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Chapter

Anomalous Origin of Coronary Arteries

Xhevdet Krasniqi and Hajdin Çitaku

Abstract

Coronary arteries supply the heart muscle with blood maintaining myocardial hemostasis and function. Coronary artery anomalies may persist after birth affecting cardiovascular system through haemodynamic impairment caused from shunting, ischaemia, especially in young children or adolescents and young adults. In patients undergoing coronary angiography the incidence of anomalous origination of the left coronary artery from right sinus is 0.15% and the right coronary artery from the left sinus is 0.92%. A recent classification of the coronary anomalies is based on anatomical considerations, recognizing three categories: anomalies of the origin and course, anomalies of the intrinsic coronary artery anatomy, and anomalies of the termination. In the setting of anomalous coronary artery from the opposite sinus, the proximal anomalous CA may run anterior to the pulmonary trunk (prepulmonic), posterior to the aorta (retroaortic), septal (subpulmonic), or between the pulmonary artery and the aorta itself (interarterial). Among them, only those with an interarterial aorta-pulmonary course are regarded as hidden conditions at risk of ischaemia and even sudden death. We presented two cases with anomalous origin of coronary arteries from opposite sinus, and two other cases with anomalous origin of left circumflex artery. The atherosclerotic coronary artery disease leads to the need of coronarography which can find out the presence of coronary artery anomalies. Anomalous origin of coronary artery that is present with atherosclerotic changes continues to exist as a challenge during treatment in interventional cardiology.

Keywords: coronary arteries, anomalous origin, opposite sinus

1. Introduction

Coronary arteries (CAs) are the blood vessels that supply the heart muscle with blood.

Intact coronary circulation is therefore important for myocardial hemostasis and function, thus enabling rest of body to function. The disruption of coronary development during embryogenesis results in coronary congenital defects such as coronary mispatterning, structural vascular defects, and anomalous communication of coronary vessels, they can alter coronary artery blood flow.

Such anomalies may persist after birth, occasionally they are in association with other cardiac conditions, so can severely affect cardiovascular system through haemodynamic impairment caused from shunting, ischaemia, or even sudden cardiac death, especially in young children or adolescents and young adults.
The estimated prevalence of CA anomalies is not quite clear with variable, ranging from 0.21 to 5.79% based on angiography, computed tomography (CT), and autopsy databanks [1]. Congenital anomalies of coronary arteries have in incidence about 1% in patients undergoing coronary angiography while the incidence of anomalous origination of the left coronary artery from right sinus is 0.15% and the right coronary artery from the left sinus is 0.92% [2, 3].

2. Coronary artery anatomy

In normal anatomy, the LAD and Cx originate from an aortic area located above the upper or middle third of the left coronary sinus of Valsalva (also called the left posterior sinus). The right coronary artery originates from the upper or middle third of the right sinus of the Valsalva. Normally, the coronary ostia lead an orthogonally oriented coronary proximal stem, off the aortic wall.

Of the many coronary arteries, the “primary” (or elementary) ones are defined as the three main proximal arteries: one provide circulation to the anterior septum and anterior lateral wall (the left anterior descending or LAD), another provides blood flow to the obtuse marginal region of the left ventricle (the circumflex, or Cx), and the third provides circulation to the free wall of the right ventricle (the right coronary artery or RCA). The left main trunk may serve as a common stem that joins the LAD and Cx (a common left main stem is present in about 90% of the cases and is not essential, but the LAD and CX are essential). Normally, the LAD and Cx originate from an aortic area located above the upper or middle third of the left coronary sinus of Valsalva (also called the left posterior sinus) [4]. In Figure 1 we presented normal origin of coronary arteries.

3. Embryology of coronary artery

Coronary artery formation is a process involving vasculogenesis, angiogenesis, and arteriogenesis. The vasculogenesis is a process through which is formed the
Anomalous Origin of Coronary Arteries
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early arterial coronary vascular system via the coalescence of the endothelial precursor cells (angioblasts), and subsequent fusion of the endothelial cell clusters [5].

