A rare case of Left Main Coronary Artery Atresia presenting as Dilated Cardiomyopathy and misdiagnosed as ALCAPA

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Abstract

This case report presents an extremely rare case of left main coronary artery atresia in a 14-week-old female infant presenting with severe symptoms of dilated cardiomyopathy.

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IRB approval, consent statement and clinical trial registration are not applicable for this study.

Summary

This case report presents an extremely rare case of left main coronary artery atresia in a 14-week-old female infant presenting with severe symptoms of dilated cardiomyopathy.

Introduction

Coronary artery anomalies are relatively uncommon congenital disorders of the coronary artery anatomy. The incidence of coronary artery anomalies has been reported to be 0.6–1.3%. [1] Left main coronary artery atresia (LMCAA) is the rarest form of congenital coronary malformations, in which coronary ostium and main trunk in the left coronary artery system is absent [2]. Thus, blood flows from the right coronary artery to the left coronary artery through collateral vessels [3]. Clinical presentation, management, and prognosis of this disease depend on the characteristics of the collaterals and native vessels [4]. Hereby, we report a case

of congenital atresia of the left main coronary artery, misdiagnosed as Anomalous left coronary artery from the pulmonary artery (ALCAPA).

Case Presentation

A 14-week-old female infant, who had respiratory distress worsened after bronchiolitis, was admitted to the intensive care unit. She was intubated in another hospital before the admission. On the first evaluation, the patient was cyanotic and had a cutis marmorata appearance on her skin. Capillary refill time was 4 seconds, and her pulse was week. She had tachycardia and bilaterally crepitant rales. According to the echocardiographic examination, the left atrium and left ventricle were greatly enlarged and left ventricular contractions were markedly decreased (Shortening Fraction:13%), thus indicating a dilated cardiomyopathy. Figure 1 represents the right ventricle (RV) stuck between the sternum and the left ventricle (LV). Septum movements were better than the posterior LV wall. A moderate degree of insufficiency was observed in the mitral and tricuspid valve. Pulmonary arterial pressure was measured to be approximately 60 mmHg through the tricuspid valve insufficiency. There was also a mild degree of insufficiency in the aortic valve. Pulmonary and aortic flows were decreased due to low cardiac output. Since she had a cardiogenic shock, IVIG, hydrocortisone, and inotropic support drugs were started with the preliminary diagnosis of myocarditis. The patient was placed on extracorporeal membrane oxygenation (ECMO) due to the low cardiac output leading to multiorgan failure using neck vessels. After two weeks on ECMO support, the patient was successfully weaned and separated from ECMO support. Both Coronary Multi-detector Computed Tomography (MDCT) and heart catheterization were performed with suspicion of ALCAPA syndrome. Figure 2 shows the coronary arteries anatomy.

The shadow indicated by the black arrow may appear as the pulmonary cusp, which the left coronary artery drains, in Figure 3. Although origin of the left coronary artery from aorta or pulmonary artery, could not be visualized, the patient underwent operation for left coronary artery revascularization, with the preoperative possible diagnosis of ALCAPA. After a midline sternotomy, the thymus was resected, and the pericardium was opened. After heparinization, aortic and bicaval cannulation was performed to initiate cardio-pulmonary bypass (CPB). The pulmonary artery and aorta were dissected from each other. Diastolic cardiac arrest was provided with 30°C systemic hypothermia and antegrade tepid blood cardioplegia after cross-clamping. Tepid blood cardioplegia was repeated every 20 minutes until releasing cross-clamp. The left side of the heart was vented. Pulmonary arteriotomy and aortotomy were performed. There were no left coronary ostia neither in the pulmonary artery nor in the aorta.

The Circumflex artery (Cx) and left anterior descending artery (LAD) were visible, but small in diameter. They were merged into a small blind pouch, 2 cm away from the left coronary sinus of the aorta. There was no possibility for preparation and anastomosis to the aorta, due to very small diameter and fragile vessel. Left internal mammary artery (LIMA) was harvested. LAD, near to the pouch was opened, LIMA-LAD anastomosis was performed with 8-0 prolene. Pulmonary artery and aortotomy were closed. Cross clamp was removed after de-airing. The patient was weaned from CPB with high dose s of inotropic support including, adrenaline, noradrenaline, dopamine, and milrinone. The sternum was left open due to hemodynamic instability. Cardiopulmonary bypass time was 141 minutes, and cross-clamp time was 101 minutes.

