Coronary artery anomalies presenting with ST-segment elevation myocardial infarction

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Abstract

ST-segment elevation MI (STEMI) is a rare presentation in patients with coronary artery anomalies. In these patients, the identification of the culprit lesion and its treatment may be difficult, particularly in the emergency setting of primary percutaneous coronary intervention (PCI). From January 2008 to April 2011, 1015 STEMI patients received coronary artery angiography and primary PCI in our centre. Of these, 5 (0.4%) patients showed a coronary artery anomaly. In this paper we reported two rare cases: i) the first is a single coronary artery originating from right sinus of Valsalva; ii) the second is a separate origin of 3 coronary arteries originating from the right sinus of Valsalva. In conclusion, coronary artery anomalies presenting with STEMI are really uncommon, but often are a challenge. The integration between traditional coronary artery angiography and multidetector computerized tomography is crucial to optimize the interventional and medical management of these patients.

Introduction

Coronary artery anomalies are rare occurrences, with an incidence ranging from 0.17% in autopsy studies to 1.3% in angiographic series.[1-4] These anomalies are generally asymptomatic, but more potentially serious ones may lead to myocardial ischemia, myocardial infarction (MI) and/or sudden death. In particular, ST-segment elevation MI (STEMI) is a rare clinical presentation in patients with coronary artery anomalies. As suggested by current guidelines, all patients presenting with STEMI should receive emerging coronary artery angiography (CAA) and subsequent primary percutaneous coronary intervention (PCI) after culprit lesion identification. Unfortunately, in these patients, the identification of the culprit lesion and its treatment may be difficult and complex. From January 2008 to April 2011, 1015 STEMI patients received emergent CAA and primary PCI in our centre. Of these, 5 (0.4%) patients showed a coronary artery anomaly. In this paper we reported the clinical management, history and follow-up of two really rare cases, with a very uncommon coronary artery anatomy.

Case Reports

Case #1

A 61-year-old woman with no past medical history presented to the emergency department with typical chest pain lasting for 45 min. Hypertension is reported as only a cardiovascular risk factor. Her electrocardiogram (ECG) showed sinus rhythm, right bundle branch block and significant ST-segment elevation in V1-V3 leads. Thus, urgent CAA was immediately performed. CAA shows the presence of a single giant right coronary artery (RCA). RCA courses in all coronary sulcus and gives the posterior descending artery (encircling the apex), two posterolateral branches and three obtuse marginal branches. These vessels supply all RCA and left circumflex artery territory. From the first portion of RCA two small anomalous vessels arose (Figure 1). The first one (asterisk, Figure 1) goes into the middle portion of the anterior interventricular sulcus (AIS), sprouting diagonal branches. It shows an asymmetric stenosis (black arrow, Figure 1). The other one (arrow head, Figure 1) reaches the proximal AIS, giving side branches (Figure 1). During the procedure, spontaneous symptom relief and ST-segment resolution were observed. TIMI flow was 3 in all vessels. Thus percutaneous coronary intervention was not performed. After procedure, no recurrent angina was observed and medical therapy with aspirin, clopidogrel, ramipril, metoprolol and atorvastatin was titrated. A significant increase of myocardial necrosis markers (CK-MB peak 21.2 ng/mL and troponin I peak 0.91 ng/mL) above the upper normal range (5 ng/mL and 0.15 ng/mL, respectively) was observed. Four days after, a 256 slices ECG-gated multidetector computerized tomography (MDCT) study of coronary arteries was done to delineate the anatomy and the relationship with the great vessels. The presence of single coronary artery was confirmed (Figure 2). The MDCT study showed that the first anomalous vessel (asterisk, Figure 2) hurdles the pulmonary artery (PA) and supplies the medium portion of antero-lateral wall. On the contrary, the second anomalous vessel (arrow head, Figure 2) courses behind the PA and reaches the basal segment of antero-lateral wall. Curiously, these two anomalous vessels hug the PA and form a singular *horseshoe* variant of left anterior descending (LAD) artery (Figure 2C). Two days after the MDCT study, the patient was discharged and the 1 year follow-up was uneventful.

Case #2

A 71-year-old male with a history of hypertension and hyperlipidemia was admitted to our emergency department with the diagnosis of anterior-lateral STEMI. He complained about chest pain that he had for ≈2 h. A CCA was immediately performed and after a very complex exam the coronary artery anatomy was clarified. Three main coronary arteries originate all from the right sinus of Valsalva. The RCA is characterized by normal origin and distribution (Figure 3A). RCA presents a critical stenosis in the distal segment. Near the RCA ostium, the second coronary artery arises and reaches the mid portion of AIS (Figure 3B). This vessel shows a distribution and side branches similar to a *normal* mid-distal LAD and a lesion on proximal segment. Finally, only the first portion of a third vessel is visible (Figure 3A, red arrow). It's distal portion is visible via collateral branches arising from the LAD and supplies the territory of circumflex artery and of proximal LAD (asterisk, Figure 3B). With many difficulties, a PCI with only balloon was performed (it was impossible to deliver the stent for the lack of support). At the end of the procedure the final TIMI flow in the culprit vessel was 2 and a partial resolution of symptoms and of ST-segment elevation were observed. Medical therapy with aspirin, perindopril, metoprolol, nitrates and simvastatin was immediately titrated. No recurrent angina or complications were observed. Cardiac markers confirmed the necrosis (CK-

