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Case Reports

Coronary Ostial Acquired Occlusion or Congenital Atresia: An Ongoing Discussion

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Abstract

Coronary ostial atresia is a developmental abnormality that typically causes asymptomatic anomalies in the coronary blood supply. This case report, which presents 2 symptomatic patients with right coronary artery abnormalities, explores difficulties in diagnosing coronary ostial atresia and distinguishing it from single coronary artery and coronary artery disease–related acquired occlusion. Factors underlying management decisions are also discussed.

Keywords: Heart defects, congenital; coronary vessel anomalies; coronary angiography; diagnosis, differential

Introduction

Coronary ostial stenosis or atresia (COSA) is a collection of developmental abnormalities that involve the coronary ostia and cause coronary blood supply anomalies.[1-3] For COSA to be diagnosed, the defect must be congenital, which may be difficult to ascertain in adult patients. Thus, acquired COSA causes, such as atherosclerotic disease, coronary artery disease (CAD), and syphilitic, Kawasaki, and Takayasu arteritis, must be excluded, although this can be difficult, particularly for atherosclerotic disease and CAD.[1,4] Additionally, ostial or proximal coronary stenosis or obstruction must be present, with evidence of a coronary ostial stump or dimple on coronary computed tomography angiography (CCTA) in some cases.[1,5] In the congenital type (COSA), perfusion is generally achieved through prenatal development of collaterals from contralateral circulation, which is distinguished from the more rich networks that arise with acquired coronary artery occlusions.[2,6] Typically, COSA presents without myocardial scarring.[4,5] Notably, COSA often remains asymptomatic because of collateral circulation.[1,6] Treatment can vary based on vessel course. This case report, which presents 2 patients with right coronary artery (RCA) abnormalities and symptomatic manifestations, discusses the difficulties associated with making a diagnosis of COSA in adult patients and distinguishing it from single coronary artery–related and CAD-related acquired occlusion.[3,6] Written informed consent was obtained from all patients.

Case Reports

Patient 1

Patient 1 was a clinically stable, asymptomatic 67-year-old man who was referred to an institution in Sydney, Australia, for an elective coronary angiogram for the evaluation of stable ischemic heart disease (IHD). This referral was based on a previous stress echocardiogram that revealed inducible ischemia with suboptimal left ventricular relaxation at peak exercise and an outside center CCTA. The exact CCTA diagnosis was not made available upon referral. No anatomic abnormalities and ostial stump or dimple abnormalities were reported at the time. His electrocardiograms (ECGs) did not show any evidence of previous myocardial infarction. Investigations to rule out an acute coronary syndrome (ACS)

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were not carried out because this patient presented for an elective procedure and was clinically stable and asymptomatic at the time of presentation. There was no evidence of causes of arteritis that could present as acquired COSA, and cardiac magnetic resonance imaging (MRI) was not deemed to be clinically required because of the institution's limitations on cardiac MRI.

Access for the coronary angiogram was via the right radial artery. The patient had a right dominant circulation. In the proximal left main coronary artery, there was 30% focal stenosis. There were multiple sites of stenosis in the left anterior descending artery (LAD), with ostial 40% stenosis in the proximal LAD and 50% stenosis in the mid-LAD with a fractional flow reserve (FFR) of 0.82. There was also 20% stenosis in the proximal left circumflex artery (LCx). We observed RCA abnormalities with no aortic origin but rather origin from the LCx retrogradely (Fig. 1A, Fig. 1B). A left ventriculogram was also performed, which was normal, as was left ventricular function (LVF).

Patient 1 was diagnosed with single left coronary artery (SLCA) of RCA origin from the LCx and nonobstructive CAD; the patient had normal LVF. Medical management and regular follow-up were recommended, with surgery not deemed necessary after discussion in a departmental multidisciplinary team meeting. Outpatient investigations were arranged for chest pain.

Key Points:

- Coronary ostial atresia is a rare abnormality that operators may encounter during cardiac catheterization and coronary angiography.
- True coronary ostial atresia may be difficult to distinguish from other anatomical abnormalities, such as single coronary artery, and acquired causes of occlusion, such as CAD-related occlusion.
- Management of coronary ostial atresia may depend on the burden of any associated acquired CAD and vessel course.

