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Transfemoral Biopsy—A Routine Procedure after Orthotopic Heart Transplantation for Dilated Cardiomyopathy in a Patient with Persistent Left Superior Vena Cava and Hypoplastic Right Superior Vena Cava

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ABSTRACT

Introduction: The increasing number of end stage heart failure patients has caused a high number of transplant candidates, including patients with concomitant other cardiac abnormalities. Congenital heart failure can exhibit changes in a variety of anatomic landmarks, and performing heart transplantation in this setting can be challenging. Monitoring for possible rejection is done via intramyocardial biopsies. Here the difficulties arise from variations in anatomic structures.

Background: We present a case of a persistent left superior vena cava discovered intraoperatively during heart transplantation. The patient was a 45-year-old man who underwent transplantation for a severely reduced left ventricular function, along with a high left ventricular end-diastolic pressure and and end stage heart failure.

Discussion: In previous cases, the biopsy was performed by means of left-sided transjugular venous access. Bearing the well-known complications in mind, we chose the transfemoral access so we could take biopsies postoperatively. Biopsies in patients with persistent left vena cava should routinely be performed using the transfemoral access.

INTRODUCTION

The prevalence of adults with congenital heart disease tripled from 1980 to 2000 [Braunwald 2001]. The explanation for this increase is the improved detection of congenital heart defects and improvements in surgical and percutaneous therapies. A consequence of this progress is the increased exposure of adult patients with congenital cardiac defects to cardiac interventions. Thus the knowledge of both, the anatomy of this patient population and well-described approaches to diagnostic and therapeutic interventions are necessary.

Heart transplantation is the remaining therapy option for end-stage heart failure. We present a patient whose condition was treated with orthotopic cardiac transplantation for severely reduced biventricular function.

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CLINICAL HISTORY

The patient was a 45-year-old man who underwent cardiac transplantation for a severely reduced left ventricular function, along with a high left ventricular end-diastolic pressure and a reduced cardiac index (2.3 L/min per m2). Clinically, the patient appeared to be in New York Heart Association stage III heart failure. He had previously undergone mitral valve replacement for congenital mitral stenosis and surgical repair for a membranous ventricular septum defect.

Angiographic findings showed an increased pulmonary pressure (mean pulmonary artery pressure, 40 mm Hg; pulmonary capillary wedge pressure, 25 mm Hg), as well as coronary heart disease with occlusion of the circumflex artery. The following findings were found during the echocardiographic study: (1) a left-to-right shunt with a peak-to-peak pressure gradient of 50 mm Hg; (2) left ventricular dilatation with hypokinetic wall sections and a left ventricular ejection fraction of 10%; (3) good function of the mechanical mitral valve prosthesis, with a peak-to-peak pressure gradient of 12 mm Hg and a mean pressure gradient of 3 mm Hg; (4) minimal aortic insufficiency; and (5) severe tricuspidal insufficiency.

The persistent left superior vena cava (SVC) is frequently associated with other congenital defects in the cardiovascular anatomy. In this case, we knew that the patient had a hypoplastic right SVC and a persistent left SVC, but the exact dimensions became apparent only intraoperatively, when a persistent left SVC connected to a partially unroofed coronary sinus was noted.

The cardiac transplantation was performed according to the modified Shumway technique, which involved retaining the recipient's inferior vena cava (IVC), as well as his hypoplastic right SVC and their communication with the right atria. Thus, the donor-recipient atrial anastomosis was proximal to this communication. An anastomosis of the donor's right SVC to the recipient's persistent left SVC was then performed [Weymann ????].

In addition, we made a longitudinal incision of the donor heart's right atrium, beginning from the IVC to the level of the junction between the SVC and the anastomosis with the

