Cardiology in the Young

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A case of isolated right subclavian artery and right patent ductus arteriosus successfully treated with catheter intervention in early infancy

Makoto Yoshida, Tao Fujioka and Akio Inage

Department of Pediatrics, Japanese Red Cross Medical Center, Tokyo, Japan

Brief Report

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Corresponding author:
Makoto Yoshida;
Email: yoshimako.0821@outlook.com

Abstract

In isolated subclavian artery, abnormal aortic arch development causes a loss of continuity with the aorta. Patent ductus arteriosus is a known cause of congestive heart failure. Herein, we present a rare case of congestive heart failure caused by isolated right subclavian artery and right patent ductus arteriosus associated with left-sided aortic arch treated by early closure.

Case report

A 45-day-old female infant was referred to our hospital because of heart murmur, poor feeding, and weight gain. She was born at 37 weeks and 5 days of gestation, with a birth weight of 2464 g. No antenatal abnormalities were detected. On admission, she weighed 3300 g and exhibited depressed respiration, although no rales were heard. A serial Levine II/VI murmur was heard between the second and third ribs of the right sternal border. Blood tests further revealed elevated BNP (193.2 pg/mL) and NT-pro BNP (6598 pg/mL) levels, with no other abnormal findings. Chromosomal abnormalities were not observed in subsequent G-band or fluorescence in situ hybridisation testing. However, chest radiography revealed an enlarged heart (cardiothoracic ratio, 61%) and mild pulmonary congestion (Fig. 1a).

Echocardiography revealed a small atrial septal defect, persistent left superior caval vein, and blood flow into the right pulmonary artery (Fig. 1b). The blood flow of the duct pattern was unidirectional; left to right, with a tubular shape similar to a Krichenko type C duct. Enhanced CT confirmed the presence of a left-sided aortic arch and persistent left superior caval vein, as well as a right subclavian artery originating from the right pulmonary artery (Fig. 1c).

Cardiac catheterisation was performed, and aortography revealed no connection between the right subclavian artery and the aorta. Examination of the right common carotid angiography confirmed that blood flow retrogradely entered the pulmonary artery via the right subclavian artery and the right patent ductus arteriosus. The pulmonary blood flow-to-systemic blood flow ratio was 4.71, and the mean pulmonary artery pressure was 40 mmHg (Fig. 2a, b).

Based on these findings, the patient was diagnosed with congestive heart failure and pulmonary hypertension with pulmonary overcirculation due to isolated right subclavian artery, right patent ductus arteriosus, and atrial septal defect. Device closure of the patent ductus arteriosus by catheter treatment was preferred due to the perceived risk of right subclavian artery stenosis following surgical repair because of the potential for hypoplastic right subclavian artery due to blood flow stealing from the arm into the pulmonary artery.

Percutaneous embolisation of the patent ductus arteriosus was performed on the 39th day of admission. A 4F sheath was placed in the right femoral artery and vein, and a 4F multipurpose catheter was advanced into the right subclavian artery using a 0.035-inch Radifocus™ Guide Wire (Terumo). The reference vessel diameter of the right ductus arteriosus on RSCA angiography was 5.1 mm and an 8 mm Amplatzer™ Vascular Plug II was selected (Fig. 2c). In addition, RSCA angiography via the right pulmonary artery revealed tubular-shaped ductus arteriosus without restriction. There was no obvious difference in the shape of the right and left pulmonary arteries and only a slight stenosis of the RPA at the ductus arteriosus insertion site. The guidewire was placed in the right subclavian artery, and the sheath was replaced with a Parent Plus® 45 (Medikit) (Fig. 2d1, 2). The Amplatzer™ Vascular Plug II was advanced into the right subclavian artery and deployed. Following implantation, the pulmonary blood flow-to-systemic blood flow ratio decreased to 1.6, and mild RPA stenosis with pressure gradient of 24 mmHg between the main PA and RPA due to deployment of the first disk of the AVP in the RPA. Without any treatment, it gradually improved as the patient grew, and the pressure gradient was decreased to 10 mmHg 1-year after treatment.

Following treatment, the patient gained weight and the BNP level decreased to 12 pg/ml. The patient with a small amount of diuretics discharged 25 days after treatment.

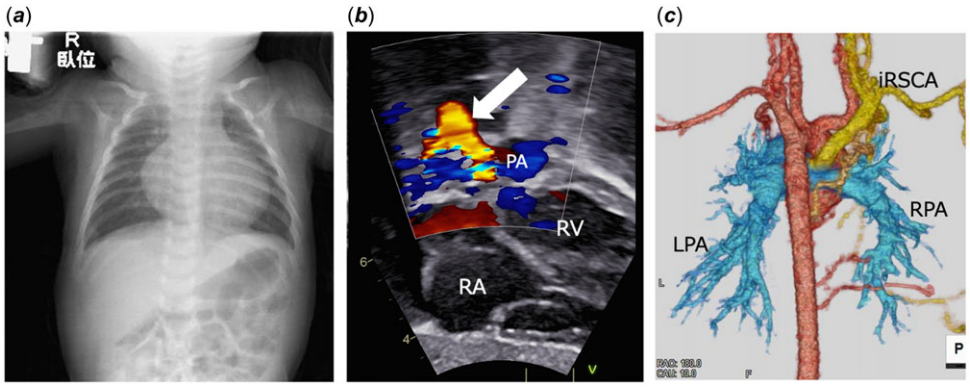

Figure 1. Multimodal imaging assessment on admission: (***a***) Chest X-ray; (***b***) Echocardiography; and (***c***) Computed tomography.

