis of preoperative chest x-ray and echocardiography.

Statistical analysis

Data were analyzed using IBM SPSS Statistics 25.0 (IBM Corp., Armonk, New York, USA) statistical package program. Descriptive statistics are given as

number of units (n), percentage (%), mean ± standard deviation. The sensitivity and specificity were used to delineate the diagnostic accuracy of CT angiography and TTE for the extracardiac and intracardiac malformations in each group.

3. Results

During the study period the data of thirty-seven patients who underwent surgery for CoA were evaluated. The patients were aged from one days to 240 months and the mean body weight ranged from 2 to 80 kg. Of this sample, 54% of 37 patients were in the neonatal period. According to the examination by gender, 35% were female and 65% were male. A murmur of the heart was found in 44% of admitted patients, followed by respiratory distress with 19% and antenatal diagnosis with 12%. Short segment coarctation was present in 81% of the patient and in 19% long segment coarctation was seen.

When we examine the accompanying intracardiac pathologies, VSD was present in three cases, ASD in seven cases, subaortic membrane and Shone complex in two cases. Pulmonary artery banding operation was performed in two of our patients with large muscular VSD. The prevalence of bicuspid aortic valve was 30%. The (patent ductus arteriosus) PDA was observed in 11 cases in the newborn period. There was one case who had Turner syndrome, one had VACTERL (vertebral defects, anal atresia, cardiac malformations, tracheoesophageal fistula, renal anomalies and extremity anomalies), one with reactive airway disease, one case with a single kidney and one had epilepsy. Aberrant right subclavian artery was seen in 10.8% of the cases (Fig.1). Also with the help of CT angiography, one patient was diagnosed with tracheal duplication, one patient with Scimitar syndrome, (Fig. 2), one patient with a pulmonary vein course anomaly, and one patient with left bronchial pressure.

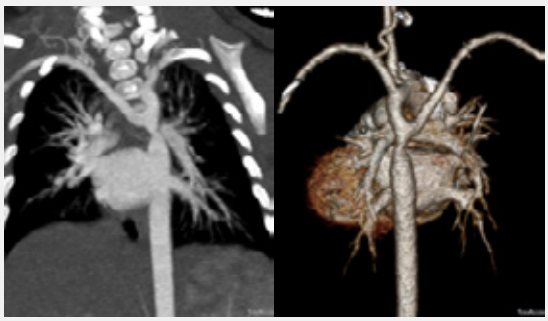

Fig. 1. CT angiography of 3 years old girl with aortic coarctation and aberrant right subclavian artery association in coronal image. The same patient 3D reconstruction image also seen on the left part of image.

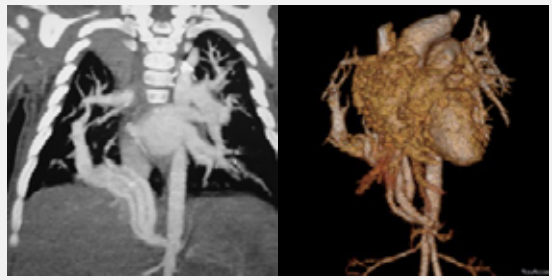

Fig. 2. Coronal CT angiography of a 6-months old case with Scimitar syndrome + recoarctation. Aorto-pulmonary collateral artery, Scimitar vein and the clips in the recoarctation region. The same patient 3D reconstruction image also seen on the left part of the image.

Enlarged end-to-end anostomosis was performed on 29 (78%) of our patients after resection, four patients were treated with tube graft interposition (10.8%) and aortoplasty with xenograft pericardial patch and glutaraldehyde treatment was administered in three cases (8.1%). If it seems too far for end-to-end anastomosis with measurements by CT angiography, a synthetic tube graft was chosen for the patient prior to the operation. In one case, an arcus hypoplasia was detected and arcus reconstruction surgery was performed (Fig. 3). Isthmus hypoplasia was detected in five cases with CT angiography and the operation plan was not altered because it was not severe. Abnormal right subclavian artery was present in 10.8% of the cases and the surgical team was more sensitive to paraplegia measures. In a15 days old case re-coarctation was encountered due to PDA clips or fibrotic stenosis at the site of surgery (Fig. 4). In one case, left bronchial compression was observed due to the abnormal course of the aortic arch, and after end-to-end anastomosis, posterior aortoplasty procedure was performed on the descendant aorta. Aortic patch plasty was preferred only in patients with re-coarctation. Tube graft interposition was favored in patients with long segment stenosis and isthmus hypoplasia in adult type aortic coarctations. In all other patients, extended end-to-end anastomosis operation was performed after resection. All patients' operations were performed using a simple cross-clamping technique. There was no complication in our patients and there was no death. Detailed information of the patients is given in Table 1.

