Development of Myocardial Ischemia after Pulmonary Embolism in Anomalous Aortic Origin of Right Coronary Artery

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Abstract

Anomalous aortic origin of the right coronary artery from the left coronary sinus (AAORCA) is a relatively rare congenital heart anomaly and often clinically silent, but one of the major causes of sudden death in young people. AAORCA patients are detected incidentally during imaging procedure for unrelated diseases. However, it remains unclear when myocardial ischemia appears in AAORCA patients who previously have no findings of myocardial ischemia.

We herein report a 34-year-old AAORCA patient who newly developed myocardial ischemia after acute pulmonary embolism (PE). The patient had presented chest pains and syncope attacks on exertion four months prior to the visit, although the thrombus had already disappeared. Although thrombus in the pulmonary artery had disappeared after anticoagulation therapy, chest pains and syncope attacks on exertion remained. Transthoracic echocardiogram showed mild pulmonary hypertension (estimated systolic pulmonary artery pressure: 47 mmHg).

We performed exercise-stress electrocardiogram (ECG) test, and the excise at 7 Mets provoked chest pain and syncope associated with ST change and sinus arrest in ECG. Coronary computed tomography angiogram revealed that the right coronary artery (RCA) was originated from the left coronary sinus, and the proximal portion of RCA was severely compressed between aorta and pulmonary artery.

Surgical re-implantation of the RCA from the left coronary sinus into the right coronary sinus was performed. Since the surgery, there have been no ischemic findings in the patient.

Introduction

Anomalous aortic origin of the right coronary artery from the left coronary sinus (AAORCA) is a relatively rare congenital heart anomaly and often clinically silent, but one of the major causes of sudden death in young people [1]. AAORCA patients are detected incidentally during imaging procedure for unrelated diseases. However, it remains unclear when myocardial ischemia appears in AAORCA patients who previously have no findings of myocardial ischemia [2, 3].

We herein report an AAORCA patient who newly developed myocardial ischemia after acute pulmonary embolism (PE).
Figure 1:
A, B: Computed tomography (CT) showed large thrombus in the right and left pulmonary arteries (red arrows).
C, D: Follow-up CT revealed the absence of a thrombus.
E, F: Coronary CT angiography showed the right coronary artery (RCA) arising from the left sinus. The RCA was compressed between the aorta and pulmonary artery (black arrow). Ao; aorta, PA; pulmonary artery, LA; left atrium, RA; right atrium.
**Figure 2**: Exercise stress electrocardiogram. At 7 Mets, chest pain with ST segment change and syncope with sinus pause were induced.

**Figure 3**: Volume rendering of computed tomography. A, B: Pre-surgery and C, D: post-reimplantation surgery.
Discussion

There are two types of anomalous aortic origin of coronary artery: anomalous aortic origin of the left coronary artery from the right sinus (AAOLCA) and AAORCA. AAORCA has lower risk of sudden death but has higher prevalence than AAOLCA [4]. Although the precise prevalence of AAORCA in the entire population remains unclear because detecting asymptomatic AAORCA is difficult, Kaku, et al reported that the prevalence of AAORCA was 0.25% in 17731 Japanese patients undergoing coronary angiography [5]. The possible pathophysiological mechanisms for the restriction of coronary blood flow in AAORCA were mechanical compression between aorta and pulmonary artery, slit-like narrowing orifice, and acute angle takeoff, all which were shown with CT angiogram in our case.

Sudden death in AAORCA patients usually occurs during or just after exercise, and one of the possible mechanisms is increased pressure of aorta and pulmonary artery associated with exercise. In our case, we considered that increasing pulmonary artery pressure caused by acute PE changed mutual positional relationship of RCA, aorta and pulmonary artery, and worsened the compression of RCA [6].

Surgical repair (re-implantation, unroofing or pulmonary artery translocation) is indicated for AAORCA patients with evidence of ischemia to prevent sudden death [3]. In this case, we decided surgery because the symptoms were fatal and the patient was a young athlete, although myocardial ischemia may have disappeared spontaneously with decreasing of pulmonary artery pressure. We chose re-implantation surgery because the orifices of RCA and LCA were separated and there was no intramural course. Disappearance of the symptoms after the surgery revealed these symptoms were associated with myocardial ischemia. Though there is not enough evidence of clinical outcome of AAORCA surgery, Law, et al. reported re-implantation surgery for AAORCA is safe and has excellent medium to long-term outcome [7].

Conclusion

We should care appearance of myocardial ischemia in AAORCA patients especially when they develop PE. In addition, it is inferred that other diseases increasing pulmonary artery pressure may also induce myocardial ischemia in AAORCA patients.

References