Journal of Zhejiang University SCIENCE B
ISSN 1673-1581 (Print); ISSN 1862-1783 (Online)
www.zju.edu.cn/jzus; www.springerlink.com
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Case Report:

Coronary arterial spasm in single right coronary artery*

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Received Apr. 23, 2009; Revision accepted July 30, 2009; Crosschecked Oct. 12, 2009

Abstract: We presented a case of anomalous single-coronary artery detected incidentally during routine coronary angiography. A 32-year-old male Chinese patient presented with recurrent pre-syncope and six episodes of syncope. Coronary angiography and coronary-computed tomography (CT)-angiography performed by a dual-source computed tomography (DSCT) revealed that the patient had a single large right coronary artery. A moderately large branch originated from the proximal part of the single right coronary artery and extended to the left, passing the anterior to the pulmonary artery, and divided into the anterior descending artery branch and circumflex branch at the base of the left auricular appendage. The episodes of the syncope were suspected to be caused by coronary arterial spasm, so this patient was on a regimen of 30 mg of diltiazem every 6 h and had no recurrence of syncope during follow-up.

Key words: Single right coronary artery, Spasm, Syncope
doi:10.1631/jzus.B0920117 **Document code:** A **CLC number:** R54

INTRODUCTION

About 0.3%~1% of healthy individuals in the world have the anomalies of the coronary arteries (Angelini *et al*., 2002). According to the modified version of a classification system developed by Greenberg *et al*.(1989), the coronary arterial anomalies have been grouped into anomalies of origin, course, and termination. Among the anomalies of the coronary arteries, the single-coronary artery is defined as only one coronary artery arising with a single ostium from the aortic trunk. This is an extremely rare congenital anomaly that is seen in only 0.0024%~0.044% of the population (Desmet *et al*., 1992).

People with a single-coronary artery may have a normal expectation of life; however, some may be at a risk for sudden death. In addition, spasm of the single-coronary artery is also suggested as a cause for syncope (Yamamoto *et al*., 1981). Here, we presented a patient with single right coronary artery, who suffered from episodes of syncope caused by the spasm of the single-coronary artery.

CASE PRESENTATION

A 32-year-old male Chinese patient was admitted to the Department of Cardiovascular Medicine, the First Affiliated Hospital of Nanjing Medical University, Nanjing, China due to syncopal episodes on Jan. 6, 2009. He had suffered from recurrent pre-syncope and six episodes of syncope since October, 2007. He was a smoker and alcohol drinker. He was 170 cm tall and weighed 60 kg. He had no fever, and his laboratory and chest X-ray examinations were within normal ranges. Electrocardiography (ECG) showed normal sinus rhythm, magnetic resonance imaging (MRI) of the brain revealed no abnormal findings, and a 2D echocardiogram showed normal left ventricular function and no valvular abnormality.

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* Project (Nos. 30400173 and 30971257) supported by the National Natural Science Foundation of China

No symptom occurred during the initial Holter recording or monitoring.

This patient was treated as an acute coronary syndrome patient and underwent coronary angiography on Jan. 12, 2009. However, the left coronary artery was not found during the operation, and a single-coronary artery was presented from the cannulation of the right coronary artery (RCA) with a JR4 6-Fr catheter. The monitor ECG demonstrated ST-segment elevation in leads II, III, and aVF with reciprocal ST-segment depression in the anterior leads. During the operation, the patient was alert and felt pre-syncope similar to the one occurred in the past. Although the right coronary artery spasm could be caused by the cannulation, we suspected that the episode might be caused by right coronary arterial spasm. Isosorbide dinitrate (ISDN; 0.5 mg/ml solution over 20 s) was injected through the Judkins catheter. One minute after the injection, the coronary arterial spasm was relieved and electrocardiographic abnormalities disappeared. The angiography was performed from several projections.

