Chronic Ischemia Induced by Woven Coronary Artery Anomaly with Typical Atrial Flutter: Insights from Multiple Imaging Devices

Akio Chikata¹, Satoru Sakagami¹, Naomi Kanamori¹, Chieko Kato¹, Wataru Omi¹, Takahiro Saeki¹, Hideo Nagai¹, Soichiro Usui², Kenichi Nakajima³ and Masayuki Takamura²

Abstract

A 75-year-old man with a 120-bpm tachycardia and typical atrial flutter was admitted. Echocardiography showed a dilated left ventricle with anterior and apical wall akinesia. Tachycardia was terminated with cavo-tricuspid isthmus ablation. Multiple imaging findings revealed a woven coronary artery anomaly (WCAA) in the left anterior descending artery. Stress myocardial perfusion imaging was performed after ablation in the sinus rhythm and revealed stress-induced ischemia and a fixed low uptake in the WCAA territory. WCAA is generally regarded as a benign condition; however, compromised blood flow within the anomaly, caused by tachycardia-related diastolic shortening, may induce ischemia.

Key words: woven coronary artery anomaly, intravascular ultrasound, computed tomography angiography

(DOI: 10.2169/internalmedicine.54.4333)

Introduction

Woven coronary artery anomaly (WCAA) is an extremely rare congenital malformation in which the epicardial coronary artery is divided into multiple thin channels which twist along the coronary artery axis and then merge to form the main lumen (1, 2). Only a few cases of WCAA have been reported in the literature. It is generally regarded as a benign condition (3), but its hemodynamic effects and natural history remain unclear. In this report, we evaluated a patient with WCAA using several imaging techniques, including coronary computed tomography angiography (CTA), coronary magnetic resonance angiography (CMRA), stress myocardial perfusion imaging, invasive coronary angiography (CAG), and intravascular ultrasound (IVUS). The results suggested that sustained tachycardia due to typical atrial flutter may have decreased the coronary flow reserve (CFR) of the left anterior descending artery (LAD), which had a WCAA, and induced chronic ischemia.

Case Report

A 75-year-old Japanese man was admitted to our hospital with palpitations. He denied any angina or dyspnea and had no history of diabetes mellitus, Kawasaki disease, vasculitis, sarcoidosis, or Chagas disease. His prescribed medications were atorvastatin (10 mg daily) for dyslipidemia along with carvedilol (10 mg daily) and verapamil (80 mg twice daily) for heart rate control. On physical examination, he had a blood pressure of 90/56 mmHg, a heart rate 120 bpm, and an oxygen saturation of 98% without oxygen. A 12-lead electrocardiogram showed a regular tachycardia of 120 bpm with incomplete left-bundle branch block. An inverted sawtooth F-wave pattern was observed in the inferior leads (II, III, and aVF) and an upright F-wave pattern was observed in lead V1 (Fig. 1). Echocardiography showed the following: a slightly dilated left ventricle (LV) with an end-diastolic dimension of 56 mm, akinesia in the anterior and apical walls, severe hypokinesia in the other LV walls, and a decreased ejection fraction of 22.5% according to the modi-
Figure 1. A 12-lead electrocardiogram (ECG) showed regular tachycardia at 120 bpm with incomplete left-bundle branch block. An inverted sawtooth F wave pattern was observed in the inferior ECG leads (II, III, and aVF), with low-amplitude biphasic F waves in leads I and aVL, an upright F wave in precordial lead V1, and an inverted F wave in lead V6. This ECG indicated typical atrial flutter.

Faced Simpson’s method. The heart rate during echocardiography was 120 bpm.

The laboratory data revealed the following values: white blood cell count, 4,700 mm$^3$; C-reactive protein, 0.3 mg/dL; brain natriuretic peptide (BNP), 563.3 pg/mL; total cholesterol, 87 mg/dL; and angiotensin-converting enzyme, 26.0 U/L. The results were negative for antinuclear antibody, myeloperoxidase-antineutrophil cytoplasmic antibody, cytoplasmic antineutrophil cytoplasmic antibody, lupus anticoagulant, anticytodiulin antibody, and anti-beta2-glycoprotein I antibody.

An electrophysiological study revealed that the tachycardia was consistent with typical atrial flutter, and there was no accessory pathway. The tachycardia was terminated during cavotricuspid isthmus (CTI) ablation.

We performed CAG to investigate the cause of cardiac dysfunction. CAG revealed multiple irregular filling defects and intraluminal haziness with the appearance of a spiral dissection lesion, ranging from the proximal to the middle segment of LAD (Fig. 2A, Supplementary material 1). TIMI-III flow was evident distal to the anomalous LAD segment. The left main (LM), the right coronary artery (RCA), and the left circumflex coronary (LCX) artery were normal.

