Case report - Coronary

Congenital bilateral coronary artery to pulmonary artery fistulas associated with left main trunk stenosis

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Abstract

A rare case of a 57-year-old patient who presented with an acute coronary syndrome with incidental discovery of bilateral coronary arteriovenous fistulas originating from both coronary arteries to the pulmonary artery trunk and coronary artery atherosclerosis.

Keywords: Coronary artery; Fistula

1. Introduction

Coronary arteriovenous fistulas (CAVF) were first described by Krauss in 1865 [1]. They are present in 0.002% of the general population [1] and they are the most common hemodynamically significant coronary lesion. In angiographic series they represent between 0.13 and 0.87% of all coronary anomalies [2, 3]. We report one case of bilateral CAVF discovered incidentally during a routine angiogram in a patient evaluated for acute coronary syndrome, who underwent transpulmonary closure of the fistulas and coronary artery bypass graft surgery.

2. Case report

A 57-year-old male presented with acute coronary syndrome. An urgent coronary angiogram was performed (Fig. 1, Videos 1 and 2) and revealed left main trunk stenosis and two fistulas arising from left anterior descending artery and right coronary artery to pulmonary artery, with coronary steal from right coronary fistula (Fig. 2). Under cardiopulmonary bypass (CPB), a double coronary artery bypass graft surgery with both left and right internal mammary arteries was undertaken. Closure of pulmonary end of both fistulas was performed through a pulmonary artery arteriotomy. Postoperative course was uneventful. The patient remains asymptomatic at eight postoperative months.

3. Discussion

Coronary artery fistulas are rare anomalies, more often originating from right coronary artery [4]. Bilateral fistulas, originating from both the coronary system, account for 5% of the total. These types of fistulas terminate more often into the pulmonary artery (56%) than unilateral fistulas (17%) [4, 5].

Clinical examination may reveal a continuous murmur and/or a thrill in the upper chest. Younger patients (under 20 years) are more often asymptomatic [6]; symptoms may develop later in life due to chronic left-to-right shunt. Liberthson et al. [6] in a review of 174 cases reported in the literature, found that 55% of the patients older than 20 years had symptoms at presentation, such as dyspnea on exertion (35%), fatigue (8%) or angina (22%). Conversely, only 9% of those <20 years of age had had such symptoms. The incidence of death before operation also increased from 1% in those patients <20 years of age to 14% in those >20 years of age. The most common fistula related complication was cardiac heart failure (12–19%) and endocarditis represented only 3–4% accordingly to age. As seen in this case, these fistulas could be the cause of the coronary steal syndrome, and when they are associated with other cardiac diseases such as coronary atherosclerosis, they may potentiate myocardial ischemia.

In asymptomatic patients, observation with endocarditis prophylaxis is recommended. In this group of patients, closure of the fistula may be recommended in those with significant shunt [7]. General agreement exists that symptomatic patients should be treated. Current treatment options include: transcatheter embolization or surgical ligation. Transcatheter approach presents a closure rate of 85% [8]. Nevertheless, vessel tortuosity and multiple drainage sites may render CAVF unsuitable for this technique [8]. Various surgical techniques were described. They include: internal closure from within the cardiac chamber or pulmonary artery, distal ligation alone or proximal and distal ligation/division with or without closure from within the...
Fig. 1. Coronary angiogram with (a) left anterior descending artery to pulmonary artery (PA) fistula and left main trunk stenosis (white arrow), and (b) right coronary artery (RCA) to pulmonary artery fistula. We may observe that almost all the contrast agent goes through the fistula.

Fig. 2. Surgical view of fistulas (arrows).

Video 1. Left coronary artery angiogram showing a left anterior descending artery to pulmonary artery fistula and left main trunk stenosis.

Video 2. Right coronary artery angiogram showing the coronary artery-to-pulmonary artery fistula. We may observe almost all contrast agent going to the pulmonary artery through the fistula.

aneurysmal coronary artery [7, 9, 10]. Surgical ligation presents a very low mortality rate (1.4%) [8]. It could be achieved with or without CPB [7, 9, 10]. The main concern is the long-term results. Cheung et al. [9] in a series of 41 patients operated on for CAVF, showed that 80.5% of the patients were asymptomatic in the follow-up (range 5.08 months to 28.09 years). Despite this good clinical result, the authors showed a recurrence rate of 19%, the latter being more important in the external ligation/division group as compared to the intracardiac closure group (22% vs. 16%, respectively). When surgical correction of bilateral fistulas to the pulmonary artery is undertaken, as in the present case, we recommend routine exploration of the pulmonary artery under CPB to avoid the technical difficulty of approaching the friable vascular network and the omission of a communication in the vascular malformation, which can cause late recurrence.

In summary, bilateral coronary arteriovenous fistulas are rare congenital anomalies, when they coexist with other cardiopathies they should be treated surgically and the closure should be achieved from within the pulmonary artery.

References