Double Right Coronary Arteries Originated from Separate Ostia with Total Occlusive Lesion in One of the Two Right Coronary Arteries

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Double right coronary arteries (RCA) are very rare congenital anomalies of coronary artery. We report a case of double RCA with total occlusion, incidentally found by collateral flows. A 71-year-old patient underwent percutaneous coronary intervention of left coronary arteries for angina, and the presence of double RCA was missed at initial coronary angiography (CAG). About 20 months later, second CAG was performed due to recurrent angina, and the CAG showed newly developed collateral flow suggesting the presence of the other missed RCA. There was a total occlusion at missed RCA and the lesion was successfully revascularized with drug eluting stent. If there was no collateral flow, the other RCA could not be found and its critical lesion could not be managed properly. Our case suggests that collateral flow can be a useful clue in detecting coronary anomaly. Besides, it is important to fully understand coronary anatomy, not to miss uncommon coronary lesion. (J Lipid Atheroscler 2017 December;6(2):97-101)

Key Words: Congenital heart disease, Acute coronary syndrome, Percutaneous coronary intervention

INTRODUCTION

The prevalence of congenital anomalies of coronary artery has been reported to be 0.6%-1.3% of the patients undergoing invasive coronary angiography (CAG). Among congenital coronary anomalies, the double right coronary arteries (RCA) are very rare. It has been reported that the incidence of double RCA was 0.01% in general population during CAG. Herein, we report a case of double RCA from different ostia, which presented as an angina due to total occlusive lesion in one of the two RCAs.

CASE REPORT

A 71-year-old male visited emergency department due to two days of exercise induced chest pain. He had hypertension and 40 pack-year of smoking history. On his physical examination, there was a grade IV mid-systolic ejection murmur at right upper sternal border radiating to right neck. At presentation, cardiac enzymes were elevated (Troponin I 1.51 ng/mL [normal range: ≤0.04 ng/mL], creatinine kinase-MB 6.4 ng/mL [normal range: ≤6.3 ng/mL]), and abnormal Q waves in V1 to V3 with nonspecific ST-T segment changes in V5 and V6 were
Fig. 1. Patient’s electrocardiogram at first presentation.

Fig. 2. Coronary angiography at initial presentation. (A and B) Total occlusion of proximal left anterior descending artery (LAD) (red arrow) and significant stenosis in left circumflex artery (LCX) (white arrows), (C) Right coronary artery with collateral flow (yellow arrows) to LAD, and (D) revascularized LAD (red arrow heads) and LCX (white arrow heads) after percutaneous coronary intervention with drug eluting stents.

shown in electrocardiography (ECG) (Fig. 1). In trans-thoracic echocardiography, left ventricular ejection fraction was 45% by biplane method, and there were regional wall motion abnormalities suggesting multi-
Fig. 3. Patient’s electrocardiogram at second presentation.

Fig. 4. Coronary angiography at the second presentation. (A and B) Left coronary angiogram showing collateral arteries to right coronary artery (RCA) territory (yellow arrows), (C) detected total occlusive lesion in distal segment of the missed RCA (red arrow), and (D) revascularized missed RCA after percutaneous coronary intervention with drug eluting stents (white arrow heads).

vessel territory ischemia: mid to apical septal and anterior wall contractions were akinetic with wall thinning, and mid anterolateral wall and apical lateral wall contractions were hypokinetic. Moderate degree of aortic valve stenosis
was also observed. As acute coronary syndrome (ACS)
was suspected, urgent invasive CAG was performed. In
CAG, there was chronic total occlusion in proximal left
anterior descending artery (LAD), and multiple significant
stenosis in distal left circumflex artery (LCX) (Fig. 2A and
2B). RCA was relatively hypo-plastic, but there was no
significant stenosis and good collateral flows to LAD was
observed (Fig. 2C). Based on ECG, echocardiography and
CAG findings, LCX was suspected as culprit vessel rather
than LAD for the event. Thus, percutaneous coronary
intervention (PCI) for LCX was done firstly, and then PCI
for LAD was also done successfully with drug-eluting stents
(Fig. 2D). The patient was discharged from the hospital
one day after PCI with free of symptom, and received
regular follow-up every three months with medications
including aspirin, clopidogrel, beta-blocker, and statin.

About 20-month later, the patient visited outpatient
department earlier than scheduled with newly developed
chest pain and dyspnea in daily activity which had begun
one week ago. ECG change was not significant compared
with previous ECG (Fig. 3), and cardiac enzyme levels
were not elevated. In transthoracic echocardiography,
there were newly developed hypokinetic motion of basal
to mid inferior walls of left ventricle, and previous
moderate degree aortic stenosis progressed to severe
degree. The left ventricular ejection fraction was decreased
from 45% to 38%. To rule out coronary lesion, invasive
CAG was performed. In CAG, left coronary angiogram
showed no significant change except new collateral flow
from LAD to RCA, and course of collateral flow was not
matched to previously observed RCA (Fig. 4A and 4B).
We reviewed previous CAG and found that the previously
observed RCA course was not matched to general RCA.
It flowed to right ventricular wall. It was variant of right
ventricular branch which originated from aorta with
separate ostium (Fig. 2C). We had misjudged this right
ventricular branch as hypoplastic RCA. Overall, these
findings suggested the existence of true RCA with separate
ostium. After cautious manipulation of catheter, the RCA
ostium was found. CAG showed total occlusion at distal
segment of newly found RCA (Fig. 4C). PCI for the RCA
was done successfully with drug-eluting stents (Fig. 4D).
As patient refused surgical aortic valve replacement or
transcatheter aortic valve replacement, the patient
discharged one day after PCI and are on regular follow-up
without symptom.

DISCUSSION

Congenital coronary anomalies are present about 1%
of the general population undergoing CAG. In one study
enrolling 126,595 patients, rate of congenital coronary
anomalies is 1.6% but, no case of a double RCA was
reported.3 Another study with 7,400 patients, 0.46% had
congenital coronary anomalies and there was just one
case of double RCA.4 Several terms were used including
“double right coronary artery”, “duplicated right coronary
artery”, “dual right coronary artery” and “split right coronary
artery”. Nonetheless, there is no scientific consensus over
the definition and correct diagnosis of a double RCA.5
Chien et al. divided double RCA into two groups based
on whether the double RCA originated from a single
ostium or from separate ostia. To our best knowledge,
a total of 18 cases of double RCA from separate ostia
have been reported since first report by Guepta et al.
in 1987.6,7 As it is very rare coronary anomaly, the correct
diagnosis of double RCA based on conventional CAG is
challenging, and thus, further imaging tests such as
computed tomography are frequently needed.8 Most of
congenital coronary anomalies are considered benign. But,
some may be associated with serious complications, such
as ACS, ventricular arrhythmias, congestive heart failure
and sudden cardiac death.9-11 In this case, the patient had
ACS due to total occlusion of one of the RCA with collateral
from LAD. Fortunately, missed RCA could be detected
by collateral vessels showing different flow direction than
we expected, and the patient escaped the danger by successful revascularization. It is important for the interventional cardiologist to keep in mind this abnormality, in order to avoid wrong diagnosis and inadequate treatment, especially in case of abnormal course of arterial flows. And if we have paid more attention to abnormal course of previously observed RCA, we could have found significant coronary lesion before occurrence of ACS. This case suggests that collateral flow can be useful as a clue to detect coronary anomaly. Besides, fully aware of coronary anatomy is important, not to miss uncommon coronary lesion.

REFERENCES