A Case of Multiple Coronary Artery-Left Ventricular Micro Fistulae Complicated With Hepatic Arteriovenous Fistulae

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SUMMARY

We present the first case of multiple coronary artery-left ventricular micro fistulae complicated with hepatic arteriovenous fistulae (AVF) in an adult patient. Multiple coronary artery fistulae originated from the left anterior descending coronary artery with aneurysmal change. Multiple coronary artery-left ventricular micro fistulae presented on the left ventricular wall and showed significant localized hypertrophic change. Stress and enhanced cardiac magnetic resonance imaging (CMR) revealed myocardial ischemia that could not be detected by stress Tl-201 cardiac scintigraphy, and late patchy gadolinium enhancement (LGE) in the mid-ventricular wall apex. This LGE pattern did not match the typical pattern observed in patients with apical hypertrophic cardiomyopathy. These observations may help distinguish multiple coronary artery-associated myocardial ischemia and hypertrophy from apical cardiomyopathy. (Int Heart J 2016; 57: 123-126)

Key words: Cardiac magnetic resonance imaging (CMR), Late gadolinium enhancement (LGE), Coronary fistula, Cardiac hypertrophy

Coronary artery fistula is a rare anomaly defined as abnormal connection between coronary arteries to cardiac chambers.¹⁾ This abnormal connection provokes myocardial ischemia by coronary steal phenomenon. A case of multiple coronary artery-left ventricular fistulae with apical hypertrophic cardiomyopathy have been reported.²⁾ This case affords the collateral evidence of association between myocardial ischemia by coronary fistulae and cardiac hypertrophy. On the other hand, hypertrophic cardiomyopathy itself also showed subendocardial ischemia.³⁾ It is difficult to distinguish coronary fistulae-associated cardiac hypertrophy from hypertrophic cardiomyopathy. Here we reported a case of multiple coronary fistulae associated with cardiac hypertrophy. In this report, we evaluated magnetic resonance images of myocardium and showed novel staining pattern of late gadolinium enhancement which is not observed in hypertrophic cardiomyopathy.

CASE REPORT

A 69-year-old woman with hepatic aneurysms and hepatic arteriovenous fistulae (AVFs) was admitted to our hospital for evaluation of perioperative cardiac risks prior to a hepatectomy and resection of hepatic artery aneurysms (Figures 1A, B). These operations were recommended to reduce the risk of aneurysmal rupture because one of the two hepatic aneurysms

had rapidly expanded during the previous year (from 34 mm to 37 mm) (Figure 1A). The patient had noticed shortness of breath 8 years prior, and her symptoms had not changed. The cardiovascular risk factors were hypertension and hypercholesterolemia. An ejection systolic murmur was detected during physical examination. Chest X-rays showed mild cardiomegaly with a cardiothoracic ratio of 56%. Electrocardiography at rest showed a sinus rhythm (59 bpm/minute) with left ventricular high voltage and deep T wave inversions (>0.2 mV) in leads I/aVL and ST segment depression in leads V4-6. Laboratory blood tests showed hyperlipidemia and mild elevated brain natriuretic peptide serum levels (135 mg/dL). Transthoracic echocardiography revealed a left coronary artery aneurysm and myocardial hypertrophy from the anterior wall to the apical wall without an intra-left ventricular pressure gradient. The maximal left ventricular wall thickness was 16 mm at the apical portion (Figure 2). The left ventricular cavity size and systolic function were normal (left ventricular ejection fraction: 62%). A computed tomography scan of the coronary artery was performed to further evaluate the coronary aneurysm. It showed a markedly dilated (diameter 20 × 18 mm) and significantly tortuous left anterior descending (LAD) coronary artery (Figures 3A, B). Multiple coronary artery-left ventricular micro fistulae arising from the LAD and left circumflex (LCX) coronary arteries were detected. The LAD was the dominant origin of multiple coronary micro fistulae. In addition, a diffuse network of multiple coronary micro fistulae from the

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Figure 1. Hepatic angiography of the common hepatic artery. **A:** Two aneurysms (37 mm and 29 mm) were observed in segment 4 of the hepatic artery; the arrow shows a 37 mm aneurysm. **B:** Micro arteriovenous fistulae originating from aneurysms were detected (circle).

