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CASE REPORT

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Congenital left main coronary artery aneurysm

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Abstract

Left main coronary artery aneurysm (LMCAA) is an uncommon coronary abnormality seen in 0.1% of patients during routine diagnostic coronary angiographies. The most common etiology is atherosclerosis in acquired cases. However, it can also be a congenital malformation. We present the case of a 26 year-old female with a large LMCAA. She was diagnosed with tetralogy of Fallot initially. (Cardiol J 2011; 18, 4: 430–433)

Key words: coronary aneurysm, congenital

Introduction

Coronary artery aneurysms are described as localized coronary artery dilations greater than 1.5–2 times the diameter of the adjacent segments [1]. The incidence of coronary artery aneurysms has been reported as 0.3% to 4.9% among all patients who underwent coronary angiography, and they usually occur in men [1, 2]. The most common cause is atherosclerosis [1]. The next most common cause is idiopathic which is often presumed to be a congenital malformation. Coronary artery aneurysms in patients below the age of 33 are considered to be congenital [3]. Left main coronary artery aneurysm (LMCAA) is the most uncommon coronary abnormality among all coronary artery aneurysms. Its incidence is 0.1% among adult patients who underwent routine coronary angiography [4]. Here we present a 26 year-old female with large congenital LMCAA. She was initially diagnosed with tetralogy of Fallot by echocardiography. Cardiac catheterization and multislice computed tomography evaluation revealed the significant LMCAA.

Case report

A 26 year-old female was admitted to our hospital complaining of shortness of breath, fatigue and palpitation. She have had dyspnea since the age of 20. Cyanosis has been present since childhood. This was the first admission of the patient to a health facility. On physical examination, the patient was active, and co-operation and orientation were normal. Central cyanosis with finger clubbing and significant cachexia were noticed. Her blood pressure was 115/70 mm Hg and heart rate was irregular (130 beat/min). Cardiovascular system examination revealed a systolic thrill at the left sternal border with 4/6 pansystolic murmur best heard at the apex and mesocardiac area. Electrocardiography (ECG) revealed incomplete right bundle branch block, right axis deviation and atrial fibrillation with rapid ventricular rate. The chest X-ray revealed enlarged cardiac silhouette, suggesting right atrial and ventricular enlargement. Her complete blood work and arterial blood gas analysis were as follows: hemoglobin: 14.7 g/dL, hematocrit: 46.7%, arterial pH:

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7.528, pCO_2: 43.1 mm Hg, pO_2: 56.2 mm Hg, sO_2: 67.4%, HCO_3^-: 36.1 mmol/L. Transthoracic echocardiography revealed the dextroposition of aorta, subvalvular pulmonary stenosis with a peak 55 mm Hg gradient, perimembranous type ventricular septal defect, right ventricular hypertrophy, mild mitral and tricuspid regurgitation and biatrial dilatation. She was diagnosed with tetralogy of Fallot and the patient was referred for cardiac catheterization to investigate the accompanying pathologies. The coronary angiography revealed a large (26 × 18 mm) fusiform LMCAA extending towards the left anterior descending and circumflex arteries. Coronary angiographic and intraoperative images are demonstrated in Figures 1 and 2. The right coronary artery was also aneurysmatic. Multislice computed tomography was considered for better imaging of the aneurysm and its relation to adjacent cardiac structures (Fig. 3). The patient was referred to cardiac surgery and a successful corrective operation was performed for the tetralogy of Fallot. LMCAA was not corrected during the surgery since a single stage operation for Fallot tetralogy and coronary aneurysm might have increased morbidity and mortality. Currently, the patient is in her second postsurgical month and is asymptomatic.

