Journal of Zhejiang University-SCIENCE B (Biomedicine & Biotechnology)
ISSN 1673-1581 (Print); ISSN 1862-1783 (Online)
www.zju.edu.cn/jzus; www.springerlink.com
E-mail: jzus@zju.edu.cn

Case Report:

Congenital atresia of the left main coronary artery in an infant

Su Ye SOHN, Gi Young JANG[†‡], Byung Min CHOI

(*Department of Pediatrics, Korea University Hospital, Ansan 425-707, Korea*)

[†]E-mail: JGYNHG@yahoo.co.kr

Received Nov. 11, 2009; Revision accepted Feb. 26, 2010; Crosschecked June 2, 2010

Abstract: Congenital atresia of the left main coronary artery is a rare occurrence, and surgical revascularization-bypass graft is required. We here report a rare case of congenital coronary anomaly in an infant. A 10-month-old male infant was admitted to the hospital with heart failure symptoms. Echocardiographic examinations revealed mitral valve regurgitation and ischemic changes of the anterolateral papillary muscle and chordae. Coronary angiography showed atresia of the left main coronary artery with a severe hypoplastic left anterior descending artery and a circumflex coronary artery. Unfortunately, sudden cardiac arrest occurred after catheterization and the infant did not recover despite of immediate cardiopulmonary resuscitation. Further studies are needed to find a newer diagnostic method to detect coronary anomaly in an infant, and coronary angiography, if necessary, has to be performed very carefully.

Key words: Congenital heart disease, Anomalous coronaries, Infant
doi:10.1631/jzus.B0900361 **Document code:** A **CLC number:** R72

1 Introduction

Congenital anomaly of the coronary arteries is a rare disease occurring in 1%–2% of all congenital heart diseases. Atresia of the left main coronary artery is one of its least-frequently observed variations, with very few cases presented in the literature (Amaral *et al*., 2000; Gebauer *et al*., 2008). The prognosis is unfavorable because it results in myocardial ischemia or infarction, and even sudden cardiac death. Surgical revascularization of the left main coronary artery is recommended in most patients with atresia of the left coronary artery because of unfavorable clinical outcome, of which only a few cases have been reported for successful surgical revascularization (Sunagawa *et al*., 2005; Gebauer *et al*., 2008). In this paper, we report a case of congenital atresia of the left main coronary artery, which was confirmed by coronary angiography and autopsy.

2 Case report

A 10-month-old male infant was admitted and evaluated at the cardiology clinic because of cardiac murmur and heart failure symptoms such as mild tachypnea and sweating. On the physical examination, a grade II/VI systolic ejection murmur was heard at the apex. Chest X-ray showed mild cardiomegaly and increased pulmonary vascularity. Echocardiographic examinations showed a dilated left atrium and a left ventricle with an ejection fraction of 70%. It also showed a mitral valve prolapse of the anterior leaflet with a grade III/IV mitral valve regurgitation and ischemic changes of the anterolateral papillary muscle, chordae, and endocardium. The right coronary artery was dominant, the left coronary artery was hypoplastic, and the ostium was not definitely visible during the echocardiographic examinations. A hemodynamic study was therefore required to confirm the coronary anomaly. Coronary angiogram revealed atresia of the left main coronary artery with a severe hypoplastic left anterior descending artery and a circumflex coronary artery (Figs. 1 and 2).

[‡] Corresponding author

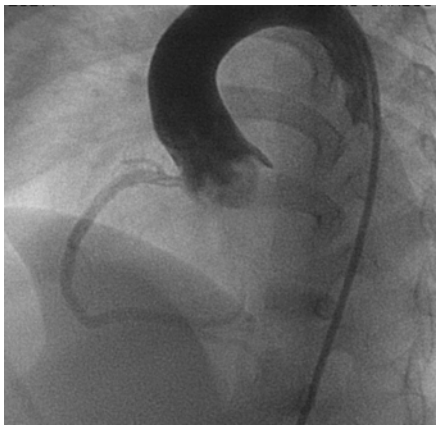

Fig. 1 Aortography demonstrating absence of any left coronary artery filling

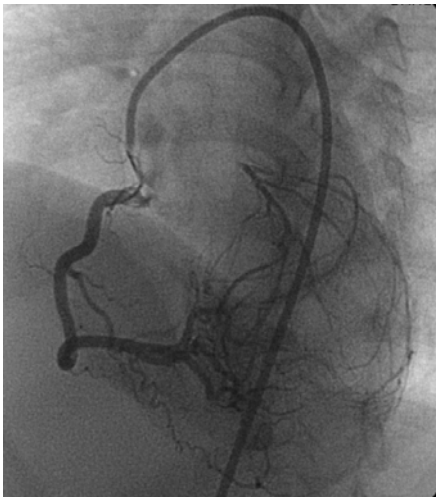

Fig. 2 Right coronary angiogram showing dilated right coronary artery with collaterals filling the branches of the left coronary artery