The results of angiography showed an absence of the left coronary ostia with a single right coronary artery, and that a moderately large branch originated from the proximal of the single right coronary artery and extended to the left, passing the anterior to the pulmonary artery and divided into the anterior descending artery branch and circumflex branch at the base of the left auricular appendage (Figs.1~2). No atheromatous lesion was seen during angiography. The results of coronary-computed tomography (CT)-angiography by a dual-source computed tomography (DSCT) (Somatom-Defintion, Siemens Medical Solutions, Forchheim, Germany) confirmed the findings of coronary angiography (Fig.3). CT scan was obtained using the following settings: collimation, 64 sections of 0.6 mm thickness; rotation time, 0.33 ms; tube voltage, 120 kV; charge, 400 mAs/rot; and pitch, 0.25. Prospective electrocardiographic tube current modulation (electrocardiographic pulsing of 28%~80%) was used to provide the optimal radiation dose throughout the whole cardiac cycle. Bolus tracking was used to trigger data acquisition as the density in the descending aorta reached 100 Housfield unit.

Because we suspected that the episode of syncope might be caused by coronary arterial spasm, we treated this patient with a regimen of 60 mg of

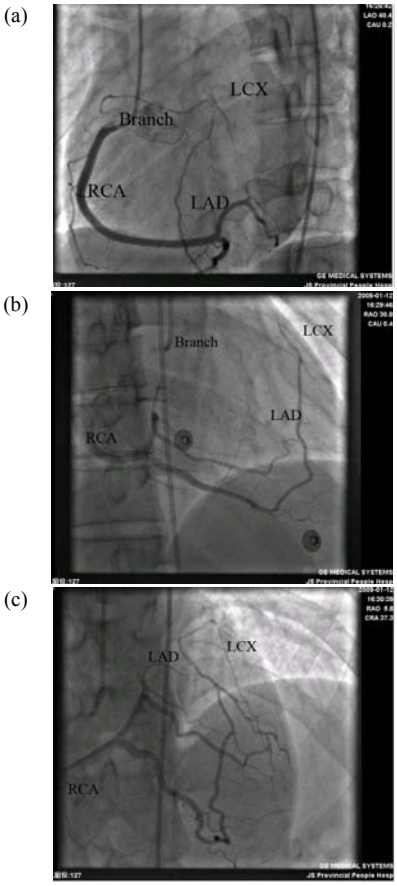

Fig.1 Coronary cineangiogram obtained by Sones technique from several angiographic views. A single large right coronary artery originated from the right aortic sinus, and approximately 2 cm from the origin of the single right coronary artery. A moderately large branch extended to the left and divided into the anterior descending artery branch and circumflex branch. (a) 40° left anterior oblique projection; (b) 30° right anterior oblique projection; (c) 37° cranial oblique projection

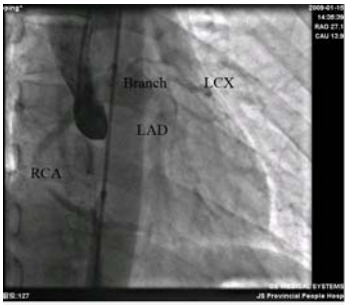

Fig.2 Coronary cineangiogram obtained by non-selective technique from 27° right anterior oblique and 14° caudal oblique projection. The results of angiography showed an absent of left coronary ostia with a single right coronary artery (RCA) continuing as the left circumflex (LCX) artery and a hypoplastic left anterior descending (LAD) artery

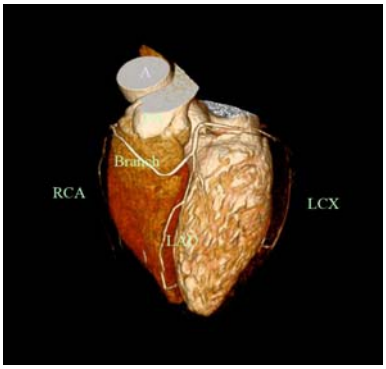

Fig.3 Anterior oblique volume rendering image showed that a single large right coronary artery originated from the right aortic sinus, and approximately 2 cm from the origin of the single right coronary artery, and that a moderately large branch extended to the left, passing anterior to the pulmonary artery, and divided into the anterior descending artery branch and circumflex branch

isosorbide dinitrate daily and 30 mg of diltiazem every 6 h. In the next 9 d, he had no recurrence of syncope, and several 24-h Holter tape monitorings showed no significant ST-T changes. This patient was discharged and was on a regimen of 30 mg of diltiazem every 6 h and had had no recurrence of syncope so far.

