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Incidentally Diagnosed Malignant Coronary Artery Anomaly: A Clinical Case

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Key Words: *congenital anomaly; left main coronary artery; right coronary sinus; coronary artery.*

Summary. *The incidence of congenital coronary artery anomalies is estimated to range between 1% and 2% in the general population. The separate types of coronary artery anomalies are even rarer – the left main coronary artery arising from the right coronary sinus and passing between the thoracic aorta and the pulmonary artery is one of them. In this case, the segment of the artery that courses between the aorta and the pulmonary artery is prone to compression, especially during heavy exercise. Outcomes may be fatal due to myocardial hypoperfusion, which is associated with sudden cardiac death especially among children, young adults, and athletes. Nowadays, innate coronary artery anomalies may be incidentally diagnosed in older age using new investigation methods such as computed tomography angiography.*

Introduction

A coronary artery originating from the atypical sinus of Valsalva is a rare innate anomaly. The left main coronary artery (LMCA), the left anterior descending branch, or the circumflex branch may arise from the right sinus of Valsalva. The right coronary artery (RCA) may also originate from the left sinus of Valsalva. Seldom, coronary arteries may arise from the noncoronary sinus of Valsalva. The LMCA arising from the right coronary sinus is the most common type and is associated with sudden cardiac death if it passes between the aorta and the pulmonary artery (1). Basso has considered that the LMCA originating from the right cusp and crossing between the aorta and the pulmonary artery is more dangerous than the RCA taking a similar path because of the larger amount of the myocardium of the left ventricle being at ischemic risk (2). This anomaly occurs in 1 of 12 500 people and has been reported to have a recognized association with ischemic symptoms and sudden death in more than 50% of patients. This risk is thought to be greatest during or just after exercise, especially among children and young adults (1). Here we report a clinical case of an innate coronary artery anomaly in a female patient.

Case Report

A 67-year-old woman presented to the outpatient department of the Clinic of Cardiology, Hospital of Lithuanian University of Health Sciences, on January 20, 2013. Her main complaints were palpitations and retrosternal chest pain, originating from the left side of the chest and irradiating to the right side. Pain developed on exertion and lasted approximately for 45–60 minutes. The patient had been experiencing pain for 6 months. However, the patient did not take any medications to relieve pain. She had a history of hypertension, type 2 diabetes mellitus, and hypothyrosis.

On hospital admission, her height was 1.63 m; weight, 78 kg; and BMI, 30 kg/m^2. Her blood pressure was 150/90 mm Hg with a heart rate of 80 beats per minute (bpm). An examination of the heart revealed no visible abnormalities. An electrocardiogram (ECG) showed a sinus rhythm with premature ventricular contractions, mild ST-segment depression in the inferior leads (II, III, and aVF), and T-wave inversion in leads III and aVF.

A transthoracic echocardiogram revealed mild concentric left ventricular hypertrophy (myocardial mass index, 116.4 g/m^2) and normal global systolic function (ejection fraction of 55%). No wall motion abnormalities were detected. Grade 1 diastolic dysfunction was documented. The right ventricle was mildly enlarged with normal systolic function. Mild mitral and tricuspid regurgitation was documented.

The patient underwent the bicycle ergometer stress test, the results of which were equivocal. There were mild ST-segment depression of 0.5 mm in leads V5 and V6 at maximal exertion and several premature ventricular contractions. The patient did not experience any chest pain or shortness of breath, and there were no signs of cardiac dysfunction.

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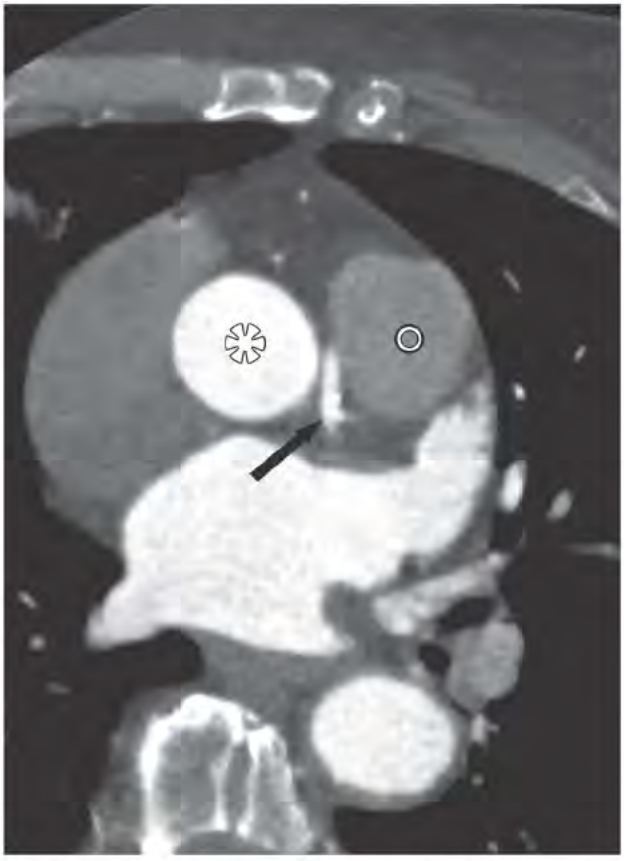

