Aberrant origin of the right coronary artery: diagnostic and surgical aspects

Abstract A 43-year-old man with angina for 15 years underwent coronary angiography, which showed an anomalous origin of the right coronary artery (RCA) from the left sinus of Valsalva with a 30-40% fibrous stenosis in the proximal part of the artery, which was presumably responsible for the patient's symptoms. Myocardial scintigraphy (Tc-99m Cardiolite) suggested reversible ischemia at the apex and the posterior wall of the ventricles. After coronary bypass and anastomosis of the right internal mammary artery (RIMA) to the middle segment of RCA, the patient was asymptomatic; however, a postoperative myocardial scintigraphy indicated that the myocardial ischemia was irreversible.

Key words Coronary artery anomalies · Aberrant origin of right coronary artery · Myocardial ischemia · Myocardial scintigraphy

Introduction

The knowledge of anomalous origins of the main coronary arteries from the aorta appears to be relatively recent. In 1974 Vlodaver et al. [10] described and classified the condition. Aberrant origin of the left main coronary artery (LMCA) from the right aortic sinus was shown to be associated with angina pectoris and increased mortality, while aberrant origin of the right coronary artery (RCA) from the left sinus of Valsalva was not regarded to be a dangerous anomaly — with no reports suggesting increased risk of sudden death. However, in 1982, a necropsy review of ten patients with this anomaly showed that three had died suddenly, indicating the clinical importance of the condition [9]. Other reports have subsequently appeared [3, 6-8] supporting the fact that an anomalous origin of the RCA is related to various signs of myocardial ischemia including angina, ventricular arrhythmias and sudden death. However, none of these cases was conclusive in showing that there was a correlation between ischemia and the anomaly.

In this case report we describe a 43-year-old man with anomalous origin of RCA and angina presumably due to the anomaly. After coronary artery bypass grafting the patient became completely free of cardiac complaints.

Case report

A 43-year-old man with disabling angina (NYHA class IV) was referred for further investigations. For 15 years the man had had daily attacks of oppressive chest pain occasionally radiating to the left or right arm and often accompanied by exertional dyspnea. In addition, there had been episodes of faintness and one incident of syncope. The heart attacks were provoked by physical and emotional stress, but could occur even during rest and sleep. The incidents lasted from a few minutes up to hours with varying relief from sublingual nitroglycerin.

The electrocardiogram (ECG) showed right bundle branch block (RBBB). An exercise test was performed with maximal load of 175 W, which the patient had to stop because of anginal pain that ceased during rest. No ECG changes besides RBBB were detected. A myocardial scintigraphy (Tc-99m Cardiolite) showed impaired myocardial perfusion at the apex and the posterior wall of the ventricles during rest. Perfusion was further reduced during exercise,
suggesting reversible ischemia at the apex and the posterior wall of the ventricles. Echo-Doppler cardiography showed dilatation of the right ventricle, but neither valve disease nor hypokinesia of the myocardium.

The coronary angiography (CAG) seemed to explain the cause of the patient's symptoms. The RCA ostium was located within the left sinus of Valsalva just anterior to the LMCA ostium. The RCA passed right and forward between the aorta and the pulmonary artery, close to the anterior wall of the aorta to reach the midpoint of the right sinus before following a normal course (Fig. 1). An insignificant stenosis (30-40%) was noticed in the proximal part of the RCA between the aorta and the pulmonary artery. The stenosis was uniform without any sign of atherosclerosis. The dimensions and course of the LMCA were normal. On the basis of the symptomatology and the scintigraphic and angiographic findings, it was decided to perform surgical correction. Coronary artery bypass was performed, anastomosing the right internal mammary artery (RIMA) to the RCA. The wall of the RCA appeared pathologically fibrous and thickened, but no stenoses or atherosclerotic plaques were detected.

At control 3 months after the operation, the patient was well and asymptomatic and working full time. An exercise test did not provoke angina and the patient had to stop at 225 W because of general exhaustion. There were no signs of ischemia on ECG. Unchanged from the preoperative state, a myocardial scintigraphy still showed an activity deficiency at the apex and the posterior wall of the ventricles – to all appearances because of irreversible ischemia unaccessible to revascularization.

**Discussion**

Four coronary artery anomalies compromising the coronary circulation are of particular significance in adults [1]: (1) The origin of one coronary artery from the pulmonary artery, (2) the origin of both coronary arteries from the right sinus of Valsalva, (3) the origin of both coronary arteries from the left sinus of Valsalva and (4) coronary arteriovenous fistula. Myocardial ischemia due to anomalous origin of the main coronary arteries from the aorta ((2)–(3)) is believed to be caused by narrowing of the proximal part of the arteries during systole inhibiting pressure storing and perfusion of the vascular bed during diastole. Several mechanisms have been suggested including compression of the coronary artery between the aorta and the pulmonary artery [1], kinking [3], closure of a slit-like orifice of the aberrant coronary artery [2, 3] and alterations of the angle of the coronary ostium with acute stretching of the intramural segment producing intraluminal narrowing [8].

In our report the CAG findings indicated that the stenosis of the RCA was persistent without any dynamical changes of the lumen during the heart cycle, and other pathophysiological mechanisms seemed to be involved. At exploration, we found that the wall of the proximal segment of the RCA was fibrously thickened without atherosclerosis. We cautiously suggest that the persistent mechanical stress resulted in fibrosis of the wall of the artery and contributed to the myocardial ischemia. Right coronary artery dominance has been considered important to the clinical outcome of the anomaly [5]. In our report there was no preferential dominance of the RCA or the left circumflex artery.

Surgical treatment should aim at restoring adequate blood flow through the aberrant coronary artery. The most expedient operation is a coronary bypass graft operation anastomosing the RIMA or a saphena vein graft to the first part of the RCA [6, 7]. More sophisticated surgical tech-
niques than aim at restoration of the normal anatomical position of the right coronary ostium have been reported [4, 8]. The results of these operations have generally been good in terms of survival and relief of symptoms. Any objective judgement of the efficiency of the operation is limited by the small number of patients and lack of knowledge of the natural history of the disease. Only in one single case report has it been demonstrated that reversible ischemia disappeared after surgical correction [4].

The decision to attempt surgical correction in our case was based on the patient's symptoms of severe angina, supported by signs of reversible ischemia in the preoperative scintigraphy. The patient was relieved of angina after the operation, but the success of the operation could not be substantiated by the scintigraphic findings. Retrospectively we may conclude that the myocardial scintigraphy reflected irreversible ischemia at the apex and the posterior wall of the ventricles. Without turning to subtle “placebo explanations”, the question of why the patient was successfully treated by operation remains unanswered.

This case report indicates that surgical correction should be considered in symptomatic patients in whom an anomalous origin of the coronary artery has been established by CAG, even though ischemia has not been demonstrated by exercise testing or myocardial scintigraphy.

References