Case Report

A CASE OF MALIGNANT RIGHT CORONARY ARTERY: FREQUENT ANGINA IN YOUNG PERSON

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ABSTRACT

We presented a case a young adult with activity-triggered atypical chest pain and diagnosed with anomalous origin of right coronary artery (RCA) from the left coronary sinus with an interarterial course between the aorta and the main pulmonary artery that was detected by CT coronary angiography. This anomaly has been called malignant RCA. Coronary artery anomaly is a congenital condition. Most of the cases remain asymptomatic. This condition is also one of the most causes of sudden cardiac death, because the coronary artery examination is not regularly done. Nevertheless, during high intensity activity, it could be symptomatic and might be lethal. Diagnosing coronary artery anomalies might be tricky and cardiologists must be aware of this. The CAAs condition is a rare situation. The CAAs condition is associated with sudden death, especially intense physical activity. There was no rigid guideline for the management of the CAAs condition, so that planning a treatment in the inter-specialist team should be done.

Keywords: Malignant RCA; chest pain; health risks; CT coronary angiography

INTRODUCTION

Coronary artery anomalies (CAAs) are still becoming an important matter to discuss in basic and clinical practice. There is extensively discussed in the literature regarding the definition of CAAs, especially in how it can be different with a normal variant of the coronary artery. Nevertheless, a previous study has proposed a comprehensive classification of CAAs based on several groups of abnormalities; anomalies of origination and course, anomalies of intrinsic coronary arterial anatomy, anomalies of coronary termination, and abnormal anastomotic vessels (Angelini 2007). Moreover, Angelini (2002) had proposed an accessible definition of the normal variant shown in Table 1. The clinical spectrum of CAAs is enormously extensive, from asymptomatic to resting ischemia. The awareness of atypical coronary anatomy could help the cardiologist in giving precise diagnosis and treatment,
including catheterization laboratory and surgical procedure (Angelini 2002).

Table 1. Purpose definition of the normal heart variant conditions

| Feature             | Range                                      |
|---------------------|--------------------------------------------|
| Number of Ostia     | 2–4 Right and left anterior sinuses (upper mid-section) |
| Location            | Only left (left anterior descending and left circumflex artery) |
| Proximal orientation| 45–90 degrees off the aortic wall          |
| Proximal common stem or trunk | Direct, from ostium to destination      |
| Mid-course          | Extramural (sub-epicardial)               |
| Branches            | Adequate for the dependent myocardium     |
| Essential territories| Right Coronary Artery (Right ventricle free wall) |
|                     | Left anterior descendent (anteroseptal)   |
|                     | Obtuse Marginal Artery (Left ventricle free wall) |
| Termination         | Capillary bed                             |

The CAAs are a rare congenital condition, and their incidence is around 0.17%–1.2% (Graidis et al. 2015). Regardless of its incidence, around 20% of the CAAs are malignant and could undergo life-threatening manifestation, such as lethal arrhythmias, syncope, myocardial infarction, or even sudden death (Datta et al. 2005). Specifically, the incidence of the malignant CAAs from the right coronary artery origins is around 0.03–0.17% from patients undertaking angiography (Satija et al. 2012). Here, we presented the right CAAs malignant type with symptomatic atypical chest pain in a 34 years old man.

CASE REPORT

A-34-years-old man has complained of vague chest pain since more than six months ago. There were no risk factors or any significant past medical history. No remarkable result from routine blood examination and rest electrocardiography (ECG). However, during exercise the treadmill test showed ST depression in the lead VI. The patient was referred to the radiology department to undergo CT coronary angiography.

CT coronary angiography was performed using a 64 slice CT scanner (Brilliance-64, Philips, The Netherland). The following parameters were used: 120 KVP, 800 mAs, rotation time: 0.4 s, and collimation: 64 × 0.625 mm. Using a dual-head pressure injector, 80 ml of non-ionic iodinated contrast was administered at the rate of 5 ml/sec, followed by 20 ml of saline. The total scan time was 8.6 seconds. Reconstruction was done with 0.6 mm slice thickness 0.5 mm increment.