Angiogenesis implies the generation of the new microvessels by endothelial proliferation and migration, mostly by means of controlled endothelial sprouting [6]. Coronary artery (CA) were originally thought to form by angiogenesis from the aortic root endothelium based on anatomical facts that ones join the systemic circulation at the aortic root, whereas cardiac veins connect to the general circulation via the coronary sinus [7].

Arteriogenesis describes the remodeling that form mature arteries by migration of supporting smooth muscle cells (SMCs) and pericytes from the epicardium during development [8, 9].

4. Congenital anomalies of the coronary circulation

Coronary anomalies are defined as those angiographic findings in which the number, origin, course and termination of the arteries are rarely encountered in general population. Coronary anomalies may occur in 1–5% of the patients undergoing coronary arteriography, depending on the threshold for defining an anatomic variant [10–12].

A recent classification of the coronary anomalies (Table 1) [1] is based on anatomical considerations, recognizing three categories:

a. anomalies of the origin and course;

b. anomalies of the intrinsic CA anatomy; and

c. anomalies of the termination [13, 14].

<table>
<thead>
<tr>
<th>I. Anomalies of origin coronary artery connection</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Anomalous origin to the aorta</td>
</tr>
<tr>
<td>a. Absent of left main trunk</td>
</tr>
<tr>
<td>b. Anomalous coronary artery ostium location</td>
</tr>
<tr>
<td>c. Anomalous coronary artery location at improper aortic sinus-wrong sinus:</td>
</tr>
<tr>
<td>- RCA to left sinus</td>
</tr>
<tr>
<td>- LCA to right sinus</td>
</tr>
<tr>
<td>- LCX to RCA/or sinus</td>
</tr>
<tr>
<td>- RCA or LCA to posterior sinus</td>
</tr>
<tr>
<td>with anomalous course: interarterial, prepulmonic, intraseptal, retroaortic, posterior atrioventricular groove or retrocardiac; postero-anterior interventricular groove</td>
</tr>
<tr>
<td>d. Single coronary artery</td>
</tr>
<tr>
<td>e. Anomalous coronary artery ostium location outside sinu-tubular aorta:</td>
</tr>
<tr>
<td>- LV</td>
</tr>
<tr>
<td>- Ascending aorta</td>
</tr>
<tr>
<td>- Aortic arch</td>
</tr>
<tr>
<td>Others (innominate artery; right carotid artery; internal mammary artery; bronchial artery; subclavian artery; descending thoracic aorta)</td>
</tr>
</tbody>
</table>
Coronary Artery Disease

4.1 Anomalous pulmonary origin of the coronary arteries (APOCA)

This syndrome is characterized by the origin of the coronary artery arising from the pulmonary artery. The most variant is an anomalous origin of the LCA from the pulmonary artery. (ALCAPA) [15, 16] although single-vessel origins of the RCA, LCx coronary, or LAD artery from the pulmonary artery have also been reported. If untreated, and in the absence of an adequate collateral network, most (95%) infants with APOCA will die within the first year. In the presence of an extensive collateral network, patients may survive into adulthood.

Aortography reveals a large RCA with absence of a left coronary ostium in the left aortic sinus of Valsalva, and with LAD and Cx branches filling through collateral circulation from the RCA branches. Still very delayed in filming sequence retrograde flow from LAD and LCx opacifies the LMCA and its origin from the main pulmonary artery. Still later in the filming sequence retrograde flow from the LAD and LCx arteries opacifies the LMCA and its origin from the main pulmonary artery. Once it is diagnosed, CABG surgery is recommended because of the high incidence of sudden death, cardiomyopathy and arrhythmias associated with APOCA.

Table 1.
Classification of coronary artery anomalies.

