CASE REPORT

Anomalous origin of the right coronary artery with concomitant myxomatous mitral valve disease: a rare coexistence

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SUMMARY

A 45-year-old man previously diagnosed with myxomatous mitral valve disease was admitted to our hospital with chest pain at rest and on effort. Contrast-enhanced CT revealed that the patient's right coronary artery originated from the left sinus Valsalva, which was compressed between the aortic and pulmonary roots. This anatomical abnormality can be associated with sudden death, syncope and chest pain. Ultrasonography showed mitral valve prolapse with severe regurgitation. Surgical repair was performed, which included coronary artery bypass graft and mitral valvoplasty with tricuspid annuloplasty. The postoperative course was uneventful and the patient remains asymptomatic. This is the first description of the coexistence of an anomalous origin of the right coronary artery with myxomatous mitral valve disease.

BACKGROUND

Anomalous origin of coronary arteries is reported in <1% of patients undergoing angiography. Anomalous right coronary arteries (RCAs) from the left sinus Valsalva can be the cause of angina or sudden death. Surgical treatments for high-risk or symptomatic patients have been reported, although indications for surgical repair remain unclear.^{1–6} The existence of an anomalous origin of a coronary artery with concomitant valve disease has been reported, but it is a rare finding. Here we report a case of an anomalous origin of the RCA with concomitant myxomatous mitral valve disease.

CASE PRESENTATION

A 45-year-old man was admitted to our hospital with chest pain at rest and on effort. The patient had already been diagnosed with severe mitral regurgitation (MR) due to myxomatous mitral valve disease, which had not been an underlying cause of symptoms or heart failure. There was no remarkable medical history, except for ileus, and no significant perinatal or childhood events had been recorded. A pansystolic murmur of Levine grade III was auscultated at the apex. Blood analyses including haemogram and differential cell count were normal, as were the patient's hepatic and renal functions and brain natriuretic peptide levels. An ECG demonstrated only an early repolarisation and chest radiograph was normal with no signs of cardiomegaly. Transthoracic ultrasonography showed prolapsing medial scallop of the posterior mitral

leaflet, with severe MR (figures 1 and 2). Contrast-enhanced CT revealed that the RCA originated from the left sinus Valsalva, which was compressed between the aortic and pulmonary roots (figure 3). Coronary angiography revealed significant proximal RCA stenosis (figure 4). Because the patient was symptomatic and stress cardiac scintigraphy suggested a mild, reversible defect in the inferior wall, we decided to provide surgical treatment.

TREATMENT

We chose coronary bypass graft (saphenous vein graft to the 4-posterior descending branch) and mitral valvoplasty with tricuspid annuloplasty.

OUTCOME AND FOLLOW-UP

The perioperative and postoperative courses were uneventful. Angina pectoris disappeared, and the patient remains asymptomatic.

DISCUSSION

To the best of our knowledge, we describe the first case of the RCA originating from the left sinus Valsalva and myxomatous mitral valve disease. The coexistence of valve diseases with anomalous coronary artery is a rare condition, but the coexistence of valve diseases with mitral valve disease is even rarer.⁷

The aetiology of anomalous coronary artery is uncertain. There is reportedly no definite inheritance pattern or sex predominance. Anomalous origin of coronary arteries is reported in <1% of the patients undergoing angiography; however, the origin of the RCA from the left sinus Valsalva is common.^{8–11} The origin of the RCA from the left

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Figure 1 Transthoracic ultrasonographic image showing a posterior mitral leaflet, medial scallop prolapse (arrow).

Figure 2 Transthoracic ultrasonographic image showing mitral valve prolapse with severe mitral regurgitation.

Figures 3 Enhanced CT scan showing the right coronary artery (arrow) originating from the left sinus of Valsalva, which was stuck between the ascending aorta and pulmonary root. LCA, left coronary artery; Ao, aorta; PA, pulmonary artery; LA, left atrium.

sinus Valsalva is a clinically important anomaly because it can be associated with sudden death, syncope and chest pain. Previous studies have used various criteria to classify anomalous coronary arteries. Kim *et al*¹² identified four patterns in which an artery crossed over to the opposite sinus: (1) an anterior course, anterior to the pulmonary trunk or right ventricular outflow; (2) an interarterial course, between the pulmonary artery and aorta; (3) a septal course, through the interventricular septum and (4) a retroaortic course, posteriorly between the aortic root and left atrium. The interarterial course was considered of greatest clinical importance because it was believed to be associated with angina and sudden death.

Although there is no conclusive, established theory explaining angina and myocardial ischaemia in anomalous origin of coronary arteries, the origin of the RCA from the left sinus Valsalva, compressed by aortic and pulmonary roots, can result in these conditions. Flattening of the artery due to angulation of the coronary artery ostium may be another explanation for myocardial ischaemia, particularly aortic root dilation, on exercise. These presumptions, however, can explain chest pain on effort but not at rest.

Coronary CT angiography is useful for the diagnosis of anomalous origin of coronary arteries. We performed the same in the present case, and it was useful for understanding the relationship between the aortic and pulmonary arteries. Mitral valve

Figure 4 Coronary angiographic image showing significant stenosis in the proximal right coronary artery alone.

repair, performed at a Heart Valve Center of Excellence, is reasonable in chronic severe primary MR with preserved left ventricular function, if the patient is asymptomatic.^{13–15} In our case, the patient had angina confirmed by stress cardiac scintigraphy, and angiography and CT scan showed an interarterial course between the pulmonary artery and aorta; therefore, we decided to provide surgical treatment.

Several cases of anomalous coronary artery and mitral valve regurgitation that include concomitant congenital heart disease have been reported,^{16–20} and there have been some reports on concomitant anomalous coronary artery with rheumatic mitral disease.^{21–24} In these reports, almost all patients underwent surgical valve repair with bypass grafting uneventfully.

Patient's perspective

The patient would prefer to undergo the operation while he is young.

Learning points

- The coexistence of valve diseases with anomalous coronary artery is a rare condition.
- In anomalous coronary arteries, right coronary artery between the pulmonary artery and aorta is considered of greatest clinical importance because it can be associated with angina and sudden death.
- We can consider surgical treatment for patients with angina or structure heart disease.

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Competing interests None.

Patient consent Obtained.

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