Original Article Anomalous origin of bilateral coronary arteries: a case report and review of literature

Jianyi Liao1*, Xin Li1*, Qiuqin Xu², Xudong Ran¹, Yonggeng Xu¹

¹Department of Cardiothoracic Surgery, Children's Hospital of Soochow University, Suzhou, China; ²Department of Cardiology, Children's Hospital of Soochow University, Suzhou, China. ^{*}Equal contributors.

Received May 28, 2018; Accepted August 4, 2018; Epub November 15, 2018; Published November 30, 2018

Abstract: Anomalous origin of the coronary arteries is a rare congenital disorder which can lead to myocardial ischemia, valvar disorder, congestive heart failure, and sudden cardiac death in early childhood. It is an extremely rare type of congenital disorder. We report a case of a patient with anomalous origin of the left coronary artery from pulmonary artery along with anomalous aortic origin of right aortic coronary who was successfully corrected by surgical intervention.

Keywords: Congenital heart disease, anomalous origin of coronary arteries, anomalous origin of bilateral coronary arteries

Introduction

Anomalous origin of the coronary arteries is a rare congenital disorder which can lead to myocardial ischemia, valvar disorder, congestive heart failure, and sudden cardiac death in early childhood. Thorough preoperative testing is required to make correct diagnosis. Without adequate and timely surgical treatment, it can cause high infant mortality. Anomalous origin of bilateral coronary arteries is an extremely rare type of this congenital disorder. Here we report a case of patient with anomalous origin of the left coronary artery from pulmonary artery (ALCAPA) along with anomalous aortic origin of right aortic coronary (AAORCA). The patient was successfully corrected by surgical intervention to re-implant the left coronary arteries to the corresponding aortic cusp and un-roof the inner wall of the right coronary artery. Postoperative echocardiogram demonstrated antegrade flow in the bilateral coronary arteries and no evidence of stenosis of coronary arteries. The patient had an uneventful post-op course and was discharged from the hospital without any symptoms. The left ventricular systolic function was noted to be later improving on post-op Doppler echocardiography.

Case report

A 5-month-old male infant was admitted to our institution with a 3-day history of fever with non-productive cough with difficulty breathing and wheezing. Physical examination confirmed the presence of a 39 C body temperature, pulse 175/min, respiratory rate of 45/min, blood pressure 50/28 mmHg, and oxygen saturation on room air of 91%. He was awake but lethargic and had peripheral cyanosis involving the lips but no jaundice, rash, or bleeding. Lung examination revealed positive suction concave sign, wheezing, and rale in bilateral lung fields. Cardiovascular examination revealed that the heart rhythm and rate were normal, and there was a grade 3/6 systolic murmur best heard at the left lower sternal border. The abdomen was soft without distention, tenderness, and hepatosplenomegaly. Edema or cyanosis was not found on the extremities. However, the extremities were cool. The patent was initially diagnosed as bronchial pneumonia, and treated with oxygen, steroid, and antibiotic.

On the day of admission, laboratory testing revealed significant elevation of cardiac enzymes including CK-MB 10.3 ng/mL, troponin (cTnI3) 4.24 ng/mL and myoglobin 39.2 ng/



Figure 1. ECHO showing (A) Right coronary artery (RCA) originated from the left coronary sinus of Valsalva and coursed to right through the aortic wall between the aorta (AO) and PA at pre-operation. The flow rate of the right coronary accelerated (B, C) The position and shape of bilateral coronary arteries were good at post-operation. (D, E) Color Doppler showing the flow of blood in coronary artery at postoperation. (F) The shape and size of the LCA opening at 3 months after operation.

Anomalous origin of bilateral coronary arteries



Figure 2. Cath angiograms showing (A, B) Right coronary artery (RCA) origin from the left coronary sinus. (C, D) Left coronary artery (LCA) origin from the pulmonary artery (PA).



Figure 3. The operative findings.

mL. Electrocardiogram (ECG) demonstrated sinus tachycardia, left atrial and ventricular enlargement, left anterior fascicular block, ST depression in inferior leads II, III, aVF, and abnormal Q waves in I, V4 and V5 leads. The

chest X-ray showed moderate cardiomegaly and increased pulmonary vascular markings. Therefore, cardiac ischemia with congestive heart failure was diagnosed, and congenital coronary artery abnormality was suspected.



Figure 4. Changes of mitral regurgitation before and after operation: (A) pre-operation, (B) 2 weeks after operation, (C) 1 month after operation, (D) 3 months after operation, (E) 6 months after operation, (F) 12 months after operation.