The patient needed ECMO support due to the low cardiac output on the second postoperative day. The patient died of massive cranial hemorrhage on ECMO, despite some improving ventricular function in the 10th postoperative day.

Discussion

LMCAA is a quite rare condition with unclear etiology. In LMCAA, the left coronary system receives blood only by collateral arteries from the RCA. Thus, the heart may eventually be unable to cope with collateral circulations and develop myocardial ischemia. Patients mostly present with nonspecific symptoms depending on their age group and the formation of collateral vessels from the right coronary artery to the left coronary artery. Adult patients usually present with angina pectoris at an advanced age when collateral flow cannot keep pace with myocardial demands. Children and adolescents often present with chest pain,

dyspnea, syncope and tachyarrhythmia, and sudden cardiac death [2, 3]. Infants mostly present with growth retardation and myocardial infarction [2]. Catastrophic situations like sudden cardiac arrest, low cardiac output, and cardiomyopathy, as seen in our case, can also be the first symptoms of infants with LMCAA. [5, 6] Since these symptoms are not specific to LMCAA, the clinical diagnosis might be neglected. Thus, other coronary anomalies should be excluded to reach a correct diagnosis. As stated in a retrospective study done by Yildiz et al., LMCA was the most common anomalous vessel [1]. Separate origins of LAD and CX from the left coronary sinus of Valsalva were the most seen anomaly and should be excluded in patients presenting with symptoms of myocardial infarction. ALCAPA is one situation that LMCAA can be confused with and should be differentiated from [2,7].

Although congenital atresia of the left main coronary artery usually occurs as an isolated cardiac lesion, concomitant anomalies including bicuspid aorta, supravalvular aortic stenosis, right coronary ostial stenosis, pulmonary stenosis, ventricular septal defect, and mitral valve prolapsus secondary to myocardial ischemia can be seen [4, 8, 9].

The diagnosis of LMCAA can be done by coronary angiographic findings, which usually show no left coronary ostium and left coronary artery filled in a retrograde manner via the RCA instead of antegrade blood flow. In recent years, MDCT has also played an essential role in diagnosing LMCAA in older children and adults and can be used in patients suspected of congenital coronary artery abnormalities [3, 10]. According to some researchers, MDCT provides more precise details in a less invasive way than coronary angiography and is thus recommended for evaluation of congenital coronary abnormalities. [11] MDCT not only defines the anatomic course and the ostium shape, but also has less complications as coronary spasm than conventional coronary angiography. However, there is no reliable research reporting the use of MDCT in infants with coronary anomalies. [9]

The prognosis of LMCAA is poor [2]. Due to the symptomatic nature of LMCAA and the risk of sudden cardiac death, patients in the pediatric population with LMCAA should undergo surgical intervention to restore the antegrade flow to the left coronary system. Various surgical interventions have been described and coronary artery bypass grafting using the internal mammary artery or saphenous vein has been identified as the treatment of choice, regardless of the caliber of left sided vessels [4, 8, 12]. A LIMA graft for LMCAA appears to be a reasonable early interventional approach with successful results 1 year postoperatively. Nevertheless, due to the rarity of this disease, long-term postoperative outcomes have not been reported yet [2, 4, 5, 8, 9, 11]. Alternative to bypass grafting, direct surgical reimplantation or reconstruction of the left main coronary artery using azygos vein have been described to provide the shortest and most efficient way for blood to the myocardium. [6, 12, 13]. The advantages they offer relative to bypass grafting are prevention of the occlusion of main trunk, provision of the antegrade flow and avoidance of competitive flow. After the application of these techniques, reoperative surgery can be easily performed. However, application of this technique for LMCAA is not common and the long-term outcomes of the surviving patients were unknown [6, 12].

Conclusion

LMCAA is an extremely rare congenital coronary anomaly in which the left coronary ostium and left the main trunk in the left coronary artery system are absent. Considering the severe symptoms such as myocardial infarction or sudden cardiac death, surgical revascularization should probably be the treatment of choice.