4. Discussion

In our study detection rate of transthoracic echocardiography for intracardiac anomalies and CoA were 100% while the rate for extracardiac anomalies such as an aberrant right subclavian artery (ARSA) and Scimitar syndrome were 86.5% (using surgery as the gold standard). With the advances

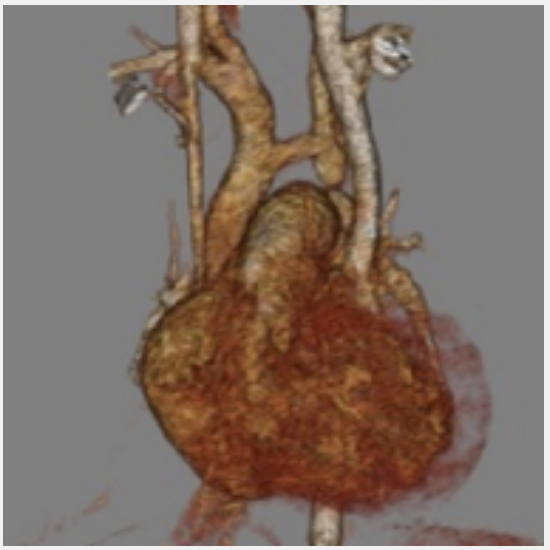

Fig. 3. CT Angiography 3D reconstruction image. four-month old; aortic coarctation + arcus hypoplasia.

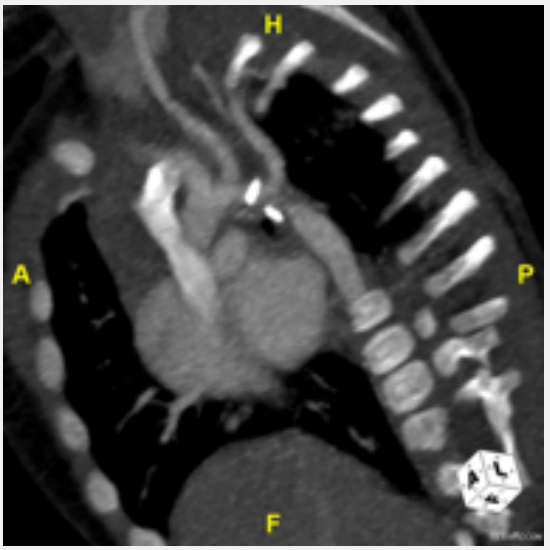

Fig. 4. Sagittal multiplanar examination showed re-coarctation due to PDA clips or fibrotic stenosis at the site of surgery in the 15-days-old case operated for aortic coarctation.

in transthoracic echocardiography devices and the increased experience of the clinicians performing it, it is now one of the best noninvasive and easily applied methods in the diagnosis of congenital heart diseases in infants and young children. Other commonly used methods are catheter, CT and MR angiography. There are some studies comparing CT angiography with TTE in diagnosis of congenital CoA and accompanying malformations (Al-Azzazy et al., 2014). In the diagnosis of CoA; sensitivity of CT angiography was found to be 100%, while the sensitivity of transthoracic echocardiography was 91%, and for the diagnosis of VSD, the specificity of CT angiography was found as 91.7% (Turkvatan et al., 2009; Rose-Felker et al., 2017). Didier et al., observed MR, MR angiography and transthoracic echocardiography results were evaluated preoperatively and postoperatively and it was observed that especially in postoperative patients transthoracic echocardiography was less effective in showing pseudo-aneurysms, isthmus hypoplasia, and re-coarctation. (Didier et al., 2006). Lee et al., reported 14 newborn cases referred for catheter angiography were correctly diagnosed with CT angiography and catheter angiography was no longer required. (Lee et al., 2006).