Table 1. Changes of left ventricular ejection fraction (LVEF) after operation

Subsequently echocardiography was completed which identified a grossly hypokinetic and dilated left ventricle with estimated ejection fraction (LVEF) of 39%, mild-to-moderate mitral regurgitation, Echocardiography showed that the right coronary artery originated from the left sinus (Figure 1), and no direct sign of LCA originating from pulmonary artery was seen. The echo of the mitral valve papillary muscles was enhanced and rigid. To better understand, the origin and course of the coronary arteries, CT angiogram of coronary arteries was performed on the fourth day of admission. Interestingly, CT angiogram revealed origin of the RCA from the left sinus of Valsalva. On day 7 of admission, the patient underwent aortic root angiography that also confirmed the origin RCA from originated from the left coronary sinus. In addition, pulmonary artery angiography was also performed, confirming a small artery originating from the left side of the pulmonary artery with proximal diameter of 1.1 mm (Figure 2). Based on the results from CT and Cath angiograms, the patient was diagnosed as anomalous origin of bilateral coronary arteries that is extremely rare congenital coronary artery abnormality.

The patient was taken to the operating room for surgical correction of the coronary arteries one

month after fever and respiratory symptoms resolved. The surgery was performed through a median sternotomy. Obvious cardiomegaly and severe tortuous epicardial vessels were noted. After cardiopulmonary bi-caval bypass was established, the bilateral pulmonary arteries were snared tightly to prevent perfusion steal into the pulmonary bed. After mild hypothermia, a left side vent was placed through the right upper pulmonary vein. The aortic cross clamp was placed high, and cardioplegic spikes were inserted into both the ascending aorta and the pulmonary trunk (PT). Cardioplegic solution (HTK Custodiol 50 mL/kg, 7 min, single dose) was delivered simultaneously into the aortic root and the PT. The ascending aorta was then tran-

sected at a high level more than 10 mm above the sinotubular junction and the main PA was transected just below the bifurcation.

During intra-operation (Figure 3), the left coronary artery was identified as originating from the distal end of the main pulmonary artery just proximal to right pulmonary artery. It had a short trunk which bifurcated into left anterior descending branch and the left circumflex artery. Anomalous right coronary artery was found to arise from the left sinus of Valsalva and ascend from the right coronary sinus after coursing through the aortic wall between the aorta and pulmonary artery for about 5 mm. As such, the inner wall of the right coronary artery was un-roofed for about 5 mm. The coronary button was harvested with a generous cuff of the pulmonary wall, and the proximal portion of the coronary artery was mobilized with preservation of its branches. The right aortic wall was longitudinally incised from the opening of the right coronary artery up to the middle part of the left coronary sinus. A trap-door incision was made in the ascending aorta higher than the aortic commissure and the coronary cuff was sewn with a 7-0 polypropylene suture at the left aortic sinus. The proximal aortic stump was anastomosed to the ascending aorta with a 6-0 polypropylene suture. After rewarming, aortic roots attracted exhaust and the aortic clamp was opened and the heart regenerated into sinus rhythm. The defect in the main PA was reconstructed with a generous fresh autologous pericardial patch with 7/0 prolene suture and an end-to-end anastomosis of pulmonary artery with 6/0 prolene (**Figure 4**). Weaning from the bypass was performed without any problems, and no ischemic change or arrhythmia was encountered in the EKG.

The patient was transferred to cardiac ICU with ventilator support and low dose infusion of epinephrine to maintain adequate cardiac output. Postoperative echocardiogram demonstrated antegrade flow in both left and right coronary arteries and no evidence of stenosis of coronary arteries (**Figure 1**). ECHO also revealed mild to moderate mitral regurgitation was stable without change (**Figure 4**), The change of EF after operation is shown in **Table 1**. Postoperative EKG showed no evidence of ischemic change, and cardiac enzymes resolved to normal limit. The patient was extubated on postoperative day 7, and was discharged from the hospital without any symptoms.

