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CASE REPORT

Congenital Atresia of the Left Main Coronary Artery With Noncompaction of the Ventricular Myocardium in an Asymptomatic Young Child

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Abstract Congenital atresia of the left main coronary artery (LMCA) is an extremely rare cardiac anomaly, and no cases have been reported from the mainland of China. The diagnosis for the 20-month-old boy in the reported case highlights the essentiality of comprehensive diagnostic measures. To avoid a misdiagnosis, electrocardiographic and echocardiographic evidence should be vigilantly explored in young children suspected of having dilated cardiomyopathy. This is the first case report of LMCA atresia associated with noncompaction of the left ventricular myocardium.

Keywords Congenital atresia · Left main coronary artery · Noncompaction of ventricular myocardium · Electrocardiography · Echocardiography

Introduction

Congenital atresia of the left main coronary artery (LMCA) is an extremely rare cardiac anomaly. In this condition, the ostium or main trunk of the left coronary artery (LCA) is atretic, and the LCA with its major branches—the left anterior descending (LAD) and left circumflex (LCX) arteries—are supplied by the right coronary artery (RCA) via coronary collaterals in a retrograde direction. Only 33 cases of LMCA atresia (17 pediatric and 16 adult cases) had been reported until 2003 [3], and no cases have been reported from the mainland of China.

Frequently, LMCA atresia occurs as an isolated cardiac anomaly, but it also has been described in association with other cardiac defects such as supravalvular aortic stenosis, ventricular septal defect with pulmonary stenosis, and stenosis of the RCA ostium [7]. We present the case of a young Chinese boy with LMCA atresia and noncompaction of the ventricular myocardium (NVM) and review the clinical features, diagnostic measures, and therapeutic strategies of LMCA atresia.

Case Report

In January 2012, a 20-month-old boy was referred to Fuwai Hospital for treatment of mitral insufficiency. He had experienced a respiratory tract infection at 2 months of age and was found to have a heart murmur. When he was 6 months old, echocardiography at his local hospital showed left ventricular (LV) dilation with significant mitral regurgitation. He had been asymptomatic and unlimited in physical activity since that time.

At his physical examination, the boy had a normal heart rate (110 bpm), blood pressure (94/52 mmHg), and growth (11 kg). He had normal S1 and S2 without S3 or S4 gallop. A grade 3/6 systolic murmur was heard at the cardiac apex. His lungs were clear to auscultation, and he had no peripheral edema.

Chest x-ray exhibited an enlarged cardiac silhouette with a cardiothoracic ratio of 0.62. An electrocardiogram (ECG) demonstrated deep and wide Q waves with inverted or biphasic T waves in leads I, aVL, and V3–V5 (Fig. 1),

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which raised a high suspicion for anomalous origin of the LCA from the pulmonary artery (ALCAPA).

Echocardiography showed prominent LV dilation, myocardial noncompaction, echogenic mitral chordae, and moderate to severe mitral regurgitation (Fig. 2a, b). The LV ejection fraction was 58 %, with an end-diastolic dimension of 45 mm.

Color Doppler imaging showed small coronary collaterals within the ventricular septum and LV free wall (Fig. 2c). The ratio of the proximal RCA to the aortic (Ao) annulus diameter (RCA/Ao) was 0.18. A presumed "LCA" was connected to the left aortic sinus (Fig. 2d).

Based on these noninvasive findings and uncertainty of diagnosis, we performed multidetector computed tomography (MDCT), which demonstrated the absence of the LCA ostium and left main trunk. Subsequent aortic root angiography showed only a dilated RCA originating from the aorta. A diminutive LAD and LCX showed delayed filling via small coronary collaterals, which ended blindly at their confluence without a shunt into the pulmonary artery (PA) (Fig. 3a, b). Left ventriculography confirmed the presence of NVM (Fig. 3c). We considered the LMCA atresia to be congenital because the reported child had no history or signs of Kawasaki disease or autoimmune diseases and because an atherosclerotic lesion was deemed rare in very young children.

After consulting with cardiac surgeons, we considered the boy's LCA too diminutive for a graft. Considering his preserved LV function, we chose standard medical therapy for heart failure at the time including digoxin, diuretics, and angiotensin-converting enzyme inhibitor. The boy was asymptomatic without further deterioration of LV function at the 6-month follow-up evaluation.

