Double outlet right ventricle with giant pulmonary artery aneurysm and severe aortic coarctation: diagnosis with multislice CT

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Complex cardiovascular pathologies in the pediatric population are usually evaluated with echocardiography and catheter angiography as initial and advanced imaging of choice, respectively. Echocardiography may pose some difficulties in the diagnosis of complex cardiovascular pathologies. Due to short acquisition times, detailed imaging by the use of post-processing techniques, reduced radiation exposure compared to catheter angiography, and additional information obtained on lung parenchyma, multi-slice computed tomography (CT) is the advanced imaging method of choice in selected cases. The present report describes a 14-year-old symptomatic case with complex cardiovascular pathology, whose vascular architecture could be properly demonstrated by multi-slice CT.

Key words: congenital heart disease, pediatric patients, imaging.

Recent advances in imaging technologies have led to the introduction of computed tomography (CT) angiography. Higher resolution images obtained by multi-slice CT (MSCT) with increasing number of slices compete with diagnostic catheter angiography. We aimed to present CT angiography findings of a rare and complex cardiovascular pathology.

Case Report

A 14-year-old boy had been treated for pulmonary infection in various centers without any response. He was then referred to the pediatrics clinic of our institution with a possible diagnosis of pulmonary tuberculosis. Physical examination revealed clubbing, cardiac murmur and heart failure. Heart rate was 98 beats/min at apex, respiratory rate was 21/min, and brachial blood pressure was 100/65 mmHg and 110/70 mmHg at right and left arm, respectively. He had low levels of albumin together with leukocytosis and neutrophilia. Tests for tuberculosis (acid resistant bacillus [ARB] and polymerase chain reaction [PCR]) were negative. On chest X-ray, the mediastinum was enlarged with increased opacity at the left middle and lower zones and a less prominent, sparse and limited increase in opacity at the lower zone of the right lung. Due to the presence of heart failure, murmur and possible findings of pulmonary hypertension, echocardiography was performed. Echocardiography demonstrated the presence of a ventricular septal defect (VSD) with bi-directional shunting and findings consistent with truncus arteriosus type 3 with subvalvular stenosis and a giant truncal aneurysm. He was referred to the Radiology Department to clarify the vascular morphologic condition and to explain clinical and radiological findings suggestive of pulmonary infection. Thoracic MSCT angiography was performed using a 64 slice scanner (Philips Brilliance), with 60 mAs and 120 kV parameters. A total of 35 cc (300 mg/ml) iodinated non-ionic contrast material was given intravenously at a rate of 4 ml/sec. Bolus tracking was placed on the descending aorta and the area from the

pulmonary apex to the base was scanned with 1.5 mm slice thickness, in 4 seconds with a delay of 4 seconds. Reconstruction interval was 0.75, pitch factor was 1.72, rotation time was 0.75 seconds and collimation was 64x0.625. During scanning, dose-length-product (DLP) was 99.5 mGy x cm and estimated effective dose was 1.28 mSv. Post-processing evaluations of axial, multiplanar, 2-dimensional MIP, 2-dimensional Min-IP and volume rendering were done at 0.5 mm thickness on the workstation (Philips Extended Brilliance Workspace, Philips Medical Systems, Best, The Netherlands). CT angiography demonstrated a double outlet right ventricle (DORV) with a giant pulmonary artery. Main pulmonary artery was aneurysmatic from the origin, and normal caliper pulmonary artery segment was not observed. Ascending aorta was displaced rightwards and anteriorly by the giant aneurysm of the pulmonary artery, which caused misinterpretation of the pulmonary artery as truncus arteriosus during the initial echocardiographic evaluation. Aortic arch was hypoplastic with advanced narrowing distal to the subclavian artery. The narrow lumen was communicating with the descending aorta at the level of the patent ductus arteriosus (PDA) in concordance with severe aortic coarctation. The descending aorta, which was almost filling via the PDA, was larger in caliper relative to the aortic arch (Fig. 1). Moreover, the PDA was also aneurysmatic as continuation of the giant pulmonary artery and the segment of coarctation was localized at the PDA orifice (Fig. 2).

CT angiography also revealed VSD, and different density of contrast material mixture at the level of septum wall defect was observed (Fig. 3). Due to the compression of the giant pulmonary aneurysm, moderate (right) and severe (left) narrowing of the main bronchi were seen. Consolidation and atelectasis due to the compression were found in the inferior lobe of the left lung (Fig. 4). Radiological findings of pulmonary hypertension were present. As surgical intervention was no longer an option in this patient due to the development of heart failure and pulmonary hypertension, he was followed with medical treatment.

