

Case Report

Adolescent presentation of interrupted aortic arch with extensive collateral flow: a case report diagnosed by computed tomography and echocardiogram

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Abstract: Interrupted aortic arch is a rare congenital cardiac anomaly which commonly detected in the perinatal period or during infancy. Here, we present the case of a 14-year-old adolescent with type A, interrupted aortic arch, which identified by computed tomography angiography and echocardiogram. This interesting case have small ventricular septal defect, small patent ductus arteriosus but extremely rich collateral circulation.

Keywords: Echocardiography, aortic arch, congenital heart defects, computed tomography

Introduction

Interrupted aortic arch (IAA) is a rare congenital cardiac anomaly characterized by interruption between the ascending and descending aorta. Typical IAA is often combined with large ventricular septal defect (VSD) and large patent ductus arteriosus (PDA) which supplies blood to the abdomen and lower limbs. Here, we reported a rare case of IAA with small VSD and PDA but with extremely rich collateral circulation.

Case report

A 14-year-old boy presented with cough, palpitation, and 12-year history of heart murmur. On admission, physical examination revealed a bruit over bilateral carotid artery, subclavian artery, back, chest and abdominal aorta and a grade of 3/6 systolic murmur at precordial region. His respiration rate was 20/min, heart rate 92 beats/min. Blood pressure was 120/93 mm Hg in the right arm, 116/90 mmHg in the left arm, 114/91 mmHg in the right lower limb, and 118/94 mmHg in the left lower limb. The oxyhemoglobin saturation of this patient between upper limb and lower limb was not significantly different.

A transthoracic echocardiogram revealed thickened of left and right ventricular wall with nor-

mal left ventricular function (ejection fraction of 70%). The suprasternal notch view showed the aortic arch and descending aorta were discontinuous, and there was no blood flow between them (**Figure 1**). Meticulous echocardiographic revealed ventricular septal echo interruption of 3 mm and shunt signal from left to right (**Figure 2**), arterial duct of 4 mm connect with pulmonary artery and the descending aorta with shunt signal from descending aorta to pulmonary artery both systolic and diastolic (**Figure 3**). No other abnormality except mild aortic valve regurgitation was noted.

Computed tomography angiography confirmed interruption of the aortic arch 13 mm distal to the origin of left subclavian artery (**Figure 4**). Three-dimensional reconstruction showed arterial duct put pulmonary artery connected to the descending aorta (**Figure 5**), numerous tortuous collateral vessels were seen including prominent intercostal arteries and internal mammary arteries (**Figure 6**). The patient refused surgical repair, but in order to control the progress of pulmonary hypertension, he accepted transcatheter closure of ventricular septal defects.

Discussion

IAA is a rare congenital cardiovascular disease, its estimated incidence is 3 per million live

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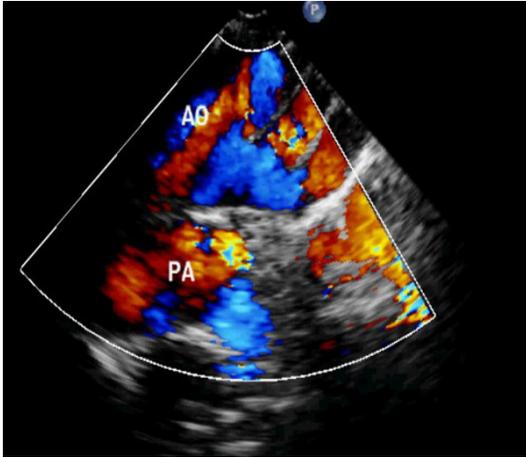


Figure 1. Transthoracic echocardiogram in supra-sternal notch view showing interrupted aortic arch with no blood flow between aortic arch and descending aorta.

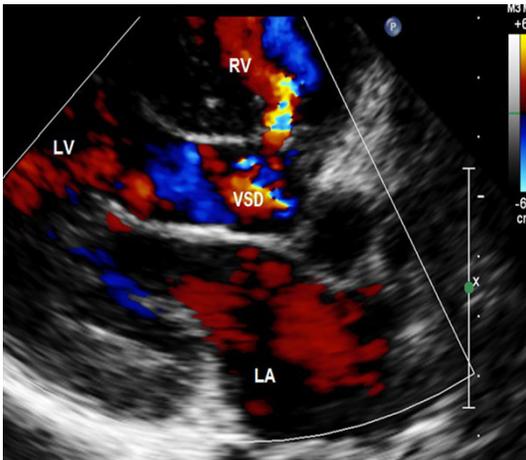


Figure 2. Transthoracic echocardiogram in parasternal long-axis view showing ventricular septal echo interruption of 3 mm with shunt signal left to right.

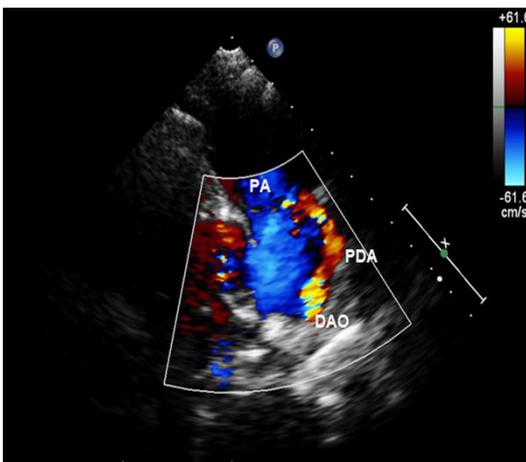


Figure 3. Transthoracic echocardiogram in parasternal short-axis view showing patent ductus arteriosus with shunt signal from descending aorta to pulmonary artery.

