Case Reports

Sudden Cardiac Death

Associated with Anomalous Origin of the Left Main Coronary Artery from the Right Sinus, with an Intramural Course

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Anomalous origin of the left main coronary artery from the right sinus of Valsalva is extremely rare and can lead to sudden cardiac death. We report a case in which an 18-year-old college student collapsed immediately after a long-distance run of 10 km. After cardiopulmonary resuscitation and electrical shock for ventricular fibrillation, she experienced a return of spontaneous circulation. Cardiac catheterization and cardiac computed tomographic angiography revealed an unusually long intramural course of the left main coronary artery from the right sinus of Valsalva. The young woman underwent a successful unroofing operation for coronary artery correction. She remained asymptomatic upon exercise during 2.5 years of follow-up. ***(Tex Heart Inst J 2015;42(6):554-7)***

Key words: *Coronary vessel anomalies/radiography/surgery; death, sudden, cardiac/pathology; exercise, strenuous; heart defects, congenital; left main coronary artery/abnormalities; sinus of Valsalva/abnormalities; sports*

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Congenital anomalies of coronary arteries are rare, insofar as they are seen upon coronary angiography at a rate of approximately 1%.[1] Anomalous origin of the left main coronary artery (LMCA) with subsequent intramural coursing between the ascending aorta and the main pulmonary artery (PA) is certainly even rarer.[2] In young people engaged in strenuous exercise, this condition commonly results in myocardial ischemia, ventricular arrhythmia, and sudden cardiac death. We report such a case in which the result was favorable.

Case Report

In March 2011, an 18-year-old woman suddenly lost consciousness and collapsed upon completion of a 10-km run at her university's annual campus marathon. She had not been trained for running in accordance with any protocol, and this was her first time to expend such a level of effort. She had not experienced any symptoms before the event.

In our emergency department, a 15- to 20-min course of cardiopulmonary resuscitation and electrical shock for ventricular fibrillation (Fig. 1A) enabled a return of spontaneous circulation (ROSC), and the patient was admitted to our medical intensive care unit (MICU).

In the MICU, she was unconscious and her systemic pressure remained low (~80/50 mmHg), despite a high intravenous dose of an inotropic agent. Her jugular venous pressure was elevated, and her body temperature was 36 °C. A 12-lead electrocardiogram (ECG) obtained 20 minutes after ROSC showed sinus tachycardia with nonspecific ST-T changes and no prolonged QTc interval (QTc, 446 ms) (Fig. 1B). Her initial arterial blood gas analysis showed hypoxia with respiratory and metabolic acidosis. The chest radiograph taken immediately after admission to the MICU showed a normal heart size with bilateral pulmonary edema, similar to a pattern of acute respiratory distress syndrome. A transthoracic echocardiogram (TTE) taken 2 hours after her admission to the MICU revealed a normal left ventricular (LV) chamber size with diffuse hypokinesia and an LV ejection fraction (LVEF) of approximately 0.30. No pericardial effusion was found. The initial complete blood counts, the blood chemistry results, and the toxicology screen were all normal. An intra-aortic balloon pump (IABP)—inserted about 3 hours after admission to the MICU—subsequently assisted, over the course of 4 days, with extracorporeal membrane oxygenation (ECMO) for cardiogenic shock.

The patient's general condition improved substantially after therapeutic hypothermia, and she awoke on the 3rd day of hospitalization. On the 4th and 5th days, she was successfully weaned from the ECMO, the IABP, and the mechanical ventilator.

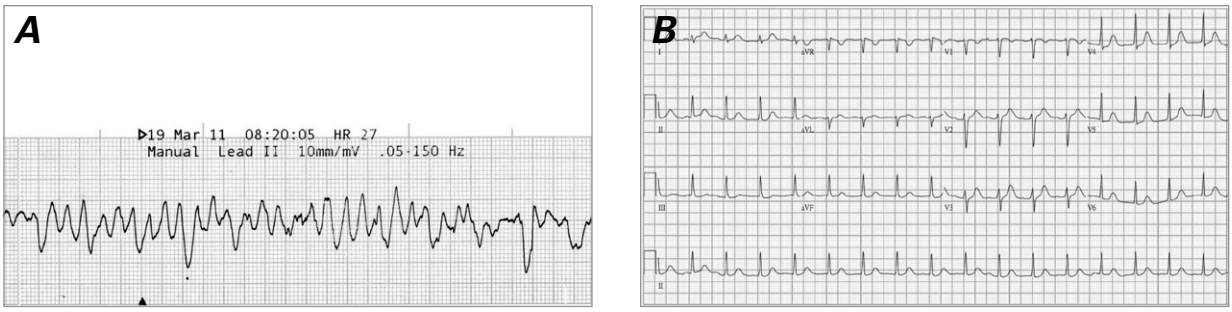

Fig. 1 ***A****) Single-lead II electrocardiogram rhythm strip shows ventricular fibrillation, in the emergency department.* ***B****) Twelve-lead electrocardiogram, taken 20 minutes after return of spontaneous circulation, shows sinus tachycardia with nonspecific ST-T changes and no prolonged QTc interval (446 ms).*

