Cardiovascular Revascularization Medicine 16 (2015) 362–366

Contents lists available at ScienceDirect

Cardiovascular Revascularization Medicine

Anomalous right coronary artery: case series and review of literature☆

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ARTICLE INFO

Article history:
Received 22 December 2014
Received in revised form 14 March 2015
Accepted 26 March 2015

Keywords:
Anomalous coronary artery
Percutaneous coronary intervention
Radial access approach

ABSTRACT

Anomalous right coronary arteries (ARCA) are extremely rare in general population. Although mostly asymptomatic and recognized incidentally on cardiac catheterizations, they can be catastrophic and can cause sudden cardiac death. Sudden cardiac deaths are more common when the anomalous vessel runs an inter-arterial course between the aorta and the pulmonary artery. Asymptomatic patients with malignant course of anomalous coronaries can pose clinical dilemmas. Based on prior experience, management of asymptomatic ARCA with malignant course should be subjected to a risk–benefit analysis.

This case series begins with a brief description of four separate cases of ARCA. They had their origin in the left coronary sinus or off left anterior descending artery (LAD). Three of them had anterior course between aorta and pulmonary trunk, confirmed by coronary CT angiography (CTA). Whereas two of our patients presented with chronic symptoms, two presented as acute cases with electrocardiographically proven STEMI. These cases were managed differently; by conservative, surgical or interventional approaches. All four cases had good final outcomes. This goes to show how different treatment options can be employed in management of complications associated with anomalous coronary arteries.

It is also interesting to note that the radial access provides better guide support that is needed to tackle complex lesions. Many operators have been using radial approach for anomalous coronary interventions. We have successfully employed radial technique after failed trans-femoral attempts and also in STEMI situations. Based on our experience, right radial approach appears to be safer and quicker.

1. Introduction

Anomalous right coronary artery (ARCA) is an uncommon congenital anomaly with varied clinical outcomes. Many of the coronary anomalies are asymptomatic, but there is also an association with serious outcome such as sudden cardiac death. We present four cases of ARCA, as well as a brief review of the literature.

2. Case 1

A 55-year-old male presented with exertional chest pain of two years duration. Physical examination and lab studies were unremarkable. Coronary angiography documented the presence of moderate LV dysfunction with EF of around 30–35%. Coronary angiogram performed via femoral approach showed single coronary artery with ARCA originating from LAD and non-obstructive CAD. Since the anomalous artery was detected during left coronary injection using JL4 catheter, no further attempts were made to identify a separate right coronary artery ostium. A non-selective aortogram was performed to confirm these findings. Coronary CT angiogram (CTA) was suggestive of right dominant circulation and anomalous RCA arising from LAD, coursing between aorta and pulmonary artery (Fig. 1). The caliber of the vessel in this region appeared to be narrow with more normal caliber distally. He was managed conservatively with suggestion to stop alcohol consumption. Since there was no significant improvement in LV systolic function despite optimal therapy, ICD was implanted. He has been doing well after two years of follow up.

3. Case 2

A 47-year-old male with hypertension and dyslipidemia presented with sudden onset of chest pain. Initial EKG showed minimal ST-elevation in the inferior leads. The patient was taken emergently to the catheterization lab in view of ongoing chest pain, although only subtle ST elevation was noted on the EKG. Emergent cardiac catheterization from femoral approach revealed an ARCA arising from left coronary sinus. Despite multiple attempts with different catheters (JR4 initially, followed by AL1 and multi-purpose catheters), there was difficulty in engaging the RCA ostium, and hence no intervention could be performed). However, follow up troponin levels were only minimally elevated (maximum value of 0.15 ng/ml). Echocardiogram showed normal LV systolic function with LVEF of 60% and no regional wall motion abnormalities. Coronary CT-angiography was subsequently done

☆ There are no conflicts of interest.

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http://dx.doi.org/10.1016/j.carrev.2015.03.006
1553-8389/

Fig. 1. Cardiac CT-scan of case 1. This coronary CT with 3D reconstruction shows narrowed segment of anomalous RCA (arrow) at the site where it is coursing between the aorta and pulmonary artery.

which showed an ARCA with tortuous takeoff from the left coronary sinus that coursed between the aorta and pulmonary artery (Fig. 2). There was 75% narrowing of the proximal segment in reference to the mid-segment of the vessel, but no plaques or obstructive lesions were present. The patient underwent a mini-sternotomy, unroofing of ARCA and aortic valve commissural suspension (commissure between left and right cusps of the aortic valve). Post-operatively, the patient was well and was symptom free on discharge and subsequent follow-up.