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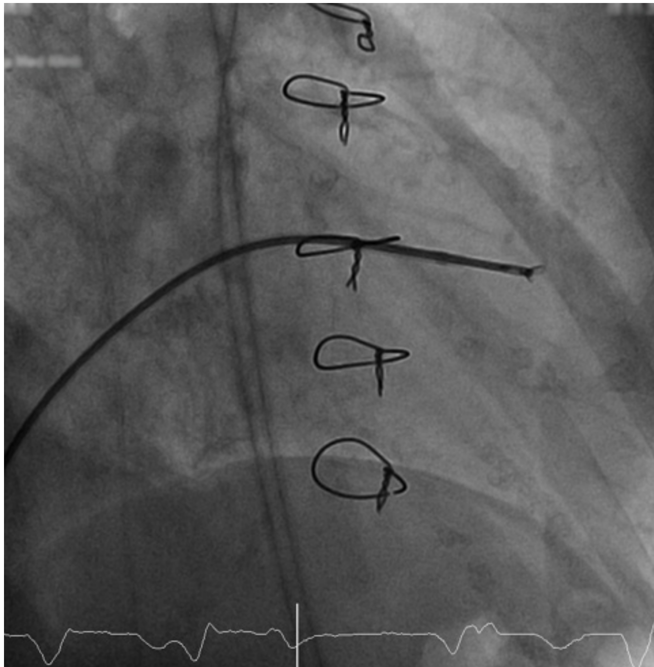

Postoperative transfemoral biopsy. Biopsy catheter in the right ventricle.

persisting right atrial cuff of the recipient. Adjustments in lengths of the pulmonary artery and the aorta along with subsequent anastomosis were accomplished with running suture.

The attempt to perform a transjugular biopsy was not successful. The biopsy catheter could not be advanced through the left SVC owing to the unusual anatomy; however, our attempt to take biopsies via transfemoral access was completed without complications (Figure). The catheter access was the right vena iliaca externa. A 9F sheath was placed with ultrasound guidance as a venous access. The balloon-tipped catheter was advanced through the inferior vena cava to the right atrium, further into the right ventricle, and then into the pulmonary artery. A wire was advanced instead of the placed catheter, and a 7F long introducer sheath was introduced. During the exchange of the catheter for the bioptome sheath, the exchange wire remained in the pulmonary artery to reduce the risk of right ventricular perforation and ventricular arrhythmias.

The advancement of the bioptome sheath was performed with hemodynamic monitoring and radiographic control to assist in appropriately positioning the sheath prior to biopsy. A long biopsy forceps (Baxter, Deerfield, IL, USA) was used to obtain 4 to 6 biopsies from the right ventricular wall under radiographic control.

DISCUSSION

In 1850, the first case of a persistent left SVC was described [Marshall 1850]. Ninety-two percent of cases of left SVC present with a drain into the coronary sinus and then into the right atrium. The drainage pattern can differ, however, and it can lead into the left atrium [Higgs 1998]. Eighty percent of cases present with a right SVC that empties into the right atrium, thereby providing uncomplicated access to the right heart. This explains why the left SVC in our case was identified only intraoperatively [Josloff 1995; Higgs 1998]. The frequency of a persistent SVC in healthy patients is 0.3% to 0.5%; it increases to 4.3% in patients with congenital heart disease [Josloff 1995; Higgs 1998]. Typically in embryonic life, the left SVC lumen is obliterated by the left lung and the left atrium. Reduced compressive forces and a persistent left SVC may be due to several defects, such as atrial septal defect, cor triatratum, and mitral atresia, which can occasionally lead to an association with these defects [Higgs 1998].

A number of case reports have described central venous access in patients with a persistent left SVC [Sweitzer 1993; Josloff 1995; Higgs 1998; Tripathi 2004; Fuchs 2006; Innasimuthu 2007; Shyamkumar 2007; Nair 2008]. Complications of central venous access and right heart catheterization via a persistent left SVC have been reported and include an increased tendency to knot a pulmonary arterial catheter, paradoxical hypotension with volume loading through the coronary sinus (which presumably compresses the left atrium, reducing the left ventricular preload), angina, and cardiac arrest [Schelling 1991; Ho 2006; Ranatunga 2007]. Liberal use of fluoroscopy (particularly oblique views), venography, and blood gas analysis have been proposed to help circumvent complications by better defining the venous anatomy and more accurately locating catheter position [Sweitzer 1993; Josloff 1995; Schummer 2002; Innasimuthu 2007].

We point out that routine transfemoral biopsy can help reduce complications in patients with persistent left SVC.

In summary, invasive procedures in adults with congenital heart disease will become increasingly frequent as this patient population grows. Transfemoral access is a good option for preventing the previously described complications. We have described a protocol for safe and effective myocardial biopsy after orthotopic heart transplantation with the modified Shumway technique.

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