CASE REPORT

Aborted sudden cardiac death associated with an anomalous right coronary artery

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SUMMARY

Coronary artery anomalies arising from the opposite sinus of Valsalva and having an interarterial course between the aorta (AO) and pulmonary artery (PA) are the second most common cause of sudden cardiac death among young athletes, after hypertrophic cardiomyopathy. The right coronary artery (RCA) originating from the AO above the left sinus of Valsalva (LSV) is an extremely rare anomaly. We report the first case of a RCA arising from the AO above the LSV that subsequently runs between the AO and the PA, discovered by a 64-slice multidetector coronary CT, in a patient who was successfully resuscitated from ventricular fibrillation (VF) cardiac arrest while running in a marathon race.

BACKGROUND

This case clearly illustrates how a coronary anomaly can lead to sudden cardiac death (SCD) due to ventricular fibrillation (VF) induced by exercise and that a multidetector coronary CT (MDCCT) should be considered to evaluate patients experiencing cardiovascular symptoms during exercise and routinely in competitive athletes.

CASE PRESENTATION

Background

Coronary artery anomalies are found in 0.6–1.3% of patients undergoing coronary angiography[1 2] and represent the second most common cause of SCD in young athletes who died during or shortly after strenuous physical activity.[3 4] Coronary arteries arising from the opposite sinus of Valsalva and having an interarterial course between the aorta (AO) and the pulmonary artery (PA) are considered to be life-threatening anomalies.[5–7] A right coronary artery (RCA) originating from the AO above the left sinus of Valsalva (LSV) is an extremely rare anomaly, scarcely reported in the literature, and represents fortuitous findings at autopsy[8] or during coronary angiography.[9 10] We report the first case of a RCA arising from the AO above the LSV that subsequently runs between the AO and the PA, discovered by a 64-slice MDCCT in an athlete who had a VF cardiac arrest while competing in a marathon.

Case presentation

A 38-year-old woman, an athlete, was admitted to the intensive care unit at Clinica Reñaca after collapsing with loss of consciousness while competing in a marathon race. Two months before this event, she had begun to experience dyspnoea on exertion. She underwent a medical check-up with an exercise ECG and a two-dimensional echocardiography because of the symptoms; the tests were normal except for the diagnosis of exercise-induced asthma. She was prescribed treatment in the form of a salbutamol inhaler.

At the scene of her collapse, the patient was immediately resuscitated with use of cardiopulmonary resuscitation, which was continued during ambulance transportation to our hospital. An automated external defibrillator was unavailable. On admission, the patient was in VF, which was successfully converted to sinus tachycardia with 120 J. After cardioversion, she regained consciousness and her blood pressure was 130/85 mm Hg. Pulmonary and cardiac auscultations were normal. A 12-lead ECG showed sinus rhythm without signs of ischaemia. Chest X-ray and a 2D echocardiogram were normal. Initial laboratory investigations showed arterial blood gases (using FiO_2 of 0.50): pH 7.1, PaO_2 67.0 mm Hg, $PaCO_2$ 50.1 mm Hg, HCO_3 15.2 (base excess −14.5), oxyhaemoglobin saturation 85.6%, arterial lactate 57.4 mg/dL, troponin <0.03 ng/mL, creatine kinase-myocardial band 2.8 ng/mL and blood urea nitrogen 32.1 pg/mL. We considered that further diagnostic evaluation was warranted. A MDCCT depicted an anomalous origin of the RCA originating from the left side of the AO anteriorly and above the LSV (figure 1), with an acute angle take-off along the aortic wall that subsequently courses between the AO and the PA, resulting in compression and narrowing of the proximal RCA lumen (figure 2). We felt that the RCA likely became compressed during the patient's vigorous activity and that she subsequently suffered VF arrest due to RCA-associated myocardial ischaemia.

