Coronary Artery Fistulae In Jeddah

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SUMMARY

During a nine-year period, three patients underwent surgical treatment for congenital coronary arteriovenous fistulae. In two patients, the right coronary artery (RCA) was draining into the right atrium. In the third patient, the left coronary artery drained into the right atrium. Distal filling of the coronary arteries involved was good as seen in the cineangiography.There was no associated aortic or pulmonary valve disorders in the three patients. The fistulous openings were oversewn from within the right atrium (the recipient chamber) using cardiopulmonary bypass. Preservation of the coronary artery continuity and distal myocardial perfusion was achieved.

INTRODUCTION

Congenital coronary arteriovenous fistula is not common (1), since Krause first described coronary arteriovenous fistula in 1865 (2), and Bjork first reported on its surgical treatment in 1947 (3), 300 cases have been reported. Surgical corrections have been documented for fistulous connections between the coronary arteries and the cardiac chambers (1). The majority of the fistulae originate in the coronary artery and terminate on the right side of the heart (right ventricle, right atrium, pulmonary artery, in that order of frequency), and rarely into the left atrium (1). Cases of rarer sites of communication such as peripheral pulmonary arteries have been reported (4), and this occurred with left coronary artery.

The following cases illustrate our experience in treating patients with congenital right coronary fistulous opening into the right atrium.

CASE REPORTS

Patient 1

A15-year old girl with a history of dizziness, exertional syncope, dyspnoea and palpitation on moderate exertion since early childhood was admitted to the hospital for investigation and treatment. She gave a history of recurrent epistaxis but did not admit rheumatic fever attack.

Examination revealed an underweight patient in a fairly good general condition. The rhythm was sinus with a heart rate of 90/min. Blood pressure was 100/70 mmHg. A systolic thrill was palpable over the aortic area. An ejection systolic murmur (grade 4/6) was audible in the aortic area with radiation to the neck. A systolic murmur (grade 5/6) was heard over the apex.

Electrocardiography revealed tachycardia and left ventricular hypertrophy. Roentgenogram showed mild cardiomegaly with clear lung fields. Echocardiographic examination revealed subaortic stenosis of the discrete type.

Cardiac catheterization was performed which showed left ventricular pressure 234/10 mmHg, left ventricular outflow tract (LVOT) 134/10 mmHg. There was no gradient across the aortic valve. Pulmonary artery pressure was 42/22 (29), and aortic pressure was 108/69 (87). There was a mild elevation of the mean pulmonary artery capillary wedge pressure

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20 mmHg. A gradient of 100 mmHg existed between the left ventricle body and the left ventricle outflow tract. Aortic root injections revealed a discrete subaortic stenosis with aortic regurgitation. There was also proximal dilatation of the right coronary artery but no other arteriovenous fistula was discernible. Oximetry was normal.

Using a median sternotomy and cardiopulmonary bypass with cold cardioplegia, a subaortic membrane was resected. Right atriotomy was done which showed a fistulous opening between the dilated right coronary artery and the right atrium. This fistulous connection was closed using pledgeted sutures from within the right atrium. This patient also had a persistent left superior vena cava which was not mentioned at the time of the initial cardiac catheterization. The patient has been followed up for eight years and she is in good health.

Patient 2

A 5-year old girl with a history of easy fatigability, cyanosis on moderate exertion, dyspnoea, cough and recurrent upper respiratory tract infection since infancy was referred to our Heart Centre for further investigations and management.

On examination, there was no growth retardation, pulse rate was 80 beats/min. and regular, blood pressure was 100/60 mmHg. There was grade 3/6 systolic murmur heard over the left second intercostal space with early short diastolic murmur. Electrocardiography showed sinus tachycardia, left axis deviation with left ventricular hypertrophy. A chest roentgenogram was normal. Phonocardiogram revealed a continuous murmur which was louder during systole.

Cardiac catheterization with aortic root injection revealed a very large and dilated left coronary artery draining into the coronary sinus near its opening into the right atrium (Fig. 1) There was moderate left to right shunt with oxygen step up between mid right atrium (68.7%) and right ventricle (76.8%). A small patent foramen ovale was found. Selective coronary angiography revealed a dominant circumflex artery with a small distal left anterior descending artery.

Operation was performed through a median sternotomy using cardiopulmonary bypass with moderate hypothermia (28°C). A small opening about 4 mm in diameter was found just above the coronary sinus opening which was draining the left coronary artery, a patent foramen ovale (PFO) 1/2 cm in diameter was also found. Both were closed with a running 5/0 prolene sutures. The patient made an adequate recovery. She has been followed up for nine years and she is doing well.

Patient 3

A 22-year old boy with a history of atypical chest pain of 5 years duration, started to have palpitation and easy fatigability six months back. He was diagnosed as having rheumatic valvular heart disease and was started on long-acting penicillin.

Examination revealed a well built, healthy looking boy. Heart rate was 75/min. in sinus rhythm, blood pressure was 120/60 mmHg, increased P_2 and a continuous murmur grade 3/6 maximal over the right sternal border (1st and 2nd interspace).

Electrocardiogram revealed biventricular enlargement. Chest roentgenogram showed cardiomegaly with a cardiothoracic ratio of 16/29 cm, normal lung vasculature and a prominent pulmonary artery.

Pressure measurements at cardiac catheterization revealed R.V. pressure 40/12 mmHg, P.A. 33/9 mm Hg. A step up in oxygen saturation was detected between the mid-cavity right atrium (86.2%) and the right ventricle (91.3%). Left ventricular injections revealed a dilated ventricle with good performance. Selective coronary cineangiography revealed a fistula that originated from the dilated proximal part of the coronary artery and draining into the right atrium (Fig. 2). Distal right coronary artery (RCA) was normal. Left coronary artery (LCA) was normal.