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MB peak 42.8 ng/mL and troponin I peak 1.4 ng/mL). Five days after, a MDCT study of coronary arteries was done (Figure 4). In particular, MDCT permitted to better clarify the relationship between great vessels and coronary arteries. One of the two anomalous vessels, to reach the mid-distal anterior wall, runs behind the pulmonary artery (asterisk, Figure 4). Contrarily, the other anomalous vessel with the culprit lesion shows a retro-aortic course (arrow head, Figure 4). Considering the residual stenosis in the three vessels, the impossibility to obtain a percutaneous revascularization of coronary artery with retro-aortic course, and the relationship between anomalous vessels and aorta and pulmonary artery, surgical revascularization is recommended to the patient. Eight days after surgical revascularization was performed without any complications. The six month follow-up was uneventful.

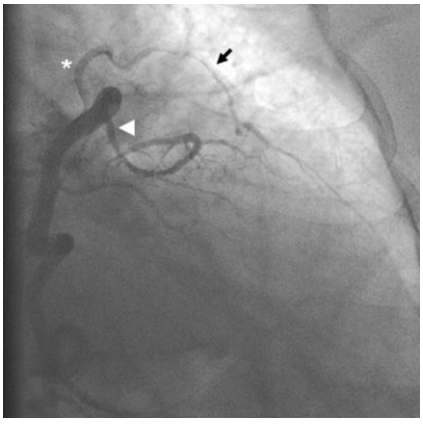

Figure 1. Case #1. Coronary artery angiography of single coronary artery. *Anomalous vessel for the middle portion of the anterior interventricular sulcus. Black arrow: probable culprit lesion. White arrow head: anomalous vessel for the proximal portion of the anterior interventricular sulcus.

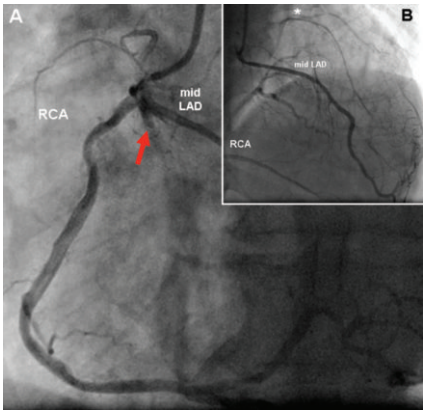

Figure 3. Case #2. Coronary artery angiography. Red arrow: culprit lesion. *collateral branches for distal segments of culprit vessel.

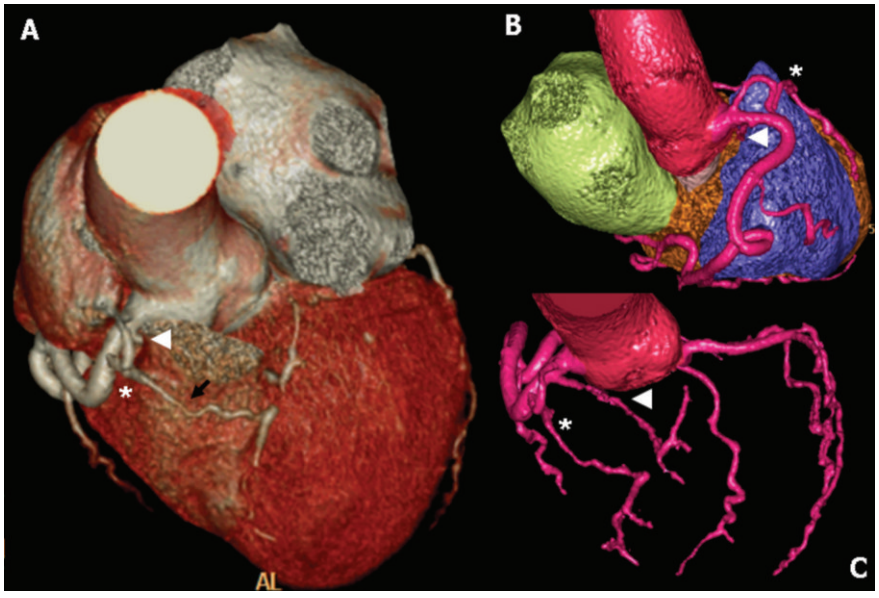

Figure 2. Case #1. 256-slice electrocardiogram -gated computed tomography of single coronary artery. *anomalous vessel for the middle portion of the anterior interventricular sulcus. Black arrow: probable culprit lesion. White arrow head: anomalous vessel for the proximal portion of the anterior interventricular sulcus.