Fig. 1. Computed tomography angiography (axial view) Aorta, pulmonary artery, left main coronary artery passing between the aorta and the pulmonary artery.

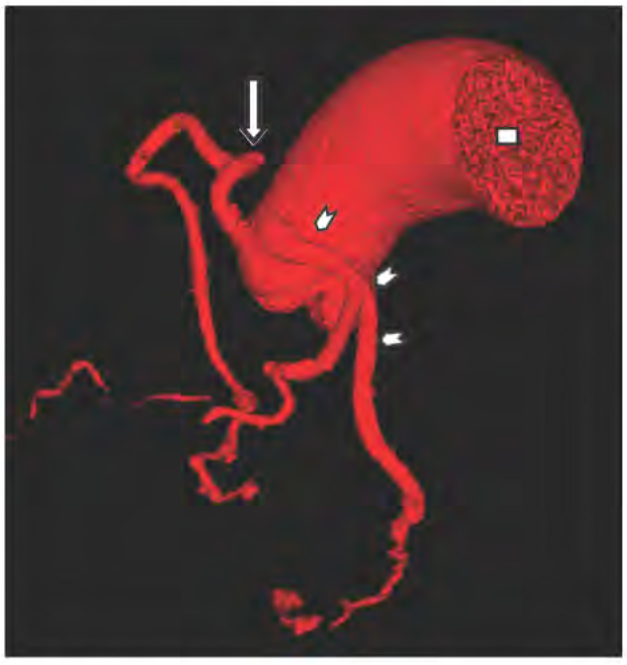

Fig. 2. Computed tomography angiography 3D reconstruction of the coronary arteries and the ascending aorta

Aorta, right coronary artery, the trunk of the left coronary artery passing close to the anterior wall of the ascending aorta.

Due to an intermediate probability of coronary artery disease (Forrester 29.5%), the cardiologist decided to perform computed tomography (CT) coronary angiography. It was performed with a 320-slice CT scanner (Toshiba Aquillion One) with single heart beat acquisition. CT revealed right coronary blood flow with a malignant coronary artery anomaly – the trunk of the left coronary artery branching from the right coronary sinus (with a separate ostium) and passing between the pulmonary trunk and the ascending aorta. No significant stenosis (>50%) was found in the coronary arteries (Figs. 1–3).

The patient consulted the cardiologist, and she was advised to avoid strenuous exercise and to ensure her blood pressure within the reference range. She was also prescribed the following medications: metoprolol (100 mg/day), acetylsalicylic acid (100 mg/day), and trimetazidine (35 mg, 2 times a day).

Discussion

The incidence of congenital coronary artery anomalies is estimated to be between 1% and 2% in the general population (3). They are usually diagnosed incidentally during coronary artery angiography, CT coronary angiography, or postmortem evaluations. An anomalous path of coronary arteries is observed in 0.3%–1.3% of patients undergoing

Fig. 3. Computed tomography angiography 3D reconstruction of the coronary arteries, the pulmonary artery, and the ascending aorta

Aorta; pulmonary artery; the trunk of the left coronary artery branching from the right coronary sinus (with a separate ostium) and passing between the pulmonary trunk and the ascending aorta.

routine coronary angiography and approximately 1% of cases of autopsies (4); 0.5% of the detected anomalies have the highest-risk lesions of the LMCA or the left anterior descending branch originating from the opposite sinus of Valsalva (5).