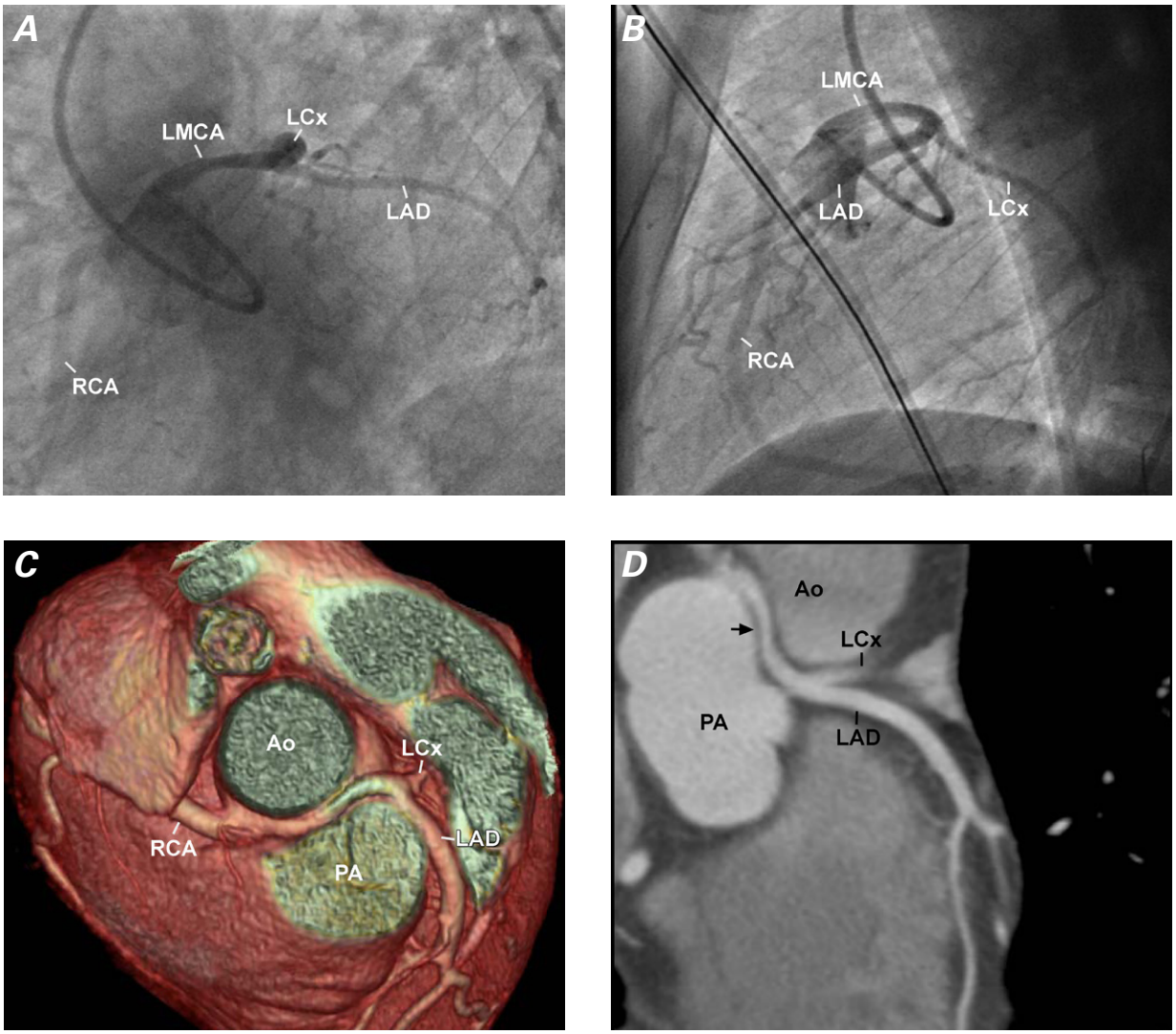

Fig. 2 *Selective left coronary arteriograms in* ***A****) right anterior oblique and* ***B****) lateral views show an anomalous origin of the left main coronary artery (LMCA) from the right sinus of Valsalva. Seen on 64-slice multidetector computed tomography are* ***C****) the anomalous origin of the LMCA from a separate ostium of the right sinus of Valsalva and* ***D****) the intramural course of the LMCA (arrow), which is compressed between the aorta (Ao) and the main pulmonary artery (PA).*

