Case report - Congenital
Successful correction of congenital giant right coronary artery aneurysm with fistula to left ventricle

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Abstract

A 26-year-old male presented with radiographical evidence of enlargement of the right side of the heart. Echocardiography and computed tomography-scan revealed a diffuse, hugely enlarged right coronary artery (RCA) aneurysm, with its distal portion flowing directly into the left ventricle (LV). A radical correction operation was performed successfully. The RCA was reconstructed with an autograft of a greater saphenous vein after the aneurysm was removed and the RCA-to-LV fistula was closed. The postoperative course was uneventful and the patient was discharged on 12th day after operation.

Keywords: Coronary artery aneurysm; Coronary artery fistula

1. Introduction

Congenital coronary artery fistulas are rare and unusual coronary artery abnormalities in which blood is shunted between RCA and left ventricle (LV). Diffusely dilated RCA is still considerably rare. The following case we present is a congenital giant RCA aneurysm with fistula to LV with impressive radiographical and echocardiographical presentations, as well as a good result of surgical correction.

2. Case report

A 26-year-old male, complaining of paroxysmal palpitation for two weeks visited our hospital. Plain radiograph revealed an enlarged shadow on the right side of the heart (Fig. 1e). On auscultation, a mild to-and-fro heart murmur could be found. Precordial echocardiography revealed a huge RCA, diffusely dilated, with a direct communication with the LV (Fig. 1f).

To evaluate the heart in more detail, contrast-enhanced multislice computed tomography (CT) was performed. Three-dimensional (3D) volume rendered images clearly revealed a huge, enlarged RCA aneurysm from the proximal to the distal portion of the RCA, with its distal portion terminating abruptly into LV without showing distal branchings (Fig. 1a and b). No wall thickness, calcification, or luminal stenosis could be observed in the entire thoracic aorta, RCA or major side branches, suggesting absence of arteritis.

To evaluate the status of each coronary artery, coronary angiography was performed and there was no obvious lesion on its coronary arteries and their branches, except this tortuous dilated giant RCA aneurysm, which drained directly into the posterior of the LV (Fig. 1c and d). As the patient did not have a past history of Kawasaki disease or Takayasu’s arteritis, and had no evidence of aortitis or arteritis, the RCA aneurysm resulted from the RCA-to-LV fistula.

After thorough preoperative preparation, this patient received radical surgical correction via median sternotomy approach. After the pericardium was opened, a hugely dilated RCA, about 2.5 cm in diameter, was exposed (Fig. 2a), with a giant aneurysm, approximately 5×5 cm in size, located in its distal portion, whose extremity was draining directly into the LV. The RCA was carefully inspected, with its three distributions noted and no calcification or atherosclerosis lesions were found. With the support of cardio-pulmonary bypass (CPB), the dilated RCA was opened longitudinally and its huge ostium was revealed, about 3 cm in diameter, which almost occupied the whole right valsalva sinus. The termination site was also identified, approximately 1.5 cm in size, directly communicating with the LV. There was no luminal stenosis, no clot or atherosclerotic lesion within the tortuous dilated RCA and its giant aneurysm. However, the aneurismal wall was a bit thinner than the dilated coronary wall. There was an obvious fiber ring covering the edge of the fistula orifice. The fistula...
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Fig. 1. Plain radiograph revealed the enlarged shadow of the heart, especially the right side of the heart (e). Precordial echocardiography revealed a large dilated RCA, with a fistula to LV. Three-dimensional CT image showed a diffuse, hugely enlarged RCA aneurysm, communicating directly with LV; coronary angiography showed the aneurysmatically dilated RCA and normal left coronary artery. RCA, right coronary artery; LV, left ventricle; CT, computed tomography; Aneu, aneurysm.

was closed with the same sized pericardium Dacron patch, the smooth side of auto pericardial patch toward the left ventricular cavity. An aortic root plasty was also performed by using a little bigger hemashield Dacron patch to repair the ‘defect’ of the huge right coronary ostium (Fig. 2b). After the patient was weaned from CPB, transesophageal echocardiography displayed normal function of the aortic valve and echocardiogram (ECG) did not reveal the existence of myocardial ischemia. The histological examination of the surgical specimens revealed the fibrocellular dysplastic change and mucoid degeneration in the wall of the RCA aneurysm (Fig. 2d).

The postoperative course of this patient was uneventful and he was discharged on the 12th day after operation. A close follow-up was performed postoperatively and this patient has recovered very well. Recent postoperative 3D-CT heart reconstruction images on this patient showed the RCA bypass graft at a nice position, with excellent blood flow (Fig. 2c).

Discussion

Congenital coronary fistulae are really uncommon abnormalities, most of which drain into a right heart chamber or into the pulmonary artery [1, 2]. Such a congenital RCA-to-LV fistula with a diffusely giant RCA and RCA aneurysm formation in an adult patient is even rarer.

Coronary artery aneurysm is also an uncommon disease, with an incidence of between 1.5% and 5% [3]. The most common cause of coronary artery aneurysm is atherosclerotic coronary artery disease [4], and it is also caused by other diseases, such as Kawasaki’s disease [5], Takayasu’s arteritis, infectious endocarditis, or iatrogenic complications, such as stent implantation [6]. In our patient, no such clinical findings were revealed. So the existence of RCA–LV fistula was thought to be the most likely pathogenesis [7]. The development of a RCA aneurysm associated with an abnormal RCA to LV connection might be due to the abnormal coronary flow. The RCA flow was turbulent to-and-fro flow between the systolic and diastolic phases. This abnormal coronary flow could overstretch the vessel wall of the RCA, and its associated shear stress could lead to dysplastic change of the media in the RCA.

Clinical presentations of patients with coronary artery-to-ventricular fistula depend on the size of fistula and the pressure of its terminal chamber. Most coronary artery fistulae are small and consequently myocardial blood flow is not compromised, and these patients are usually asymptomatic. This differs from coronary artery to right ventricle fistulae, which are often diagnosed by findings of continuous heart murmur on auscultation, to-and-fro murmur can only be heard for most coronary artery-to-LV fistulae [8],

Fig. 2. Intraoperative view of dilated RCA. The aneurysmal RCA was removed and a new one was reconstructed with an autograft of greater saphenous vein (b); postoperative three-dimensional CT image showed a nice position of the bypass graft, with excellent blood flow (c); pathological examination revealed the fibrocellular dysplastic change and mucoid degeneration in the wall of the RCA aneurysm (d). RCA, right coronary artery; CT, computed tomography; Ao, ascending aorta.
as in the present patient. Occasionally, the blood steal phenomenon of the coronary artery may occur and cause myocardial ischemia in patients with advanced coronary artery fistula [9], even when the shunt volume is a bit lower. But, in this case, the only symptom of this patient was paroxysmal palpitation.

There are several possible treatments of coronary artery-to-ventricular fistula, including surgical correction and transcatheter coil embolization [10]. With regard to this special case, due to the existence of huge RCA aneurysm, surgical correction was more optimal, with satisfactory result.

References