Coronary artery to pulmonary artery fistula with dual origin — embryological, clinical and surgical significance

Karthik Ramakrishnan Vaidyanathan*, Sanjay A.C. Theodore, Madhu Nainar Sankar, Kotturathu Mammen Cherian

Department of Cardiac Surgery, International Center for CardioThoracic and Vascular Diseases, R-30-C Ambattur Industrial Estate Road Mogappair, Chennai 600101, India

Received 15 September 2006; received in revised form 30 October 2006; accepted 14 November 2006; Available online 11 December 2006

Abstract

Coronary artery fistulae are rare anomalies encountered in 0.1—0.2% of angiographic series. We recently encountered a patient evaluated for mitral valve disease who incidentally had bilateral coronary artery fistulae detected on preoperative angiogram. These fistulae drained into the pulmonary artery. She underwent successful transpulmonary closure of the fistula along with mitral valve repair. We discuss the embryological basis of this anomaly and the clinical as well as surgical significance.

Keywords: Embryology; Fistula; Congenital; Heart disease

1. Introduction

Coronary artery fistulae are very rare anomalies with fistulous communication to the pulmonary artery constituting 17% of them [1]. Dual origin of the fistula from both the coronary systems is an extremely rare occurrence. We report one such case discovered incidentally during routine preoperative angiogram in a patient evaluated for mitral valve replacement who underwent transpulmonary closure of the fistula and valve replacement.

2. Case report

A 59-year-old female presented to us with a long standing history of dyspnoea on exertion NYHA Class II, worsening to Class III for the past few months and recent onset of palpitations. She also gave history of paroxysmal nocturnal dyspnoea. Physical examination revealed a low-volume irregular pulse rate of around 90/minute, and blood pressure of 100/70 mm Hg. Auscultation showed first heart sound of varying intensity, an opening snap and a mid diastolic murmur at the apex. Transthoracic echocardiography confirmed the diagnosis of severe calcific mitral stenosis of rheumatic etiology. Both leaflets were thickened and calcified with severe subvalvular fusion. The mitral valve area was 0.6 cm² and she had severe pulmonary artery hypertension. We decided to replace her mitral valve. Before surgery the patient underwent coronary angiogram which revealed two fistulous communications, one from each of the two main coronary systems to the main pulmonary artery (Figs. 1 and 2). The patient was operated through a midline sternotomy. On opening the pericardium, a tortuous vessel was seen to course from the right coronary artery across the right ventricular outflow tract to the main pulmonary artery. The fistulous communication from the left coronary artery was seen to run behind the main pulmonary artery and seemed to end at the same place as that of the vessel from the right side. Cardiopulmonary bypass was established with aorto bicaval cannulation. Aorta was cross-clamped and antegrade cold blood cardioplegia was administered while occluding the main pulmonary artery to prevent runoff through the fistula. After replacing the mitral valve, the main pulmonary artery was opened through a transverse incision just above the pulmonary valve. Cardioplegia was administered to identify the internal opening, which was directly obliterated with a pledgetted prolene stitch; there were no other openings in the pulmonary artery or the right ventricular outflow tract. The pulmonary artery was closed and the patient weaned off bypass.

Postoperative recovery was uneventful and the patient was discharged on the eighth postoperative day.
3. Discussion

The incidence of coronary artery fistulae is reported to be around 0.1—0.2% in angiographic series [1]. The proximal communication is usually the right coronary artery and the distal communication varies with pulmonary artery being the second most common site following the right ventricle [1]. Dual origin of the fistula, as seen in our patient, has only rarely been reported [2,3].

The embryological basis of coronary artery to pulmonary artery fistula can be founded on Hackensellner’s involution-persistence hypothesis [4]. This theory proposes that there are six anlages in the truncus of which two that are seen in the aortic sinuses persist and give rise to the coronary arteries while the others involute. Accordingly, the normally involuted anlage from the pulmonary sinus persists and connects with the anlage from the aortic sinus giving rise to these fistulae. In view of their unique embryological origin, we propose that this abnormality should be classified separately under coronary artery abnormalities rather than clubbing it together with other coronary cameral fistulae.

Clinically, these fistulae are mostly asymptomatic and are incidentally detected although a large shunt may present with features of myocardial ischemia or congestive heart failure. Our patient was totally asymptomatic as far as the fistula was concerned.

Although conservative treatment has been recommended for asymptomatic patients [1], we decided to repair the fistula in our patient for two reasons. First, the patient required a surgical procedure on cardiopulmonary bypass and aortic cross clamp and cardioplegia.

Second, the lowering of PA pressure following mitral valve replacement would increase the left to right shunt causing coronary steal or congestive cardiac failure due to volume overload. Further, the long duration of dyspnoea might be related to the presence of a large shunt prior to the development and progress of mitral valve disease.

A few surgical points need to be emphasized. During administration of cardioplegia, a significant fistula will cause run off into the pulmonary artery leading to inadequate myocardial protection. Thus it is imperative that a clamp be placed across the pulmonary artery while cardioplegia is being delivered. Surgical techniques that have been described include internal closure of the fistula from within the PA, distal ligation alone or proximal and distal ligations and closure from within the aneurysmal coronary artery [5,6].

Identification of the internal opening in the pulmonary artery through a pulmonary arteriotomy and over sewing it is the surest way of obliterating the shunt without any adverse effects. Thus, inadvertent injury to a major coronary branch in the vicinity can be avoided. Further, the fistula may sometimes take the form of a leash of vessels after origin and thus simple ligation of a large fistulous vessel may not completely resolve the condition.

References