Grayscale IVUS was performed. Interestingly, the arterial lumen was divided into multiple thin channels in the proximal to middle segment of LAD, which then reassembled into a single lumen in the distal LAD segment after a slightly twisted course (Fig. 2B, Supplementary material 2). Although these multiple small channels rejoined the main lumen, they did not communicate with each other. We observed no thrombus or dissection flaps. Therefore, based on the IVUS and CTA findings, we diagnosed WCAA and did not perform percutaneous coronary intervention (PCI).

In 64-slice coronary CTA, curved multiplanar reconstruction of LAD also showed that the proximal LAD was divided into two major branches that bisected each other and fused distally in a “figure-eight” pattern. Cross-sectional images at the lesion site showed multiple, twisting small channels (Fig. 3). The length of the woven segment was 37 mm. The Agatston coronary artery calcium score was as follows: LM, 0; LAD, 0; LCX, 6.10; RCA, 24.41; total, 30.52. 3.0-T contrast-enhanced, whole-heart CMRA was less clear than CTA (Fig. 4). Cardiac stress myocardial perfusion testing was performed after CTI ablation in the sinus rhythm. Adenosine stress $^{99m}$Tc-tetrofosmin single-photon emission computed tomography (SPECT; single-day protocol) revealed stress-induced ischemia and a fixed low uptake in the anterior and apical walls (Fig. 5). The BNP level decreased to 108.2 pg/mL 1 month after CTI ablation, and the ejection fraction slightly improved to 32.2% of that estimated by the modified Simpson method.

**Discussion**

WCAA is characterized by the branching of a major epicardial artery into thin channels, which then reassemble to reform the main lumen after twisting along the coronary artery axis. Additionally, they typically have a TIMI-III blood flow distally (1, 2). Few cases of WCAA have been reported and data on this anomaly are limited (3). The differential diagnosis of WCAA includes spontaneous coronary artery dissection, thrombus formation, and chronic total occlusion (CTO) with recanalization and the development of vasa vasorum bridging collaterals (3). Although the diagno-
We presumed that if the FFR wire can be passed through causing coronary stenosis with an accuracy of more than (FFR) value of 0.80 or less is able to identify ischemia-associated with a fixed low uptake would be similar to the present patient’s condition. We performed CTI ablation without PCI. Based on these facts, we speculate that sustained tachycardia due to typical atrial flutter may have reduced the CFR of the LAD with WCAA, which resulted clearly from coronary dissection because of its limited resolution.

According to previous findings that patients with WCAA generally demonstrate normal coronary reserve during stress tests, it has been proposed that WCAA is a benign condition (3); however, the hemodynamic effect and natural history of this anomaly remain unclear. Soylu et al. reported a patient with WCAA who presented with myocardial infarction (7) and Oylumlu et al. reported a case with reversible ischemia visualized via dobutamine stress echocardiography (8). In our patient, adenosine stress myocardial perfusion imaging performed after ablation revealed stress-induced ischemia and a fixed low uptake in the WCAA territory. Although the degree of stress ischemia was milder than in typical cases with severe coronary artery stenosis, the ischemic location at the anterosetal territory near the apex was in alignment with a lesion in the peripheral part of LAD. Stress ischemia indicates decreased CFR, and a fixed low uptake indicates myocardial infarction. Therefore, mild ischemia associated with a fixed low uptake would be similar to the present patient’s condition. We performed CTI ablation without PCI. Based on these facts, we speculate that sustained tachycardia due to typical atrial flutter may have reduced the CFR of the LAD with WCAA, which resulted in chronic ischemia.

In the present case, we evaluated CFR by ATP-stress SPECT; other modalities may provide more accurate evaluation. It has been reported that a fractional flow reserve (FFR) value of 0.80 or less is able to identify ischemia-causing coronary stenosis with an accuracy of more than 90% and FFR-guided PCI improved patient outcomes (9). We presumed that if the FFR wire can be passed through
the complex structure of WCAA, a more accurate evaluation would be possible. CFR assessment by ATP-echocardiogram and FFRCT have been reported as novel, noninvasive methods for determining the physiological significance of coronary artery disease (10, 11). Future studies using these methods will provide more accurate structural and functional assessments in cases of WCAA.

Considering that the coronary blood flow is dominant in the diastolic phase and that tachycardia shortens the diastolic phase, we believe that tachycardia could compromise the coronary blood flow in some patients with WCAA. Although long-standing tachycardia is a well-recognized eti-
ological factor for diffuse LV dysfunction, our findings of anterior and apical wall asynergy, stress-induced ischemia, and a fixed low uptake in the LAD area suggest that additional chronic ischemia existed in the LAD area. We therefore speculate that thin channels in WCAA with a compromised blood flow into LAD, due to diastolic shortening by atrial flutter, likely induce ischemia.

The authors state that they have no Conflict of Interest (COI).

References