Abbreviations and Acronyms

ACS	acute coronary syndrome
CAD	coronary artery disease
CCTA	coronary computed tomography angiography
COSA	coronary ostial stenosis or atresia
ECG	electrocardiogram
FFR	fractional flow reserve
IHD	ischemic heart disease
LAD	left anterior descending artery
LCx	left circumflex artery
LVF	left ventricular function
MRI	magnetic resonance imaging
RCA	right coronary artery
SLCA	single left coronary artery
SRCA	single right coronary artery

Patient 2

Patient 2 was also a 67-year-old man referred to the institution for an elective coronary angiogram for evaluation

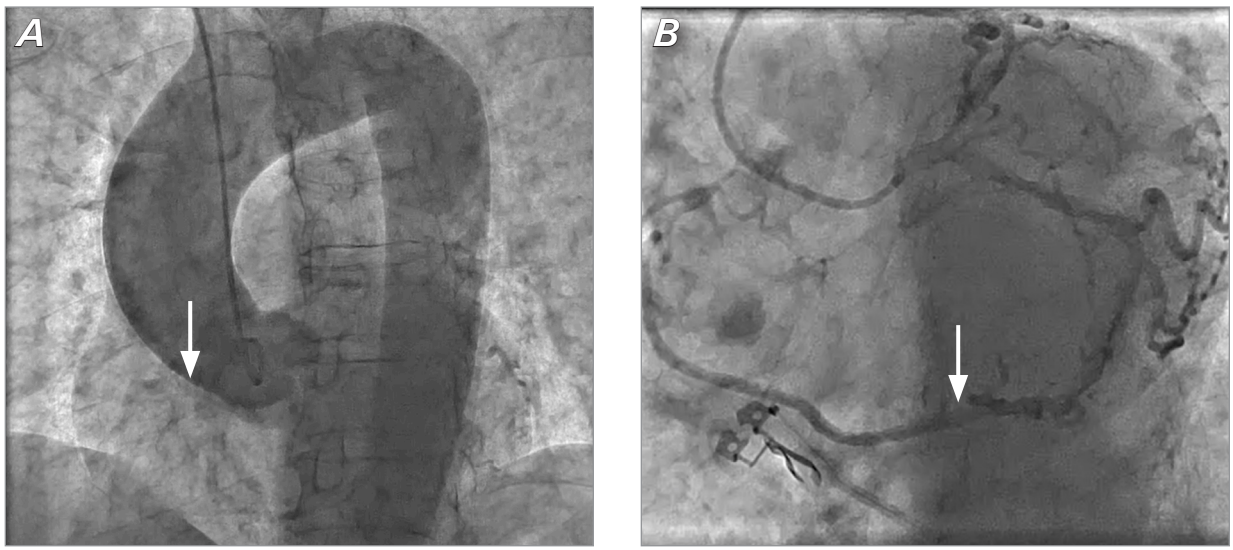

Fig. 1 Aortograms and angiograms showing coronary blood supply abnormalities for patient 1. **A**) Aortogram in the left anterior oblique projection showing no right coronary artery ostia (arrow) and **B**) angiogram spider (left anterior oblique/caudal) projection showing right coronary artery arising as a continuation of the left circumflex artery (arrow).

Supplemental motion image is available for Figure 1A and Figure 1B

of stable IHD. He had anteroseptal ST depression on exercise stress testing in the community and a CCTA calcium score exceeding 300. Investigations to rule out ACS were not conducted because this patient presented for an elective procedure and was clinically stable at the time of presentation. There was also a previous history of multivessel disease impacting the LAD and LCx and a family history of IHD. However, his ECGs showed no evidence of prior myocardial infarction. There was also a known anomalous RCA with dual origins from the LCx and a course between the aorta and pulmonary artery, as well as one from the LAD, detected during previous CCTA in the community. No ostial stump or dimple abnormalities were recognized. Cardiac MRI was not deemed to be clinically required because of the institution's limitations on cardiac MRI. There was no evidence of causes of arteritis that could present as acquired COSA.

Access for the coronary angiogram was initially obtained via the left radial artery because of a prior right ulnar artery injury. During the procedure, access was also obtained via the right femoral vein owing to radial artery spasm and inability to get coaxial engagement. The patient had a right dominant circulation. Mild left main coronary artery and ramus irregularities were observed. Considering the LAD, 40% proximal, 60% mid-eccentric, and 50% distal stenosis (with an FFR of 0.75 with adenosine) was observed. Additionally, the second diagonal artery branch had 60% stenosis. Considering LCx, 30% proximal, 90% mid, and 40% distal stenosis was observed. Additionally, the first obtuse marginal artery branch had 50% ostial stenosis. Both the LAD and second obtuse marginal artery were observed to be graftable. Aforementioned RCA anomalies were observed with dual origins from the LCx and a course between the aorta and pulmonary artery, as well as one from the LAD. The RCA origin was supplied by a branch of the proximal LCx, and the right marginal branch was supplied by the LAD septal artery (Fig. 2A, Fig. 2B, Fig. 2C, Fig. 2D). Unfortunately, an aortogram could not be performed at the time. Collaterals to the RCA were diseased despite minor disease in the RCA itself. The LVF was normal.

