Case report

Unusual presentation of myocardial ischemia after arterial switch operation

Gürkan Çetin, Levent Saltık, Ahmet Özkara, Kadir Babaoğlu

*Department of Cardiovascular Surgery, Institute of Cardiology, Istanbul University, Istanbul, Turkey
bDepartment of Pediatric Cardiology, Medical School of Cerrahpaşa, Istanbul University, Istanbul, Turkey

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Abstract

Ischemic changes and acute or subacute cardiac failure after arterial switch operation generally results from imperfect transfer of coronary arteries to the neoaorta. Peroperative and early postoperative myocardial ischemia is the main cause of death in these patients. We present an unusual cause of myocardial ischemia and cardiac failure after arterial switch: a congenital coronary artery fistula between the circumflex artery and the right ventricle. The symptoms imitate coronary translocation failure. In differential diagnosis of the coronary perfusion problems encountered after the arterial switch operation, coronary artery fistula should not be forgotten, although it is rare.

Keywords: Arterial switch; Coronary arteriovenous fistule; Ischemia

1. Introduction

Currently, arterial switch operation (ASO) is the procedure of choice for surgical repair of transposition of the great arteries (TGA). Ischemia and ventricular dysfunction are usually due to translocation failure of the coronary arteries following ASO. With the development of surgical techniques and experience, almost all kinds of coronary artery pattern can be translocated successfully. Myocardial ischemia, which frequently occurs due to coronary ostial narrowing that might result from technical failure in translocation of the coronary arteries to the neoaorta, can be seen in ASO patients after discharge from hospital and even at long-term follow-up.

Congenital coronary artery fistula is a rare anomaly, accounting for approximately 0.2–0.4% of all congenital cardiac anomalies [1]. In 50–55% of cases, the fistula originates in the right coronary artery or its branches and more than 90% drain into the right heart chamber or its connecting vessels [1]. Here, we report a case of myocardial ischemia associated with coronary artery fistula following TGA after ASO.

2. Case report

A 3-day-old girl (3.5 kg) was admitted to our institute with severe cyanosis. Physical examination showed no pathological signs other than cyanosis. The echocardiographic diagnosis was d-TGA, restrictive atrial septal defect and a narrowed ductus arteriosus. Prostaglandin E1 infusion was started and the patient was taken to the theater for urgent ASO. Cardiopulmonary by-pass (CPB) was initiated by cannulation of the distal ascending aorta and the superior and inferior vena cava. Moderate hypothermia (25–28 °C) was used during CPB. The left anterior descending artery was arising from sinus I, and the circumflex and right coronary arteries were arising from sinus II as a single and large ostium. There was no indication of any other cardiac anomaly when inspecting the heart. Coronary artery transfection was achieved by implantation of the coronary buttons to the previously anastomosed neoaorta. Neopulmonary artery anastomosis was performed after removal of the aortic cross-clamp. A single piece of fresh pericardium was used to reconstruct the neopulmonary artery. CPB and aortic cross-clamp times were 138 and 72 min, respectively. The patient was taken to the intensive care unit with infusion of low dose dopamine (5 μg/kg per min) and adrenaline (0.02 μg/kg per min). Postoperative electrocardiogram (ECG), troponin-T and other cardiac enzyme levels were normal. She was extubated on the first postoperative day and discharged on the seventh postoperative day without any problem.

After 10 days, the patient was brought to the hospital with congestive heart failure. Heart rate was 190 per min and ischemic ST segment depression on ECG was present. There was a continuous murmur that was maximal at the lower left sternal edge, which did not exist preoperatively. Color Doppler echocardiographic examination revealed a systolic-diastolic color flow at the mitral annulus area. Minimal aortic and pulmonary valve insufficiency was also detected.
Coronary angiography showed a coronary arteriovenous fistula (AVF) arising from the circumflex artery, which drains into the right ventricle (Figs. 1 and 2a).

After management of congestive heart failure with digital and diuretics, transcatheter coil embolization of the A-V fistula was performed. Three Microplex 10 endovascular embolization coils (Microvention, length, 11 cm; diameter, 5 mm) were deployed and complete occlusion of fistula was documented on angiography through the guiding catheter (Fig. 2b). Control echocardiographic examination showed normal left ventricular function and the child was discharged 2 days after intervention.

3. Discussion

Although coronary artery fistulae usually occur as isolated lesions, any type of congenital or acquired cardiac disease may associate with this anomaly [2]. Coronary AVF is rarely seen in TGA patients [3]. Gomes and co-workers presented the first case of TGA with left and right coronary artery fistulae [4]. This is the first case of coronary artery fistula causing myocardial ischemia and ventricle failure after ASO.

Successful transfer by using the correct surgical technique for any type of coronary artery pattern is crucial in ASO. Ischemic changes and acute or subacute cardiac failure after ASO usually indicate imperfect transfer of the coronary arteries to the neoaorta. The risk is high especially in TGA with abnormal coronary artery patterns. However, by using appropriate translocation techniques, this high risk is decreased to very low levels even in patients with intramural coronary arteries.

Ischemic changes also appear in the mid- or long-term follow-up period. The etiology of the ischemia is still the subject of debate. Residual tension, injury at the wall of the coronary artery during surgical dissection, and minimal ostial distortion, which are potential reasons for myocardial ischemia, are the most common predisposing factors for proximal narrowing of the coronary arteries [5]. There was no such anatomical problem in our patient, who was readmitted to hospital with symptoms of myocardial ischemia on the 17th postoperative day when she was at 20 days old. Misdiagnosed coronary AVF was the cause of the myocardial ischemia in our patient.

The severity of the signs and symptoms related to coronary AVF depends on the blood volume load of the fistula. The blood flow through the fistula was minimal in the preoperative and early postoperative period due to high pulmonary vascular resistance (PVR) of the neonate. In our opinion, this was the reason for the misdiagnosis of the fistula preoperatively because when PVR falls during the first few weeks of life, the right ventricle pressure also falls. Furthermore, the ligature of the ductus arteriosus during the operation causes a decrease in volume load and pressure in the pulmonary artery. As a result, there is an increase in blood flow through the fistula, which leads to myocardial ischemia and cardiac failure as seen in our case.

In conclusion, in the differential diagnosis of coronary perfusion problems encountered after ASO, coronary artery fistula should not be forgotten, although it is rare. Cardiac catheterization, aortography and selective coronary angiography are necessary for definitive diagnosis.

References


