ABSTRACT

Coronary artery fistula is the most common of the coronary artery malformations. Ischaemic heart disease and fistulous communication between the LAD artery and the main pulmonary artery was diagnosed in two brothers aged 48 and 49. They had coronary artery bypass grafting and also the fistulae were ligated in both. This seems to be the first report of congenital coronary artery fistula and obstructive coronary artery disease in two brothers.

Key Words: Congenital coronary artery fistula(e), Pulmonary artery, Coronary artery disease

INTRODUCTION

Congenital coronary artery fistula is an uncommon but well-known entity with an incidence of 0.15% to 0.20% in adults who undergo coronary arteriography (1,2). Coronary-pulmonary artery communication comprises about 15% to 66% of coronary artery fistulae in different reports (1-4). Although patients with congenital coronary artery fistula over 20 years of age have more symptoms and complications (5) they can also be asymptomatic.

Here we report two adult patients with coronary-pulmonary artery fistula with associated coronary artery disease. The unusual point is that "these two men are brothers".

CASE 1

A 49 year old male (elder brother) was referred for the investigation of his hypertension and non-specific complaints. He had been known to be diabetic for the last year and had had hypertension for three years and also a family history of coronary artery disease. He had been a heavy smoker for 30 years. Blood pressure was 165/100 mmHg and no bruit was heard. Colour Doppler echocardiography showed systolic turbulence in the main pulmonary artery. Coronary angiography revealed a fistula rising from the proximal LAD and draining to the main pulmonary artery (Fig.1). There were multiple stenoses ranging from 40% to 60% in the RCA, the first obtuse marginal and the first diagonal arteries. No atherosclerotic disease was seen in the LAD artery. Left ventriculogram was normal with a LVEDP of 8 mmHg. With these findings surgery was planned for the future, and yearly follow-ups with coronary angiography besides medical treatment was recommended.

Eight months later the patient presented with angina of two weeks duration and coronary angiography was performed. Ventriculography was again near normal. Although there was no change in the right coronary artery lesions, left coronary angiogram showed a great difference. There was a 70% stenosis in the LAD artery just after the fistula, which had been free of disease 8 months before. And an 80% stenosis was detected in the second obtuse marginal artery. The fistula was patent.

He had coronary artery bypass with a configuration of LIMA-LAD, and saphenous bypass to first diagonal, first and second obtuse marginal, right posterior descending arteries and also the LAD-pulmonary artery fistula was ligated in three sites under cardiopulmonary bypass. He is doing well without signs of ischaemia after thirty five months.

CASE 2

A 48 year old male (younger brother) presented with angina of three weeks duration. He had had diabetes mellitus for 11 years, hypertension for 6 years and a
positive family history of coronary artery disease. Clinical examination was normal apart from elevated blood pressure of 190/100 mmHg. There was no clinical evidence of a continuous murmur. Selective coronary angiogram showed stenoses of the LAD in two sites and there was insignificant stenosis in the right coronary artery. A tortuous fistula was detected between the LAD and the main pulmonary artery proximal to the first LAD stenosis (Fig. 2).

The patient underwent surgery, the fistula was confirmed to be draining to the posterior aspect of the main pulmonary artery. Oxygen saturations of blood samples from the right atrium and pulmonary artery elevated from 61% to 65%. The fistula was ligated in two sites and he also had coronary artery bypass with a configuration of LIMA-LAD (distal segment), and saphenous bypass to proximal segment of LAD and first diagonal arteries in beating heart without cardiopulmonary bypass. He continues to do well after forty one months and has no sign of ischaemia.

**DISCUSSION**

Coronary artery fistula, although rare is the most common of the coronary artery malformations. It was first described in 1865 by Krause (6). These fistulas are known to originate mostly from the right coronary artery but in a more recent report LAD artery is the commonest site of the origin with an incidence of 63.15% (2). The low pressure chambers are the usual drainage sites, so communication to pulmonary artery may be up to 57% in different series (1-3). Coronary-pulmonary fistulas are believed to arise due to the supernumerary implantation of the developing coronary artery into the pulmonary arterial division of the embryonic truncus arteriosus (7).

Symptoms are usually angina, dispnea and fatigue. Angina is thought to be a consequence of "coronary steal phenomenon" which is well demonstrated with stress thallium tomography (8). Thallium imaging was not performed on either of our patients as it would be impossible to document the cause of myocardial ischaemia. (Whether it is because of the coronary lesions or the result of the "Coronary steal phenomenon"). Dispnea and fatigue are probably due to long term cardiac volume overload with advanced age.

As age has a great role in the beginning of the symptoms and complications, most authors (1, 3-5) suggest surgical closure of the fistula at the time of diagnosis even if the patient is asymptomatic. Transcatheter micro embolization or coil embolization are also known methods of fistula closure (9). We agree that the treatment of the coronary artery fistula must be performed at the time of diagnosis, to prevent the appearance of later symptoms and potential complications. We preferred surgery because of associated atherosclerotic coronary lesions. Gilbert et al. (2) controversially comment the opposite. They presented 19 cases of congenital coronary artery fistulas in adult age group and pointed out that surgery can only be planned in the follow up period if clinical deterioration occurs. Spontaneous closure of the coronary fistula is very exceptional and only three cases have been reported (1, 10).

In our first case, surgery was planned for the future because the fistula was small, the patient had no clinical symptoms due to the fistula and the coronary artery disease did not involve the LAD artery. Although
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it was thought that he would undergo an operation a few years later when he would present with a LAD lesion or with symptoms of fistula, he presented with a 70% lesion in the LAD artery just 8 months after his first angiography. The relation of the coronary artery fistula and atherosclerotic disease has been discussed in many papers (11, 12).

Finally, coronary - pulmonary artery fistulas in two cases of brothers both with obstructive coronary artery disease do not seem to have been previously reported. Although we could not find any familial aspect in the related literature, it is hard to say whether the fistulas in these two brothers are coincidental or whether there is a familial diathesis.

REFERENCES