Case report

Orthotopic heart transplantation in a patient with situs invs, transposition of the great arteries and Mustard operation

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Abstract

Orthotopic heart transplantation has become standard treatment for end-stage cardiomyopathy, but experience with this technique for complex congenital heart diseases is limited. We report a patient with visceral atral situs invs, transposition of the great arteries and previous Mustard correction, who successfully underwent orthotopic heart transplantation.

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Keywords: Heart transplantation; Situs invs; Transposition of the great arteries; Mustard operation

1. Introduction

Since Barnard performed the first heart transplant operation in 1967, the technique has proved highly effective in end-stage heart disease. However, heart transplant as treatment for complex congenital heart diseases is limited. Several authors have performed heart transplantation in patients with situs invs (SI) [1—3] and in patients with transposition of the great arteries (TGA) [4—6]. However, we have not found any reports concerning patients with SI, TGA and a previous Mustard procedure as presented here. We discuss the anatomical considerations and technical challenges that led to a successful heart transplantation in this complex patient.

2. Case report

A 24-year-old woman was diagnosed at birth of visceral atrial SI and a complex congenital cardiac defect: dextrocardia, atrioventricular concordance, ventriculoarterial discordance, D-transposition of the great arteries, anterior and right-sided aorta, anterior and left-sided right (systemic) ventricle, posterior and right-sided left (pulmonary) ventricle, short perimembranous ventricular septal defect, atrial septal defect, superior and inferior vena cavae on the left and joined to the left atrium, and aortic arch on the right. At the age of nine months, surgical correction of the TGA was performed using the Mustard technique. At 19 years old she was admitted to our hospital for palpitations, dyspnea on exertion and fatigue with exercise. A Holter recording showed frequent premature ventricular contractions, bigeminy and runs of supraventricular tachycardia. Cardiac-gated magnetic resonance and echocardiogram revealed dilated systemic ventricle (tele-diastolic diameter 54 mm), right atrioventricular valve regurgitation, and no baffle obstruction. At 24 years of age the patient was readmitted to the hospital with increasing shortness of breath, orthopnea and paroxysmal nocturnal dyspnea. The electrocardiogram showed atrial fibrillation. The echocardiogram revealed a dilated systemic ventricle (60 mm) and severely depressed contractile function. Because of this tendency to arrhythmia and signs of failing myocardial function, she was placed on the heart transplantation waiting list. A suitable cardiac donor was identified at 10 months.

The procedure was performed through a midline sternotomy. Cardiopulmonary bypass was established using an arterial cannula in the right femoral artery, and direct cannulation of the superior and inferior left-sided vena cava. The aorta was cross-clamped and cardiotomy was performed, leaving the atria and taking care not to damage the Mustard intra-atrial baffle (Fig. 1). The left atrium of the donor heart was anastomosed to the left atrial chamber that contained the drainage of the pulmonary veins. The right atrium of the donor heart, opened from the inferior vena cava to the right atrial appendage, was anastomosed to the right side of the atrial

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chamber that contained the drainage of the venae cavae through the Mustard baffle. Finally, the aorta and pulmonary arteries were anastomosed in end-to-end fashion with the corresponding vessels of the recipient. Following implantation, the apex of the donor heart was positioned in the midline, lying frontwards, due to the particular anatomical location of the recipient’s atrial chambers (Fig. 2). The patient was weaned off bypass with a low dose of dobutamine and atrio-ventricular pacing. The postoperative course was uneventful. Immunosuppressive therapy consisted of lymphoglobulin and corticosteroid treatment before and during surgery, and triple-therapy with azathioprine, methylprednisolone and tacrolimus after the procedure. The patient made rapid progress and was discharged 20 days after cardiac transplant. She continued to improve in the 48 months post-surgery. Cardiac-gated magnetic resonance and echocardiography confirmed the patency of the baffle and functioning of the anastomoses. All cardiac biopsies performed to date have been negative.

3. Discussion

Heart transplantation can be performed for almost any cardiac anomaly. Transplantation of a normal heart to a patient with SI, however, is a significant technical challenge [2,3], and has been previously reported by Tkebuchava and colleagues [7]. There are some anatomic constants that facilitate transplantation of the heart in all patients, including those with complex congenital cardiac anomalies [1]: the pulmonary artery is a midline structure at some point, the aorta is always anterior and to the right of the pulmonary artery at the point of exit from the pericardium, and the left atrium is a midline structure that receives the right and left pulmonary veins. Three main heart structures, therefore, can easily be anastomosed to a normal donor heart. The real complexity of a transplant such as that described here is the complete reversal of the anatomic position of the systemic venous return [1,8]. The key to successful surgery is generally systemic venous reconstruction using autologous tissues or prosthetic conduits [1,3,8].

The particular condition in our patient was that she had previously undergone Mustard operation for correction of TGA. Therefore, the presence of an intra-atrial reconstruction of the systemic venous return allowed us to perform the cardiac implant in a standard fashion. When performing cardiectomy in these circumstances it is crucial to take great care not to damage the intra-atrial baffle. To perform atrial anastomoses it was necessary to place the donor heart with the apex tipped frontwards, so that it remained in the midline of the mediastinum. Although this special positioning of the heart could produce obstruction of the anastomoses due to distortion, these have remained patent and unobstructed throughout the postoperative period.

Heart transplantation today can be considered a safe and highly effective treatment for patients with complex congenital heart defects [9]. According to the literature, the outcome in patients with congenital heart disease is similar to that of patients without such anomalies [10]. For the successful preoperative and postoperative management of these patients, thorough assessment of the cardiac anatomy and detailed surgical planning are essential [1,9]. Multislice computed tomography and magnetic resonance imaging are the most reliable techniques for assessing the cardiac anatomy in these complex patients.

References