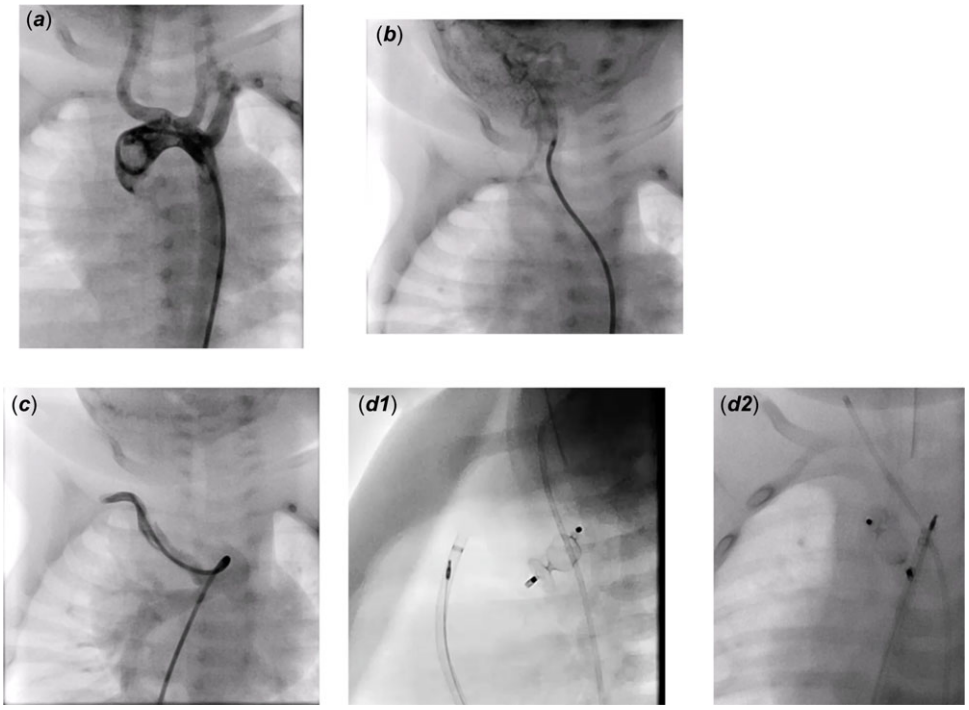

Figure 2. Angiographic imaging before and after device deployment: (***a***) Aortography; (***b***) Right common carotid artery angiography; (***c***) Right subclavian artery angiography; and (***d, e***) Post-deployment views.

Discussion

Isolated subclavian artery is a rare congenital aortic arch anomaly commonly associated with complex cardiac malformations such as tetralogy of Fallot, aortic arch dissection, transposition of the aorta, and a variety of other syndromes.[1,2] The majority of existing reports have involved isolated left subclavian artery associated with a right-sided aortic arch, while isolated right subclavian artery is relatively rare, with an incidence of approximately a quarter that of its left-sided counterpart.[3]

In one prior study, Edwards hypothesised that abnormalities of the vasculature arising from the aortic arch result from the abnormal retraction of arches I to VI during embryonic development.[4]

According to this hypothesis, during normal development, the distal components of the right IV and VI arches retract, the left IV arch develops into the aortic arch, and the left VI arch survives as a patent ductus arteriosus. However, in the present case, the distal end of the right VI arch, which forms the right ductus arteriosus and pulmonary artery, remained intact, while the aorta of the right IV arch disappeared; as such, the right subclavian artery separated from the aorta and became continuous with the pulmonary artery via the right ductus arteriosus. As a result, blood steal from the right subclavian artery to the pulmonary artery occurred through the right ductus arteriosus as the pulmonary vascular resistance decreased. Although the clinical symptoms of isolated right subclavian artery with patent ductus arteriosus vary according to the presence or absence of the ductus arteriosus and the degree of patency, heart failure may occur in early infancy; as such, it is important to keep this condition in mind if a continuous murmur along the right sternal border or a decreased right brachial pulse is observed in patients with heart failure.

In cases of isolated subclavian artery with concomitant CHD, treatment is often performed simultaneously with intracardiac repair, and there have been some reports of catheter treatment for isolated subclavian artery.[5,6] Supplementary Table 1 presents a summary of all previously reported cases, including the present case, of percutaneous closure of the patent ductus arteriosus in a left-sided aortic arch.

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In the present case, catheter treatment was selected instead of surgery due to concerns regarding the risks of anastomotic stenosis following transplantation, while heart failure dramatically improved following treatment without any severe complications. Device closure of the ductus arteriosus is an effective option for patients with no complications of complex intracardiac anomalies, or those with a small body size at the time of intervention and a high risk for surgical treatment. However, if percutaneous embolisation is selected for isolated right subclavian artery, the risk of postoperative complications should be considered. For example, even after percutaneous embolisation of the patent ductus arteriosus, the isolated right subclavian artery is still supplied retrogradely from the vertebral artery, and blood flow to the ring of Willis continues to be reduced as a phenomenon of blood theft, which may cause syncope during exercise,[7] requiring long-term follow-up.[3,6]

Supplementary material. The supplementary material for this article can be found at https://doi.org/10.1017/S104795112500040X.

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Competing interests. The authors declare no competing interests.

Ethical standards. The authors confirm that the ethical standards of the relevant national guidelines on care were observed while evaluating this case.

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