Discussion

The clinical manifestations of LMCA atresia are diverse in the literature due to variation in collateral circulation development and different presenting ages. The presenting symptoms in infants and young children include failure to thrive, heart failure, and sudden cardiac death [7]. The children often had significant LV dilation and dysfunction due to insufficient collateral circulation, and dilated cardiomyopathy (DCM) was a common initial diagnosis [6]. In the reported case, heart failure symptoms were absent despite significant LV dilation and dysfunction.

Comprehensive diagnostic measures are essential. In infants and young children presenting with DCM, the characteristic infarct-like pattern (deep and wide Q waves with inverted or biphasic T waves in the anterolateral leads, especially lead aVL) on ECGs could be suggestive of ALCAPA or LMCA atresia [1, 2, 7, 12]. This pattern also provides a valuable clue for ultrasound inspection. Indirect echocardiographic signs portending the presence of ALCAPA or LMCA atresia involve enhanced echogenicity of the mitral apparatus, a dilated RCA, and coronary collaterals within the ventricular septum and LV free wall [2, 10, 12].

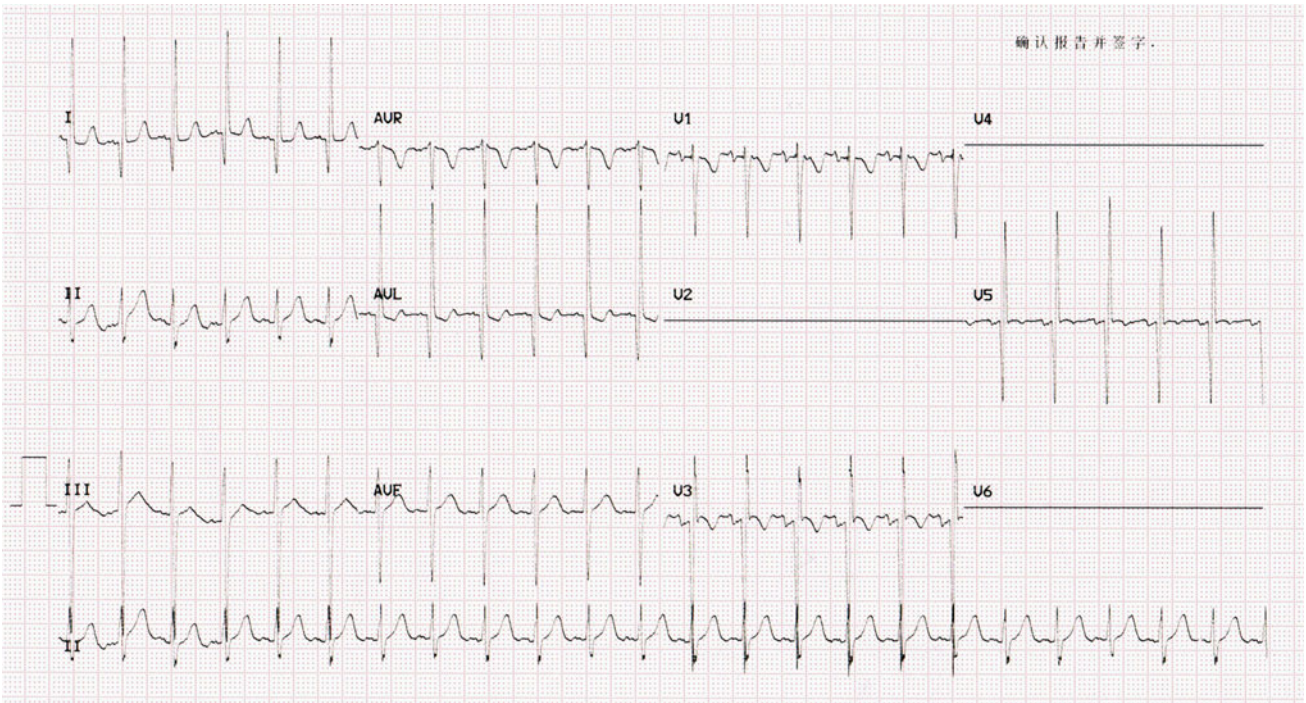

Fig. 1 ECG showing deep and wide Q waves with inverted or biphasic T waves in leads I, aVL, and V3–V5

