Case Report

A Resuscitated Adult With Left Main Coronary Artery Ostial Atresia and Graves' Disease: 10-Year Follow-up

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ABSTRACT

This is the first report of a resuscitated adult with left main coronary artery ostial atresia (LMCAOA), with long-term follow-up for 10 years. A 57-year-old woman with untreated Graves' disease presented with resuscitated cardiac arrest, and her computed tomography coronary angiography showed a string-like left main without significant atherosclerosis, which led to the diagnosis of LMCAOA. Noninvasive and invasive testing revealed extensive myocardial ischemia because of LMCAOA with concomitant coronary spasm. After successful revascularization with coronary artery bypass grafting, the patient has remained stable for 10 years, which highlights this treatment as being highly effective and durable in patients with LMCAOA and cardiac arrest.

RÉSUMÉ

Il s'agit du premier rapport d'un adulte réanimé présentant une atrésie ostiale de l'artère coronaire principale gauche (atrésie ostiale de l'ACPG), comportant un suivi à long terme sur une période de 10 ans. Une femme de 57 ans atteinte de la maladie de Basedow non traitée a subi un arrêt cardiaque en réanimation. La coronarographie par tomodensitométrie a montré une artère principale gauche en forme de cordon sans athérosclérose importante, ce qui a conduit au diagnostic d'atrésie ostiale de l'ACPG. Des tests non invasifs et invasifs ont révélé une ischémie myocardique étendue due à l'atrésie ostiale de l'ACPG avec spasme coronaire concomitant. Après une revascularisation réussie par pontage aorto-coronarien, la patiente est restée stable pendant 10 ans, ce qui montre que ce traitement est très efficace et durable chez les patients atteints d'atrésie ostiale de l'ACPG et d'arrêt cardiaque.

Left main coronary artery ostial atresia (LMCAOA) is an exceedingly rare coronary artery malformation, with less than 100 cases reported to date.¹ Only a few cases of LMCAOA in resuscitated adults have been reported previously.² There have been no previous reports of long-term follow-up cases in resuscitated patients.

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Ethics Statement: The research reported has adhered to the relevant ethical guidelines.

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See page 1194 for disclosure information.

Case

A 57-year-old woman with no significant past medical history collapsed while tidepooling. After immediate bystander cardiopulmonary resuscitation, endotracheal intubation and electrical defibrillation for initial rhythm of ventricular fibrillation were performed by the emergency medical service team. The initial electrocardiogram showed sinus tachycardia with downsloping ST depression and T-wave inversions in lateral leads with a corrected QT interval of 416 ms, and the echocardiography showed mild hypokinesia of the left ventricular lateral wall. A repeat echocardiogram several hours later revealed normal wall motion with a hyperkinetic left ventricle. Her serum creatinine kinase level peaked at 421 IU/L, and she had an uneventful recovery. During the admission, she was also diagnosed with Graves' disease with high levels of

Novel Teaching Points

- LMCAOA is an exceedingly rare form of coronary artery malformation, and there are only a few previously reported cases of resuscitated adults with LMCAOA.
- Our patient is unique, as the mechanism of ventricular fibrillation was likely myocardial ischemia associated with LMCAOA and coronary spasm. In addition, our case is the first with long-term follow-up for over 10 years.
- We have confirmed the patency of the left internal mammary artery graft and resolution of ischemia by 10-year follow-up CT coronary angiography and adenosine stress test, which highlights CABG as a highly effective and durable treatment option in patients with LMCAOA and cardiac arrest.

free triiodothyronine and free thyroxine, undetectable thyroid-stimulating hormone, and positive thyroid-stimulating hormone receptor antibodies, and treatment was initiated with methimazole and metoprolol. Coronary angiography, 2 weeks after her initial presentation showed absent left main coronary artery with the distal left anterior descending artery supplied via the large collateral vessel from the posterodescending artery (Fig. 1A). The retrograde flow to the proximal left anterior descending artery further supplied the left circumflex artery. Computed tomography (CT) coronary angiography showed a string-like structure at the site of the left main coronary artery (Fig. 1B). She did not have a history of smoking, Raynaud's phenomenon, or migraine headaches. However, as there were minimal atherosclerotic findings by both invasive and CT angiography, coronary spasm was suspected as a cause of the cardiac arrest, and she was started on diltiazem. As adenosine stress thallium-201 myocardial perfusion

scintigraphy confirmed large ischemia in the anterior and lateral walls (Fig. 1C), revascularization was performed with off-pump coronary artery bypass grafting (CABG) of the left internal mammary artery to the left anterior descending artery. Postoperative angiography confirmed the graft patency, and a subsequent acetylcholine stress test induced diffuse severe spasm of the right coronary artery (Fig. 1, D and E). Medical therapy with antithyroid medication and diltiazem was continued, and she has remained asymptomatic for 10 years after the CABG. A follow-up CT coronary angiography confirmed graft patency, and adenosine stress thallium-201 myocardial perfusion scintigraphy showed no ischemia (Fig. 2).

Discussion

This is the first report with long-term follow-up for over 10 years in a resuscitated adult with LMCAOA. This patient had untreated Graves' disease, a known cause of coronary artery spasm.³ Her acetylcholine test confirmed the diagnosis with extensive spasm in the right coronary artery, the sole donor vessel for the left coronary artery. Therefore, the mechanism of ventricular fibrillation was likely due to extensive myocardial ischemia associated with LMCAOA and coronary spasm.

Although anatomic repair has been reported as an effective method for revascularization of LMCAOA, we selected CABG. The reported advantage of anatomic repair over CABG is access for future percutaneous coronary intervention if necessary.⁴ However, given the minimal atherosclerosis findings, and with lack of significant cardiovascular risk factors, the possibility of the need for a future coronary artery operation was expected to be low. As there was concern that the graft might not mature, owing to extensive and brisk competitive flow from the right coronary artery, the adenosine stress test was performed prior to CABG to confirm ischemia

Figure 1. (A) Angiography with extensive collaterals from right coronary artery to the left anterior descending artery, which further supplies the left circumflex artery. (B) Multiplanar reconstruction view showing a string-like structure at the site of the left main coronary artery. (C) Thallium-201 myocardial imaging showing ischemia in the anterior and lateral walls. (D) Coronary angiography after coronary artery bypass grafting showed patent left internal mammary artery. (E) Provocation test using 50 μ g of acetylcholine showed diffuse coronary spasm throughout the right coronary artery, accompanied with chest pain (left), which resolved after nitroglycerin injection (right).

Figure 2. (A, B) Computed tomography angiography at 10 years after coronary artery bypass grafting showed patent left internal mammary artery to left anterior descending artery. (C) Follow-up thallium-201 myocardial imaging without ischemia.

in the left coronary artery territory. At 10 years, the patency of the left internal mammary artery graft and resolution of ischemia was confirmed by follow-up CT coronary angiography,

and an adenosine stress test, which highlights CABG as a highly effective and durable treatment option in patients with LMCAOA and cardiac arrest.

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Disclosures

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