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Multiple fistulas between coronary and pulmonary arteries

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Congenital anomalies of the coronary arteries occur in 1–2% of the population, the most common and sometimes hemodynamically significant is the coronary arteriovenous fistula [3, 7]. These fistulas were first described by Krause [5]. A single fistula originating from the right coronary artery is most common, bilateral fistulas (involving both coronary arteries) are rare [4]. Surgical correction, ligation, of a coronary arteriovenous fistula was first performed by Björck and Crafoord in 1947, and nowadays surgical intervention is recommended [1–7].

An unusual case of a patient with multiple coronary arteriovenous fistulas originating from both coronary arteries draining into the pulmonary artery is presented with a review of the literature.

Case report

The patient, a 62-year-old white female, consulted a cardiologist in 1992, because of atypical anginal complaints. Physical examination revealed a continuous murmur I/IV. The chest X-ray and electrocardiogram (sinus rhythm) showed no abnormalities. Treadmill test showed no signs of ischemia.

In 1993, the patient complained of angina New York Heart Association Class III. Thallium exercise testing showed signs of diffuse ischemia. Cardiac catheterization revealed normal right- and left-sided pressures. Oximetry showed a saturation of 96% in the aorta, 67% and 62% in the superior and inferior caval veins, 66% in the right atrium, 69% in the right ventricle and 79% in the pulmonary artery. The pulmonary to systemic flow ratio was calculated to be 1.8. The coronary angiogram showed fistulas from the left anterior descending coronary artery and from the right coronary artery, going to the pulmonary artery (Figs. 1, 2).

At operation a slightly enlarged heart with vascular convolutions and/or arteriovenous malformations on the right ventricular outflow tract and on the pulmonary artery was found. Extracorporeal circulation was established and myocardial protection was ensured by intermittent antegrade infusion of St. Thomas’ Hospital cardioplegia (4°C). The right coronary artery was prepared, the fistula identified and ligated. The left anterior coronary artery was identified and all visible arteriovenous fistulas ligated. An arteriomy of the pulmonary artery was performed, and multiple “vascular structures” in the wall were sewn up. The postoperative course was uneventful. A right heart catheterization performed postoperatively did not show any shunt. One year after the operation, the patient is still without angina.

Discussion

Arteriovenous fistulas occur in only 1–2% of the population. The location of the fistula is nearly equally distributed between the right and left coronary arteries, nearly 90% of these fistulas empty into a right heart low-pressure chamber [3, 7]. Bilateral fistulas are rare, 4–5% of the total reported arteriovenous fistulas; 56% of them terminate in the pulmonary artery, in contrast to the unilateral fistulas (17%) [1]. The etiology of the arteriovenous fistula is unclear. It is suggested that they are the result of an anomalous development of intra-trabecular spaces through which blood is supplied to the myocardium during intra-uterine life. Normally these spaces shrink immediately after birth and become capillaries or the besian vessels [8]. This theory can explain fistulas between coronary arteries and cardiac chambers. An accessory coronary artery originating from a cardiac chamber or sinus of Valsalva has also been proposed in the case of a fistula as a single vessel with a single origin and termination [6].
Most of the patients are asymptomatic but present a continuous murmur. A palpable thrill is rather rare. Chronic cardiac volume overload due to the left-to-right shunt, and angina pectoris due to blood shunting through the fistula and away from the myocardium (coronary steal) occur, but infrequently [4]. Bacterial endocarditis and rupture of an aneurysm associated with a fistula, myocardial ischemia and infarction are rarely described complications [2, 6, 9].

Chest X-rays and electrocardiograms are non-specific. Transesophageal echocardiography may be helpful as a diagnostic procedure [10]. Left and right cardiac catheterization, with an oximetry to confirm the left-to-right shunt are the only procedures for establishing the differential diagnosis with other causes of continuous heart murmurs. The precise diagnosis, however, cannot be made without a selective coronary angiography. This investigation defines the origin and termination of the fistula and the coronary artery system. This latter is important should a (vein) graft interposition be needed.

While surgical intervention is clearly indicated in symptomatic patients, with angina, cardiac decompensation or complications, the continued threat of potential complications and circulatory overload warrants surgical treatment even in symptomless patients [7]. A general technique for surgical correction does not exist. Ligation with or without cardiopulmonary bypass is the most simple surgical intervention. However, surgical treatment should be adapted to the anatomy of the fistula. Normal coronary circulation might have to be reconstructed by graft interposition, lateral arteriovenous grafts can be necessary in cases of multiple lateral fistula or, as in our case, an arteriotomy of the pulmonary artery. Surgical complications are low, and occur mostly in elderly patients [2, 4].

In summary, coronary arteriovenous fistulas are rare congenital anomalies. Most patients are asymptomatic, but surgical treatment is advised to avoid later complications.

References