Figure 2. Transthoracic echocardiography showed localized myocardial hypertrophy in the apical and anterior walls of the left ventricle without intra left ventricular pressure gradient. The maximum wall thickness of the apex was 16 mm.

Figure 3. Coronary artery – cardiac chamber multiple micro fistulae by angiography. **A:** Computed tomography of the coronary artery. Coronary artery fistulae extending from the left anterior descending artery (LAD) and left circumflex (LCX) artery to all cardiac chambers (left ventricle (LV), right ventricle, left atrium, and right atrium) and a giant LAD aneurysm were detected. **B:** 3-dimensional reconstruction of the coronary artery using a surface rendering method. LAD-LV multiple micro fistulae were dominant. **C:** Coronary angiography of the coronary arteries. A giant aneurysm of the left anterior descending artery and multiple fistulae were detected.

LAD and LCX coronary arteries that drained to all cardiac chambers (left ventricle (LV), left atrium, right ventricle, and right atrium) were detected by curved multi-planar reconstruction. Selective coronary angiography showed no significant obstructive coronary disease (Figure 3C). While there was significant hypokinesia of the anterior wall, left ventriculography showed normal global systolic function (LV ejection fraction: 58%). Left ventricular end-diastolic pressure was slightly elevated (18 mmHg), and cardiac output was 3.81 L/minute (cardiac index: 2.44 L/minute/m²). The pulmonary to systemic flow ratio (Qp/Qs) was 1.01, which suggested that there was no significant right-to-left shunt. Myocardial ischemia and LV diastolic volume overload are commonly reported hemodynamic changes provoked by multiple coronary fistulae, hence thallium (TI)-201 exercise scintigraphy was performed. However, there was no myocardial ischemia in the left ventricular

myocardial wall (Figure 4). Adenosine stress perfusion cardiac magnetic resonance imaging (CMR) can assess cardiac structure, function, and inducible ischemia. Compared to nuclear scintigraphy, CMR can provide higher spatial resolution and better detect delineation of both subendocardial and transmural perfusion defects. CMR can show perfusion defects on the LV anterior and septal walls during a stress perfusion test (Figure 5A) and late gadolinium enhancement (LGE) at the anterior, septal, and apex walls of the LV (Figure 5B). LGE was detected in the mid-myocardial wall of the hypertrophic lesion, where micro fistulae existed. This LGE pattern can distinguish cardiomyogenic changes from coronary artery disease.^{1,2)} Common LGE patterns in regions of ischemia are subendocardial or transmural enhancement, while patchy, multifocal, and mid-myocardial fibrosis patterns are typical within hypertrophic lesions.¹⁾ In this case, the perfusion defect indicated myocar-

Figure 4. Thallium (TI)-201 exercise stressed cardiac scintigraphy showed no stress induced myocardial ischemia in the territory of LAD coronary artery.

Figure 5. Stress and contrast-enhanced myocardial perfusion magnetic resonance image. **A:** The perfusion defect of the hypertrophied lesion on the left ventricular wall was detected using stress (adenosine) perfusion magnetic resonance imaging. The arrows show perfusion defects on the anterior and septal walls. **B:** A patchy and multifocal mid-myocardial late gadolinium-enhancement (LGE) was detected on a hypertrophied lesion on the left ventricular wall. The arrow shows the LGE lesion on the left ventricle.

dial ischemia, but the LGE pattern was not typical of ischemia. In some cases, coronary artery-LV fistulae can be candidates for surgery. However, the diffuse and complex structures of coronary-LV fistulae make resection very risky. Consequently, surgical ligation and catheter embolization were avoided. The left-lobe hepatectomy and aneurysm excision were successful. The patient was discharged on the 11th postoperative day without complications.

