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Original Research

A Case of Malignant Course of Right Coronary Artery: Frequent Angina in Young Person

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ABSTRACT

Congenital anomalous coronary artery is a rare condition, but it might be the biggest pitfall for cardiologist. We present a case of young adult with activity-triggered atypical chest pain and diagnose with anomalous origin of right coronary artery (RCA) from the left coronary sinus with inter-arterial course between the aorta and the main pulmonary artery that was detected by CT coronary angiography. This anomaly has been called malignant course RCA. Coronary artery anomaly is a congenital condition. Most of the cases are remain asymptomatic. This condition also one of the most cause for sudden cardiac death because the coronary artery examination is not regularly done. Nevertheless, during high intense activity, it will be symptomatic and might be lethal. Diagnose coronary artery anomalies might be tricky and cardiologist must be aware with this. More devastating, no firm guideline in treatment of right anomalous coronary artery from opposite sinus.

Introduction

Coronary artery anomalies (CAAs) are still becoming a noteworthy matter to discuss in basic and clinical practice. There is extensively discussed in the literature regarding definition of CAAs, especially in how it can be different with normal variant of the coronary artery. However, the “normal” terminology in CAA means commonly observed and more frequent variant (normal variant) and the “abnormal” terminology comes from less than 1% of the general human coronary course [1]. Normal course of CAA related to Valsalva sinuses in one point and dependent area of myocardium in the other point. Nevertheless, Angelini [2] had proposed assessable of the definition of the normal variant shown in the table 1.

Historically, coronary artery was studied by gross anatomic inspection during autopsy examination. It was provided adequate examination especially in locating ostia with respect to aortic root references structure, evaluating artery’s course, distal distribution, termination and also intrinsic anatomy features. Nowadays we has modern medical technology like catherization laboratory for diagnostic and treatment, however coronary angiography is not appropriate as a primary screening test for ruling out coronary anomalies. Otherwise, non-invasive clinical imagine techniques (like CT angiography) consider more safer, convenient for the patient, and suitable cost [3].

Table 1. Purpose definition of the normal heart variant conditions [2]

Feature	Range
Number of ostia	2 – 4
Location	Right and left anterior sinuses (upper mid-section)
Proximal orientation	45-90 degree off the aortic wall
Proximal common stem or trunk	Only left (left anterior descendent and left circumflex artery)
Proximal course	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 its incidence around 0.17%-1.2% [4]. Regardless its incidence, around 20% of the CAAs are malignant and could undergo life-threating manifestation such as lethal arrhythmias, syncope, myocardial infraction, or even sudden death [5]. There are four most common coronary anomaly; right coronary artery (RCA) arises from left coronary sinus, left coronary artery (LCA) originate from right coronary sinus, or LCA or RCA arising from the non-coronary sinus. However, RCA that originate from anterior might associate with acute coronary syndrome and CT-angiography could be better in diagnosing CAA than intraluminal coronary angiography [6]. Specifically, the incident of the malignant course of the CAAs from the right coronary artery origins is pretty rare, around 0.03-0.17% [7]. The clinical spectrum of CAAs is enormously extensive, from asymptomatic to resting ischemia. The awareness of atypical coronary anatomy could aid the cardiologist to give precise diagnosis and treatment, including catherization laboratory and surgical procedure [2]. Here we are presenting right malignant course of CAAs with symptomatic atypical chest pain in the young adult.

Case Presentation

A 34 years old man complained of having vague chest pain since more than six months ago. There were no risk factors (diabetes, hypertension, family history of sudden death or similar abnormality) or any past medical history that is significant to the symptom (chest pain dyspnea on effort, dyspnoea on effort, dyspnea at rest, syncope, palpitation either at rest or during activity). Physical activity was within normal limit and he had good physical fitness. There is no remarkable result from routine blood examination and rest electrocardiography (ECG). However, during the treadmill test exercise, it showed ST depression in lead VI-VII although there is no chest pain. The patient was then 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). We used these parameters: 120 KVp, 800 mAs, rotation time: 0.4 s, and collimation: 64 × 0.625mm. 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 at 0.5 mm increment.

CT coronary angiography showed at the right coronary artery originated from the left coronary sinus, coursing between the aortic root and pulmonary artery (Figure 1, a). The rest of the coronary arteries, left coronary artery and its branches, have normal course (Figure 1, b and c).

