

Percutaneous closure of a fistula from the left circumflex coronary artery to the coronary sinus in an infant

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Abstract

Coronary artery fistula is an abnormal direct connection between the coronary artery and any of the four chambers of the heart or great vessels. A fistula from the left circumflex coronary artery to the coronary sinus is a relatively rare situation. We report a case of 12-month-old infant with coronary artery fistula from the left circumflex coronary artery to the coronary sinus that was identified incidentally. The N-terminal pro-brain natriuretic peptide level was elevated. Additionally, the proximal segment of the left circumflex coronary artery was dilated. On the basis of these findings, percutaneous closure of the fistula was performed with a vascular plug. This procedure achieved no residual flow and good hemodynamics were observed during follow-up.

Keywords

Fistula, transcatheter closure, N-terminal pro-brain natriuretic peptide, vascular plug, left circumflex coronary artery, coronary sinus

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Introduction

Coronary artery fistula (CAF) is an abnormal direct connection between the coronary artery and any of the four chambers of the heart or great vessels. CAF is a common coronary arterial malformation with a reported incidence of 0.002% in the general

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population.¹ In cases of CAF, the left circumflex coronary artery (LCx) accounts for 4.8% of cases as the site of the fistula, while 7% of these cases drain into the coronary sinus. Therefore, a CAF from the LCx to the coronary sinus is a rare situation.² The origin of CAF is considered to be congenital. Complications secondary to CAF include dilation of the coronary artery, congestive heart failure, and infective endocarditis, the incidence of which increases with age.² The treatment options of CAF are conservative treatment, surgery, and percutaneous occlusion. Development of transcatheter devices offers more choices for treatment, particularly asymptomatic pediatric CAF cases, because the risk and trauma from interventions and benefits from preventing future complications are more balanced than those with surgery. We report here percutaneous closure of CAF from the LCx to the coronary sinus in an infant.

Case report

A 12-month-old girl was referred to our department after cardiac murmurs were

found and she was diagnosed with coronary artery fistula at another institution. Her mother reported that her daughter was healthy overall, except easily catching a cold and gaining weight more slowly than other infants. A general physical examination was unremarkable. A cardiovascular examination showed an increased area of cardiac dullness on percussion. Routine laboratory tests showed an elevated N-terminal pro-brain natriuretic peptide (NT-proBNP) level of 528 ng/L and an elevated white blood cell count of $13.25 \times 10^9/L$. Cardiomegaly was observed on a chest X-ray. There were no ST-T wave changes on an electrocardiogram. Two-dimensional transthoracic echocardiography (TTE) showed an aneurysmally dilated coronary sinus (inner diameter of 10 mm) with the presence of a CAF originating from the LCx with a normal left ventricular ejection fraction (Figure 1a). The diameter of the fistula measured 6 mm. After discussion with the heart team, cardiac catheterization and an attempt to percutaneously occlude the fistula were planned.

On catheterization, a dilated LCx was visualized and this fed a distal fistula with

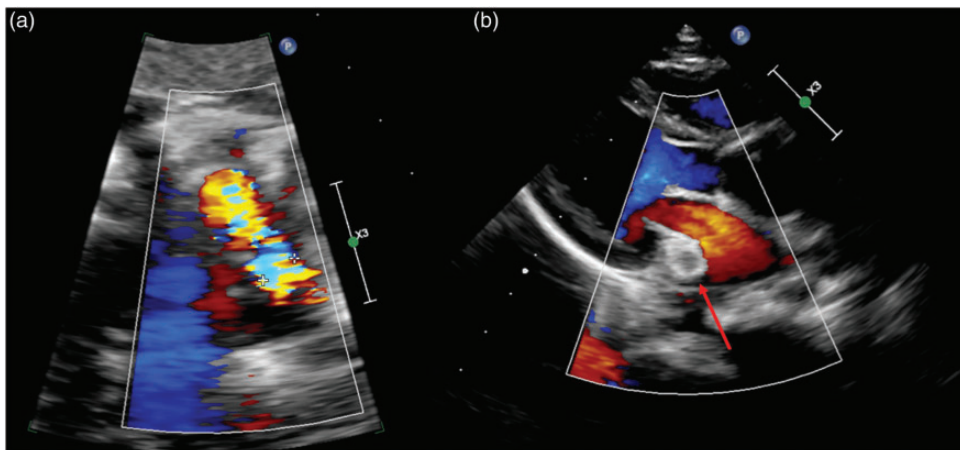


Figure 1. Transthoracic echocardiography before and 3 months after the procedure. (a) Enlarged view of the jet through a fistula before the procedure. (b) Follow-up echocardiography 3 months after the procedure. The red arrow indicates a plug.

