INTRODUCTION

Coronary artery fistulas (CAF) are rare congenital defects of the coronary circulation system that cause communications between the coronary artery and cardiac chambers or a primary vessel (vena cava, sub-pulmonary veins, pulmonary artery, mediastinal vessels, or coronary sinus). The reported incidence range was 0.2% to 1.2%, rising over the last decade due to the wide use of echocardiography and angiography.  

Most cases were asymptomatic and diagnosed incidentally during coronary catheterization. Some patients, especially with larger fistulas, may present with signs and symptoms of heart failure, myocardial infarction, pulmonary hypertension, and other cardiopulmonary functional abnormalities. Coronary angiography or noninvasive modality, such as coronary computerized tomography angiography (CTA), is highly reliable for diagnosing CAF. The transcatheter technique is a safe, minimally invasive management approach with excellent outcomes. Reports about percutaneous closure using Amplatzer duct occluder II device in PDA and VSD cases stated that the device is as effective and less invasive than open surgery with a shorter stay. This procedure also does not leave a profound scar superior to surgery in the cosmetic aspect. Based on those mentioned above, we present a case of a successful transcatheter approach to CAF closure using an Amplatzer duct occluder II device.

CASE PRESENTATION

A 10-month-old, 8.6 kg girl was referred to our pediatric cardiologist clinic with a diagnosis of coronary artery fistula to plan for transcatheter closure of the fistula due to shortness of breath and weakness. Based on medical history, the patient was hospitalized at three months old and diagnosed with bronchopneumonia when the pediatrician found a continuous murmur. Furthermore, the doctor said there was a suspicion of a congenital heart abnormality, and then she was consulted by a pediatric cardiologist. An echocardiographic examination shows coronary fistula disease. She has taken lisinopril 1 x 0.6 mg and spironolactone 2 x 6.25 mg.

According to her mother, the patient’s growth and development look normal, like children of her age. The patient never suffers from jaundice or cyanosis. None of the family members have a history of heart disease. She was born spontaneously, assisted by a midwife, full term, and immediately cried after being born. Her birth weight was 3300 grams with a body length of 50 cm.

On physical examination, the patient presentations are alert. Vital signs show a normal body temperature of 36.6 °C, pulse rate of 110 times per minute with a regular rhythm, respiratory rate of 26 times per minute, blood pressure of 90/60 mmHg, and oxygen saturation of 98%. The anemia
sign, jaundice, cyanosis, or dyspnea were absent on head and neck examination. No lymphadenopathy was found. On chest examination, there was no chest wall retraction. The first and second heart sounds are normal and regular. Grade 3/6 continuous murmur was present without gallops. Stridor, crackles, or wheezing were not present. The abdomen was within normal limits. No edema was found on the limbs, with a capillary refill time of fewer than 2 seconds. There was no abnormality on neurological examination.

The laboratory test reported hemoglobin 12.3 mg/dL, leukocytes 14,080 /μL, platelets 124,000/μL, PPT 11.1, APTT 27.1 sec, sodium 142 mmol/L, potassium 4.9 mmol/L, chloride 104 mmol/L, calcium 10.75 mg/dL, blood glucose 96 mg/dL, total protein 7.4 mg/dL, albumin 4.8 g/dl, BUN 6 mg/dL, serum creatinine 0.25 mg/dL, AST 12 U/L, ALT 40 U/L.

An examination of the chest X-ray shows the impression of cardiomegaly with the configuration of right ventricular enlargement accompanied by increased pulmonary vascularity in both lung fields (Figure 1). The electrocardiographic examination showed sinus tachycardia, with a heart rate of 135 beats per minute and a normal axis (Figure 2). On echocardiographic examination, found a situs solitus, AV-VA concordance, normal pulmonary venous drainage, RA-RV heart chambers were dilated, normal valves, PDA was not found, there was a secundum ASD with a diameter of 0.76 cm, a distal RCA fistula to the right atrium, intact ventricular septum. In conclusion: Fistula RCA to moderate RA and ASD secundum (Figure 3). The patient was diagnosed with a coronary artery fistula and planned for cardiac catheterization examination for definitive diagnosis and management.

A pediatric cardiologist consultant performed the cardiac catheterization. The operative steps are described below:

1. The patient is placed in a supine position.
2. The sterile drape was placed on the area after disinfection of the left and right inguinal regions.
3. Perform right femoral artery and vein puncture using a 20-gauge intravenous catheter with the Seldinger technique.
4. Using Teflon wire, a 5Fr sheath was inserted into the right femoral artery and right femoral vein.
5. Insert the 5Fr pigtail to ascended the aorta-left ventricle and measure the aortic pressure.
6. Perform angiography on the left ventricle. The fistula was found to communicate the distal right coronary artery to the right atrium with an ampulla size of 7.29 mm, waist of 3.11, and length of 4.71 mm (Figure 4).
7. Insert the 5Fr multipurpose angiographic catheter into the inferior vena cava-right atrium-right ventricle-pulmonary artery. Measure the pressure and saturation on the pulmonary artery, right ventricle, and right atrium. Perform venography and innominate vein (+). PLSVC was found (+).
8. Measure the saturation on the superior vena cava and inferior vena cava.
9. Insert the 0.89 guidewires Radifocus into the aorta.
10. Insert the delivery catheter TorqVue then the Amplatzer Duct Occluder II 6mm/4mm LOT 8353342. Expand...
both sides (Figure 5).