At surgery, a median sternotomy approach under cardiopulmonary bypass and moderate hypothermia was used. The fistula was repaired through the right atrium. The patient had an uncomplicated post operative period. This patient has been followed up one year and he is well.

DISCUSSION

All the three cases described represent a common

form of coronary arteriovenous fistula formation into the right side of the heart (mainly the right atrium). Half of the congenital coronary arteriovenous fistulae originate from the right coronary artery and 92% of all coronary fistulae enter the right side of the heart (1,5).

The clinical picture depends upon the length and calibre of the fistulae, as well as the degree of blood flow. Patients may become symptomatic during infancy or in later life usually after the 4th decade. They may present with recurrent bronchitis as in Patient 2, right heart failure and atrial fibrillation. Clinical or electrocardiographic signs of coronary insufficiency, mitral regurgitation or occasionally paroxysmal atrial tachycardia, as well as myocardial infarction is not uncommon.

Complications include bacterial endocarditis and coronary thrombosis. Occasionally, the patient is asymptomatic or his symptoms may be dominated by another cardiac anomaly, like subaortic membrane stenosis as in Patient 1, and fistulous connections with the coronary arteries are most frequently found in cases with either aortic or pulmonary atresia with an intact ventricular septum (6). Where this occurs, the coronary arteriovenous fistula may be a coincidental finding and the diagnosis is difficult and could be missed.

Cardiac catheterization, aortography and selective coronary arteriography, if feasible, are essential for proper diagnosis of this anomaly. Chest roentgenogram and electrocardiographic changes are usually non-specific and depend on the size of the shunt.

The management of coronary arteriovenous fistula is rather controversial, particularly regarding operative intervention in the asymptomatic patients. 19% of patients under 20 years of age are symptomatic preoperatively, and fewer that 1% have operative complications. 63% of patients over 20 years of age are symptomatic preoperatively and 23% have surgical complications (7). Also, the risk of bacterial endocarditis has been put at 4-10%. We agree with Liberthson, et al (5) that fistulous connections should be repaired in early childhood even in the asymptomatic patient.

Various surgical techniques have been used depending on the anatomical picture (8). Some prefer ligation of the main coronary vessel proximal to the fistulae. Others do a recurrent obliteration of the fistulae with subcoronary mattress sutures, a technique that preserves distal coronary artery flow. Also, direct closure of the fistulous opening (9) through the receiving chamber like the right atrium, as in our cases, could be done safely with the use of cardiopulmonary bypass. The latter method was applied in our three patients and it worked well.

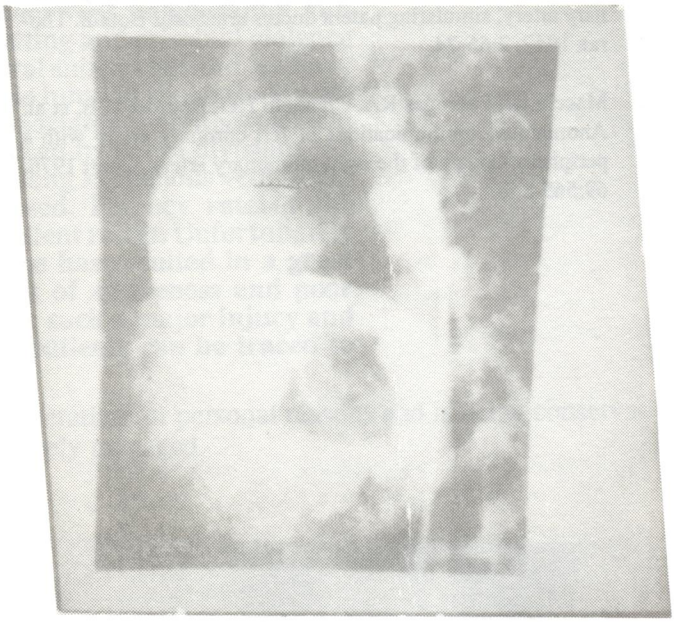

Fig. 1: Left coronary artery fistula draining into the coronary sinus in a 5-year old girl.

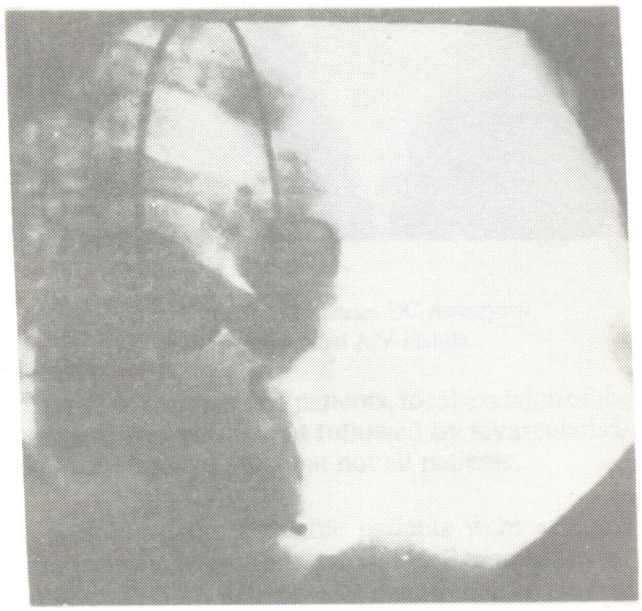

Fig. 2: Right coronary artery fistula draining into high right atrium in a 22-year old male.

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