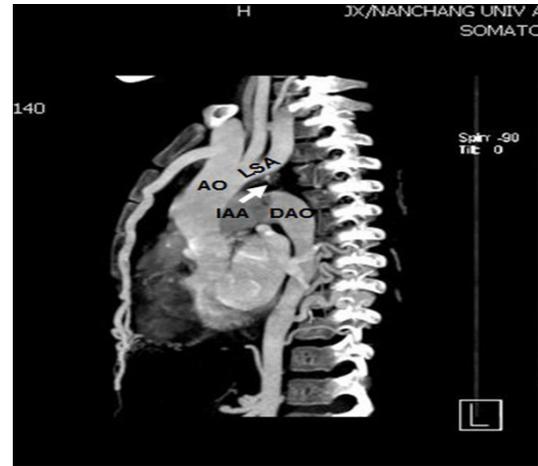


Figure 4. Computed tomography angiography showing isolated type A interrupted aortic arch (arrow) distal to the left subclavian artery.



Figure 5. Three-dimensional reconstruction showing aortic arch and descending aorta are discontinuous (←), and arterial duct puts pulmonary artery connected to the descending aorta (→).

births [1]. It is commonly presented in the perinatal period or during infancy, 80% of them died of heart failure within 1 month, less than 10% can survive to 1 year old without treatment [2]. IAA in an adolescent or adult is extremely rare.

Typical IAA can be associated with large VSD and PDA, called interrupted aortic arch triad. Also, other anomalies can be associated such

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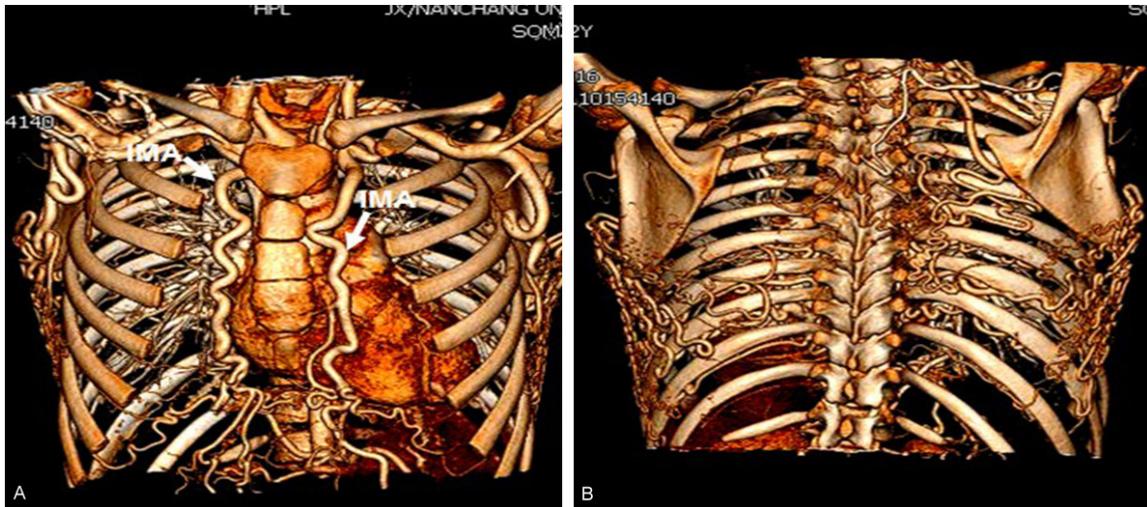


Figure 6. Three-dimensional reconstruction showing there exists extensive collateral vessels. A. Anterior view. B. Posterior view.

as transposition of great arteries, bicuspid aortic valve, mitral stenosis, persistent left superior vena cava [3, 4]. Usually, for purpose of making the blood supply to abdominal and lower limb, the direction of blood flow as the follows: Left ventricular -VSD- right ventricular -pulmonary artery -PDA- descending aorta. Because of the blood supplied by the channel is limited, patients often have more severe symptoms and signs, such as dizziness, chest congestion, cyanotic, hypertension. However, in this case, there are a lot of collateral vessels supply the abdominal and lower limbs, blood flow direction is no longer as the above, because blood pressure of descending aorta is higher than pulmonary artery, the direction of blood flow is from descending aorta to pulmonary artery. Also, the patient have no hypertension, cyanosis, and arterial pulse weakened since the enough collateral vessels supply to the body.

IAA can be confirmed by computed tomography angiography, magnetic resonance angiography and ardiac catheterization. Compared with them, echocardiogram can provide a wealth of information which other imaging techniques can not provide such as color doppler flow and associated with intracardiac anomalies. Several surgical means have been used for repair of IAA. The end-to-end anastomosis is the most commonly performed repair in neonates and infants, also it can be performed in older children and adults, graft interposition is often used in the older population [5]. Patients like

ours who refuse surgical or interventional repair should control the blood pressure and follow-up to assess the disease progression and prevent complications.

In summary, IAA is a rare congenital cardiovascular disease, computed tomography angiography can provide extracardiac anomalies while echocardiogram can provide color doppler flow and intracardiac anomalies. Combination of these two methods, diagnosis of IAA will be more comprehensive.

Disclosure of conflict of interest

None.

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