CASE REPORT

Usefulness of multidetector CT angiography for anomalous origin of coronary artery

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SUMMARY

A 17-year-old man underwent clinical evaluation of exercise-induced syncope. Routine exercise stress test did not show any myocardial ischaemic changes or arrhythmias on the ECG recording. However, multidetector CT (MDCT) angiography of the coronary arteries revealed an abnormal origin of the left coronary artery from the right coronary sinus. The participants' symptoms were diagnosed as cardiogenic syncope possibly due to transient stenosis of the left main coronary artery caught between the functionally distended aortic root and the pulmonary trunk during exercise. After successful patch coronary angioplasty, his symptoms disappeared completely even during a similar degree of strenuous exercise. It is important for clinicians not to overlook possible coronary artery anomalies during management of patients with exercise-induced syncope. MDCT coronary angiography may be a useful and non-invasive tool to establish diagnosis and a surgical approach to rectify congenital coronary artery anomalies.

BACKGROUND

Congenital anomalies in coronary artery origin are observed in 0.2–1.2% of the population¹ and detection of these abnormalities by coronary angiography (CAG) is rare at about 1–5%.² Some coronary artery origin anomalies are an important cause of sudden death during exercise in young athletes. However, less than half of the cases show symptoms such as chest pain, syncope or palpitations before the condition becomes evident, and the first symptom is sometimes sudden death.³ We have reported a juvenile case with an abnormal coronary artery origin who reported of syncope without any cardiac symptoms during exercise. His condition was diagnosed using multidetector CT (MDCT) CAG, and complete recovery was achieved after coronary artery reconstruction surgery.

CASE PRESENTATION

A 17-year-old man presented in March 2011 reporting of exercise-induced syncope. He had no history of smoking or drug misuse, and had no medical history or familial history of cardiovascular disease or sudden death. From October 2010, he had experienced occasional sudden syncope during strenuous physical exercise, such as playing basketball, without precursor symptoms such as chest pain, palpitations, dyspnoea or dizziness. He was unconscious for approximately 1–5 min, and did not report of chest pain or dyspnoea after regaining consciousness. He underwent brain MRI and EEG

at the division of neurology of our hospital. These neurological tests did not observe any abnormal findings.

On physical examination, the participant had regular heart rhythm and no heart murmur in auscultation. His laboratory tests including lipids, glucose levels and blood pressure were within the normal range. On a routine transthoracic echocardiographic study, left ventricular systolic function and dimensions were within the normal range (left ventricular ejection fraction=67%, end-systolic left ventricular dimension/end-diastolic left ventricular dimension=51/31 mm). In addition, there was no evidence of any valvular disorder.

Baseline ECG showed negative T wave inversion only in lead V1, and no other abnormalities were found in ST-T segments, axis or P wave (figure 1A). He underwent a treadmill exercise test, and this achieved an average maximum heart rate of 175 bpm, 17 metabolic equivalents and a peak Borg scale of 17. There was no evidence of ischaemic ECG changes or arrhythmias during peak exercise (figure 1B). Exercise stress myocardial perfusion scintigraphy during ergometer exercise also showed no myocardial ischaemic changes.

Cardiac catheterisation showed no evidence of fixed coronary stenosis (figure 2A, B) or abnormal left ventricular wall motion. At the routine CAG, the left coronary ostium could not be found at the normal site, and the left coronary artery (LCA) originated from the right sinus. However, the exact course of the left main coronary artery and its localisation related with the aortic root and the pulmonary trunk were entirely unknown from the CAG.

However, 320-row MDCT revealed an abnormal origin of the LCA from the right coronary sinus (figure 3A). The right coronary artery (RCA) had a normal origin from the right coronary sinus. The left main coronary artery coursed aberrantly between the pulmonary trunk and the aortic root (figure 3B). After consideration of the cardiac imaging tests, his symptoms were diagnosed as cardiogenic syncope possibly due to transient stenosis or obstruction of the left main coronary artery caught between the distended aorta and the pulmonary trunk during exercise.

The participant underwent open-heart surgical operation at our hospital in 16 May 2011. During surgery it was confirmed that the LCA originated from the commissure part of the right aortic sinus, while the RCA was located in the normal position. The left main coronary artery and the left Valsalva sinus wall were sutured using pulmonary wall

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Figure 1 Standard 12-lead ECG.
(A) Standard 12-lead electrocardiogram at rest. (B) Standard 12-lead electrocardiogram after exercise. Scale=10 mm/mV.

covered with autopericardium (patch plasty of LCA with roofing over pulmonary arterial wall). The patient's postoperative course was uneventful, and he was extubated on the same evening and discharged 11 days postoperatively with an antiplatelet regimen. Follow-up coronary MDCT did not show any abnormalities in the reconstructed LCA and the RCA (figure 4A, B).