Discussion

Coronary artery anomalies are the result of changes during the third week of fetal development. Their overall prevalence ranges from 0.3% to 1.3% in the previously reported angiographic series.[2-4] The coronary artery anomalies are classified into anomalies: i) of origin ii) of courses iii) of termination. These anomalies include numerous different types. Although their incidence is low, the potentially fatal consequences of coronary artery anomalies mean that it should always be taken into consideration by clinicians. As this is the second most common cause of sudden death in young adults, after hypertrophic cardiomyopathy.[5-6] The diagnosis can be challenging because patients are often asymptomatic and have a normal physical examination. Most patients are asymptomatic for a large portion of their lives, and an atypical chest-pain syndrome is the most common reason for which they are referred to for medical attention. The milder cases are more likely to be identified fortuitously (because of a falsely positive stress test and/or coincidental atherosclerotic disease). Coronary artery anomaly does not appear to be associated with an increased risk for the development of coronary atherosclerosis.[7] Some Authors reported an increase in stenosis of the anomalous LCx, but survival was not adversely affected within seven years.[8] Conventional CAA is regarded as the gold standard to diagnose coronary artery disease. However, identification of coronary artery anomalies is frequently difficult with conventional CAA. It is particularly difficult to be selective in the ostium of anomalous vessel, and for the lack of 3D information, the relationship between coronary arteries and great vessels remain unclear. In the emergency setting all these difficulties are enhanced. During primary PCI it is frequently difficult to identify the culprit lesion. When the course of anomalous vessel and the culprit lesion are clear, often the revascularization procedure hides several pitfalls. Frequently the lack of support by guiding catheter limits the possibility to cross culprit lesion with guide-wire and/or to deliver balloon and stent. Analysing the last four years of emergent activity of our cath-lab, we found five (0.4%) patients with STEMI and coronary artery anomalies. This data confirms that coronary artery anomalies are really rare and infrequent in daily clinical practice. Even in a high-volume centre with skilled interventional cardiologists (>60 primary PCI by year for operator), coronary artery anomalies presenting with STEMI are a complex occurrence. Of our five cases, two were really complex and in one patient (Case #2) the reperfusion obtained was partial, whereas in the other patient (Case #1) was luckily, spontaneous.

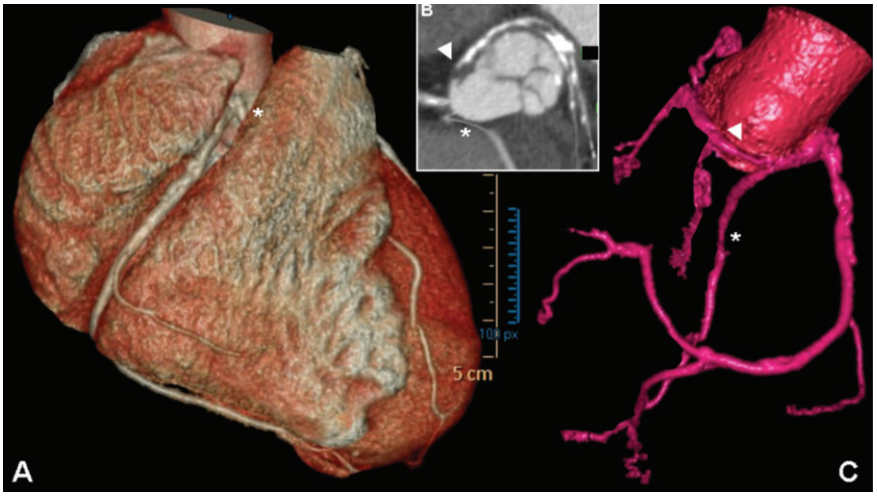

Figure 4. Case #2. 256-slice electrocardiogram -gated computed tomography showing the relationship between great vessels and coronary arteries and their distribution to myocardial tissue. *anomalous vessel arising from right coronary sinus and running behind pulmonary artery. Arrow head: anomalous vessel with retro-aortic course and culprit lesion.

However, considering the coronary anatomy of this last patient, it was very difficult to predict a successful mechanical intervention. In the first patient (Case #2) we observed a 3-vessel disease with separate origins of three coronary arteries originating from the right sinus of Valsalva. As far as we know this anomaly is very uncommon and there are only few reports previously published.[9-10] Additionally, the course of these anomalous vessels is peculiar, being one retro-aortic and the other between pulmonary artery and aorta. The second patient (Case #1) presents a single coronary artery. A single coronary artery is the rarest coronary anomaly and is an isolated finding that occurs in approximately 0.03-0.09% of the population, with a prevalence reported to be <3% of all major coronary anomalies.[2-4] Currently, the ideal imaging tool for the diagnosis and delineation of coronary artery anomalies is angiography supported by MDCT.[11-12] MDCT is particularly essential to clarify the relationship between coronary arteries and great vessels and the correct position of coronary arteries ostia. These informations are crucial to select the correct revascularization procedure (surgical *vs.* percutaneous) and to correctly plan the intervention, if PCI is preferred. In the primary PCI setting, the aim of each operator should be the reperfusion, also using only balloon angioplasty or thrombo-aspiration. Thereafter a MDCT study may be planned and the final strategy of revascularization may be chosen.

In conclusion, coronary artery anomalies presenting with STEMI are really uncommon, but often are a challenge. The integration between traditional CAA and MDCT is crucial to optimize the management of these patients.

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