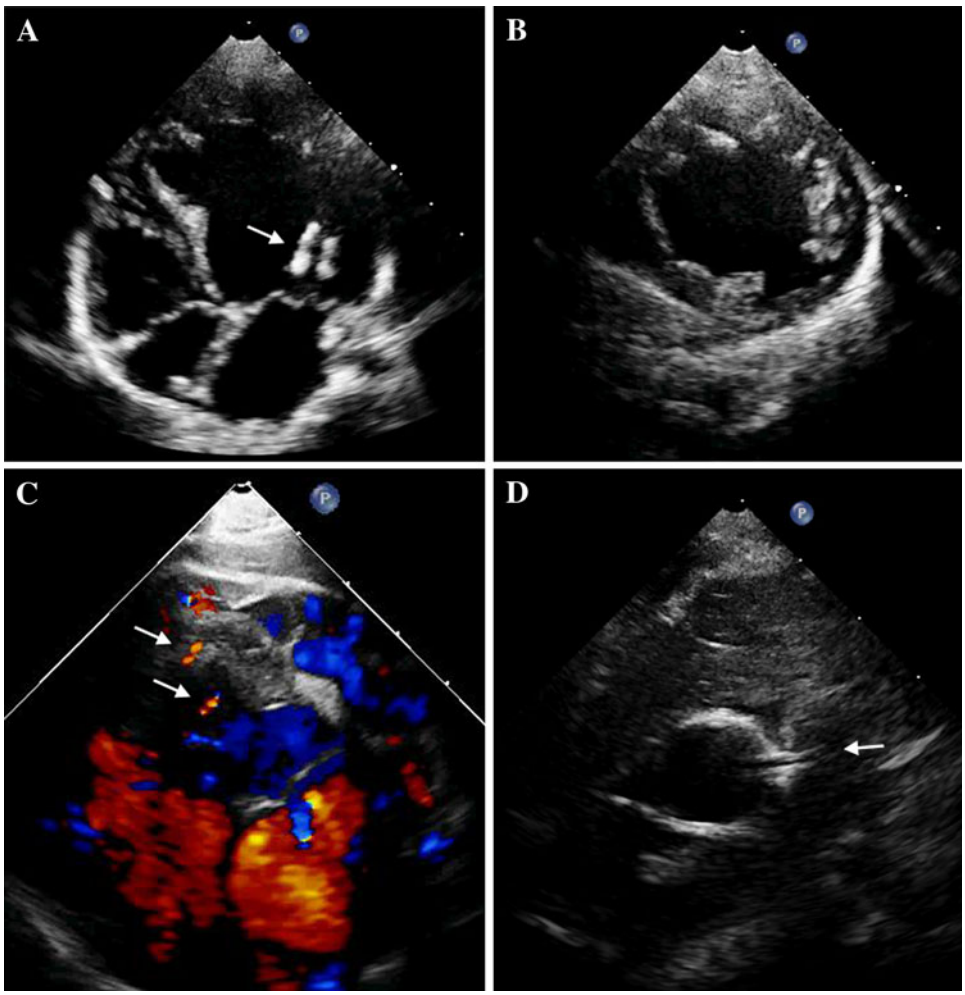

Fig. 2 Echocardiogram showing **a** prominent LV enlargement (apical aneurysmal dilation) and echogenic mitral chordae (*arrow*), **b** noncompaction of the LV myocardium, **c** small coronary collaterals (*arrows*), and **d** a presumed left coronary artery (*LCA*) seemingly arising from the left aortic sinus (*arrow*)