<table>
<thead>
<tr>
<th>2. Anomalous origin to the pulmonary artery</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. LCA to posterior facing sinus (ALCAPA)</td>
</tr>
<tr>
<td>b. LCX to posterior facing sinus</td>
</tr>
<tr>
<td>c. LAD to posterior facing sinus</td>
</tr>
<tr>
<td>d. RCA to anterior right facing sinus</td>
</tr>
<tr>
<td>e. Ectopic connection (outside facing sinuses) of any CA to PA left sinus, trunk, or branch</td>
</tr>
<tr>
<td>f. RV</td>
</tr>
</tbody>
</table>

II. Anomalous intrinsic coronary artery anatomy

1. Congenital ostial stenosis or atresia (LCA, LAD, RCA, Cx)
2. Coronary ostial dimple
3. Coronary ectasia or aneurysm
4. Absent coronary artery
5. Coronary hypoplasia
6. Intramural coronary artery (myocardial bridge)
7. Subendocardial coronary course
8. Coronary crossing
9. Anomalous origination of posterior descending branch or septal penetrating branch
10. Absent PD or split RCA
11. Absent or split LAD
12. Ectopic origination of first septal branch

III. Coronary artery interaction

1. Inadequate arteriolar/capillary ramifications
2. Fistulae from RCA, LCA, or infundibular artery to: RV, RA, coronary sinus, superior vena cava, PA, PV, LA, LV, multiple

4.1 Anomalous pulmonary origin of the coronary arteries (APOCA)
4.2 Anomalous coronary artery from the opposite sinus (ACAOS)

Anomalous origin of either the RCA to the left coronary sinus or the LCA to the right coronary sinus, the proximal anomalous coronary artery (CA) may run anterior to the pulmonary trunk (prepulmonic), posterior to the aorta (retroaortic), septal (subpulmonic), or between the pulmonary artery and the aorta (interarterial). Only those with an interarterial (aorta-pulmonary) course can increase risks of myocardial ischemia, arrhythmia, syncope, and sudden cardiac death considering life threatening and clinical guidelines recommend surgical correction [17].

Numerous mechanisms of ischaemia particularly during exercise have been suggested: (1) the compression of the anomalous vessel coursing between the aorta and the pulmonary artery during increased cardiac output and expansion of the great vessels; (2) the acute angle takeoff of the anomalous vessel with further stretch during exercise, possibility accounting for a flap-like closure of the coronary ostium; (3) spasm or kinking of the anomalous vessels; and (4) the course within the aortic wall (“intramural”) of the proximal segment of the anomalous vessel [13, 17]. The intramural aortic course can explain the imaging feature (angiography and echo) of CA intussusceptions into the aortic wall: the proximal segment of the anomalous vessel (segmental hypoplasia) is narrowed, and the asymmetrical lateral compression of the anomalous vessel with a silt-like or ovoid rather than circular lumen, particularly during systole and stress.

In Figure 2 (A and B) we present anomalous Coronary Artery from the Opposite Sinus (ACAOS).

4.3 Coronary artery fistulas

Coronary artery fistulas are defined as abnormal communications between a coronary artery and a cardiac chamber or major vessel, such as to the vena cava, right or left ventricle, pulmonary vein or pulmonary artery [18, 19]. Coronary artery fistulas are rare findings, identified in 10 (0.05%) of 18,272 diagnostic cardiac catheterizations [20].

4.4 Myocardial bridging

The three major coronary arteries generally course along the epicardial surface of the heart. On occasion, however, short coronary artery segments descend into the myocardium for a variable distance. This abnormality, termed myocardial bridging occurs in 5–12% patients and usually is confined to LAD [21].

5. Clinical presentation

5.1 Anomalous pulmonary origin of the coronary arteries (APOCA)

Coronary steal syndrome results where an alteration of circulation patterns leads to a reduction in the blood directed to the coronary circulation. The low pressure in the pulmonary artery causes blood from the abnormal LCA to flow towards the pulmonary artery instead of towards the heart resulting in ischaemia and collateral growth.