References

1. Yildiz, A., Okcun, B., Peker, T., Arslan, C., Olcay, A., & Bulent Vatan, M. (2010). Prevalence of coronary artery anomalies in 12,457 adult patients who underwent coronary angiography. *Clinical cardiology* , *33* (12), E60–E64. https://doi.org/10.1002/clc.20588
2. Tian, M., Wang, X., Gao, H., Wang, L., & Hu, S. (2018). Left main coronary artery atresia with concomitant mitral regurgitation in an adult: A case report. Medicine, 97(41), e12367. https://doi.org/10.1097/MD.0000000000012367

3. Fujita, S., Sato, A., Nagata, Y., Usuda, K., Murata, A., & Hatasaki, K. (2017). Congenital left main coronary artery atresia presenting as syncope and generalized seizure during exercise in a 13-year-old boy. *Journal of cardiology cases* , *16* (4), 126–130. https://doi.org/10.1016/j.jccase.2017.06.004
4. Tanawuttiwat, T., O'Neill, B. P., Schob, A. H., & Alfonso, C. E. (2013). Left main coronary atresia. *Journal of cardiac surgery* , *28* (1), 37–46. https://doi.org/10.1111/jocs.12044
5. Yajima, S., Toda, K., Nishi, H., Yoshioka, D., Nakamura, T., Miyagawa, S., Yoshikawa, Y., Fukushima, S., & Sawa, Y. (2017). Redo coronary bypass grafting for congenital left main coronary atresia: a case report. Journal of cardiothoracic surgery, 12(1), 26. https://doi.org/10.1186/s13019-017-0588-2
6. Lin, Ying-Jui, et al. "Left Main Coronary Artery Atresia Masquerading as Dilated Cardiomyopathy Treated with Aortic Reimplantation." The Journal of Thoracic and Cardiovascular Surgery, vol. 130, no. 4, Oct. 2005, pp. 1210–11. DOI.org (Crossref), doi:10.1016/j.jtcvs.2005.06.011.
7. Gay, F., Vouhé, P., Lecompte, Y., Guarnera, S., Tamisier, D., Kachaner, J., & Neveux, J. Y. (1989). Atrésie de l'ostium coronaire gauche. Réparation chez un nourrisson de deux mois [Atresia of the left coronary ostium. Repair in a 2-month-old infant]. Archives des maladies du coeur et des vaisseaux, 82(5), 807–810.
8. D'Souza, Travis F., et al. "Surgical Treatment of Neonate With Congenital Left Main Coronary Artery Atresia." The Annals of Thoracic Surgery, vol. 101, no. 1, Jan. 2016, pp. 352–55. DOI.org (Crossref), doi:10.1016/j.athoracsur.2014.12.104.
9. Sohn, S. Y., Jang, G. Y., & Choi, B. M. (2010). Congenital atresia of the left main coronary artery in an infant. *Journal of Zhejiang University. Science. B* , *11* (7), 539–541. https://doi.org/10.1631/jzus.B0900361
10. Ten Kate, G. J., Weustink, A. C., & de Feyter, P. J. (2008). Coronary artery anomalies detected by MSCT-coronary angiography in the adult. *Netherlands heart journal : monthly journal of the Netherlands Society of Cardiology and the Netherlands Heart Foundation* , *16* (11), 369–375. https://doi.org/10.1007/BF03086181
11. Saedi, S., Pouraliakbar, H. R., Ghaderian, H., & Saedi, T. (2018). Congenital atresia of left main coronary artery. The Egyptian heart journal : (EHJ) : official bulletin of the Egyptian Society of Cardiology, 70(4), 451–453. https://doi.org/10.1016/j.ehj.2018.10.005
12. Musiani, A., Cernigliaro, C., Sansa, M., Maselli, D., & De Gasperis, C. (1997). Left main coronary artery atresia: literature review and therapeutical considerations. European journal of cardio-thoracic surgery : official journal of the European Association for Cardio-thoracic Surgery, 11(3), 505–514. https://doi.org/10.1016/s1010-7940(96)01121-9
13. Takeuchi, D., Mori, Y., Kishi, K., Nakajima, T., Nakazawa, M., Tsurumi, Y., Kurosawa, H., & Nakanishi, T. (2009). Percutaneous transluminal coronary angioplasty for postoperative left coronary artery stenosis following surgical reconstruction of congenital atresia of the left main coronary artery. Circulation journal : official journal of the Japanese Circulation Society, 73(12), 2360–2362. https://doi.org/10.1253/circj.cj-08-0136