

Surgical Treatment & Long-term Follow Up of Giant Spherical Coronary Artery Aneurysm with Fistula to Left Ventricle: A Case Report

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ABSTRACT

Congenital fistula between the first diagonal branch of the coronary artery and left ventricle with a giant coronary artery aneurysm is extremely rare. We present the case of a 50-year-old asymptomatic male patient with such a condition that was diagnosed by transthoracic echocardiography, coronary computed tomography angiography, and coronary angiography. The patient was treated by surgical fistula closure under cardiopulmonary bypass. The postoperative coronary computed tomography angiography showed the patient got a complete cure, and the patient remains asymptomatic after 5-year follow up.

INTRODUCTION

Congenital coronary fistula (CAF) is an abnormal connection between the coronary artery and any of the four chambers of the heart or great vessels. It is a relatively rare condition, occurring in 0.18% of unselected patients undergoing diagnostic coronary angiography [Yamanaka 1990]. Coronary artery aneurysm (CAA) is a coronary dilatation that exceeds 1.5 times the diameter of a normal adjacent coronary artery or the largest coronary vessel. CAF originating from the diagonal branch of the left coronary artery and draining into the left ventricle is rare, and it is even rarer when it is accompanied with an associated spherical CAA. In this article, we present such a case to provide a better understanding of CAF.

CASE REPORT

A 50-year-old man presented to our department for evaluation and treatment of an abnormal cardiac silhouette found on chest X-ray during a routine physical examination after a hand trauma. He didn't complain of chest pain, palpitation, or other symptoms with unremarkable past medical history. Upon physical examination, the blood pressure was 152/88 mmHg, and the pulse rate was 76 beats per minute. No murmur was heard on precordial auscultation.

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The results of the blood test and electrocardiography (ECG) were normal. Transthoracic echocardiography revealed the left coronary artery markedly was dilated with a giant aneurysm. Color Doppler revealed blood flow communication between the aneurysm and left ventricle through a fistulous connection with a normal ejection fraction (EF) of 57%. Multi-slice spiral computed tomography angiography (MSCTA) revealed both the left main coronary artery and first diagonal branch of the coronary artery were tortuous and dilated (up to 14 mm). The distal part of the dilated coronary

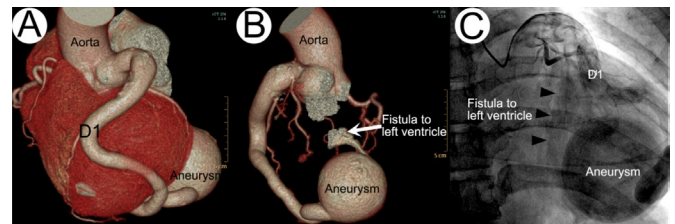


Figure 1. A, B) Three-dimensional CT scanning revealed the dilated left main coronary artery and the first diagonal branch of the coronary artery (14mm in diameter) with a giant coronary artery aneurysm (50mm in diameter), and the fistulous connection (light arrow) between the CAA and left ventricle. C) The flow from the first diagonal branch of the coronary artery drained into the coronary artery aneurysm, then emptied into the left ventricle rapidly (dark arrows). LM, left main coronary artery; LAD, left anterior descending; LCx, left circumflex artery; CAA, coronary artery aneurysm; D1, the first diagonal branch of coronary artery; Ao, aorta

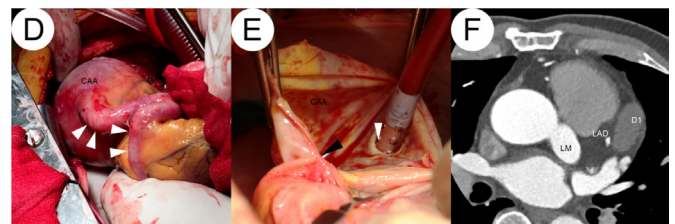


Figure 2. D) Intraoperative view, after incision of the pericardium, the dilated first diagonal branch of the coronary artery with a huge aneurysm was exposed clearly. E) After dissection of the aneurysm, the inlet (dark arrow) and outlet (light arrow) of the giant aneurysm were seen, and the aneurysmal wall was thin. F) 5 years after surgery, the first diagonal branch of coronary artery has been occlusion. LM, left main coronary artery; LAD, left anterior descending; D1, the first diagonal branch of coronary artery; CAA, coronary artery aneurysm

