Congenital anomalous origin of the right coronary artery from the left coronary sinus is rare and many patients often present with sudden death or myocardial ischaemia. Thus, aggressive management is usually recommended at diagnosis. This case report describes a 19-year-old man with an anomalous origin of the right coronary artery from the left coronary sinus who underwent successful surgical management.

Case Report

A 19-year-old morbidly obese male patient was referred from a peripheral hospital to the cardiology service at the Sultan Qaboos University Hospital, Muscat, Oman, in 2015 with a seven-month history of intermittent substernal exertional chest pain (Canadian Cardiovascular Society grade II) associated with diaphoresis, palpitations and dizziness. In addition, he also suffered from insulin-dependent diabetes mellitus and adult-onset Still's disease. At the previous hospital, he had initially been treated as a case of acute coronary syndrome as his echocardiography results were reportedly normal. However, 24-hour Holter monitoring had shown seven premature supraventricular ectopic beats and an exercise stress test had been prematurely terminated due to the patient’s poor exercise tolerance.

Since the findings were inconclusive and the patient had a history of recurrent chest pain, a cardiac computed tomography (CT) scan was performed which showed an anomalous origin of the right coronary artery (RCA) from the left coronary sinus. The RCA passed between the root of the aorta and the main pulmonary artery. Moderate narrowing of the proximal segment and the ostium was noted [Figure 1]. Considering the CT findings and the patient’s history of ischaemic heart disease, a coronary angiogram and a myocardial perfusion scan were carried out. The coronary angiogram showed a codominant system with no other apparent abnormalities apart from the anomalous RCA. The patient subsequently underwent a technetium-99m stress test which showed normal myocardial uptake. However, the anomalous course...
of the RCA, together with the narrowing of the proximal segment, made the patient more susceptible to ischaemic events and sudden death.

The patient was referred to the Sultan Qaboos University Hospital for further management. An intraoperative transoesophageal echocardiogram was done in which the RCA was clearly visualised as arising from the left coronary sinus [Figure 2]. Accordingly, an unroofing ostioplasty of the anomalous RCA was undertaken. The intraoperative period went smoothly. The aortic cross-clamp and cardiopulmonary bypass time was 45 and 144 minutes, respectively. The postoperative period was uneventful and the patient was discharged five days after the surgery. At a one-year follow-up visit to the outpatient clinic, he denied any further episodes of chest pain and a cardiac CT scan showed that the proximal part of the RCA was wide open with no signs of narrowing.

**Discussion**

To the best of the authors’ knowledge, this is the first reported case from Oman of an anomalous origin of the right coronary artery from the left coronary sinus. In a prospective angiographic study of 1,950 consecutive adult cases, Angelini *et al.*, reported the incidence of anomalous coronary arteries to be 5.6%; of these, the incidence of the RCA arising from the left coronary sinus was 0.92%.\(^1\) However, even though the incidence is relatively low, the consequences of this type of anomaly are often fatal.\(^1\)

As with the current case, the diagnosis of an anomalous RCA is often made incidentally as physical examinations or tests like electrocardiograms, stress tests and perfusion scans are usually non-revealing.\(^2\) A precise diagnosis is often made as the result of cardiac CT imaging which can provide numerous three-dimensional image reconstructions that allow precise evaluation of the origin and course of the anomalous coronary artery.\(^3\) Other diagnostic modalities, such as coronary angiography, are of limited value because they provide only two-dimensional pictures and cannulation of the aberrant coronary artery can be difficult.\(^4\) In view of these limitations, cardiac CT is recommended as the imaging modality of choice in diagnosing such anomalies.\(^2,3\)

Various treatment modalities have been suggested for treating coronary artery anomalies. Kaku *et al.* proposed conservative therapy, such as
limited exercise and drug therapy (i.e. nitrates, calcium or β-blockers and antiarrhythmic drugs), with 69.2% of cases treated using this approach reporting symptomatic improvement. However, the surgical correction of an anomalous RCA is generally indicated when the anomalous artery traverses interarterially between the aortic root and the pulmonary artery. This is commonly termed the ‘malignant’ course, with the artery often passing proximally within the aorta wall (i.e. the intramural segment). During exertion, this intramural segment can transiently narrow due to changes in the pressure and diameter of the pulmonary artery and aorta. The surgical method of correcting such anomalies remains controversial, with many techniques recommended over the years, including a coronary artery bypass, coronary reimplantation and unroofing of the intramural segment. In 1981, Mustafa et al. first described the use of an unroofing procedure to correct an anomalous left coronary artery anomaly. This technique has since become the procedure of preference for many surgeons, especially if the course of the anomalous coronary artery is intramural.

Using the unroofing approach, a neo-ostium is created in the correct sinus and the course between the pulmonary artery and aorta is eliminated, as well as the intramural course within the aortic wall. This moves the coronary lumen away from the interarterial position and effectively relieves the stenosis. Other techniques such as coronary artery bypass grafting using a saphenous vein or ipsilateral internal mammary artery have been proposed; however, they may be more suitable for older patients with associated atherosclerotic changes. Performing this type of surgery on younger patients, such as in the present case, may be counterintuitive because the long-term patency of the graft has not been established and the graft may theoretically compete with the normal antegrade flow of the anomalous artery. Instead, a coronary reimplantation can be considered in which the anomalous coronary artery is ligated and transected proximally and anastomosed in the correct sinus of Valsalva.

Conclusion

The anomalous origin of the RCA is a potentially fatal congenital condition. An unroofing surgery is the procedure of choice when the course is malignant and intramural. In the current case, a one-year postoperative follow-up indicated that the procedure was successful, with the patient reporting the complete resolution of his symptoms.

References