Preoperative knowledge of the extracardiac anomalies and arcus hypoplasias by the surgeon provide significant benefit in terms of operation selection (Stone et al.,1990; Adaletli et al., 2011). In our cases of long and hypoplastic segment coarctation the surgical team prepared for patch enlargement of arcus aorta and patchplasty procedure can be done safely when the PDA was open. In addition if the surgical team consider the case too far for end to end anastomosis they may choose a syntetic tube graft for the patient. It may be difficult to decide tube grafts size intraoperative moment but if the team was prepared on images for the case this is not an issue for procedure.

Our cases who were diagnosed with ARSA were between the ages of 0-3 years and all of them were localized behind esophagus and caused esophagus compression. In these patients, surgical division performed on ARSA to release the aorta in order to anastomose the tips and decompress the pressure on the esophagus. When aortic coarctation is accompanied by ARSA the rate of morbidity and surgical strategy change. If the patient have an aberrant artery associated with coarctation then the surgeons awareness is very important because we know that these patient groups have higher paraplegia ratio (Kieffer et al., 1994). At this situation the surgical team may be more sensitive for paraplegia precautions. Complications did not develop in our patients.

Computerized tomography angiography was helpful in the diagnosis of some patients. One of these patients (case 31) who was referred to our unit from another center with re-coarctation diagnosis. The echocardiographic evaluation revealed significant enlargement on the right side structures and pulmonary hypertension. The patient had obvious respiratory distress, CT angiography was planned hence the general condition of the patient was not appropriate for catheter angiography. Our case with right lung hypoplasia was found to have Scimitar vein and large aorta pulmonary collateral arteries. In another patient (case 19), partial tracheal duplication was detected by CT angiography. This anomaly was an uncommon and

Table 1. Demographic characteristics, symptoms and results of various imaging methods in 37 patients with CoA.

No	Age	G	Symptoms	Type of CoA	BAV	Accompanying intracardiac pathology	The defect detected in CT-angiography	Comorbidity	Type of operation
1.	1 M	M	Murmur	Short		PFO			End-to-end
2.	20 Y	F	Murmur	Long				Turner Syndrome	Tube Graft
3.	1 M	M	Tachypnea	Long		PFO, PDA, hypoplasia of arcus aorta	Hypoplasia of İsthmus, PDA		End-to-end
4.	1 M	M	Antenatal diagnosis	Short		PFO, large muscular VSD	VSD		End-to-end, pulmonary artery banding
5.	5 M	M	Murmur	Short					End-to-end
6.	17 Y	M	Control	Long	+	Shone complex, subaortic membrane			Tube Graft
7.	8 M	M	Murmur	Short	+	Shone complex			End-to-end
8.	2 Y	F	Tachycardia	Short					End-to-end
9.	1 M	F	Murmur	Short			Moderate hypoplasia of arcus	Epilepsy	End-to-end
10.	5 D	M	Dyspnea	Short		PDA,PFO, Arcus hypoplasia	Hypoplasia of isthmus, PDA	Meningo-myelocele	End-to-end
11.	12 Y	M	Growth retardation	Long	+	PDA	PDA	Left renal agenesis	Tube Graft
12.	2 D	M	Dyspnea	Short		PDA, ASD	PDA		End-to-end
13.	10 D	M	Murmur	Short	+	PFO			End-to-end
14.	2 D	F	Dyspnea	Short		PDA	PDA		End-to-end
15.	1D	F	Antenatal diagnosis	Short		PDA	PDA		End-to-end
16.	5 D	M	Dyspnea	Short	+	PDA	Hypoplasia of isthmus, PDA		End-to-end
17.	1 D	F	Newborn congenital anomaly	Short		PDA, ASD	Hypoplasia of isthmus, PDA	VACTERL	End-to-end
18.	1 D	M	Murmur	Short	+	PDA, ASD	PDA		End-to-end
19.	10 D	F	Murmur	Short		Moderate hypoplasia of arcus aorta		Tracheal duplication	End-to-end
20.	1 M	F	Murmur	Short	+	LPSVC	ARSA, LPSVC		End-to-end, division of ARSA
21.	3 M	M	Murmur	Short	+		ARSA		End-to-end, division of ARSA
22.	4 M	F	Murmur	Long		Large muscular VSD, hypoplasia of arcus aorta	Hypoplasia of arcus aorta, VSD		Reconstruction of arcus aorta, pulmonary artery banding
23.	3 D	M	Dyspnea	Short		PDA	PDA		End-to-end
24.	5 D	F	Dyspnea	Short	+	ASD,TAPVD?	pulmonary vein course anomaly		End-to-end
25.	15 Y	M	Hypertension	Short	+				Tube Graft
26.	3 Y	F	Murmur	Short		VSD,subaortic membrane	ARSA		End-to-end, division of ARSA
27.	1 D	M	Antenatal diagnosis	Short		Right pulmonary agenesis? ASD	Hypoplasia of right PA, ARSA		End-to-end
28.	1 D	F	Antenatal diagnosis	Short		PFO,PDA	PDA		End-to-end
29.	2 M	F	Murmur	Short		ASD			End-to-end
30.	1 M	M	Murmur	Short		ASD			End-to-end
31.	6 M	M	Dyspnea	Short		Recoarctation,Pulmonary hypertension	Scimitar syndrome, APKA	Scimitar syndrome, APKA	End-to-end, Scimitar vein anastomosis, ligation of APKA
32.	3 Y	M	Murmur	Long				Reactive airway disease	Patch aortoplasty
33.	12 Y	M	Control	Long		Recoarctation			Pericardial patch
34.	8 Y	F	Control	Short		Recoarctation			Pericardial patch
35.	19 D	M	Dyspnea	Short		PDA, PFO			End-to-end
36.	4 M	M	Murmur	Short	+				End-to-end
37.	3,5 M	F	Murmur	Short			Compression of arcus to left main bronchi		End-to-end