11. Perform echocardiography evaluation.
   The residual flow was not found (Figure 6).

12. End of the procedure, remove all devices.

The intra-procedural went smoothly. The post-procedural course was uneventful, and the patient was discharged home after three days. One week and one-month post-procedure, the echocardiography evaluation showed an in-situ device with no residual flow.

DISCUSSION

The coronary artery fistulas can be grouped into two broad categories: the cameral coronary fistula, which is an abnormal connection between coronary arteries with one of the heart chambers, and the coronary arteriovenous fistula, which is an abnormal connection that occurs between coronary arteries and parts of the systemic/pulmonary circulatory vessels. The coronary cameral fistula is considered the most common type of CAF, which accounts for less than 1% of the population and is often detected incidentally using coronary angiography. The coronary cameral fistula's further classification is arterioluminal (direct communication through the cardiac chamber) and arteriosinusoidal (indirect communication through a cardiac sinusoidal network). The fistula may be single (72%) or multiple (28%) and may originate from the RCA (55%), the LAD (35%), or bilaterally (5%). The most involved chamber is the right ventricle (41%), which leads to increased right heart volume and pulmonary hypertension. CAFs can be classified as small, medium, or large if the fistula diameter is <1, 1 to 2, or >2 times the largest diameter of the coronary vessel not feeding the coronary fistula, respectively. Small CAFs likely close spontaneously over time, while larger CAFs may require surgical or transcatheter closure. Our patient suffers from a single coronary cameral fistula with an arterioluminal subtype, as the fistula was found between the coronary artery and directly draining into the right atrium.

Most CAF is asymptomatic. The clinical manifestations highly depend on the factors such as age, fistula size, the volume of flow, and the degree of coronary steal. A small fistula is asymptomatic, while a large fistula is commonly associated with the sign of congestive heart failure, especially in infancy. In an asymptomatic infant patient, the suspicion of congenital heart abnormality was accidentally detected as a continuous murmur during auscultation of the heart. The classic physical finding is a soft continuous murmur that tends to be crescendo decrescendo in both systole and diastole but louder in diastole.

The physical examination was later supported by imaging findings that consisted of chest X-ray, echocardiography, and coronary angiography. The chest x-ray show cardiomegaly found in this patient. The echocardiography should be done pre-intervention and post-intervention. During pre-intervention, echocardiography is critical in evaluating the CAF hemodynamic, excluding other coexisting cardiac anomalies, pulmonary hypertension screening, and providing an anatomic evaluation of adjacent cardiac structures for the clinician to consider the CAF closure technique. A combination of two-dimensional and color Doppler echocardiography in CAF cases visualized a dilated coronary artery and turbulent flow in the fistula and the recipient chamber, known as the steal phenomenon. In this case, the suspicion of congenital heart abnormality arose from a physical examination at three months of age when she was admitted to the hospital due to bronchopneumonia, in which the pediatrician noticed a continuous murmur. This finding is supported by echocardiography which shows RCA fistula leading to the right atrium and dilatation of the right atrium and ventricle, and supported by Doppler echocardiography examination, which shows a steal phenomenon on the right atrium. Secundum ASD was also detected on the echocardiography. Intraoperative transesophageal echocardiography helps to identify the precise location of the site of drainage of the fistula. Post-intervention echocardiography requires confirming the CAF closure and monitoring cardiac hemodynamics after the procedure.

Cardiac catheterization with coronary angiography remains the gold standard.
or dissection may occur during wire and catheter delivery. In some cases, the patient may develop device embolization. The post-procedural echocardiography shows the residual flow was absent, and no complications were found after closing the fistula in this patient.

The limitation of this study is the echocardiography evaluation should be performed in one week, one month, three months, and six months after the procedure. However, we only have the echocardiography data from one week and one-month post-procedure because the patient’s family choose to continue the medical care in another hospital near their home. We do not perform computed tomography angiography on this patient.

CONCLUSION

CAF is a very rare cardiac abnormality. The transcatheter approach is preferred according to the fistula origin and size. The procedure can be done safely using an Amplatzer duct occluder II device. The ADO II device is recommended for small infants due to its small-size delivery catheters and a softer shape.

CONFLICT OF INTEREST

The authors have no conflicts of interest to declare.

ETHICAL CONSIDERATION

The patient’s birth mother consents information about the patient to appear in a journal article. The data will be published without her child’s name attached, and every attempt will be made to ensure anonymity.

FUNDING

No funding or grant support this case study.

AUTHOR CONTRIBUTION

AF involved in writing the manuscript. TH, MAR, AU supervising and revising the manuscript. All authors prepare the manuscript and agree for this final version of manuscript to be submitted to this journal.

REFERENCE