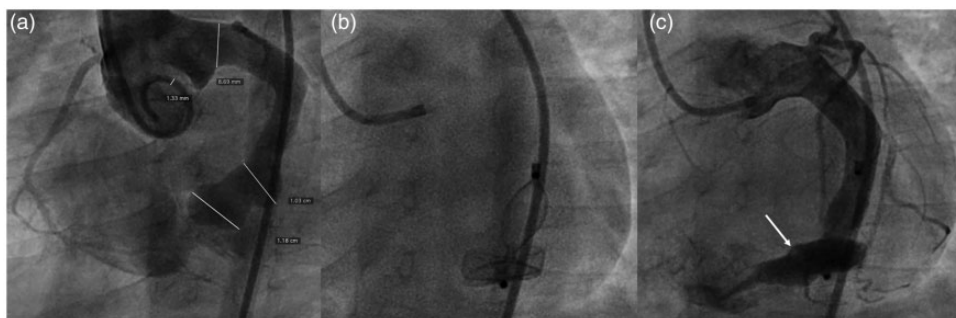


Figure 2. Angiography showing the coronary artery fistula. (a) Angiographic aortic root injection showing the course of a dilated left circumflex coronary artery to the coronary sinus. (b) Deployed device on fluoroscopy. (c) Post-procedural angiography showing trace residual flow through the coronary artery fistula. The white arrow indicates a plug.

the coronary sinus. The fistula at the entrance to the coronary sinus showed stenosis, while the left main coronary artery was dilated to 8.7 mm and the LCx was aneurysmally dilated to 10.3 mm and 11.8 mm at two segments (Figure 2a). The degree of shunting from the fistula (pulmonary-to-systemic flow ratio) was 1.31:1. Taking into consideration the size of the fistula, the considerable left to right shunt, and the possibility of blood flow steal in this young patient, we then decided to occlude the fistula percutaneously. This attempt was successfully performed using a 12-mm-diameter AMPLATZER™ Vascular plug II (AVP II; St. Jude Medical, St Paul, MN, USA), sized to fit the LCx aneurysm. The plug was advanced retrogradely from the LCx to avoid injury to the coronary sinus, and deployed through a 6 Fr right coronary guiding catheter (Cordis, Miami Lakes, Florida, USA), which met the minimum internal diameter requirements of a 12-mm AVP II. The device was successfully deployed at the proximal LCx aneurysm (Figure 2b) without causing insufficient blood flow supply or dissection of the LCx. The left anterior descending and obtuse marginal arteries were believed to compensate myocardial blood supply. Only trace residual flow was

observed at the final angiography (Figure 2c) and a TTE before discharge showed no residual flow. The patient was discharged uneventfully 2 days after the procedure on aspirin 25 mg/day. At her 3-month follow-up, no residual flow was detected on TTE (Figure 1b). The dilatation of the left coronary artery was improved. The diameter of the left main coronary artery was 6.6 mm and the proximal segment of the LCx was 5 mm. She remained in sinus rhythm. The NT-proBNP level had dropped to 43 ng/L. The troponin T level was 6.5 ng/L and no signs of ischemia were found.

Discussion

Our case represents an uncommon anatomy of CAF where the fistula originated from the LCx and connected to the coronary sinus. This type of CAF is mainly diagnosed in elderly people. Therefore, the findings of our case add further evidence for diagnosing and treating CAF from the LCx to the coronary sinus in neonates and infants.³

After identification of CAF, and its feeding vessel and termination site on TTE, the treatment strategy depends on the anatomy of the CAF and its clinical course if it

remains uncorrected in patients. When the fistula is small, CAF can be asymptomatic, but its existence gradually leads to vessel dilation and subsequent heart failure.⁴ A larger fistula can result in myocardial steal by draining blood flow from the coronary artery to veins or chambers of lower pressure, leading to ischemia of the coronary artery distal to the fistula. In neonates, CAF is usually treated conservatively as long as the clinical status is stable.⁵ In our case, although the infant was growing, her NT-proBNP level was increased with a dilated LCx and coronary sinus. Therefore, correction of the fistula was considered. If correction was not performed, this left-to-right shunt might have caused pulmonary hypertension and volume overload in both ventricles in the long term.⁶ Accurate assessment of the origin, drainage site, complexity, and the number and size of fistulous tracts is crucial in procedural planning. Measurement of malformations is performed with selective coronary angiograms, but it can also be performed with computed tomography angiography to better characterize three-dimensional structures in adult patients.

Various vascular plugs or occluders have been used to close percutaneous shunts, venous anomalies, and congenital intra- and extracardiac defects, offering wider choices of treatment to pediatric patients to avoid the trauma from surgery. The AVP was created for peripheral embolization, but was chosen in our case because of the shape and size of the LCx aneurysm and the availability of the device at the time of catheterization. An AVP is especially useful in treating high-flow situations, such as arteriovenous fistulas.⁷ In case of CAF, an AVP offers the advantage of a single device for effective closure and better stability compared with coils.⁸ However, the need to use relatively large guiding catheters requires assessment of the size and tortuosity of the access and the choice between

antegrade or retrograde delivery. The optimal antiplatelet or anticoagulation treatment for closure of CAF remains unclear. When thrombosis formation is suspected during follow-up, sufficient anticoagulation can be initiated.⁹

In conclusion, in infants with a moderate- to large-sized CAF, transcatheter closure is feasible and effective. The choice of transcatheter devices should be weighed in relation to local resources.

Ethics statement

Publication of this study was approved by the local ethics committee of West China Hospital. The patient provided written informed consent for publication.


Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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