Anomalous Right Coronary Artery Arising from the Left Sinus of Valsalva in a Young Athlete

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Abstract

Cardiac events associated with congenital coronary abnormalities are rare but potentially life-threatening in a young population. Most of these patients are not diagnosed before their initial cardiac event. Amongst such coronary artery anomalies, sudden death is frequently seen in an anomalous origination of a coronary artery from the opposite sinus. We herein present the case of a patient who presented with sudden cardiac arrest associated with an anomalous right coronary artery originating from the left sinus of Valsalva. Surgical treatment was selected because there was evidence of reversible ischemia based on the findings of a stress test.

Key words: anomalous coronary artery, sudden death, surgical therapy

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Introduction

An anomalous origination of a coronary artery is the second most common cause of cardiac death after hypertrophic cardiomyopathy in athletes (1). In previous studies, the incidence of coronary anomalies discovered in patients undergoing coronary angiography is approximately 1% (2). Angelini reported that the rate of ectopic right coronary artery (RCA) originating from the left sinus is 0.92% (3). Although sudden death occurs less frequently in patients with anomalous coronary arteries, an anomalous origination of the coronary artery from the opposite sinus (ACAOS) is a relatively frequent cause of cardiac events in the younger population. 55% of all patients who die suddenly of a coronary anomaly disease have no forewarning manifestations (4). Exercise-related sudden death in patients with coronary anomalies was seen most frequently when the left coronary artery arose from the right coronary artery, followed by an anomalous origin of the RCA from the left sinus of Valsalva (5).

Case Report

A 38-year-old man with a history of smoking but without any other significant medical history or previous history of chest pain presented with a loss of consciousness immediately after running a marathon. Ventricular fibrillation was detected with the automated external defibrillator (AED) and after two electrical defibrillation prompts by the surrounding medical staff, the patient's sinus rhythm was restored. He was subsequently transported to the emergency department, and he thereafter maintained consciousness without requiring further cardiopulmonary resuscitation. His electrocardiogram (ECG) on admission demonstrated a normal sinus rhythm without ST and T wave changes. In addition, his echocardiogram demonstrated a normal left ventricular function (the ejection fraction was 68%) with no organic abnormality, such as myocardial hypertrophy or valvular heart disease. A coronary angiogram was performed to diagnose the presence of cardiovascular disease and he had no significant coronary artery stenosis. A spasm provocation test with acetylcholine was performed and the findings were negative. However, the angiogram did show that the RCA originated

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Figure 1. Coronary angiogram showed no significant stenosis in both right coronary artery (A) and left coronary arteries (B).

Figure 2. Preoperative coronary CT angiography (A, B) demonstrated that the RCA was originating from the left sinus of Valsalva and that the proximal RCA passed between aorta and pulmonary artery trunk (arrowheads). Postoperative coronary CT angiography (C, D) showed the reimplanted RCA to right sinus of Valsalva with good patency (arrows). (Ao: Aorta, PA: pulmonary artery)

from the left sinus of Valsalva (Fig. 1). Coronary computed tomography (CT) angiography revealed an anatomical abnormality where the proximal RCA passed between the ascending aorta and the pulmonary artery trunk (Fig. 2A, B). To identify the relationship between the coronary anomaly and ischemia, exercise stress tests were performed. Exercise treadmill test demonstrated a significant ST segment depression in II, III, aVF (Fig. 3). This was further confirmed with

exercise stress myocardial perfusion imaging which demonstrated an inferior reversible ischemia (Fig. 4). In order to prevent a recurrence of the previous cardiac event and along with the patient's desire to continue his active lifestyle, the patient underwent surgical treatment. The ostium of the RCA was ligated and detached from the left sinus. Then, the ostium of the RCA was successfully reattached to the proximal RCA using an end-to-side aortocoronary anastomosis to

Figure 3. Significant ST segment depression in II, III, aVF (arrows) was seen during the treadmill stress test.

Figure 4. Myocardial perfusion imaging with exercise stress test demonstrated a reversible inferior ischemia.

the right sinus of Valsalva. Exercise stress myocardial perfusion imaging was performed one month postoperatively and the findings were negative for myocardial ischemia. One year postoperatively, follow-up coronary CT angiography demonstrated a patent RCA (Fig. 2C, D) and the patient remained asymptomatic during physical activity.

Discussion

Sudden death due to anomalous coronary arteries is believed to occur in association with several different mecha-

nisms. The primary ischemic mechanism with ACAOS is caused by an increase in the intraaortic pressure upon heavy exertion which compresses and collapses the proximal RCA running between the aorta and pulmonary artery trunk (5). Other possible mechanisms for ischemia are the acute take-off of the RCA orifice from the left sinus of Valsalva; stretching or severe angulation of the RCA; and coronary artery spasms. The treatment options for ACAOS include 1) a restriction of any high intensity physical exercise with medical therapy, 2) the deployment of a stent into the compressed vessel, or 3) surgical therapy. Although medical

therapy may be effective and the least invasive, high intensity physical activity would have to be avoided due to a potential risk of sudden cardiac death. Several cases have been reported on stent implantation therapy for compressed proximal RCA (6, 7). However, the long term clinical outcome and stent patency after percutaneous coronary intervention has not yet been confirmed. In addition, stent durability is a concern since the implanted stent would be continuously exposed to high pressure arising from the great vessels.

On the other hand, surgical treatment provides both a good anatomical and functional outcome. Several surgical techniques have been suggested for ACAOS, such as unroofing the coronary artery, reimplantation of the coronary ostium, and coronary artery bypass graft (CABG). There are advantages and disadvantages to each surgical option. If an intramural course of the RCA is identified, then the unroofing procedure may be quite reliable and a safe approach to correct the anomaly. This anomalous vessel had a slit-like ostium and followed a supracommissural intramural course, and therefore coronary unroofing was considered to be a viable treatment option. In contrast, if the patient did not have an intramural segment, then vessel translocation and reimplantation would be the recommended treatment strategy (8). CABG has a potential risk of early graft failure due to the dynamic competitive flow from native artery. Cho and colleagues suggest that if the patient is over 60 years old with concomitant atherosclerotic stenosis of the RCA as detected by angiography, then CABG may be the best option (9). In this case, surgical treatment was selected because there was evidence of reversible ischemia based on the findings of a stress test and the desire of the patient to resume marathon training. In addition, the strategy of performing a reimplantation of the RCA was selected because the patient was relatively young, there was no atherosclerotic lesions in the RCA, and no intramural course of the proximal RCA was identified. Postoperative coronary CT angiography showed a well-preserved blood flow of the reimplanted RCA.

An anomalous coronary artery causing ischemia is frequently not easy to identify during clinical stress testing. In this case, partial reversible myocardial ischemia was identified during an exercise stress test preoperatively, and a resolution of such myocardial ischemia was identified after surgical therapy. We selected exercise stress myocardial perfusion imaging instead of drug provocation stress testing in order to closely mimic the actual situation that provoked the presenting symptoms. Since there are no established treatment options for ACAOS due to the high variability of the

anomalies and their clinical relevance, the treatment strategies should therefore be carefully considered for each individual case. We herein reported a rare case of an RCA arising from the left sinus of Valsalva which was identified by CT angiography and was successfully treated with RCA reimplantation therapy.

Anomalous coronary artery disease should always be considered when a young patient presents with a cardiac event or sudden death. In addition, for these young patients, an appropriate treatment strategy should be selected to meet the various symptomatic anatomical coronary artery anomalies.

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

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