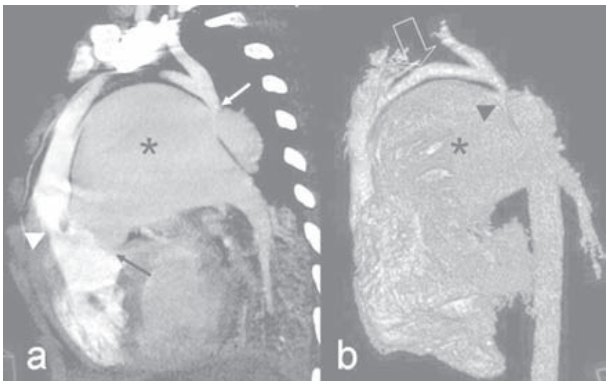

Fig. 1. Oblique sagittal 2-dimensional MIP image (a) and 3-dimensional volume rendered left lateral oblique projection (b) demonstrate double outlet right ventricle, hypoplastic aortic arch (open arrow), aortic coarctation, giant pulmonary artery (asterisk) and patent ductus arteriosus (PDA). The origin of the aorta (white arrowhead) and pulmonary artery (black arrow) is indicated. Aortic coarctation (white arrow) is at the level of the large PDA (black arrowhead) after the origin of the left subclavian artery.

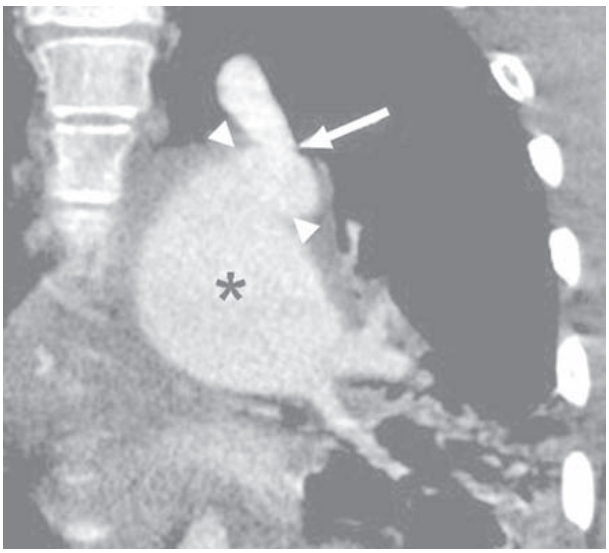

Fig. 2. Oblique coronal 2-dimensional MIP image demonstrates communication of the proximal aortic lumen (arrow) with the descending aorta at the level of the patent ductus arteriosus (PDA). Arrowheads indicate the aneurysmatic dilatation of the PDA originating from the pulmonary artery aneurysm (asterisk).

Discussion

Double outlet right ventricle (DORV) is among a group of complex cardiovascular pathologies that share the common feature that great vessels predominately originate from the right ventricle[1]. It is quite a rare anomaly with a reported frequency of 156 cases in one million live births[2]. VSD is of vital importance in these cases[3]. Localization of VSD and the presence and degree of ventricular outlet obstruction determine the physiology of DORV after birth[1]. In the present case, subvalvular pulmonary stenosis, severe aortic coarctation

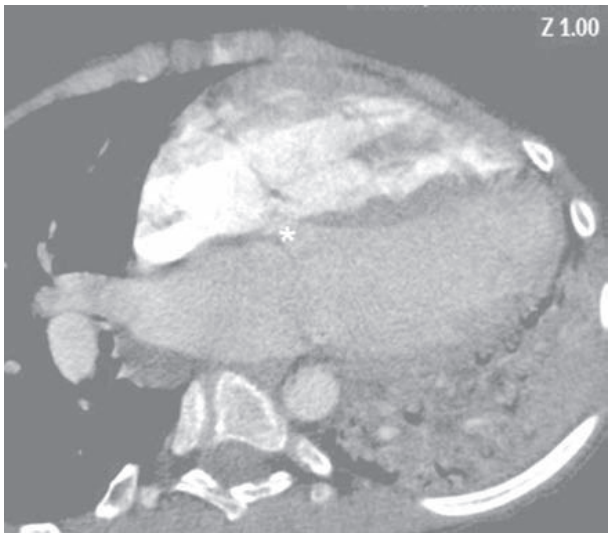

Fig. 3. Ventricular septal defect (asterisk) is shown on axial oblique reformat image.