The calibre of RCA was small with no sign of stenosis in the RCA. The RCA routes through right atrioventricular sulcus and vascularized the right part of the heart. The proximal one distributed through ascending aorta and pulmonary artery. According to the normal course of the rest coronary artery's courses and no stenosis was found, we conclude CAA was the most possible aetiology from the ischemic sign and symptoms. However, we not yet decide the treatment for this patient and still need follow up for reassurance.

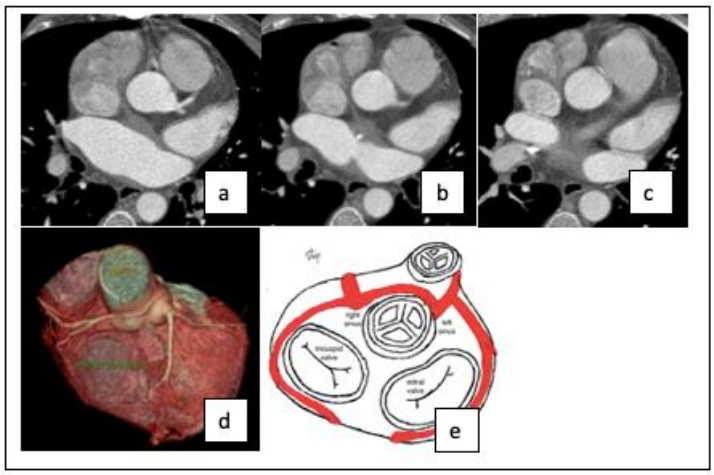

Figure 1. CT coronary angiography and schematic figure of malignant RCA. a-c. Maximum intensity projection of the heart showing RCA and LCA originating from LCS. RCA courses between PA and Aorta. d. Volume rendered image showing malignant RCA of this patient with interarterial course of RCA. e. Schematic figure of malignant course of RCA originate from left sinus. RCA: right coronary artery; LCA: left coronary artery; PA: pulmonary artery.

Discussion

The CAAs are frequently found as cause of the sudden death (SD) case in the young. Even though it is congenital condition, many subject survive asymptomatic until young adult. Previous coronary angiography study found that 0,95% people with range 26-68 years old had CAA and RCA was the commonest anomalous vessel (48.74%) [8]. However, anomalous coronary artery (CA) origin, either left main artery from right sinus or vice versa, has been found 0.17% during autopsy [7]. Specific incidence of RCA originate from left sinus is 0.019-0.49% [9]. Even-though most CAA case is benign, but it could potentially become catastrophic [3,9]. Concerning clinical manifestation about RCA that originate from contralateral sinus are sudden death (SD), myocardial ischemia, arrhythmia, and syncope [10]. Multi-detector CT (MDCT) is preferred method in diagnose anomalous RCA from the left coronary sinus with inter-arterial course.

There are two common types of clinical picture from anomalous origin of RCA from left sinus, first one is sudden death in the young after persistent physical activity and the other is atypical clinical picture of chest pain [10,11]. In some occasion, the only clues for anomalous coronary artery from the opposite sinus (ACAOS) either positive sign of ischemic due stress test or resting electrocardiogram (ECG) [10]. The clinical picture could be as accumulation of several patho-mechanical condition such as acute take-off angle, slit-like orifice, and compression of the intramural segment by the aortic valve commissure thought to narrow the orifice [9]. The anomalous origin of RCA from opposite Valsalva

doesn't change its function and nomenclature of the coronary artery [3]. However inter-arterial course, coronary artery course runs between aorta and pulmonary artery, might associated with ischemic episodes. During high intensity activity, especially in young people or athletes who has capability aorta's dilatation, the aorta and pulmonary could compress the coronary artery [12]. Meanwhile, RCA has responsibility to nutritive free wall of right ventricle.

One clinical study of 42 consecutives right ACAOS study shows that 69% are male with 57% has non-exertional chest pain symptoms [13]. From this study we know that fractional flow reserve (FFR) in right ACAOS is lower compare to normal course of coronary artery; 0.90 vs 0.98 respectively. More extensively, this study also elaborate about hypoplasia in intramural segment relative to distal vessel and luminal narrowing from intravascular ultrasound (IVUS) are the important cause of symptomatic (myocardial ischemic) right ACAOS. Histologic examination proposes that chronic ischemia as the one principal mechanism of myocardial fibrosis in CAA [14]. Other mechanisms that might be involved such as; artery intussusception, coronary hypoplasia and lateral compression of the coronary wall by the aorta [15]. Furthermore, myocardial ischemic could leads into malignant ventricular arrhythmias, supraventricular tachycardia, and Takotsubo in right ACAOS [16]. Unfortunately, management of right ACAOS is still become debatable issue [17].