LAD = left anterior descending coronary artery; LCx = left circumflex coronary artery; RCA = right coronary artery

There was no obvious neurologic complication. Her LVEF improved substantially (to 0.60), as shown by an echocardiogram taken 20 days after the cardiac arrest. Cardiac catheterization with coronary angiography was performed before the consideration of an electrophysiologic study 20 days after admission, by which time the patient was fully recovered both neurologically and psychologically. The coronary angiogram showed anomalous origin of the LMCA, which arose from a separate ostium of the right sinus of Valsalva (Figs. 2A and B). The coronary arteries were patent, without obstruction. Subsequently, 64-slice multidetector computed tomography (MDCT), performed to better delineate the course of the anomalous LMCA before cardiac surgery, showed that the LMCA was embedded intramurally, passing between the ascending aorta and the main PA (Figs. 2C and D).

Two months later, the patient underwent an unroofing operation for coronary artery correction. Upon aortotomy, both the normally positioned right coronary artery orifice and the anomalous LMCA orifice could be seen arising from the right sinus of Valsalva (Fig. 3A). Without additional coronary transfer, the long intramural segment of the anomalous LMCA was unroofed (Fig. 3B) to create a neo-orifice in the left sinus of Valsalva (Fig. 3C). The base of the commissure between the right and left cusps needed resuspension, because it was involved in the unroofing procedure.

The patient recovered well and was discharged from the hospital 9 days after surgery. Twelve months later, 64-slice MDCT showed the corrected LMCA without stenosis (Fig. 4). The patient resumed normal activities, including running, without symptoms. In addition, she underwent a treadmill exercise test in which she completed 10 minutes of a Bruce protocol without exertional dyspnea or chest tightness. In the meantime, the follow-up TTE showed a substantially improved LVEF (0.65) with normal LV wall motion. She participated in her campus's annual long-distance run of 10 km again, 2½ years after the surgery, without symptoms.

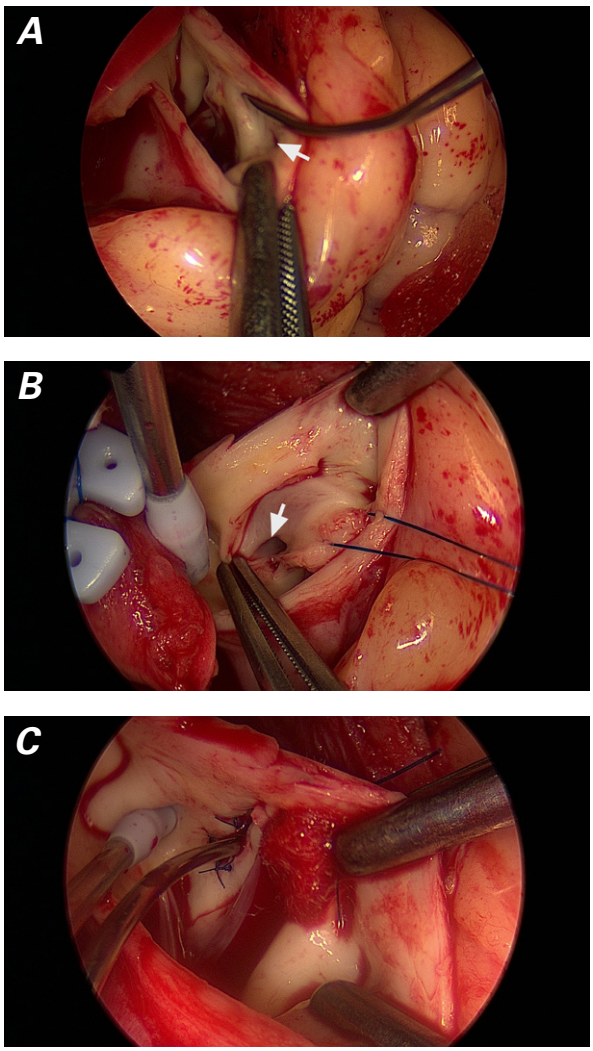

Fig. 3 *Intraoperative photographs.* ***A****) Upon aortotomy, the anomalous left main coronary artery (LMCA) orifice (indicated by the probe) and the normally positioned right coronary artery orifice (arrow) are seen arising from the right sinus of Valsalva.* ***B****) The intramural segment of the anomalous LMCA (the white curved "band" above the arrow that points to the orifice of the right coronary artery) has been unroofed to create* ***C****) a neo-orifice in the left sinus of Valsalva (indicated by the probe).*