CT coronary angiography showed the right coronary artery originating from the left coronary sinus, coursing between the aortic root and pulmonary artery (Figure 1a). The rest of the coronary arteries, the left coronary artery and its branches, a normal course (Figure 1b, and Figure 1c). However, we did not yet decide on the treatment for this patient, and still need to follow-up for reassurance (Figure 1).

Figure 1. The schematic representation of malignant right coronary artery

DISCUSSION

The CAAs were frequently found as the cause of sudden death (SD) cases in the young. Even though it is a congenital condition, many subjects survive asymptomatic until young adult (Basso 2005). The anomalous coronary artery (CA) origin, either left the main artery from the right sinus or vice versa, had been found 0.17% during autopsy (Basso 2005). Moreover, recent studies found four children from 2,388 children underwent angiogram diagnosed with either anomalous origin of the left main coronary artery (ALMCA) from the right sinus or anomalous origin of the right coronary artery (ARCA) from the left sinus (Davis et al. 2001). Two and half week-old patients with ARCA had normal serial resting ECG after repeated 12 ECG exams one, a half months later and at 35 months. However, the 12 years old patient showed uniform ventricular ectopy with a left bundle configuration consistent with right ventricular origin.

The ectopy diminishes during exercise ECG. In exercise ECG, there was also abnormal repolarization of the lateral chest lead with T-wave inversion and ST elevation. The treadmill exercise perfusion study exhibited inferior wall ischemia of moderate intensity in the RCA perfusion area. The patient persisted
asymptomatic during the test. In our case, the patient never underwent either resting or exercise ECG before and remained asymptomatic. Until the patient reached 34 years old, when he was busy with his job, he suffered atypical chest pain. The resting ECG resulted in no remarkable result. On the other hand, treadmill ECG examination showed notable change with suspicion from right ventricular origin.

Anomaly aortic origins of coronary arteries account for 14-17% of sudden cardiac death for healthy children or young athletes during or immediately after physical exercise (Silva et al. 2018). The lethal mechanisms of the CAAs are considered through malignant arrhythmias and ischemia-myocardial dysfunction (Lluri & Aboulhosn 2014). Those mechanisms are triggered by vigorous physical exercise (Hill & Sheppard 2014). Anomaly aortic origins of coronary arteries further classified into two subgroups; anomalous origin of coronary arteries from the opposite sinus (ALMCA and ARCA), and anomalous left coronary artery from the pulmonary artery (ALCAPA). Despite the classification of the two subgroups, the correlation with sudden cardiac death is quite high (Lluri & Aboulhosn 2014). The physiological supply and demand of the heart depend on its exercise type.

During isotonic and isometric exercises, the venous return, and left end-diastolic volume will increase. The adrenergic system stimulation also increases the heart rate, blood pressure, cardiac output, and myocardial contractility. The purpose of the mechanism is to increase oxygen supply to the heart. Unfortunately, the CAAs condition is contrary to that purpose (Wasfy et al. 2015; Cheitlin & MacGregor 2009). In this patient, asymptomatic condition (until he grew up to adult), seemed rational since the patient was not an athlete, and he was probably not doing high-intensity exercise until he became very busy with his work now. Nonetheless, this type of CAAs are associated with a high risk of sudden cardiac death based on previous evidence (Basso 2005). We strongly suggested the patient to reduce his work intensity and scheduled a follow-up meeting to plan the treatment.

In ALMCA or ARCA conditions, the proximal segment of the anomalous CA may course anterior to the pulmonary trunk, posterior to the aorta, or between the pulmonary artery and the aorta (Basso 2005). At some points, especially during exercise, the CA squeezes due to the increased cardiac output with a diastolic expansion of great vessels. As a result, myocardial ischemia might be happening (Basso 2005; Silva et al. 2018; Cheitlin & MacGregor 2009). The ALMCA might have a bigger risk of sudden cardiac death compared to ARCA (Basso 2005). In this case, our patient had an ARCA condition, and this condition might have a lower risk than ALMA’s risk of sudden cardiac death. However, this condition could still be life-threatening.

**CONCLUSION**

In conclusion, the CAAs condition is a rare situation. However, cardiologists need to be aware of congenital anomalies because it might help in clinical practice. Even though the incidence is low, the CAAs condition is associated with sudden death, especially intense physical activity. There was no rigid guideline for the management of the CAAs condition, so that planning a treatment in the inter-specialist team should be done.

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