Silent Progression of Coronary Artery Thrombosis in a Pregnant Woman With Anomalous Origin of Left Coronary Artery From the Pulmonary Artery

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Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA; or Bland-White-Garland syndrome) is the most common congenital coronary artery anomaly, occurring in 0.5% of children with congenital heart disease. The myocardial perfusion of the left ventricle (LV) is dependent on the development of collateral vessels from the right coronary artery (RCA) in patients with ALCAPA.1,2 Coronary arteries are usually significantly dilated because of the increased flow from collateral vessels,3 but there is no standard antplatelet or anticoagulation therapy to prevent the formation of thrombosis in the giant coronary arteries in this disease.

A 34-year-old woman visited the obstetric clinic at 7 weeks' gestation. At age 27, on diagnosis of ALCAPA, she had undergone reimplantation of the anomalous left main coronary artery to the aorta without complication. She had no coronary risk factors such as hypertension, diabetes mellitus, or dyslipidemia. She had warfarin after the operation to prevent thrombosis of the dilated coronary arteries. Her first pregnancy was managed with aspirin alone. Delivery by caesarean section was chosen considering her cardiac condition, and was uneventful. She restarted warfarin after the delivery. One year later, she converted it to aspirin following the advice of her attending physician. Coronary angiography at age 31 showed complete occlusion of the left circumflex coronary artery, and warfarin was restarted. Computed tomography (CT) angiogram prior to the second pregnancy showed dilated left anterior descending coronary artery (LAD) with thrombus formation and dilated right coronary artery (RCA) (Figure 1). Ultrasonography showed preserved LV function (ejection fraction, 66%). We converted warfarin to aspirin and monitored LV function and coronary artery carefully on ultrasonography and cardiac magnetic resonance imaging (CMR). CMR at 33 weeks' gestation showed good LV function and no exacerbation of the thrombosis. The patient underwent caesarean section under aspirin alone at 37 weeks' gestation and had a healthy male infant. D-dimer was within the normal range (<1.0 µg/mL).
before delivery, but was slightly elevated (2.4 µg/mL) 4 days after delivery. The patient continued to take aspirin and was discharged uneventfully 5 days after delivery. There were no symptoms, and rest electrocardiogram did not show any change. CT angiogram 3 months after delivery, however, showed the progression of thrombosis and narrowing of the LAD with pale contrast in the peripheral lesions (Figure 2). Exercise stress ²⁰¹thallium myocardial scintigraphy showed exercise-induced hypoperfusion in the anterior and septal walls, and persistent hypoperfusion in the anterior wall and apex (Figures S1, S2). Aspirin was converted to warfarin again, and improvement of the thrombosis was observed (coronary artery diameter increased from 7.1 to 17.0 mm) on CMR 3 months later.

ALCAPA and Kawasaki disease (KD) are the major causes of giant coronary artery. In general, pregnancy promotes thrombosis formation and cardiovascular stress increases during pregnancy. Ten patients with KD and coronary artery aneurysm were reported to have had successful pregnancy and delivery with aspirin alone.⁴ In a retrospective review of the literature for pregnancy-related acute myocardial infarction (AMI) from Japan, a patient with KD developed AMI 10 days after delivery, and then underwent coronary artery bypass graft surgery.⁵ In contrast, there have been only a few reports of ALCAPA in pregnancy.⁶⁷ and no adequate antiplatelet or anticoagulation therapy has been established for this disease. In the present case, the pregnancy and delivery were uncomplicated, and the patient had no symptoms before or after the pregnancy with aspirin. Exacerbation of coronary thrombosis, however, occurred without any change on rest electrocardiogram or any symptom. Thrombosis improved 3 months after restarting warfarin. Aggressive anti-thrombotic or anticoagulation therapy along with careful imaging and laboratory monitoring may be beneficial to maintain coronary perfusion in ALCAPA patients, especially during pregnancy.

Disclosures
The authors declare no conflict of interest.

Grants
None.

References

Supplementary Files

Supplementary File 1
Figure S1. Exercise stress ²⁰¹thallium myocardial scintigraphy showing exercise-induced hypoperfusion in the anterior and septal walls and persistent hypoperfusion in the anterior wall and apex.

Figure S2. Three-dimensional image fusion of coronary computed tomography angiography and myocardial perfusion scintigraphy, indicating that thrombotic narrowing of the left anterior descending coronary artery is relevant to the exercise-induced hypoperfusion (arrow) and persistent hypoperfusion (dotted arrow).

Please find supplementary file(s);

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