Case report

Anomalous coronary artery fistula with simultaneous drainage to the left atrium and the coronary sinus

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Abstract

One case of an extremely rare congenital aberrant coronary artery fistula, with simultaneous drainage to the left atrium and the coronary sinus, is described in a 50-year old male patient, creating a combination of an arterio—arterial and arterio—venous fistula. © 1997 Elsevier Science B.V.

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1. Introduction

Congenital coronary vasculature anomalies are being diagnosed more frequently since the extended use of cardiac catheterization. Beside anomalous origin of coronary arteries from the pulmonary artery and ectopic origin from the aorta, fistulisation to cardiac chambers or large veins may be present. Fistulas may originate from normal coronary arteries with normal pattern of distribution, from coronary vessels with ectopic origin or rarely as aberrant vessels originating from the aorta, with or without connections to the coronary arterial system.

Over 90% of coronary fistulas originate from the right coronary artery, drainage is generally to the right heart or the pulmonary artery. Fistulous drainage to the left heart or the coronary sinus occurs only in a minority of cases. The combination of connections to both is extremely rare.

2. Case report

In the 50-year old male patient, a pathological cardiac murmur had been diagnosed in childhood, without any further cardiac workup, however. Working as a butcher, he experienced throughout his life only light dyspnea on exertion.

Prior to surgery he was admitted on an emergency basis in congestive heart failure with acute dyspnea and tachyarrhythmia. Electrocardiography showed atrial flutter and atrial fibrillation. Electrical cardioversion was only temporarily effective in converting to sinus rhythm. An ECG, 8 years prior, had shown sinus rhythm. Auscultation showed a continuous murmur with a maximum in the left 4th intercostal space parasternally. In the chest X-ray there was a global dilatation of the right and left atria.

Workup consisted of echocardiography, cardiac catheterisation and coronary angiography. Echocardiography showed dilatation of the left atrium, from dorsally a large vessel entered the left atrium. The left ventricle was enlarged but with good contractility. There was a minimal relative insufficiency of the aortic and mitral valves. A persistent left superior vena cava

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could be ruled out. Following recompensation cardiac catheterisation showed normal pressure in the right and left heart. (PC 10 mm Hg, LVEDP 5 mm Hg, Cardiac Index 6.02 l/min/m²). A left to right shunt was measured oxymetric with 66%.

In coronary angiography, a normal origin and distribution of the right coronary artery was noted. The left coronary vessels had separate origins from the aorta with a normal Left anterior descending artery (LAD). The circumflex branches were rudimentary with a thin atrial branch and a small marginal branch. Between the LAD and the circumflex branches a large fistulous anomalous vessel could be demonstrated, with connection to the left atrium and the coronary sinus (Fig. 1).

Surgery was performed with extracorporeal circulation in moderate hypothermia (27°C). Following aortotomy, a normal tricuspid aortic valve was found. The ostium of the RCA was normal in size and position. On the left two small orifices representing two circumflex branches were identified. Further, ventrally an approximately 10 mm wide ostium leading to the aberrant fistulous vessel was found, followed by a normal ostium of the LAD.

Cardioplegic solution was applied selectively through the RCA and LAD. The dilated, calcified, tortuous fistulous vessel continued on the dorsal side of the left atrium to the coronary sinus, with dilatation of the vena cordis magna to about 5.5 cm. The fistulous vessel entered the vena cordis magna in the coronary sulus and continued to the coronary sinus (Fig. 2).

The dilated vessel was opened and a 4-cm large lateral connection to the left atrium found; to identify its position relative to the mitral valve the left atrium was opened from the right. The mitral valve showed no pathology. The fistula to the left atrium was closed with a running 4-0 suture from within the left atrium, dorsolateral to the mitral valve. The aortic ostium was closed with a dacron patch.

The postoperative course was uneventful. There were no electrocardiographic or enzymatic signs of myocardial ischaemic damage. On follow-up at 17 month the patient was in NYHA II.

3. Discussion

A coronary artery fistula is an abnormal communication between a coronary artery and a cardiac chamber, great vessel, or other vascular structure [7]. Since the first description of a coronary arteriovenous fistula by Krause 1865 [2], well over 400 cases have been reported. Over 90% of the fistulas originate from the right coronary artery and drain to the right heart chambers and pulmonary artery [4]. Only a small percentage drain to the left heart in 11% or to the coronary sinus in 7% [3]. A combination of simultaneous drainage to the left atrium and coronary sinus must be extremely rare and we have not found a case with this combination in the literature. The origin of the fistulous vessel may in rare cases be anomalous, without connection to a normal coronary artery.

Coronary fistulas may be well tolerated and patients come to diagnosis usually at adult age, when symptoms of congestive heart failure due to volume overload develop, precipitated by acute atrial fibrillation, ventricular arrhythmia [6], acute myocardial infarction [8], or ischemia [5]. Surgical closure of the fistulas is indicated.

Fig. 1. Angiography in form of an aortic bulbugraphy (LAO view) demonstrates large aberrant fistula to left atrium and coronary sinus (No. 1 = aortic bulbus, No. 2 = fistula).

Fig. 2. Course of the fistula with latero-lateral connection to the left atrium and termination in the coronary sinus (LAO view); arrows indicate fistulation.
to prevent progressive congestive heart failure, endocarditis, coronary aneurysm formation with rupture or embolisation [1]. In aberrant fistulous vessels simple closure of the abnormal connections with patch or direct suture is the treatment of choice [9].

References