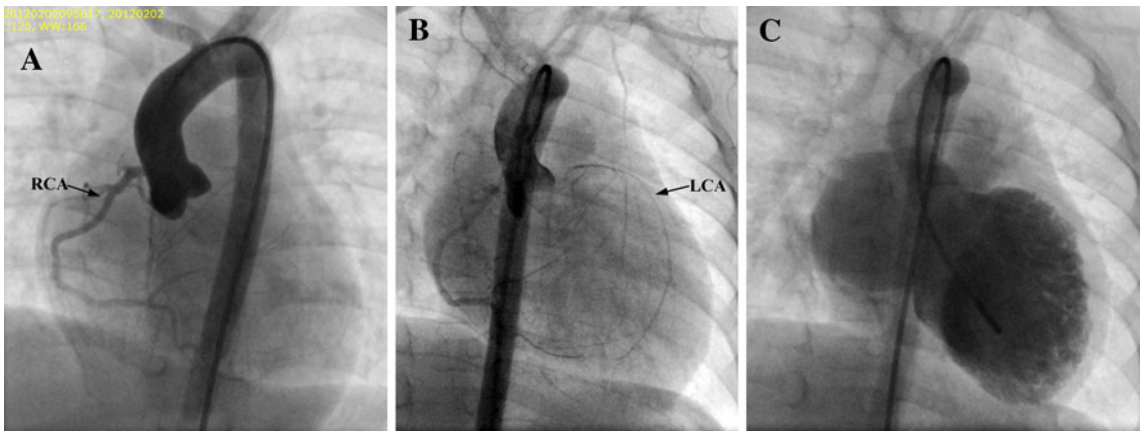

Fig. 3 Aortic root angiogram showing **a** a dilated right coronary artery (*RCA*) originating from the aorta and **b** delayed filling of the diminutive left coronary artery (*LCA*) via small coronary collaterals and its blind ending. **c** Left ventriculography showing myocardial noncompaction

The RCA/Ao ratio (0.18) was mildly increased in this case compared with that of DCM (<0.14) [2, 5] but lower than that of ALCAPA (0.26 ± 0.05 reported by Han and co workers [12], and 0.21 to 0.29 reported by Koike et al [5]). Detection of a red blood flow from the LCA to the PA is confirmative for the diagnosis of ALCAPA [2, 12] and is not seen in LMCA atresia.

Interestingly, a presumed LCA was noted to be connected to the left aortic sinus in the reported case, whereas subsequent MDCT and invasive angiography did not

demonstrate this connection. This phenomenon also was mentioned in previous reports and could lead to a false diagnosis [1, 4, 10]. Because it is unknown why the LCA seemed to arise from the aortic root, to avoid a misdiagnosis, ECG and echocardiographic evidence for LMCA atresia should be vigilantly explored in young children suspected of having DCM.

In older children and adolescents, tachycardia and syncope have been the main complaints [3, 7]. Most adult patients often had a normal and unrestricted childhood and began showing symptoms (angina) only at an advanced age [7]. They usually had preserved LV function due to a markedly dilated RCA and extensive collaterals. The ECG changes were nonspecific and indistinguishable from those of atherosclerotic coronary heart disease. The defect often was shown incidentally by coronary angiography or MDCT [8].

In recent years, MDCT has been adopted as a noninvasive measure to delineate coronary artery abnormality. Many authors have reported a successful diagnosis of LMCA atresia in adults using MDCT, which showed the absence of the LCA ostium and the left main trunk [8]. However, coronary angiography often is the confirmative diagnostic method of choice for infants and very young children [1, 6, 10]. The LCA system usually is hypoplastic and ends blindly [1, 6, 10]. These angiographic features are helpful in differentiating this condition from ALCAPA, in which the LCA system has a normal or increased caliber with retrograde flow into the PA. The difference in the LCA and RCA sizes might be attributable to flow-mediated dilation and growth in ALCAPA as well as insufficient flow in LMCA atresia.

The peculiarity of the reported case was the association with NVM, an uncommon unclassified cardiomyopathy thought to be caused by premature arrest of normal embryogenesis of the endocardium and myocardium [11]. During early embryonic development, the myocardium is a loose network of interwoven fibers separated by deep recesses that link the myocardium with the ventricular cavity. These recesses facilitate myocardial supply directly from the ventricular cavity by diffusion before arteriogenesis of the coronary arteries. Gradual compaction of this spongy meshwork occurs at weeks 5 to 8 of embryonic life, from the epicardium to the endocardium and from the base to the apex. The coronary circulation develops concurrently in the process to adapt to ventricular growth, and the intertrabecular recesses are reduced to capillaries. Thus, myocardial compaction and coronary arteriogenesis are inherently interconnected in theory, and coronary artery anomalies might coexist with NVM, as presented in several cases of ALCAPA and RCA atresia [9]. It has been suggested that in these cases, severe myocardial ischemia does not allow normal regression of embryonic myocardial sinusoids (recesses), so the embryonic pattern of trabeculated myocardium persists [11].