4. Case 3

A 73-year-old male presented with sudden development of chest pain while exercising. EKG showed ST-segment elevation in inferior leads, and the patient was taken directly for emergent cardiac catheterization. The procedure was performed using the radial access approach. Once the present anomalous right coronary artery was recognized during the initial injection of left coronary system using Tiger 4 guide catheter, we decided to switch out to AL1 guide catheter. Although ARCA

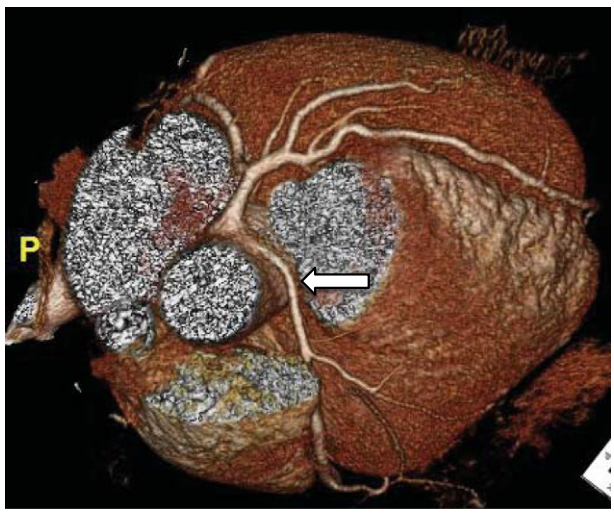

Fig. 2. Cardiac CT-scan of case 2. This cardiac CT image with 3D reconstruction demonstrates the interarterial course of anomalous RCA (arrow) between the root of pulmonary artery and aorta.

was engaged with AL1 guide catheter, it had to be changed out to a 6 F Barbeau guide catheter for better guide support during the intervention. During diagnostic angiogram, an ARCA with takeoff adjacent to the left coronary artery was noted with 99% stenosis in its mid-portion with thrombus (Fig. 3). Export catheter was used for thrombus aspiration. The stenotic segment of the ARCA was then stented. Echocardiogram performed on the following day demonstrated borderline LV systolic function with EF of 50–55%. No definite regional wall motion abnormalities were seen. Cardiac enzymes were within normal limits at admission (prior to the procedure) and increased to a maximum value of 4 ng/ml. Subsequent coronary CT-angiography revealed anomalous origin of the RCA from the left coronary sinus with an inter-arterial course (Fig. 4). The patient was well post-PCI with no in-hospital complications.

5. Case 4

A 66-year-old man presented with chest pain upon exertion. His past medical history was significant for stents in the LAD, left circumflex artery and RCA five years ago. During the previous cardiac catheterization six months earlier, femoral approach was employed. Although diagnostic images revealed anomalous origin of RCA from anterior aspect of the left coronary sinus, no intervention could be performed due to extreme technical difficulty secondary to inadequate guide support. When AL1 diagnostic catheter could not engage the anomalous vessel, attempts were made using various guide catheters such as JL4, JL 4.5, multi-purpose EBU 3.5 and AL3 guide catheters. Due to ongoing symptoms, the procedure was reattempted at a later date via right radial approach, and the focal lesions within the ARCA were stented easily (Fig. 5). A Tiger 4.5 catheter was used for diagnostic study, and 6 F Barbeau guide catheter was used for the intervention. There were no post-PCI complications, and he has been doing well on subsequent follow-up (Table 1). (See Fig. 6.)

6. Discussion

6.1. Epidemiology

Anomalous coronary artery is a rare clinical entity with varied clinical outcomes, ranging from a totally asymptomatic course to sudden cardiac death. This has been a subject of an array of clinical as well as autopsy studies so far. Studies from across the globe show a relatively constant incidence of coronary anomalies at around 1–1.5% [1], with many studies documenting right coronary artery (RCA) as the commonest anomalous coronary artery [2–4]. The prevalence of ARCA, as determined from coronary angiography studies, ranges from 0.06% to 0.5 [3,5].

6.2. Clinical presentations and pathophysiology

A variety of anomalous origins of RCA have been reported in literature, which include origin from descending thoracic aorta, left main coronary, left circumflex, above/from left sinus of Valsalva, the pulmonary arteries or even below the aortic valve [1,2,6–13]. After the anomalous take off, the artery may run anterior, posterior or in between the major vessels at the base of the heart, the last course constituting what is famously known as “malignant” type of anomaly due to risk of extrinsic compression and sudden death.

Most ARCA are asymptomatic and are seen as incidental findings on coronary angiography. They may also be responsible for a whole host of clinical manifestations that include angina pectoris, myocardial infarction or even sudden cardiac death. ARCA originating from the contralateral aortic sinus that runs between pulmonary artery and aorta has received much attention because of its association with sudden death [14–16]. However, the exact pathophysiological basis for such association is unclear. The most common theory is the mechanical compression of the ARCA, especially during exercise [1,13,17]. Other proposed mechanisms include presence of valve like ridges, acute angulation of the