The extent of the acquired circulation between the two CAs is the major determinant of the degree of ischaemia, severity of clinical presentation, and outcome. Depending collateral vessels clinically are presented: a) the adult-type with well-established collateral vessels, and b) the infant-type without or with few collaterals, with early onset of symptoms when pulmonary arterial pressure decreases [22].
5.2 Anomalous coronary artery from the opposite sinus (ACAOS)

Origin of LCA from the proximal RCA or the right aortic sinus with subsequent passage between the aorta and the right ventricular outflow tract has been associated with sudden death during or shortly after exercise in young persons.

The increased risk of sudden death may be due to a silt-like ostium, a bend with acute takeoff angles of the aberrant coronary arteries, or arterial compression between the pulmonary trunk and aorta when there is increased blood flow through these vessels with exercise and stress.

The RCA originated from the LCA or left aortic sinus with passage between the aorta and right ventricular outflow tract is also associated with myocardial ischemia and sudden death [23]. In rare cases of the LCA originated from the right sinus myocardial ischemia may occur even if the LCA passes anterior to the right ventricular outflow tract or posterior to aorta, not through a tunnel between the two great vessels [24].

The revascularization approach in patients with ACAOS has been CABG surgery, although coronary stenting has been reported with acceptable medium term success.

5.3 Coronary artery fistulas

The clinical presentation associated with coronary artery fistulas is dependent on the type of fistula, shunt volume, in situ of the shunt and presence of other cardiac
conditions, although patients (50%) often remain asymptomatic [25]. Dyspnea on exertion, fatigue, congestive heart failure, pulmonary hypertension, bacterial endocarditis and arrhythmias are common presentations in symptomatic patients. Myocardial ischaemia may also occur, but the mechanism remains speculative [25]. Symptomatic patients or those with severe shunts may be treated with surgical closure, although percutaneous closure with coil embolization may also be tried.

5.4 Myocardial bridging

A myocardial bridge occurs when one of the coronary arteries takes a tunneled intramuscular course under a bridge of overlying myocardium. The myocardial fibers pass over the involved segment of the LAD, and each contraction of these fibers can cause narrowing of the artery. On angiography, the bridged segment is of normal caliber during diastole and abruptly narrows with each systole.

Although bridging is not thought to have any hemodynamic significance in most cases, myocardial bridging has been associated with angina, arrhythmia, depressed left ventricular function, myocardial stunning, early death after cardiac transplantation, and sudden death [21, 26]. Intracoronary Doppler studies have shown that diastolic flow abnormalities may be present in patients with myocardial bridging. Medical treatment generally includes beta blockers, although nitrates should be avoided because they may worsen symptoms. Intracoronary stent and surgery have been attempted in selected patients, but the results have been mixed.

5.5 Percutaneous coronary intervention and anomalous coronary artery

Several problems may be encountered during the angiography and angioplasty of anomalous origin of culprit coronary artery (AOCCA), including precise diagnosis, selection of an appropriate guiding catheter, insufficient backup force, and difficulties in balloon or stent delivery. The final success of the procedure is depended from the careful assessment of the AOCCA configuration, proximal angulation, vessel course and subsequent selection of an appropriate guide catheter and guide wire.

5.6 Femoral versus radial approach

In case of AOCCA femoral access may offer better options allowing for easy, and multiple catheter exchanges [27]. Although, in the setting of ACS, the operator is usually unaware of AOCCA presence, having to make the best use of the chosen access site. Also, it seems best to use the approach one is most comfortable with as there is usually a way to perform successful PCI of AOCCA regardless of access site.

5.7 Right versus left radial approach

In a meta-analysis of 12 prospective randomized trials comparing above-mentioned approaches there was a small but statistically significant difference in terms of contrast use and fluoroscopy time in favor of coronary procedures performed via left radial approach compared to the right radial approach, but without any difference in access site or other procedural complications [28].

5.8 Additional tools

Anchoring balloons or anchor wire techniques may be helpful tools [29]. The latter maneuver was used to treat one of the present patients. Still, this culprit was
not proven for AOCCA. Extension catheters, such as Guideliner or Guidezilla often allow for safe and stable intubation and facilitate stent placement.