If left untreated, LMCA atresia carries an unfavorable prognosis. Surgical reconstruction of a two-coronary-artery system often is the therapeutic strategy of choice. Coronary artery bypass grafting (CABG) is the preferred procedure for adults, but its long-term results in pediatric patients are unknown, although good results have been reported with internal mammary artery grafted onto the LAD [1, 7, 10]. Successful reconstruction of the LMCA with a baffle from the ascending aorta and direct reimplantation also have been reported [6, 7].

Considering the technical difficulty of CABG due to the diminutive size of the LCA, we switched to medical therapy for heart failure in the reported case. Several infants with uncorrected LMCA atresia have been reported to survive into adolescence with effective medical treatment of heart failure but still had a high risk of sudden cardiac death [1, 6]. Therefore, surgical correction should be reconsidered for an older age when the caliber of the LCA is larger.

In summary, this is the first case report to describe LMCA atresia in association with NVM. This case suggests that comprehensive diagnostic measures are essential and that ECG and echocardiographic evidence should be vigilantly explored in young children with LV enlargement and suspected DCM. Medical therapy for heart failure is mandatory and could be effective for survival to surgery if the LCA is too diminutive for a graft procedure.

References

1. Blasco PB, Comas JG, Ferre MM, del Munoz RA (2006) Angor in a child: left main coronary artery atresia. Int J Cardiol 108:109–110
2. Chang RR, Allada V (2001) Electrocardiographic and echocardiographic features that distinguish anomalous origin of the left coronary artery from pulmonary artery from idiopathic dilated cardiomyopathy. Pediatr Cardiol 22:3–10
3. Elian D, Hegesh J, Agranat O, Guetta V, Har-Zahav Y, Rath S, Chouraqui P, Di Segni E (2003) Left main coronary artery atresia: extremely rare coronary anomaly in an asymptomatic adult and an adolescent soccer player. Cardiol Rev 11:160–162
4. Furuyama H, Kinugawa Y, Nakajima M, Okajima M, Okajima S, Nakayama T, Sawada H, Ohta Y (2010) Congenital atresia of left main coronary artery followed up for ages as a sequela of Kawasaki disease. Pediatr Int 52:e131–e133
5. Koike K, Musewe NN, Smallhorn JF, Freedom RM (1989) Distinguishing between anomalous origin of the left coronary artery from the pulmonary trunk and dilated cardiomyopathy: role of echocardiographic measurement of the right coronary artery diameter. Br Heart J 61:192–197
6. Lin YJ, Liang CD, Ko SF, Huang CF, Chang JP (2005) Left main coronary artery atresia masquerading as dilated cardiomyopathy treated with aortic reimplantation. J Thorac Cardiovasc Surg 130:1210–1211

7. Musiani A, Cernigliaro C, Sansa M, Maselli D, De Gasperis C (1997) Left main coronary artery atresia: literature review and therapeutical considerations. Eur J Cardiothorac Surg 11:505–514
8. Srinivas B, Suryawanshi SP, Seshagiri RD (2011) Left main coronary artery atresia: the role of computed tomographic coronary angiography. Pediatr Cardiol 32:1262–1263
9. Stollberger C, Finsterer J (2004) Left ventricular hypertrabeculation/noncompaction. J Am Soc Echocardiogr 17:91–100
10. Sunagawa M, Shimabukuro T, Kado H, Ushinohama H, Ohta T (2005) Congenital atresia of the left main coronary artery: successful surgical treatment (myocardial revascularisation and mitral valve repair) in a 1-year-old boy. Eur J Pediatr 164: 461–462
11. Weiford BC, Subbarao VD, Mulhern KM (2004) Noncompaction of the ventricular myocardium. Circulation 109:2965–2971
12. Zheng J, Ding W, Xiao Y, Jin M, Zhang G, Cheng P, Han L (2011) Anomalous origin of the left coronary artery from the pulmonary artery in children: 15 years experience. Pediatr Cardiol 32:24–31