Patient 2 had multivessel disease confirmed, along with an anomalous course. Considering his notable CAD burden and concerns related to the significant stenosis from the RCA course between the aorta and pulmonary artery, he was referred for coronary artery bypass graft surgery. Outpatient investigations were arranged for exertional dyspnea.

Discussion

Distinguishing COSA from SLCA or single right coronary artery (SRCA)–related and CAD-related acquired occlusion is important. Single left coronary artery or SRCA occurs when one of the conducting systems arises directly from the other, meaning there is a single ostium and trunk for the entire arterial blood supply.[1,4,5] As such, any stenosis can result in subsequent contralateral ischemic symptomatology.[7] Both SLCA and SRCA differ from COSA when the second criterion outlined earlier, proximal stenosis or obstruction, is not fulfilled; however, if there is stenosis, the SLCA or SRCA is also COSA.[1,5] Notably, this stenosis or obstruction must be congenital and not the result of acquired causes, which can be difficult to ascertain in the case of atherosclerotic or CAD-related occlusive disease.[1,5] This is also related to vessel course; for instance, a malignant course between the aorta and pulmonary artery intramurally causes significant stenosis and is therefore a type of COSA.[1,4,8] This malignant course is associated with increased compression risk and, subsequently, increased risk of angina, myocardial infarction, and sudden death syndromes.[9] The modified Lipton scale is used for categorizing SLCAs.[7,10]

Patient 1, whose primary diagnosis was SLCA with a single left coronary ostium, with the RCA arising from the LCx with a retrocardiac course, was evaluated for COSA because of proximal LCA stenosis. However, it could not be determined whether this was congenital in nature because the presence of CAD confounds the second criterion outlined earlier, namely, the presence of proximal stenosis or obstruction that is not acquired; hence, whether this could also be classified as COSA remains unclear. This situation is indicative of the broader difficulties in diagnosing COSA in adult patients with atherosclerotic disease and distinguishing congenital atresia from CAD-related acquired occlusion.[1,4,5]

Patient 2 had 1 branch following a malignant course, which implied a congenital origin to the stenosis, as outlined above, thus fulfilling definitions for both COSA and SLCA.[1,4,8] There is also a possibility that the RCA in this patient began from the left sinus of Valsalva and that some abrupt acquired occlusion caused the observed stenosis, which would not be COSA owing to the stenosis being acquired. However, given that the patient was being evaluated for stable IHD with no history of ACS and there were no ECG changes to suggest previous inferior infarction, this would appear to fulfill the defini-