Discussion

Coronary artery aneurysms usually involve the right coronary artery, followed by the left anterior descending and the left circumflex coronary arteries [1, 4]. Left main coronary artery is the least frequently involved artery (with a prevalence of 0.1%) [4]. Limited numbers of cases have been reported so far [5]. Almost half of cases occur as a consequence of atherosclerosis. Other causes include congenital heart diseases, Kawasaki disease, polyarteritis nodosa, systemic lupus erythematosus, traumatic injury, Ehler-Danlos syndrome, Behcet disease, scleroderma, Marfan syndrome, and Takayasu arteritis [2]. Morphologically, aneurysms may be fusiform or saccular, single or multiple.

In addition to possible extension of the intimal atherosclerotic process to the vessel media resulting in erosion, ulceration and hemorrhage, nitric oxide has also been suspected in the development of aneurysms. Chronic overstimulation of the endothelium-derived relaxation factor may lead to coronary dilation and potentially detrimental results [6]. On the other hand, chronic hypoxia may have an important role in this process. Faller reported that short-term exposure of endothelial cells to hypoxia results in the release of predominantly vasoconstricting factors, while longer-term and more severe hypoxic exposure generates substances that can induce smooth muscle proliferation and remodeling of the vasculature and surrounding tissues and fibrosis [7]. Fibrosis and vascular remodeling may lead to aneurysmal dilation of coronary arteries under high intracardiac pressure states, such as congenital heart diseases. Chronic hypoxia and elevated intracardiac pressures as a result of tetralogy of Fallot may be the cause of LMCAA in our patient.

The clinical presentation of a coronary artery aneurysm varies. Atherosclerotic aneurysms usually present with ischemic symptoms such as angina and myocardial infarction [8]. Coronary angiography is the gold standard in the diagnosis of coronary aneurysm. However, multislice computed tomography is an excellent alternative diagnostic method, especially in determining the exact anatomy and its relation to adjacent structures. Myocardial ischemia, infarction, thrombosis, distal embolization, dissection, and rupture are the significant complications of coronary artery aneurysms [1, 3, 8]. Because of the limited number of LMCAA cases, the optimal treatment has not been established. There is no significant difference in survival rates of coronary artery aneurysm patients treated medically or surgically when compared to patients who had similar degrees of obstructive coronary disease, but no aneurysms [2].

Conservative therapy includes anti-platelet and anti-coagulant agents to prevent the formation of thrombus in the aneurysm and distal embolization. The presence of coronary aneurysm per se is not an indication for surgery. Usually, surgical treatment is preferred for patients who have evidence of embolization from the aneurysm to the distal coronary bed and large aneurysms to avoid the potential complications. Surgery is also indicated in cases of coronary artery aneurysm enlargement, as documented by serial angiographic measurements [9]. There are various options of surgical therapy for LMCAA such as ligation, resection, reconstruction and distal bypass. Covering the aneurysm with a graft stent is an invasive treatment [10]. Our patient was treated with corrective surgery for tetralogy of Fallot only. We decided to manage LMCAA conservatively at this point, since its distal extension was significant and it might have been hazardous for her to have a single stage procedure with this anatomy. The patient is doing well now and under appropriate anti-aggregant therapy.

Conclusions

LMCAA is an uncommon coronary abnormality. Conservative therapy is the preferred treatment

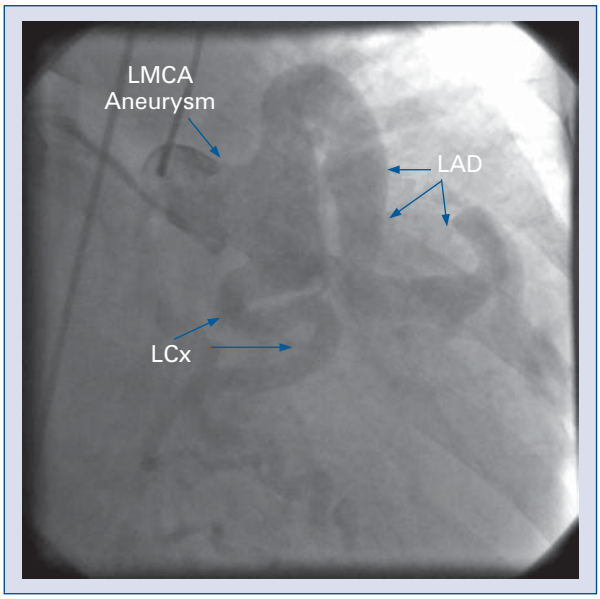

Figure 1. Coronary angiography from right caudal position demonstrating large left main coronary artery (LMCA) aneurysm and dilated left circumflex (LCx) and left anterior descending (LAD) arteries.

