Endoscopic Repair of Obstructive Cor Triatriatum

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INTRODUCTION

Cor triatriatum sinister (CTS) presents uncommonly in adults and is characterized by an anomalous fibromuscular membrane separating the left atrium into upper and lower chambers with a communicating orifice. The size of this orifice determines the severity of obstructive symptoms experienced, which often mimic mitral stenosis. Larger communications are often asymptomatic or become symptomatic in adulthood. Definitive management requires surgical resection, traditionally approached through median sternotomy. Minimally invasive cardiac surgery offers a number of potential benefits, including quicker recovery time, reduced hospitalization time, and more cosmetically appealing results. We present the case of a patient with CTS, atrial fibrillation, and mitral regurgitation who underwent successful minimally invasive repair through an endoscopic, right minithoracotomy approach.

CASE PRESENTATION

A 72-year-old female marathon runner presented with a 1-year history of intermittent dyspnea that occurred unpredictably both on exertion and at rest. She had a history of mitral valve prolapse with mild to moderate regurgitation and previous breast augmentation surgery. An implantable loop recorder showed episodes of paroxysmal atrial fibrillation, and repeat echocardiography ordered by her primary care physician demonstrated progression to moderate to severe mitral regurgitation, trace tricuspid insufficiency, and a tricuspid annular diameter measuring 35 mm. She was referred for possible mitral valve repair. Physical examination demonstrated a pansystolic murmur best heard over the apex of the heart and a high-pitched diastolic murmur. Respiratory examination was unrevealing, with a normal view of the pulmonary veins and mitral valve apparatus (Video 6). A left-sided cryomaze ablation was performed, and the mitral valve was repaired with a 32-mm remodeling annuloplasty ring (Figures 6 and 7). Despite mild tricuspid prolapse, the tricuspid valve was not repaired, as the patient had prolapse of the mitral valve with severe regurgitation (Figure 2, Video 3). Left atrial enlargement, and surprisingly a large membrane separating the left atrium into upper and lower chambers (Figure 3, Video 4).

At the time of operation, the patient was placed in a 20° left lateral decubitus position. Her previous inframammary crease incision was reopened, and her previous breast implant was temporarily removed. A port incision was made through the fourth intercostal space, midaxillary line, and cardiopulmonary bypass was initiated through jugular and femoral access. Intraoperative TEE again demonstrated the CTS membrane dividing the left atrium into two compartments. The proximal chamber was continuous with all four pulmonary veins, with the distal chamber containing the mitral valve. Color flow Doppler demonstrated a small calcified communication within the membrane with turbulent blood flow between the proximal and distal chambers. Three-dimensional TEE localized this communication close to the posterior wall of the left atrium (Figure 4). A transverse left atriotomy was used, and a 5-mm endoscope provided optimal exposure of the CTS membrane (Figure 5, Video 5). After complete resection of the membrane, the left atrial chamber was completely visualized, with a normal view of the pulmonary veins and mitral valve apparatus (Video 6). A left-sided cryomaze ablation was performed, and the mitral valve was repaired with a 32-mm remodeling annuloplasty ring (Figures 6 and 7). Despite mild tricuspid prolapse, the tricuspid valve was not repaired, as the patient had trace tricuspid insufficiency, a nondilated tricuspid annulus, and no symptoms of right-sided heart failure. TEE confirmed successful resection of the CTS and effective repair of the mitral valve (Video 7). After closure of the intercostal space, she underwent bilateral revision breast augmentation.

Postoperatively, the patient recovered well and was discharged home on the fourth postoperative day in normal sinus rhythm. At 1-year follow-up, she was well and without dyspnea (New York Heart Association functional class I). Electrocardiography revealed sinus rhythm. Transthoracic echocardiography showed normal atrial blood flow without obstruction or mitral insufficiency.

DISCUSSION

Cor triatriatum occurs in <0.1% of patients with congenital heart disease.1,2 In CTS, the left atrium is typically divided into a proximal chamber (containing the pulmonary venous return) and a distal chamber (containing the atrial appendage and mitral valve). The proximal chamber communicates with the rest of the heart either through a number of fenestrations in the CTS membrane (most common) or through a number of anomalous venous connections.2 Many cases present in infancy with symptoms of
congestion, but CTS with milder degrees of obstruction may go unnoticed until adulthood. A small fenestration in our patient allowed the passage of blood into the distal chamber. The orifice was large enough to allow reasonable compensation of the obstruction. However, as her mitral regurgitation progressed, it is likely that the regurgitant flow reduced the pressure gradient between the proximal and distal chambers and effectively reduced the driving force for flow through the fenestration. We postulated that this resulted in worsening obstruction and along with the development of paroxysmal atrial fibrillation led to the progression of symptoms in our patient. The concomitant presentation of CTS and Barlow’s disease is a rare occurrence and has been previously suggested that the obstructive membrane in CTS may in fact mask or hide the degree of mitral regurgitation in Barlow’s disease.

Multimodal imaging, including two-dimensional and three-dimensional echocardiography and cardiac-gated computed tomography played a critical diagnostic role in our patient but also helped inform the intraoperative guidance of resection and repair. Preoperatively, transthoracic echocardiography and three-dimensional TEE are the mainstay of workup for patients with CTS at our center. Computed tomography was used to further delineate the left atrial anatomy, such as identification of unrecognized anomalous venous pathways, and to assess for minimally invasive approaches to repair. Intraoperatively, we rely on TEE to guide repair and provide a real-time assessment of its effectiveness. The 5-mm endoscope aids exposure and complete visualization of the anatomy that would be otherwise impossible through the port incisions. Finally, echocardiography is relied upon for follow-up during both the immediate and long-term postoperative period.

**CONCLUSIONS**

Minimally invasive approaches to repair of cor triatriatum are effective and beneficial in appropriate patients. Multimodal imaging is essential in both planning and execution of these complex repairs and enables the entire team to fully participate in, understand, and visualize the repair without the need for a sternotomy incision.
Figure 2 Multiplanar reconstruction of the mitral valve shows Barlow’s pathology with significant billowing or prolapse of the mitral valve.

Figure 3 Echocardiography demonstrating the left atrium (LA) divided by the membrane (arrow). Ao, Aorta; LV, left ventricle.

Figure 4 Color Doppler echocardiography demonstrating flow through a fenestration (arrow) in the CTS membrane (Mn). Three-dimensional echocardiographic image demonstrating a fenestration (arrow) in the membrane. LA, Left atrium; MV, mitral valve.
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