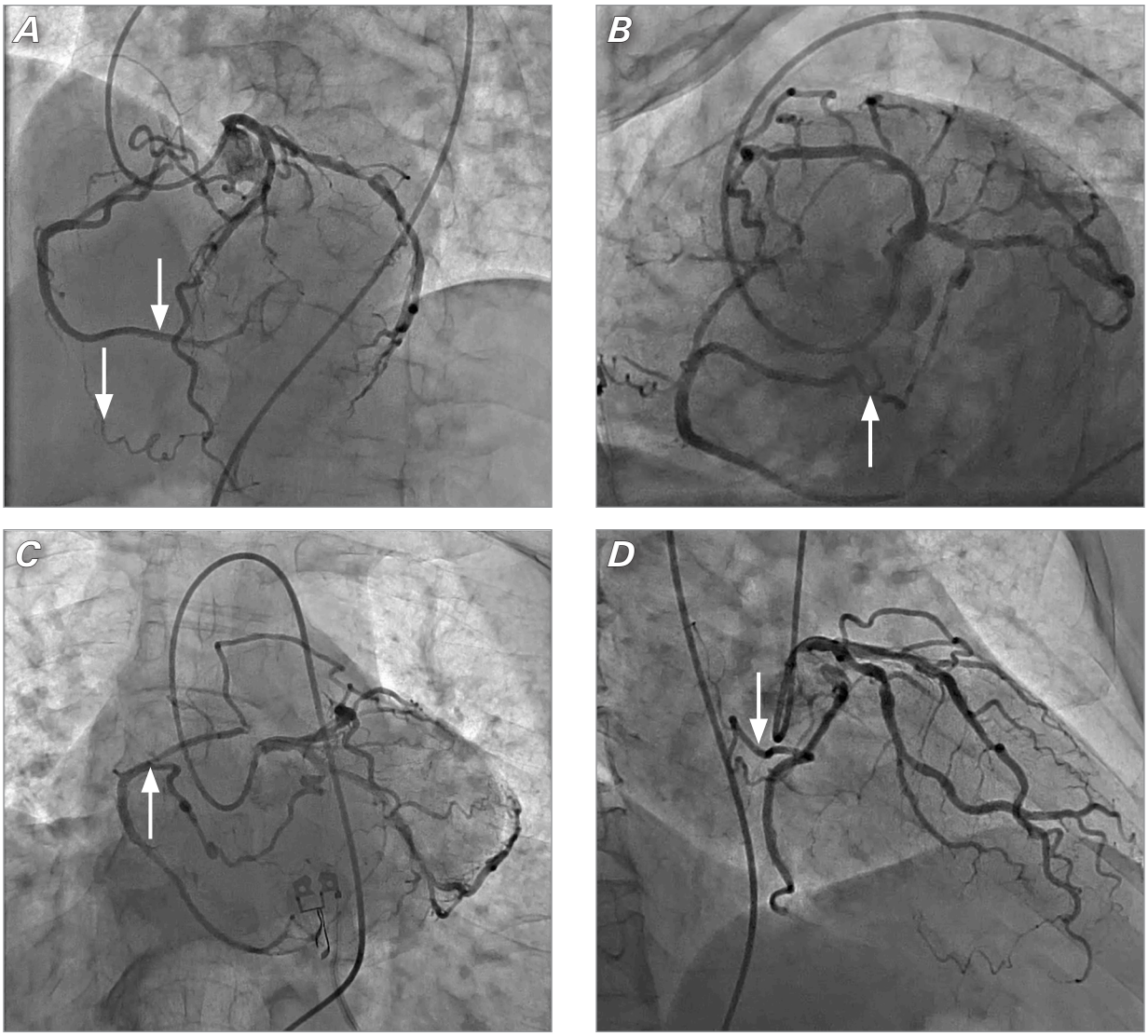

Fig. 2 Angiograms showing coronary blood supply abnormalities for patient 2. **A**) Left anterior oblique/cranial projection shows the RCA arising from both LAD and LCx (arrows) and **B**) the left main coronary bifurcating into LAD and LCx (arrow). Also seen is RCA arising from the left coronary system. **C**) Anteroposterior/caudal projection shows left main coronary bifurcating into LAD and LCx (arrow). Also seen is RCA arising from the left coronary system. **D**) Right anterior oblique projection shows the RCA taking a malignant course between the aorta and pulmonary artery (arrow).

LAD, left anterior descending artery; LCx, left circumflex artery; RCA, right coronary artery.

Supplemental motion image is available for Figure 2A and Figure 2B

tion of COSA. Although it was not possible in this case, performing an aortogram would have been ideal as this hypothesis would have been further proven if there was no anterograde RCA filling. Again, this is indicative of the difficulties in making an absolute diagnosis of COSA in adult patients.[1,4,5] Surgical findings such as RCA ostial atresia may assist in addressing the diagnostic uncertainties associated with distinguishing between COSA and CAD-related acquired occlusion.[8] The surgical report did not comment on these differences.

Because of the vastly greater incidence of left COSA, treatment strategies and long-term outcomes for right COSA are unclear.[6,11] However, surgical bypass is generally not necessary because of collateral circulation development.[6] Considering SLCA, a systematic review by Yurtdas et al[3] showed that patients with an anomalous RCA arising from the left coronary system were treated both medically and surgically, although patients with a higher CAD burden more commonly received percutaneous coronary intervention or surgical management,

and all patients with a malignant course in this review received surgical management.

In the present case series, patient 1 received medical management given the anomalous but nonmalignant course and FFR greater than 0.75, which typically does not require surgical management. In contrast, patient 2 was referred for coronary artery bypass graft surgery, considering the malignant course of the vessel and subsequent associated risk profile along with the high CAD burden.

Conclusions

This case report presents 2 rare cases of RCA abnormalities. These cases highlight difficulties associated with diagnosing COSA in adult patients, particularly in the presence of atherosclerotic disease and potential CAD-related acquired occlusion, and with distinguishing COSA from SLCA. This is a rare abnormality, and operators should be aware of its existence and how vessel course can influence management decisions.

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