DISCUSSION

Coronary artery fistula is an abnormal direct connection between a coronary artery and either a cardiac chamber (coronary-cameral fistula) or a cardiac vein (coronary arteriovenous fistula). Coronary artery fistulae draining into the cardiac chamber have been reported in 0.2% of patients undergoing

cardiac catheterization, and the incidence in the overall population is estimated to be approximately 0.002%.³⁾ Sixty percent of these fistulae arise from the right coronary artery, and 90% terminate in the right side of the heart.³⁾ Multiple coronary micro fistulae that drain into the LV, like this case, are rare.⁴⁾ This abnormality is considered a hypothetical genetic failure that causes a partial persistence of the Thebesian vein network, which supplies the subendocardial myocardium during embryogenesis.^{5,6)} Multiple coronary micro fistulae can cause coronary steal phenomenon and provoke relative myocardial ischemia.³⁾ Cardiac stress scintigraphy or stress perfusion CMR has been reported to detect coronary steal phenomenon and myocardial ischemia.^{7,8)} Although TI-201 exercise scintigraphy showed no ischemia in the myocardium in this case,⁹⁾ a significant perfusion defect could be detected by CMR. As previously reported, first pass stress perfusion CMR^{10,11)} provides greater sensitivity and specificity to detect tiny myocar-

dial ischemia compared to stress TI-201 scintigraphy. Several studies have reported that multiple coronary artery-left ventricular fistulae were coincidentally found in cases of apical hypertrophy (APH).^{5,6,12} In these reports, the authors speculated that the progression of hypertrophic cardiomyopathy (HCM) might be associated with myocardial ischemia by coronary fistulae. In general, myocardial ischemia in HCM results in increased oxygen demand by the hypertrophied myocardium, impaired coronary circulation, and increased diastolic filling pressures.¹³⁻¹⁵ An association between APH, myocardial ischemia, and LGE has also been reported.¹⁶⁻¹⁸ In these reports, CMR detected LGE in hypertrophic lesions within the apex in 48-75% of patients. However, in these cases, the LGE showed a subendocardial pattern quite unlike the patchy or focal mid-wall LGE pattern frequently observed in asymmetrical septal HCM patients.¹⁷ Only one case reported both perfusion defect and delayed enhancement being performed to evaluate patients with APH. LGE also revealed a subendocardial pattern usually known as coronary artery stenosis or APH associated ischemic type.¹⁸ There are no reports of multiple coronary micro fistulae associated with APH being evaluated by CMR; our report is the first report to show CMR findings. In our case, perfusion defects in a hypertrophic lesion, which could not be detected by TI-scintigraphy, and LGE in the same lesion were detected. Moreover, this LGE was detected in the mid-myocardium among this hypertrophic lesion. This LGE pattern did not resemble the common results observed in APH patients. A large number of micro fistulae were distributed throughout the entire thickness of the myocardial wall (Figure 3); therefore, we speculate that the LGE did not form a typical ischemic type, like subendocardial enhancement in APH, but rather a mid portion of the ventricular wall type. Given these observations, CMR might be useful to address whether apical myocardial hypertrophy is derived from coronary artery fistulae or idiopathic apical hypertrophy in patients with cardiac hypertrophy and coronary artery fistulae.

Furthermore, it is worth mentioning that this patient had hepatic AVF, which are extremely rare.¹⁹ Etiologies for hepatic AVF are generally comprised of precipitating trauma, malignancy, and Osler-Weber-Rendu syndrome. Idiopathic AVF, like this case, are usually due to congenital vascular abnormalities and thus present in the pediatric period. In general, less than 10% of all AVF are idiopathic and congenital.²⁰ In addition, Osler-Weber-Rendu syndrome is known to cause multiple AVF and aneurysms, but these aneurysms are typically found in the lungs, brain, and liver. This patient did not meet the international consensus clinical criteria for Osler-Weber-Rendu syndrome and thus was not diagnosed with the syndrome.

Conclusion: This is the first reported adult case of multiple coronary micro fistulae that show apical hypertrophy complicated with hepatic AVFs.

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