She was counselled about this risk and discussions were held with her about options for treatment, including reimplantation of the RCA into its typical native location or implantation of an implantable cardioverter-defibrillator coupled with restrictions on exercise activity levels. She elected to consider these options and requested hospital dismissal. Our patient was discharged from hospital free of symptoms at day 4.

OUTCOME AND FOLLOW-UP

After our patient was discharged from hospital, we lost the follow-up.

DISCUSSION

The anomalous origin of a coronary artery from the opposite aortic sinus and coursing between the

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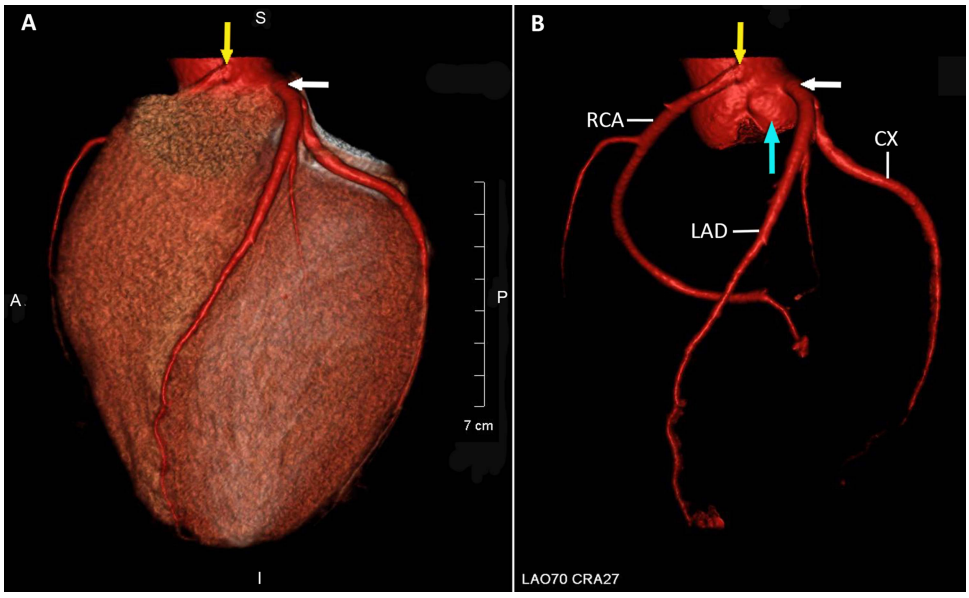

Figure 1 (A) Three-dimensional volume rendered MDCCT of the coronary arteries, showing an anomalous RCA (yellow arrow) arising from the left side of the aorta anteriorly and superiorly of the (LMCA, white arrow) coming from the LSV. (B) Volume rendered tree view of the coronary arteries. This clearly depicts that the RCA arises ectopically from the left side of the aorta (yellow arrow) anteriorly and above the LSV (blue arrow) and the LMCA (white arrow; LAD, left anterior descending coronary artery; LCX, left circumflex artery; LMCA, left main coronary artery; LSV, left sinus of Valsalva; MDCCT, multidetector coronary CT; RCA, right coronary artery).

AO and PA is one of the most common causes of SCD in the young.[3 4 11] Although SCD may be the first manifestation of this heart condition, almost half of these patients might experience some warning with cardiovascular symptoms such as palpitations, syncope, chest pain or dyspnoea on exertion,[11–13] as occurred in our patient. Certainly, the diagnosis of this disease requires a high index of suspicion and access to more advance cardiovascular imaging modalities such as a MDCCT. The tests that were chosen to initially evaluate our patient were reasonable and appropriate from a cost-effective evaluation perspective. However, they were unable to detect her congenital aortic abnormality. These tests are frequently normal in this population of patients.[11 13]

This case also highlights another unusual aspect of an anomalous origin of a RCA. Usually, the anomalous origin from the contralateral side of the AO is from within the sinus of Valsalva or from the proximal normal left coronary artery.[6 14–17]