Conclusion

In conclusion, the CAAs condition are a rare situation, especially right ACAOS. However, cardiologist need to be aware with congenital anomalies because it might help in clinical practice. There are some imaging modalities in diagnosis CAA, however CT angiography might be preferred. Even though right ACAOS might concealed for long period of time, but it might potentially lethal. Until now, there is no rigid guideline for management of the right ACAOS condition. Well plan treatment in inter-specialist team collaboration should be done.

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References

1. Villa ADM, Sammut E, Nair A, Rajani R, Bonamini R, Chiribiri A. Coronary artery anomalies overview. World J Radio. 2016;8(6):537-55.
2. Angelini P. Coronary artery anomalies: an entity in search of an identify. Circulation. 2007;115:1296-1305.
3. Angelini P, Villason S, Chan AV, Diez JG. Normal and anomalous coronary arteries in humans. In; Angelini P. Coronary artery anomalies; a comprehensive approach. Philadelphia: Lippincott Williams & Wilkins; 1999. p.27-79.
4. Angelini P. Coronary artery anomalies – current clinical issues: definitions, classification, incidence, clinical relevance, and treatment guideline. Tex Heart I J. 2002;29:271-8.
5. Graidis C, Dimitriadis D, Karasavvidis V, Dimitriadis G, Argyropulou E, Economou F, et al. Prevalence and characteristic of coronary artery anomalies in an adult population undergoing multidetector-row computed tomography for the evaluation of coronary artery disease. BMC Cardiovasc Disord. 2015;15:112.
6. Hothi SS, Ali A, Cox D. Acute coronary syndrome with a malignant course of an

unobstructed coronary artery. Heart. 2013;99:1880.

7. Datta J, White CS, Glikeson RC, Meyer CA, Kansal S, Jani ML, et al. Anomalous coronary arteries in adults: depiction at multi-detector row CT angiography. Radiology. 2005;235(3):812-8.
8. Garg N, Tewari S, Kapoor A, Gupta DK, Sinha N. Primary congenital anomalies of coronary arteries: a coronary: arteriographic study. Int J Cardiol. 2020;74(1):39-46.
9. Lee BY. Anomalous right coronary artery from the left coronary sinus with interaterial course: is it really dangerous? Korean Circ J. 2009;39(5):175-9.
10. Setianto BY, Hartopo AB, Gharini PPR, Taufiq N. Anomalous origination of right coronary artery from left sinus in asymptomatic young male presenting with positive ischemic response on treadmill test. Case Rep Cardiol. 2016;2016:4.
11. Narayanan SR, Shamkhani WA, Rajappan AK. Anomalous origin of RCA from left coronary sinus presenting as PSVT and recurrent acute coronary syndrome. Indian Heart J.2016; 68 (2):208-10.
12. Tsujita K, Maehara A, Mintz GS, Franklin-Bond T, Mehran R, Stone GW, et al. In vivo intravascular ultrasonic assessment of anomalous right coronary artery arising from left coronary sinus. Am J Cardiol. 2009; 103(5):747-51.
13. Lee SE, Yu CW, Park K, Park KW, Suh JW, Cho YS, et al. Physiological and clinical relevance of anomalous right coronary artery originating from left sinus of valsava in adults. Heart 2016;102(2):114-9.
14. Penalver JM, Mosca RS, Weitz D, Phoon CKL. Anomalous aortic origin of coronary arteries from the opposite sinus: critical appraisal of risk. BMC Cardiovasc Disord. 2012;12:83.
15. Angelini P, Walmsley RP, Libreros A, Ott DA. Symptomatic anomalous origination of the left coronary artery from the opposite sinus of valsava. Clinical presentation, diagnosis, and surgical repair. Tex Heart Inst J. 206;33(2):171-9.
16. Driessen BW, Warmerdam EG, Sieswerda GJT, Schoof PH, Meijboom FJ, Haas F, et al. Anomalous coronary artery originating from the opposite sinus of Valsalva (ACAOS), fractional flow reserve- and intravascular ultrasound guided management in adult patients. Catheter Cardiovasc Interv. 2018;92(1):68-75.
17. Barriales-Villa R, de la Tassa CM. Congenital coronary artery anomalies with origin in the contralateral sinus of Valsava: which approach should we take? Rev Esp Cardiol. 2006;59(4):360-70.