Discussion

Congenital anomalies of the coronary artery occur in 0.2%-1.2% of the general population. Congenital anomalous origin of coronary artery usually includes two main sub-categories. The first one includes origin of coronary from pulmonary artery, the majority of which is anomalous origin of left coronary artery from pulmonary artery (ALCAPA). ALCAPA occurs in approximately 1/300,000 live births or 0.5% of children with congenital heart disease [1]. The mortality associated with untreated ALCAPA has been estimated to range from 35% to greater than 85% in the first year of life [2-4]. The second subtype is the anomalous aortic origin of the coronary arteries of which anomalous RCA coming from the left coronary sinus of Valsalva is the most common [5, 6] with prevalence of around 0.17% [7, 8]. These anomalous arteries can have intramural course through the aortic wall. The Congenital Heart Surgeons Society (CHSS) investigated 113 patients with AAOCA who received surgical intervention in 29 cardiovascular centers throughout North American from 1998 to 2012. Approximately 98% of these anomalous arteries had interarterial and/or intramural course [9]. The intramural course with angle can lead to kinking of the vessel which can cause compromised coronary perfusion. This can result in myocardial ischemia or infarction with or without congestive heart failure in children. Without adequate and timely surgical treatment, it can cause high infant mortality. The initial diagnostic tests, besides thorough physical examination, include 12-Lead ECG, chest X-ray, and echocardiography. Further investigation with Computed Tomography Angiography (CTA), cardiac catheterization with coronary angiography or Magnetic Resonance Imaging (MRI) is often required to delineate the correct anatomy [10]. Although CTA has been widely used in diagnosis of coronary artery anomalies, majority of CTA data were obtained from the adult patients [5, 11]. The typical cases of ALCAPA exhibit RCA thickening and dilatation with demonstration of visualized reverse blood flow from left coronary artery to pulmonary artery [12]. However, preoperative CTA didn't show typical ALCAPA change in this case. We considered the possibility of anomalous origin of bilateral coronary arteries based on other direct and indirect evidences.

Since the first surgery in 1960, the technique of ALCAPA has undergone significant improvement. Initial surgeries included ligation of the origin of LCA with bypass of arteries in order to create a single coronary artery system [13, 14]. Currently the most widely used technique is reimplantation of coronary arteries to the corresponding aortic cusp in order to achieve anatomical and physiological correction of dual coronary artery system. This approach has been reported to have less operative mortality with good overall long term patency rates [15, 16]. If the coronary arteries in the ALCAPA are not long enough for reimplantation, a technique with diploic and pulmonary trunk tissue or pericardial tissue cuff extending technology can be used. In patients with AAOCA, due to different in location of anomalous arteries, the surgical approach is variable [9]. Un-roofing of anomalous coronary artery is recommended [17, 18] and most scholars believe that surgery usually does not require mitral valve repair. After coronary artery replantation, the mitral regurgitation of children will be significantly improved [19, 20].

To the best of our knowledge, there have been no case reports in which the patient had both ALCAPA and AAORCA. In order to make corrective diagnosis and render effective treatment, attentions to patient's medical history, careful physical examination, and auxiliary diagnostic testing is crucial [21]. If necessary, the surgeon should be well prepared to use creative repair methods to correct this complicated congenital disorder. In our case, the surgery was successful and the patient had an uneventful post-op course. The left ventricular systolic function was noted to be later improving on post-op Doppler echocardiography. We observed that the LV ejection fraction decreased early after operation (Table 1), which might be related to surgical trauma. LV ejection fraction further increased to 45% and 50% after 6 months and 12 months of surgery respectively.

Conclusion

Anomalous origin of bilateral coronary arteries is extremely rare. A through preoperative testing is required to make correct diagnosis. Long term outcome post-surgical correction is favorable.

Address correspondence to: Xin Li, Department of Cardiothoracic Surgery, Children's Hospital of Soochow University, Suzhou, China. E-mail: lixin. home@yahoo.com

References

- Keith JD. The anomalous origin of the left coronary artery from the pulmonary artery. Br Heart J 1959; 21: 149-161.
- [2] Driscoll DJ, Nihill MR, Mullins CE, Cooley DA, McNamara DG. Management of symptomatic infants with anomalous origin of the left coronary artery from the pulmonary artery. Am J Cardiol 1981; 47: 642-648.
- [3] Wesselhoeft H, Fawcett JS, Johnson AL. Anoma lous origin of the left coronary artery from the pulmonary trunk. Its clinical spectrum, pathology, and pathophysiology, based on a review of 140 cases with seven further cases. Circulation 1968; 38: 403-425.
- [4] Mongé MC, Eltayeb O, Costello JM, Sarwark AE, Carr MR, Backer CL. Aortic implantation of anomalous origin of the left coronary artery from the pulmonary artery: long-term outcomes. Ann Thorac Surg 2015; 100: 154-161.
- [5] Lee HJ, Hong YJ, Kim HY, Lee J, Hur J, Choi BW, Chang HJ, Nam JE, Choe KO, Kim YJ. Anomalous

origin of the right coronary artery from the left coronary sinus with an interarterial course: subtypes and clinical importance. Radiology 2012; 262: 101-8.