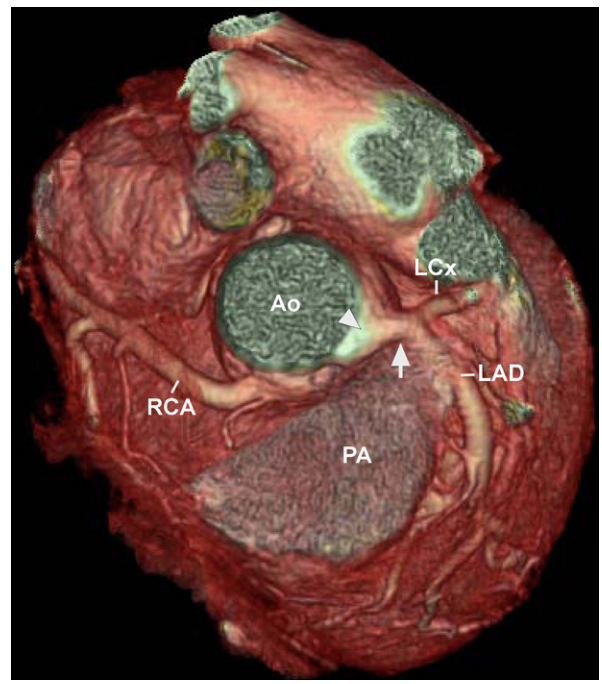

Fig. 4 *Twelve months after surgery, this 64-slice multidetector computed tomogram shows the corrected left main coronary artery (arrow) arising from the left sinus of Valsalva (arrowhead).*

Ao = aorta; LAD = left anterior descending coronary artery; LCx = left circumflex coronary artery; PA = pulmonary artery; RCA = right coronary artery

Discussion

According to autopsy examinations, the most common cause of sudden cardiac death in younger athletes (age, <35 yr) during strenuous exercise is hypertrophic cardiomyopathy; the 2nd most common is coronary artery anomaly.[1] Consequently, after a routine physical and echocardiographic examination in such patients, coronary angiography or cardiac MDCT should be the next diagnostic approach.

The prevalence of congenital coronary artery anomalies on coronary angiography stands at around 1%.[2] Anomalous origin of the LMCA from the right sinus of Valsalva is rather rare (reported prevalence, 0.017%–0.03%)[3] and is often associated with sudden cardiac death during exercise. The LMCA forms an acute angle at its origin, and this can compromise its lumen and blood flow as the LMCA follows the contour of the aorta to the left and posterior. Increased expansion of the aorta during exercise might actually occlude the already slit-like opening of the LMCA by further compressing the orifice.[4] This results in acute myocardial ischemia, fatal arrhythmia, and sudden death. For this reason, in treating young people with angina pectoris, myocardial infarction, or cardiac syncope, one should consider the possibility of this anomaly and perform coronary angiography or cardiac MDCT. However, the anomaly is not always associated with angina pectoris; rather, the presence of angina pectoris varies in accordance with the relationship between the anomalous LMCA, the aorta, and the PA. An interarterial course of the LMCA—between the aortic root and the PA trunk—correlates with a higher incidence of angina, syncope, and sudden death.[3,5] Other anatomic variants for the course of the anomalous LMCA—including retro-aortic, right ventricular free-wall, and septal courses, and a course along the floor of the right ventricle—are considered benign.[3,6] In reviewing the medical literature, we found rare cases of anomalous origin of the LMCA from the right coronary cusp.[3-8] Most reports concern the anomalous origin of the right coronary artery. To the best of our knowledge, no reported case is similar to ours in regard to clinical presentation and the presence of an unusually long intramural course of the LMCA between the aortic root and the PA trunk.

The standard diagnostic imaging tool for coronary artery anomalies is coronary angiography.[8] Other imaging methods—including MDCT or computed tomographic angiography, magnetic resonance imaging, and transesophageal echocardiography—are considered to have complementary roles.[8-10] In our patient, cardiac MDCT was further used to determine the course of the anomalous LMCA, before cardiac surgery was undertaken. That vessel was seen to follow an interarterial course, passing between the aortic root and the PA trunk. Unroofing the intramural segment without bypass grafting can reliably repair this intramural form of anomalous origin of a LMCA.[9,10] To prevent a catastrophic event, early recognition and surgical treatment are imperative.

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