artery communicated with a giant spherical aneurysm that drained directly into the posterior wall of the left ventricle (Figure 1A and 1B). Selected left coronary angiography was performed to establish a definitive diagnosis for further interventions. The result showed a giant spherical aneurysm located at the posterior wall of the left ventricle, arising from the diagonal branch of the coronary artery and rapidly emptying into the left ventricle (Figure 1C). He was then diagnosed with a coronary artery fistula originating from the first diagonal branch of the coronary artery with an associated giant spherical aneurysm draining into the left ventricle.

After thorough preoperative preparations, surgical treatment was undertaken via standard median sternotomy with assistance of routine cardiopulmonary bypass (CPB). After the cardiac arrest was achieved, the giant aneurysm and dilated coronary artery were noted (Figure 2A). Dissection of the aneurysm was performed to expose the inlet and outlet of aneurysm, which were closed with two-way simple continuous sutures (Figure 2B). Following resection of the aneurysmal wall, the remaining wall was closed with simple continuous suture to avoid the catastrophic complication of recanalization. A biopsy was taken for histological evaluation and diagnosis. After adequate warming and weaning from the cardiopulmonary bypass, TEE confirmed a complete closure of the left ventricular fistula. The postoperative pathology validated the diagnosis of coronary artery aneurysm. The patient was discharged 1 week after surgery, and he is currently on warfarin. He remains asymptomatic after 5-year follow up; the MSCTA showed thrombosis in the first diagonal branch of the coronary artery (Figure 2C).

DISCUSSION

We present a patient diagnosed with congenital coronary artery fistula to the left ventricle and giant spherical coronary artery aneurysm. The patient received surgery.

Congenital coronary fistula (CAF) is extremely rare; it is present in 0.002% of the general population and represents 0.4% of all cardiac malformations [Yamanaka 1990; Dodge-Khatami 2000]. Coronary artery fistula most commonly originates from the right coronary artery and affects the right side of the heart. Only 1.7% of CAF originates from the diagonal branch of the left coronary artery, and only 10% of CAF drains into the left-sided chambers [Mangukia 2012]. In untreated patients, 81% of patients aged younger than 20 years are asymptomatic, while the percentage of older patients is 37% [Mavroudis 1997]. The incidence of symptoms and complications rise with increased age and is influenced by the size and location of the fistula. The characteristic clinical

symptoms include chest pain, exertional dyspnea, and fatigue. Various complications have been reported, such as left ventricular hypertrophy, congestive cardiac failure, arrhythmias, myocardial infarction, infective endocarditis, and coronary artery aneurysm [Mangukia 2012]. In our case, the patient was asymptomatic, while both left ventricular hypertrophy, coronary dilation and an associated aneurysm occurred. We think the left ventricular hypertrophy may have been caused by diastolic volume overload of the left ventricle resulting from left-to-left-shunt, and the aneurysm formation was caused by hemodynamic anomaly.

Surgical closure of CAF is recommended for the patients with aneurysm because of the possibility of acute thrombosis which results from the aneurysm and the hemopericardium which results from aneurysm rupture [Liberthson 1979]. Aneurysm located on the posterior surface of the heart makes fistula inaccessible. This situation is an indication for cardiopulmonary bypass (CPB) [Mangukia 2012]. Accordingly, we decided to undertake surgical closure under CPB. Closure from within the coronary artery aneurysm was performed instead of proximal and distal ligations. Because the aneurysmal draining site with the fistula at the left ventricular wall was totally closed, it was appropriate to perform such an operation to avoid the difficulties of exposing the fistula inside the left ventricle.

In conclusion, the diagonal branch of the left coronary artery draining into the left ventricle with a giant CAA is rarely seen in congenital CAF. MSCTA is helpful for the diagnosis of CAF. Surgical treatment is recommended for patients of CAF with an associated aneurysm, which is very likely to rupture.

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