Recently, ECG-gated multidetector CT has emerged as a more accurate and noninvasive imaging method that allows depicting the origin and the course of coronary arteries. Cademartiri et al. estimated the prevalence of coronary artery anomalies to be as high as 18% in their study population of 543 patients who underwent 64-slice CT coronary angiography (6). Bazzocchi et al. found the incidence to be 5.7% in their study population of 3236 patients imaged with 64-slice CT as well (7).

Anomalies are often diagnosed retrospectively because the diagnosis of these malformations by routine screening is difficult. However, it is important to detect anomalies as soon as possible, because some of them may be associated with potentially serious events. Congenital coronary anomalies account for approximately 15% of sudden cardiac deaths in athletes (4). Moreover, in the United States, the anomalous aortic origin of a coronary artery is the second leading cause of sudden cardiac death in young individuals (1).

Coronary artery anomalies include the abnormalities of number, origin, course, termination, or structure (8). They may be benign or malignant. The latter predetermine a person to higher cardiovascular risk, early ischemia, and infarction. Malignant anomalies are far less common than benign ones and include the following:

1) Malignant RCA. It occurs when the RCA originates from the left aortic cusp and passes between the aorta and the pulmonary artery.

2) Malignant LMCA. It occurs when the LMCA originates from the right cusp and passes between the aorta and the pulmonary artery.

3) An anomalous origin of the left coronary artery from the pulmonary artery, also known as Bland-White-Garland syndrome. In this condition, reverse blood flow is observed in the LMCA as the blood enters the pulmonary artery due to low pulmonary resistance.

4) A coronary artery fistula is also one of the malignant anomalies. A fistula may form between any coronary artery and a cardiac chamber or the systemic or pulmonary circulation. Due to coronary steal, the myocardium suffers from hypoperfusion. Fistulae are more frequent between the RCA and the right ventricle, the right atrium, or the coronary sinus (9).

In patients with the anomalies of an origin of coronary arteries, pathophysiological mechanisms that lead to ischemia and sudden death are multifactorial. Based on autopsy studies involving patients with an anomalous aortic origin of a coronary artery, sudden cardiac death is attributed to the decreased flow of blood through the anomalous coronary artery (1). Hypoperfusion then results in myocardial ischemia and/or ventricular tachyarrhythmias. This diminished flow of blood may be caused by a number of anatomical malformations of coronary arteries, including acute angulation of the coronary artery originating from the aorta and creating a "slit-like" opening that easily collapses. The presence of an ostial ridge can diminish the flow of blood as well. An interarterial, intramural, or intraconal course of the proximal coronary segment leads to compression between the aorta and the pulmonary artery or within the aortic wall.

The clinical expression of a coronary artery anomaly may be arrhythmia, left ventricular dysfunction, syncope, or chest pain induced by exercise, and even sudden death (5).

Coronary angiography is one of the investigation methods for diagnosing coronary artery anomalies. However, the precise evaluation of the course of arteries is difficult as catheter angiography provides only a 2-dimensional view and a limited number of imaging planes (3). Moreover, it is an invasive technique that carries quite a few risks for the patient and cannot be used as a screening method.

Angiography can be supported by other imaging modalities including CT. Multidetector CT angiography is very accurate in depicting coronary artery anomalies as it has very good spatial resolution and rapid acquisition. CT also has the advantage of visualizing the origin as well as the course of a coronary vessel. It can also help determine its relationship to adjacent structures (such as the pulmonary trunk and the aorta). Small-thickness sections permit volumetric reconstructions of high quality. ECG gating allows minimizing or eliminating the effects of cardiac pulsation, therefore resulting in fewer artifacts (10). Using a 320-multislice CT scanner, the entire heart can be captured in one rotation for coronary analysis with a dose of about 1 mSv. Cardiac arrhythmia or heart rates up to 130 bpm are no longer an issue as opposed to helical and multislice CT for evaluating the anomalies of coronary arteries. In this way, the contraindications include only potentially nephrotoxic contrast agents and allergic reactions to them.

Magnetic resonance (MR) angiography is a noninvasive, radiation-free imaging modality. It is very accurate in clearly identifying the proximal anatomy of coronary arteries, which is usually unclear during catheter angiography. However, MR imaging has inferior spatial resolution compared with that of new-generation CT scanners. In addition, it cannot be performed in patients who have MR-unsafe pacemakers or defibrillators. ECG gating may also

be difficult to achieve in patients with certain arrhythmias.