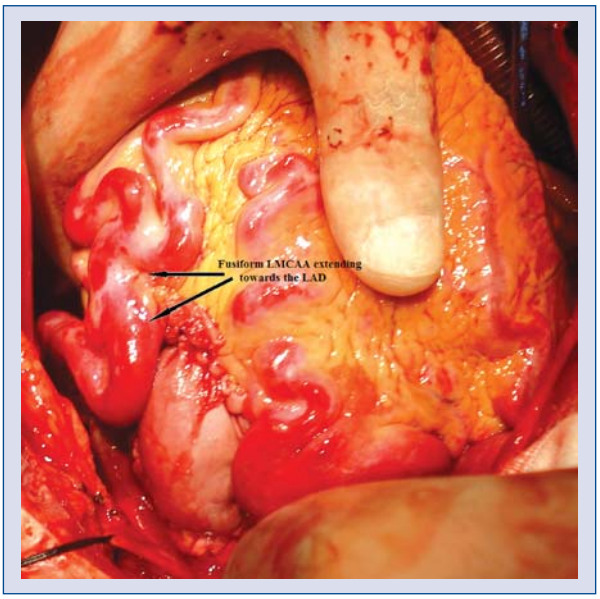

Figure 2. Intraoperative images of left main coronary artery aneurysm (LMCAA) and dilated coronary vessels; LAD — left anterior descending arteries.

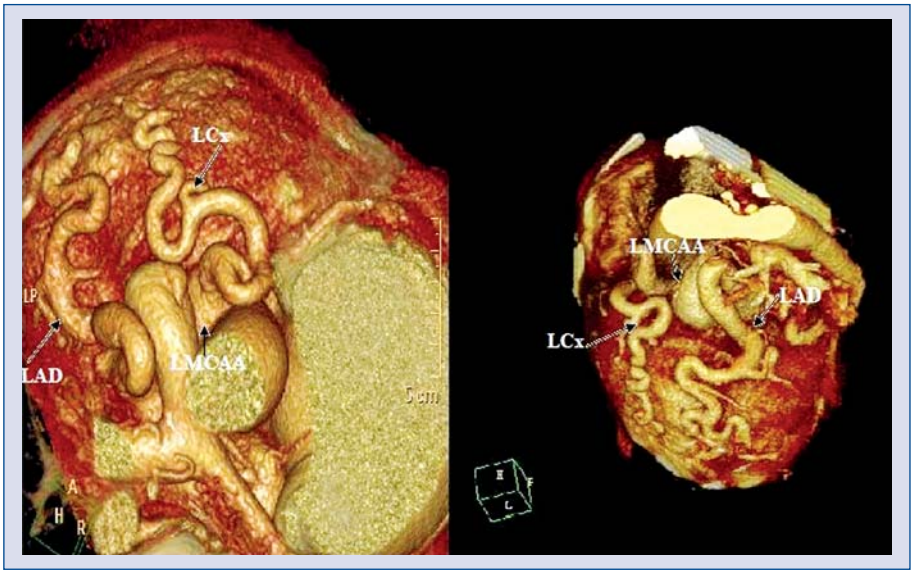

Figure 3. Multislice computed tomography images visualizing left main coronary artery aneurysm (LMCAA) and coronary vessels; LCx — left circumflex arteries; LAD — left anterior descending arteries.

in uncomplicated cases. Multislice computed tomography is the appropriate alternative diagnostic method to a conventional coronary angiogram to diagnose coronary aneurysms.

Acknowledgements

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