There were multiple collateral branches from the right coronary artery that were supporting the territory of the left ventricle (Fig. 2). So, we considered surgical revascularization-bypass graft after cardiac catheterization. However, the patient had a sudden cardiac arrest within 4 h after the catheterization, and did not recover despite of strenuous cardiopulmonary resuscitation. Coronary spasm or embolism of the coronary artery and subsequent acute myocardial infarctions were presumed to be the cause of the death. An autopsy was performed and confirmed left coronary artery ostial atresia and ischemic necrosis of the papillary muscle and endocardium of the left ventricle, but unfortunately, pathology images were not made from postmortem examination.

3 Discussion

Congenital atresia of the left main coronary artery is an extremely rare condition, and surgical correction is recommended to improve clinical outcome. Clinical diagnosis is usually delayed because of the nonspecific and broad-spectrum of symptoms ranging from asymptomatic to life-threatening. Syncope, tachyarrhythmia, failure to thrive, or a sudden cardiac arrest might present during infancy or childhood in some patients (Singh *et al*., 2005), while other patients might remain asymptomatic due to the development of a well-organized effective collateral circulation from other branch arteries. Although congenital atresia of the left main coronary artery usually occurs as an isolated cardiac lesion, this anomaly may be associated with other cardiac defects such as bicuspid aortic valve, supravalvar aortic stenosis, pulmonary stenosis, and ventricular septal defect, especially in infants and children (Vidne *et al*., 1979; Satran *et al*., 2006). In our case, there were no other associated cardiac defects except prolapse of the mitral valve. Because of the unfavorable prognosis, surgical revascularization-bypass graft is required in most patients, no matter whether there is a presence of other associated cardiac defects or not. Successful coronary revascularization cases using the internal mammary artery and saphenous vein have been reported in children, but there are no reports on the long-term outcome (Amaral *et al*., 2000; Sunagawa *et al*., 2005).

Coronary angiography is the only reliable diagnostic tool for this anomaly in infant because it is difficult to confirm a left coronary artery atresia even with excellent quality two-dimensional imaging studies. Recently, the multislice computed tomography (MSCT) has been used as a first-line evaluation for coronary anomalies in adults (Ten Kate *et al*., 2008). It is not only favorable for defining the anatomic course and the ostium shape, but also decreases the risk, such as coronary spasm, of the conventional coronary angiography. However, there are no reliable references or trials reporting on the use of the MSCT in patients with coronary anomaly in infant. Further studies are needed to find a newer diagnostic method to detect coronary anomaly in infant, and coronary angiography, if necessary, has to be performed with extreme precaution.

References

Amaral, F., Tanamati, C., Granzotti, J.A., Haddad, J.L., Leite, J.R., Barbero-Marcial, M., 2000. Congenital atresia of the ostium of the left coronary artery. Diagnostic difficulty and successful surgical revascularization in two patients. *Arquivos Brasileiros de Cardiologia*, **74**(4):339-342. [doi:10.1590/S0066-782X2000000400005]

Gebauer, R., Cerny, S., Vojtovic, P., Tax, P., 2008. Congenital atresia of the left coronary artery—myocardial revascularization in two children. *Interactive CardioVascular and Thoracic Surgery*, **7**(6):1174-1175. [doi:10.1510/icvts.2008.184317]

Satran, A., Dawn, B., Leesar, M., 2006. Congenital ostial left main coronary artery stenosis associated with a bicuspid aortic valve in a young woman. *The Journal of Invasive Cardiology*, **18**(3):E114-E116.

Singh, C., Singh, H., Kumar, A., Banerji, A.K., Aggarwal, N., Bharadwaj, P., 2005. Congenital atresia of left main coronary artery. *Indian Heart Journal*, **57**(3):255-257.

Sunagawa, M., Shimabukuro, T., Kado, H., Ushinohama, H., Ohta, T., 2005. Congenital atresia of the left main coronary artery; successful surgical treatment (myocardial revascularization and mitral valve repair) in a 1-year-old boy. *European Journal of Pediatrics*, **164**(7):461-462. [doi:10.1007/s00431-005-1677-0]

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*, **16**(11):369-375.

Vidne, B.A., Nili, M., Aygen, M., Levy, M.J., 1979. Congenital atresia of the left main coronary artery ostium. *Scandinavian Journal of Thoracic & Cardiovascular Surgery*, **13**(1):37-40.