At present, the American Heart Association Task Force on Practice Guidelines recommend coronary CT or MRI for a more definitive detection of the coronary artery course in persons suspected of having an anomalous origin of the coronary artery, especially in those younger than 50 years. These include individuals who have survived unexplained aborted sudden cardiac death or life-threatening arrhythmia, coronary ischemic symptoms, or left ventricular dysfunction (5). The accurate diagnosis of the anomalies of coronary arteries and the visualization of their course is essential in order to avoid fatal outcomes.

To date, the treatment of patients with anomalous coronary arteries remains a controversial subject. Some clinicians recommend observation and exercise restriction, while others suggest medical therapy (essentially with beta-blockers) and/or surgical repair (11). Surgical approaches include a direct repair of the coronary artery of an anomalous origin from the aorta or coronary bypass grafting. Coronary angioplasty is technically difficult, with only isolated cases reported in literature. However, to date, there are no data concerning the long-term follow-up. Coronary bypass grafting is increasingly viewed as a less favorable approach as it creates the potential for competitive flow (5).

Surgical repair is indicated when the left coronary artery arises from the opposite sinus and courses between the aorta and the pulmonary artery, and if a patient of any age presents with signs and/or symptoms of myocardial ischemia. On the other hand, the treatment of asymptomatic patients who are diagnosed with coronary anomalies incidentally is unclear. Currently, there is no system in order to adequately stratify risk to predict which patient is at higher risk for sudden cardiac death, based solely on the anatomy. Therefore, there is a critical gap in knowledge about how to treat patients and how to balance the risk of intervention against that of medical treatment.

Conclusions

The coronary artery originating from the atypical sinus and passing between the aorta and the pulmonary artery is a rare innate anomaly. The left main coronary artery arising from the right coronary sinus is a rare congenital coronary artery anomaly and is associated with the risk of sudden cardiac death. As demonstrated in our case, it can be diagnosed incidentally using new noninvasive investigation methods such as multislice computed tomography angiography with single heart beat acquisition.

Statement of Conflict of Interest

The authors state no conflict of interest.

References

1. Brothers JA, Gaynor JW, Jacobs PJ, Caldarone C, Jegatheeswaran A, Jacobs LM; Anomalous Coronary Artery Working Group. The registry of anomalous aortic origin of the coronary artery of the Congenital Heart Surgeons' Society. Cardiol Young 2010;20 Suppl:50-8.
2. Basso C, Thiene G. Congenital coronary artery anomalies at risk of myocardial ischaemia and sudden death: a report. Business Briefing: European Cardiology; 2005.
3. Mahani MG, Agarwal PP. Coronary artery anomalies on CT angiography. Appl Radiol 2011;6(4). Available from: URL: http://www.appliedradiology.com/Issues/2011/06/Articles/AR_06-11_Agarwal/Coronary-artery-anomalies-on-CT-angiography.aspx
4. Zenooz NA, Habibi R, Mammen L, Finn PJ, Gilkeson RC. Coronary artery fistulas: CT findings. Radiographics 2009; 29:781-9.
5. Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, et al. ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease). Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. J Am Coll Cardiol 2008;52:e143-263.
6. Cademartiri F, La Grutta L, Malago R, Alberghina F, Meijboom W, Pugliese F, et al. Prevalence of anatomical variants and coronary anomalies in 543 consecutive patients studied with 64-slice CT coronary angiography. Eur Radiol 2008;18:781-91.
7. Bazzocchi G, Romagnoli A, Sperandio M, Simonetti G. Evaluation with 64-slice CT of the prevalence of coronary artery variants and congenital anomalies: a retrospective study of 3,236 patients. Radiol Med 2011;116:675-89.
8. Majid Y, Warade M, Sinha J, Kalyanpur A, Gupta T. Superdominant right coronary artery with absent left circumflex artery. Biomed Imaging Interv J 2010;7:e2.
9. Rahalkar AM, Rahalkar MD. Pictorial essay: coronary artery variants and anomalies. Indian J Radiol Imaging 2009;19: 49-53.
10. Datta J, White CS, Gilkeson RC, Meyer CA, Kansal S, Jani ML, et al. Anomalous coronary arteries in adults: depiction at multi-detector row CT angiography. Radiology 2005; 235:812-8.
11. Angelini P. Coronary artery anomalies: an entity in search of an identity. Circulation 2007;115:1296-305.

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