6. Case presentations

6.1 Case 1

First case is a 62-year-old female patient hospitalized in our clinic due to chest pain with a history of arterial hypertension and diabetes mellitus. Cardiac biomarkers showed: serum creatinine kinase (CK) level of 82 IU/L, creatinine kinase-myocardial band (CK-MB) level of 33.6 IU/L, and troponin-T level of 684 ug/L. Electrocardiography (ECG) is characterized with ST segment depression in V1–V3. Transthoracic echocardiography (TTE) presented regional wall motion abnormality in the entire severely hypokinetic inferior wall.

The coronary angiography revealed the left coronary artery arising from the right coronary sinus sharing a same ostium with right coronary artery (Figure 3). The proximally and distally stenosed left anterior descending artery (LAD) (Figure 4) associates with calcified atherosclerotic medial and distal right coronary artery (RCA) stenosis (Figure 5).

6.2 Case 2

The second case is a 47-year-old male who presented to emergency department with chest pain. He also had a history of arterial hypertension and a positive history for ischemic heart disease. Cardiac biomarkers: serum creatinine kinase (CK), creatinine kinase-myocardial band (CK-MB), and troponin-T were not increased. Electrocardiography (ECG) showed atypical ST segment changes in leads V4–V6. Transthoracic echocardiography (TTE) did not present regional wall motion abnormalities.

A coronary angiogram showed an anomalous right coronary artery arising from the left Valsalva sinus from a separate ostia with the left coronary artery (Figures 6 and 7). Medial and distal segments of LAD were tortuous (Figure 8).
6.3 Case 3

The third case is 64-year-old man hospitalized to our clinic due to chest pain. Also, the patient was a smoker and had a history of arterial hypertension, obesity, and dyslipidemia. Biochemical parameters were: serum creatinine kinase (CK) level of 82 IU/L, creatinine kinase-myocardial band (CK-MB) level of 6.5 ng/mL, and troponin-I level of 0.1 ng/mL. Electrocardiography (ECG) is characterized with deep Q wave in inferior and V4–V6 leads with biphasic T in inferior and V3–V6 leads. In transthoracic echocardiography (TTE) is presented with regional motion abnormalities in the entire severely hypokinetic inferoposterior wall.

The patients underwent coronary angiography that revealed the LCx arising from the right coronary sinus (Figure 9). The mildly stenosed LCx coexists with a stenosed RCA.

6.4 Case 4

The second case is 67-year-old man presented to the emergency department with chest pain that had developed 6 h previously. That patient had a history of arterial hypertension for 10 years, diabetes mellitus type 2, and chronically hemodialyzed for 7 years. The laboratory findings showed a serum creatinine kinase (CK) level of 473 IU/L, creatinine kinase-myocardial band (CK-MB) level of 6.4 ng/mL,
and troponin-I level of 0.15 ng/mL. Electrocardiography demonstrated ST segment depression of 1–2 mm in leads V4–V6, and inverted T wave in D2, D3, aVF. The transthoracic echocardiography (TTE) revealed severely hypokinetic medioapical
segments of anterolateral wall, and hypokinetic basal segments of interventricular septum and inferior wall.

The coronary angiography revealed a left circumflex artery (LCx) as a proximal branch of the right coronary artery (Figure 10). The LAD contained an proximal lesion up to 80%. The LCx and RCA are occluded in medial segment.

7. Comments

The coronary angiography of patients with coronary ischemia determined atherosclerotic disease with possibility of the presence of coronary artery anomalies, but also coronary angiography may reveal coronary artery anomaly without the presence of atherosclerotic changes. The ectopic origin from opposite sinus of coronary artery anomalies that presents with atherosclerotic changes continues to exist as a challenge during treatment in interventional cardiology.

The atherosclerotic coronary artery disease leads to the need of coronarography which can find out the presence of coronary artery anomalies. We should think about these anomalies during coronarography knowing that based on type of these anomalies and considering the vulnerability to atherosclerosis will be determined the method of the treatment.
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