The RCA originating from the left side of the AO above the LSV is an extremely rare anomaly scarcely reported in the medical literature.[8–10] Our patient is the first case of a RCA anomaly arising from the AO above the LSV with a subsequent course between the AO and the PA, detected by a MDCCT in a competitive athlete who developed a cardiac arrest from VF during a marathon race. This non-invasive imaging technology provides exceptional quality of the 3D anatomical coronary images allowing the visualisation of the

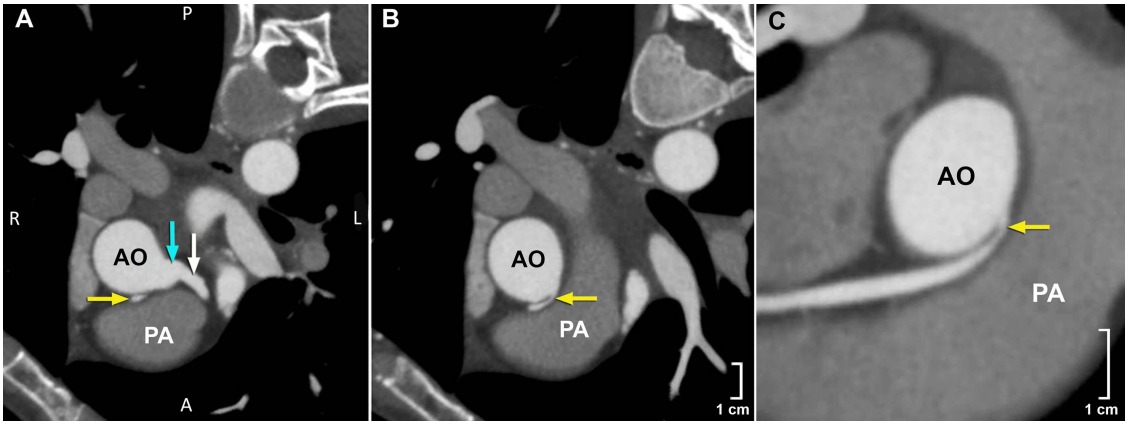

Figure 2 Axial images from a multidetector coronary CT coronary angiogram. (A) Multiplanar reconstruction at the LSV (blue arrow) showing the LMCA (white arrow) arising from it. Note the anomalous RCA (yellow arrow) located between the AO and the PA. (B) Multiplanar reconstruction through the tubular portion of the ascending AO, above the LSV, depicts the high origin of the anomalous RCA with an acute angle take-off along the aortic wall (yellow arrow), coursing anteriorly and to the right with an interarterial course between the AO and the PA. (C) Curved multiplanar reformation of the anomalous RCA depicts that the proximal portion of the RCA is narrowed and has an acute angle take-off from the AO, and follows an interarterial course between the AO and PA (AO, aorta; PA, pulmonary artery, LMCA, left main coronary artery; LSV, left sinus of Valsalva; RCA, right coronary artery).

origin, course and distal portion of these arteries.[11] Furthermore, we believe that MDCCT is likely the single best imaging technique today to evaluate patients presenting with cardiovascular symptoms during exercise and routinely in competitive athletes.[13 18]

Conclusion

We suggest that MDCCT is the diagnostic method of choice when a clinician suspects an unusual mechanism for dyspnoea or the potential for an anomalous coronary artery, owing to the increasing availability of this technology.

Learning points

- Coronary artery anomalies are rare but represent the second most common cause of sudden cardiac death in young athletes.
- Sudden cardiac death may be the first manifestation of this heart condition; however, half of the patients may experience cardiovascular symptoms such as palpitations, syncope, chest pain or dyspnoea on exertion, but often routine cardiac tests such as exercising ECG and two-dimensional transthoracic echocardiography are normal.
- A multidetector coronary CT is currently the best imaging technique to evaluate patients with cardiovascular symptoms triggered by exercise and should be considered routinely in competitive athletes.

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