Acute Coronary Syndrome in a Puerperal Patient with Coronary Artery Ectasia due to a Coronary Artery Fistula

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Abstract

Coronary artery fistulas are rare and the feeding artery is ectatic and tortuous. It is not well-known whether coronary artery ectasia (CAE) is a risk factor of acute coronary syndrome (ACS) in the puerperal periods. A 40-year-old woman with a coronary artery fistula and an ectatic right coronary artery (RCA) had delivered twins. A month later, she had chest pain and coronary angiography revealed thrombogenesis in the RCA. She had no risk factors of cardiovascular disease or thrombogenesis. We should recognize that CAE is a risk factor for ACS in women in the perinatal and puerperal periods.

Key words: acute coronary syndrome, coronary artery ectasia, coronary artery fistula, puerperal period, young woman

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Introduction

Coronary artery fistulas are abnormal connections between a coronary artery and a cardiac chamber or great vessel. They occur in 0.002% of the general population and 0.13% of patients undergoing coronary angiography (1). The feeding artery of the fistula is usually ectatic and tortuous. It has been shown that coronary artery ectasia (CAE) can lead to acute coronary syndrome (ACS) by thrombogenesis, mainly due to a stagnant blood flow at the ectatic site (2). CAE has not been previously reported to be a risk factor of ACS in the perinatal and puerperal periods, and the use of antithrombotic agents for the prevention of ACS in patients with CAE has not yet been established.

We herein report the case of a young woman with a coronary artery fistula and CAE causing ACS due to thrombogenesis in the puerperal period.

Case Report

The patient was a 40-year-old woman with a right coronary artery (RCA) to coronary sinus vein (CSV) fistula and an ectatic tortuous RCA (Fig. 1) detected by computed tomography and coronary angiography before surgery for a hydatidiform mole 7 years previously. Coronary angiography revealed slow disappearance of contrast medium in the RCA. Hemodynamic and oximetric measurements revealed a small calculated left-to-right shunt (a pulmonary blood flow/systemic blood flow ratio, 1.1). She had no symptoms, and no evidence of ischemia was detected by exercise thallium-201 single-photon emission computed tomography. Therefore, she was followed up without medication.

She presented to the emergency department of our hospital complaining of sudden chest pain that continued for 2 hours. She had no common cardiovascular risk factors such as hypertension, diabetes mellitus, dyslipidemia, smoking history, or family history of cardiovascular disorders. She had delivered twins 1 month previously. Her vital signs and cardiorespiratory findings were normal. Electrocardiography (ECG) showed sinus rhythm with ST segment elevation in II, III and aVF leads and reciprocal ST depression in I and aVL leads (Fig. 2a and b). Blood test results showed slightly elevated cardiac troponin I and D-dimer levels at 0.17 ng/mL and 0.8 μg/mL, respectively.

Coronary catheterization was performed. At that time, she...
Figure 1. Cardiac images of the patient at 33 years of age. (a) A cardiac computed tomography image and (b) angiography (left anterior oblique view) of the right coronary artery (RCA) showing a RCA-coronary sinus vein (CSV) fistula and an ectatic and tortuous RCA. The junction of the RCA-CSV was narrow (yellow arrow). RV: right ventricle.

Figure 2. Electrocardiography (a) before the delivery, (b) in emergency room, and (c) before coronary angiography. (b) ST segment elevation in II, III and aVF leads and reciprocal ST depression in I and aVL leads is observed.

had no chest pain and an ECG revealed an improvement of ST segment changes (Fig. 2c). Coronary angiography revealed a normal left coronary artery, but a stagnant coronary flow and thrombogenesis in the RCA that was present only at the RCA-CSV fistula (Fig. 3). Therefore, she was diagnosed with ACS due to thrombogenesis. It was difficult to perform coronary intervention because the RCA was ectatic and tortuous. The culprit lesion was in a peripheral site. Her symptoms and ECG findings spontaneously improved. Therefore, we elected not to perform coronary intervention. She was administered unfractionated heparin and oral warfarin therapy. Her peak CK level was 803 U/L, and transthoracic echocardiography showed wall motion abnormality in the inferior area.

We examined the primary cause of thrombogenesis before the administration of anticoagulant medications. However, no abnormalities were noted [prothrombin time-international normalized ratio (PT-INR), 1.02; activated partial thromboplastin time, 35.5 ms; fibrinogen level, 286 mg/dL; antithrombin III, 107%; protein C, 108%; protein S, 58%; antiphospholipid antibody, negative; and anticardiolipin/beta2-glycoprotein I complex antibody, negative]. Her PT-INR was
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ged at 1.6-2.6 by warfarin therapy. Her hospital course was uneventful and she was discharged 14 days after hospitalization. She did not wish to undergo repeat coronary angiography or computed tomography angiography.

Discussion

A recent report showed that nonatherosclerotic coronary artery disease (NACAD) is not rare in young women undergoing coronary angiography and accounts for 13% (66/177 cases) of myocardial infarction (MI) cases. Spontaneous coronary artery dissection commonly causes NACAD, while CAE rarely causes NACAD, and the prevalence of CAE has been reported to be 4.5% (3/66 cases) in young women with troponin-positive ACS (3).

MI is reported to be rare during pregnancy and is estimated to occur in 1 in 10,000 deliveries (4). In a previous study, coronary thrombosis without atherosclerotic disease and coronary dissection were responsible for 8% and 27% of MI cases associated with pregnancy, respectively. Pregnancy-related spontaneous coronary dissection may be related to structural changes in the vessel wall caused by an excess of progesterone and a physiologic increase in the blood volume and cardiac output that magnify shear forces in the vessels (5). It has been reported that persistent hypercoagulation occurs during the first 3 weeks after delivery and the level of coagulation returns to the normal pre-pregnancy level after 6 weeks of the puerperal period (6, 7). Moreover, it has been reported that a stagnant coronary flow may be related to coronary endothelial dysfunction (8).

The present patient had no common cardiovascular risk factors and no primary cause of thrombogenesis. The patient had an ectatic RCA and narrow RCA-CSV fistula. These features might have originally caused the stagnant coronary flow and endothelial dysfunction resulting from shear forces in the vessels. Coronary angiography showed a spherical thrombus and occlusion without a radiolucent intimal flap and contrast staining in the RCA. It was difficult to perform intravascular imaging as intravascular ultrasound and optical coherence tomography, and the patient did not wish to undergo repeat coronary angiography or computed tomography angiography. Although an association between coronary dissection and extensive thrombosis was not excluded, hypercoagulation occurring during pregnancy may have been associated with thrombogenesis in the RCA, in addition to the originally stagnant coronary flow and endothelial dysfunction resulting from CAE due to a coronary artery fistula.

Thrombogenesis with CAE in the puerperal period is a situation similar to venous thromboembolic disease (VTE) in the puerperal period, which is caused by venous stasis, endothelial injury, and a hypercoagulable state (9). The 2012 American College of Chest Physicians guidelines suggest pharmacologic thromboprophylaxis can be administered to a select population of puerperal period women considered to be at a high risk for VTE (10). Although thromboprophylaxis induced by anticoagulants in patients with CAE is not known to be effective, the present patient was treated and thrombotic recurrence was successfully prevented by anticoagulant therapy.

In conclusion, the present findings indicated that young women with CAE can develop ACS due to thrombogenesis in the puerperal period. Therefore, we should recognize that CAE is a risk factor for ACS in women in the perinatal and puerperal periods.

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

References