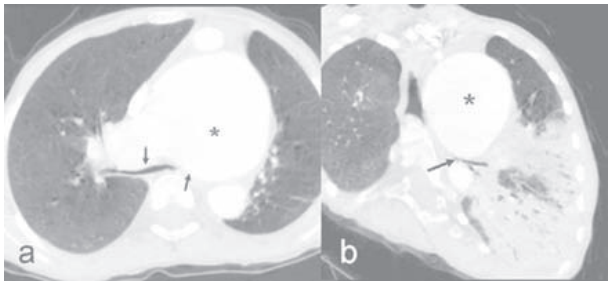

Fig. 4. Axial (a) and oblique axial (b) 2-dimensional MinIP images of lung parenchyma demonstrate narrowing of the right main bronchi and nearly total obliteration of the left main bronchi (arrows) due to giant pulmonary artery aneurysm (asterisk). There is post-obstructive parenchymal infiltration in the left lower lobe.

and large PDA probably aggravated the size of the pulmonary aneurysm. The giant pulmonary aneurysm further complicated the imaging findings of the present case. Ascending and arcus aorta, being compressed and shifted rightwards and anteriorly by the pulmonary aneurysm, was localized at the right side of the pulmonary artery. In a study of 73 cases with DORV[4], the aorta was frequently localized at the right or right anterior side of the pulmonary artery, such as in the case we report.

Coarctation of the aorta is one of the most frequent congenital anomalies of the great vessels, with an incidence around 400 cases in one million live births. As most cases are not symptomatic, the true incidence may be higher. Its frequency is approximately 6-8% among children with congenital anomalies[5]. Although varying degrees of coexistence with complex cyanotic congenital heart diseases has been reported, to the best of our knowledge, no case with the coexistence of DORV and coarctation was revealed.

Multi-slice (MS) CT angiography images of the present case revealed a severely narrowed aorta at the level of the large PDA just distal to subclavian artery, giving rise to an appearance closely resembling the quite rare condition of interrupted aortic arch. However, in interrupted aortic arch syndrome, there is no luminal or anatomical continuity between the proximal and distal segments. The highly narrow lumen of the proximal aorta in our case was in communication with the dilated PDA and descending aorta, and the condition was interpreted as aortic coarctation.

Echocardiography is frequently used as the initial imaging method. Echocardiography can differentiate tetralogy of Fallot, truncus arteriosus and DORV from other pathologies with the finding referred to as 'echocardiographic discontinuity'[7]. Additional presence of aneurysmatic dilatation of the PDA and further anterior shifting of the aorta by the giant pulmonary artery aneurysm limited the value of echocardiographic evaluation in our case. Aside from the fact that catheter angiography is an invasive imaging modality, it also requires both arterial and venous intervention for imaging the distal part of the stenosis and imaging of the heart/proximal aorta, respectively. Moreover, high flow through large VSD results in dilution of contrast material, which causes suboptimal evaluation of the heart and proximal aorta[6]. When compared with MSCT, the main disadvantage of catheter angiography, particularly in children, is the four to five times more exposure to radiation[8]. On the other hand, magnetic resonance (MR) imaging is non-invasive without any radiation risk. Three-dimensional images after contrast administration are effective diagnostic methods for anomalies of the aortic arch in children and adults[6]. However, respiratory and flow artifacts result in poor imaging of spin echo images of the thoracic aorta.

With the introduction of MSCT devices, evaluation of complex cardiovascular anomalies seen during childhood has become easier, particularly in cases in which other non-

invasive techniques are troublesome. With 64-slice CT scanner, thoracic CT angiography can be performed in as short as 4 seconds and a large area can be scanned without any need for respiratory triggering, eliminating the possibility of respiratory artifacts. The amount of contrast material used is also reduced. The main disadvantage of MSCT is radiation exposure, particularly in children. However, radiation dose is 4-5 times reduced compared to catheter angiography, when single-phase contrast examination is done. When evaluation of cardiac function and coronary imaging is not the aim, single-phase thoracic CT angiography rather than multi-phase examination with ECG triggering is preferred. In addition to the advantage of short acquisition times, easy-to-use workstations enabling post-processing techniques such as multi-planar 2-dimensional MIP, 2-dimensional Min-IP and 3-dimensional volume rendering provide highly detailed information in cardiovascular pathologies.

In conclusion, this case shows the clinical value of MSCT for non-invasive assessment of a complex congenital heart disease. Echocardiography as the initial evaluation technique may pose some difficulties in the diagnosis of complex cardiovascular pathologies. Due to short scan times, detailed imaging with the use of post-processing techniques, and additional information obtained on lung parenchyma, MSCT has become an important complementary imaging technique in the evaluation of complex cardiovascular pathologies.

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