DISCUSSION

We have reported a rare case of exercise-induced syncope in an apparently healthy high school boy. In this case, routine cardiac tests including exercise ECG and cardiac scintigraphy could not reveal any cardiogenic cause of his symptoms. However, a significant congenital anomaly of the LCA origin was found by coronary MDCT angiography. It has been reported that congenital abnormalities in the origin or course of the coronary arteries, especially the origin of the LCA from the right coronary cusp, are very rare.²⁻⁴ In 56 Japanese patients, Kaku *et al*⁵ reported that abnormal origin of the LCA occurred with only a quarter

of the frequency of abnormal origin of the RCA, and no cases were found of anomalous origin of the LCA from the right sinus of Valsalva crossing between the aorta and the pulmonary trunk. Cheitlin *et al*⁶ have reported that some patients with the LCA arising from the right coronary cusp had sudden and unexplained deaths after exercise. This report has also shown that cases with abnormal origins of the LCA and the RCA have a high risk of angina and sudden death.⁶ Maron demonstrated that causes of sudden and unexplained death in young adults (under the age of 35) were coronary artery origin anomaly (13.7%), hypertrophic cardiomyopathy (26.4%) and heart shaking (19.9%).⁷ The mechanisms underlying sudden and unexplained death in young adults due to coronary artery origin anomaly after exercise are as follows: (1) rising blood pressure and increased blood flow during exercise distend the pulmonary trunk and aortic root, and then transiently decrease blood flow in the LCA by creating a functional sandwich between the distended pulmonary trunk and the aortic root, (2) spasm is caused by a vascular disorder of the coronary artery.⁸ In the

Figure 2 Coronary angiography.
(A) Right coronary artery. (B) Left coronary artery.

Figure 3 Preoperative multidetector CT coronary angiogram. Volume-rendered image (A) and axial image (B) showing the origin and course of the left main coronary artery (LM) in relation to the aorta (Ao) and the pulmonary artery (PA). LAD, left anterior descending artery; LCX, left circumflex artery; RCA, right coronary artery.

present case, as the syncope was only induced during vigorous basketball playing and the patient had no established coronary risk factors, the second possibility may be unlikely.

Basso *et al*³ showed that 55% of sudden death cases due to coronary artery origin anomaly were asymptomatic and never underwent heart examination. On the other hand, 45% of cases had some symptoms, such as chest pain (42%), syncope (34%) or palpitations (25%),³ but had no abnormal findings even after cardiac evaluation by ECG exercise stress test or echocardiography as shown in the present case. The present case is consistent with this report, although we did MDCT CAG, which is useful for diagnosis of this type of congenital coronary artery anomaly. Several previous case reports have suggested that trans-thoracic echocardiography is useful for identification of anomalous origin of the left main coronary artery.^{9 10} In the present case, however, abnormal Doppler colour flow mapping suggested that the aberrant coronary artery would not have been amenable to detection in a routine setting. It would be important to determine whether echocardiographic evaluation may be useful for screening for this type of coronary anomaly.

The present case was judged as eligible for heart surgery as the episode of syncope occurred during exercise, making the

participant at high risk of sudden death. Several reports have described cases of coronary artery origin anomaly with severe ventricular arrhythmias or myocardial ischaemia undergoing surgical treatment such as unroofing or coronary artery bypass.^{11 12} In the present case, the LCA originated from the right coronary sinus, although the RCA originated from its normal position. It was therefore elected to create a neo-ostium in the left coronary sinus and to suture a patch between the left main trunk and the left sinus of Valsalva using pulmonary arterial tissue as a patch roof. Postsurgical coronary angiogram showed good patency of the reconstructed LCA. Several months after discharge, the patient's exercise-induced syncope was completely eliminated during a similar degree of physical activity.

CONCLUSION

We have reported a juvenile case of frequent exercise-induced syncope due to an abnormal origin of the LCA. Although this type of case is rare, it may be important for clinicians to consider coronary artery anomalies during management of patients with exercise-related symptoms including syncope. In addition, recent advances in MDCT CAG may make this a useful and non-invasive tool to establish diagnosis and surgical strategies

Figure 4 Postoperative multidetector CT coronary angiogram. Volume-rendered image (A) and axial image (B) showing the origin from the right aortic sinus and the patch graft between the aorta (Ao) and the pulmonary artery (PA). LAD, left anterior descending artery; LCX, left circumflex artery; RCA, right coronary artery; LM, left main coronary artery.

for rectifying congenital coronary artery anomalies with origin from the wrong aortic sinus.

Patient's perspective

We have reported a juvenile case of frequent exercise-induced syncope due to an abnormal origin of the left coronary artery. Although this type of case is rare, it may be important for clinicians to consider coronary artery anomalies during management of patients with exercise-related symptoms including syncope.

Learning points

- ▶ Coronary artery anomalies should be added to the list of differential diagnosis for exercised-induced syncope.
- ▶ Multidetector CT coronary angiography is a useful and non-invasive tool to establish diagnosis and a surgical approach to rectify congenital coronary artery anomalies.
- ▶ Routine cardiac tests such as transthoracic echocardiography, treadmill exercised test, exercised stress perfusion myocardial scintigraphy and cardiac catheterisation sometimes fail to reveal this abnormalities.

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