G: Gender, M:Male, F:Female, PFO: Patent foramen ovale, ASD: Atrial septal defect, VSD:Ventricular septal defect, CoA: Coarctation of the aorta, TAPVD:Total anomalous pulmonary venous, LPSVC: Left-sided persistent superior vena cava, PDA: Patent ductus arteriosus, ARSA:Aberrant retro esophageal subclavian artery, CT: Computed tomography, MRI: Magnetic resonance imaging, TTE: Transthoracic echocardiography.

asymptomatic situation that is diagnosed incidentally. As long as they do not cause respiratory problems, they do not require treatment (Karcaaltincaba et al., 2004) and we did not perform any additional intervention on our patient. But sometimes fistulas can accompany and knowing the existence of a fistula pre-operatively is beneficial in planning the anesthesia management of the patient. In another patient with re-coarctation (case 28), we found that metallic clips used for PDA closure caused re-coarctation or fibrotic stenosis at the site of surgery. In this patient, we preferred surgery instead of angiography. CT angiography is excellent at finding associated extracardiac anomalies but it is not useful for visualizing the aortic gradient or other cardiac malformations (Huang et al., 2017). In the case of recoarctation it may be vital the examining the CT images before operation. Because the if the surgeon examines CT images; dissection may be more easy and safe. At such cases the surgical team may find a reason for coarctation (large titanium clips applied to ductus, intraaortic thrombosis, pseodoaneurism etc.). We believe that the planning the operation and discussion from obtained images may help to improve safety and quality of the procedure. Maybe the clinicians use CT for the diagnose but on the other side the surgical team uses images for planning, avoiding complications. Our study has some limitations that to be considered. First, relatively small sample size is foremost limitation of our study. Secondly, this study was limited by its retrospective design and clinically obtained data.

As a result, we conclude that transthoracic echocardiography is the first choice for the coarctation diagnosis depending on the experiment of the clinician and CT angiography is useful for patients with poor acoustic window for TEE and extracardiac malformation suspicion. The location of coarctation, collateral circulation, relation with the left subclavian artery, arcus hypoplasia, and other extravascular anomalies that may accompany can be detected by CT angiography. The main purpose is planning a better management with the preoperative data and imaging and preventing possible complications. Knowing preoperative vascular anomalies may change the type of surgery. It can also prevent re-surgeries.

Conflict of interest

The authors declared no conflicts of interest.

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