DISCUSSION

The entire myocardium of the heart with a single-coronary artery is nourished by an artery, regardless of distribution, that arises by one ostium from an arterial trunk (Smith, 1950). The diagnosis of the coronary artery anomalies had depended on the coronary angiography for several decades. However, a previous study determined the origin of anomalous coronary arteries in all 16 patients, of which the coronary angiography alone detected only 53% (P=0.016) (Shi *et al*., 2004). Therefore, the conventional coronary angiography alone is not a golden standard for the detection of coronary anomalies. The ideal tool for the diagnosis of coronary artery anomalies is the conventional coronary angiography supported by CT (Sevrukov *et al*., 2002), magnetic resonance imaging (MRI) (Bunce *et al*., 2003), and transesophageal echocardiography (Dawn *et al*., 2003). Our patient was diagnosed with a single right coronary artery by the conventional angiography and was confirmed by dual-source CT angiography. The latter revealed that the single-coronary artery originated from the right Valsalva's sinus that formed the right coronary arteries (RCA) and a left-extending branch extending to be the left circumflex (LCX) coronary artery and the left anterior descending (LAD) coronary artery.

The single-coronary artery is an extremely rare coronary abnormality, and it is usually diagnosed incidentally during conventional coronary artery angiography or on postmortem evaluation. In a large series of 126595 patients undergoing coronary angiography, a single-coronary artery from the right Valsalva's sinus was found at 0.019% (Yamanaka and Hobbs, 1990). In a large retrospective study, 70850 patients undergoing coronary angiography between 1999 and 2005 were reviewed, and only 10 patients (0.014%) were found to have the single-coronary artery, of which 3 had the single-coronary artery originated from the left Valsalva's sinus and 7 from the right Valsalva's sinus (Akcay *et al*., 2008).

The current classification system of solitary coronary artery was proposed by Lipton *et al*.(1979), who incorporated two previous systems defined by Smith (1950) and Ogden and Goodyear (1970), respectively, into one final system. Based on those classifications, the first division was made between the 'R' right-type and 'L' left-type according to the site of origin of the single-coronary artery (i.e., in the right or left Valsalva's sinus). A case was then designated to Group I, II or III depending on the anatomical course of the artery. Group I had an anatomical course of either a right or left coronary arteries (RCA or LCA); for example, when the artery originates from the right Valsalva's sinus, it follows the course of a normal RCA, giving off the posterior descending artery in normal fashion near the crux and then continuing in the left atrioventricular groove to give off the posterolateral left ventricular branches. Group II anomalies arise from the proximal part of the normal RCA or LCA, and cross the base of the heart before assuming the normal position of the inherent coronary artery. The subgroup designation of Group II refers to the relationship between the anomalous coronary artery and the aorta and pulmonary artery. The letters 'A', 'B' and 'P' refer to 'anterior', 'between', and 'posterior' patterns, respectively. Group

III describes the anomaly where the LAD coronary artery and LCX coronary artery arise separately from the proximal part of the normal RCA. According to the classification system, the case we reported belongs to Right, type IIA.

The clinical significance of single-coronary artery is unknown. Although it is generally considered benign, some studied sudden death associated with isolated congenital coronary anomalies in an autopsy population and observed that sudden death occurred in patients with single-coronary artery (Frescura *et al*., 1998). Others found that 15% of patients may have myocardial ischemia directly caused by the abnormal anatomy of the arteries and not by coronary artery disease (Shirani and Roberts, 1993). In addition, coronary arterial spasm is also reported in single-coronary artery (Yamamoto *et al*., 1981). Our patient demonstrated that coronary arterial spasm may occur in single-coronary artery and may be a cause for syncope, sudden death or myocardial infarction.

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