- [6] Lee HJ, Kim YJ, Hur J, Nam JE, Kim TH, Choe KO, Choi BW. Coronary artery anomalies: detection on coronary artery calcium scoring scan. AJR Am J Roentgenol 2010; 194: W382-387.
- [7] Yamanaka O, Hobbs RE. Coronary artery anomalies in 126595 patients undergoing coronary arteriography. Cathet Cardiovasc Diagn 1990; 21: 28-40.
- [8] Roberts WC, Siegel RJ, Zipes DP. Origin of the right coronary artery from the left sinus of valsalva and its functional consequences: analysis of 10 necropsy patients. Am J Cardiol 1982; 49: 863-868.
- [9] Poynter JA, Bondarenko I, Austin EH, DeCampli WM, Jacobs JP, Ziemer G, Kirshbom PM, Tchervenkov Cl, Karamlou T, Blackstone EH, Walters HL 3rd, Gaynor JW, Mery CM, Pearl JM, Brothers JA, Caldarone CA, Williams WG, Jacobs ML, Mavroudis C; Congenital Heart Surgeons' Society AAOCA Working Group. Repair of anomalous aortic origin of a coronary artery in 113 patients: a congenital heart surgeons' society report. World J Pediatr Congenit Heart Surg 2014; 5: 507-514.
- [10] Takemoto K, Hirata K, Tanimoto T, Matsuo Y, Ino Y, Kubo T, Tanaka A, Akasaka T. Combined non-invasive doppler echocardiography and coronary computed tomography lead to diagnosis of anomalous left coronary artery from the pulmonary artery (ALCAPA) syndrome. Circ J 2015; 79: 1136-1138.
- [11] DłużNiewska N, Kostkiewicz M, Tomkiewicz-Pająk L, Krupiński M, Podolec J, Olszowska M, Podolec P. Noninvasive techniques for the diagnosis of anomalous origin of the left coronary artery from the pulmonary artery in adult patients. Acta Cardiol 2017; 72: 41-46.
- [12] Patel SG, Frommelt MA, Frommelt PC, Kutty S, Cramer JW. Echocardiographic diagnosis, surgical treatment, and outcomes of anomalous left coronary artery from the pulmonary artery. J Am Soc Echocardiogr 2017; 67: 969-969.
- [13] Naimo PS, Fricke TA, d'Udekem Y, Cochrane AD, Bullock A, Robertson T, Brizard CP, Konstantinov IE. Surgical intervention for anomalous origin of left coronary artery from the pulmonary artery in children: a long-term followup. Ann Thorac Surg 2016; 101: 1842-1848.
- [14] Lange R, Cleuziou J, Krane M, Ewert P, Pabst von Ohain J, Beran E, Vitanova K. Long-term outcome after anomalous left coronary artery from the pulmonary artery repair: a 40-year single-centre experience. Eur J Cardiothorac Surg 2018; 53: 732-739.

- [15] Kanoh M, Inai K, Shinohara T, Tomimatsu H, Nakanishi T. Outcomes from anomalous origin of the left coronary artery from the pulmonary artery repair: long-term complications in relation to residual myocardial abnormalities. J Cardiol 2017; 70: 498-503.
- [16] Kudumula V, Mehta C, Stumper O, Desai T, Chikermane A, Miller P, Dhillon R, Jones TJ, De Giovanni J, Brawn WJ, Barron DJ. Twenty-year outcome of anomalous origin of left coronary artery from pulmonary artery: management of mitral regurgitation. Ann Thorac Surg 2014; 97: 938-944.
- [17] Frommelt PC, Sheridan DC, Berger S, Frommelt MA, Tweddell JS. Ten-year experience with surgical unroofing of anomalous aortic origin of a coronary artery from the opposite sinus with an interarterial course. J Thorac Cardiovasc Surg 2011; 142: 1046-1051.
- [18] Zhang H, Cheng P, Jin G, Han D, Luo Y, Li J. Surgical strategies for anomalous origin of the left coronary artery from the right pulmonary artery with an intramural aortic course: a report of 10 cases. Thorac Cardiovasc Surg J 2017; 153: 648-653.

- [19] Sasikumar D, Dharan BS, Arunakumar P, Gopalakrishnan A, Sivasankaran S, Krishnamoorthy KM. The outcome of mitral regurgitation after the repair of anomalous left coronary artery from the pulmonary artery in infants and older children. Interact Cardiovasc Thorac Surg 2018; 27: 238-242.
- [20] Mongé MC, Eltayeb O, Costello JM, Sarwark AE, Carr MR, Backer CL. Aortic Implantation of anomalous origin of the left coronary artery from the pulmonary artery: long-term outcomes. Ann Thorac Surg 2015; 100: 154-160.
- [21] Dehaki MG, Al-Dairy A, Rezaei Y, Ghavidel AA, Omrani G, Givtaj N, Afjehi RS, Tatari H, Jalali AH, Mahdavi M. Mid-term outcomes of surgical repair for anomalous origin of the left coronary artery from the pulmonary artery: in infants, children and adults. Ann Pediatr Cardiol 2017; 10: 137-143.