INTRODUCTION

Cor triatriatum sinister is a relatively rare congenital condition in which the left atrium is bisected by a fibromuscular membrane into two distinct chambers. There are multiple hypotheses for the embryologic origin of this congenital defect. The presentation of patients can be during infancy, childhood, or adulthood, and this is due largely to variation in both the degree of obstruction to pulmonary venous return and the presence of associated lesions. We describe the case of a patient presenting in early adulthood with symptoms associated with cor triatriatum sinister and an atrial septal defect (ASD).

CASE PRESENTATION

A 21-year-old, previously very active man was admitted to our institution with peripheral edema, weight gain, and gradual onset of shortness of breath for 3 months. His symptoms had progressed to New York Heart Association functional class III with orthopnea and paroxysmal nocturnal dyspnea. Before admission he had noticed palpitations, substantial weight gain, and a cough. He was initially seen at a regional hospital several weeks prior and noted to be in atrial fibrillation. He was treated with a rate-control approach with metoprolol and anticoagulated with apixaban. He presented several more times to the emergency department over the course of the week with increasing dyspnea. He was ultimately electrically cardioverted and discharged.

Despite this management, the patient quickly noted a recurrence of his symptoms and presented to the emergency department at our institution. He had gained 9 kg from his usual weight. His blood pressure was 135/75 mm Hg and heart rate (HR) regular at 95 beats/min. His oxygen saturation was 96% on room air. His jugular venous sure was 135/75 mm Hg and heart rate (HR) regular at 95 beats/min. A secundum ASD with continuous left-to-right shunting was also noted (Figure 2 and Video 3).

Transesophageal Echocardiography

Transesophageal echocardiography (Figure 1 and Video 1) revealed a left atrial membrane, with flow convergence at the membrane on color Doppler (Video 2) and a mean diastolic gradient of 5 mm Hg at a HR of 54 beats/min. A secundum ASD with continuous left-to-right shunting was also noted (Figure 2 and Video 3). Transesophageal echocardiography also revealed pulmonary hypertension with eccentric hypertrophy of the right ventricle, with severe dilation and moderate systolic dysfunction. It was originally unclear whether the left atrial membrane represented a supravalvular mitral membrane (SVMM) associated with Shone complex or a cor triatriatum sinister—type pathology. However, on further review, the mitral valve morphology was noted to be normal with two appropriately positioned papillary muscles, there was no evidence of a parachute mitral valve, and no sub- or supra-aortic stenosis was identified.

Transesophageal Echocardiography

The patient underwent transesophageal echocardiography to confirm the diagnosis of cor triatriatum sinister (Figure 3A), with the key pathognomonic finding of the left atrial appendage arising from the left atrial chamber (Figure 3B), as opposed to the morphology of an SVMM. The cor triatriatum membrane was located immediately downstream to a secundum ASD. This ASD resulted in communication between the higher pressure pulmonary venous chamber and the right atrium (Figure 3D and Video 4). There was prominent left-to-right shunting. Four pulmonary veins were visualized connecting to the more superior pulmonary venous chamber of the left atrium. Transesophageal echocardiography also confirmed a bicuspid aortic valve with right and left coronary cusp fusion (Figure 3C), with no evidence of aortic coarctation.

Multiple three-dimensional data sets were acquired at the time of transesophageal echocardiography and confirmed a cor triatriatum sinister membrane with a single fenestration between the pulmonary venous and left atrial chamber, anteromedially in the left atrium near the aortic knuckle (Figure 4A, blue arrow, and Video 5). The secundum ASD was visualized (Figure 4B and Video 6) and was measured using 3DQ (QLAB version 10.4; Philips Medical Imaging, Andover, MA) software and found to be 1.4 × 2.0 cm in dimension with an area of 2.2 cm²; the secundum ASD was measured at its largest extent during diastole (Figure 4C).
Cardiovascular Magnetic Resonance Imaging

Cardiac magnetic resonance imaging (MRI) was performed to confirm the pulmonary vein anatomy and to assess shunt ratio. MRI confirmed a cor triatriatum sinister with a single anteriorly positioned defect (Figure 5, Videos 7 and 8). The pulmonary venous connections returned normally to the superior pulmonary venous chamber (Figure 6). Shunt ratio (Qp/Qs) calculated by phase contrast imaging was 3.6:1. Volumetric assessment of the right ventricle revealed severe dilatation and moderate systolic dysfunction (end-diastolic volume 302 mL/m², RV ejection fraction 37%).

Cardiac Catheterization

Because of suspicion of the surprisingly high shunt fraction, coupled with the mismatch between the symptoms and the relatively moderate gradient across the membrane, the patient underwent left and right cardiac catheterization. This revealed pulmonary hypertension (55/19 mm Hg), with mean pulmonary artery pressure of 32 mm Hg and normal pulmonary vascular resistance (PVR) of 2 Wood units. The secundum ASD was easily traversed, revealing a mean pressure of 17 mm Hg (at an HR of 56 beats/min) in the pulmonary venous chamber of the left atrium, a mean pressure of 13 mm Hg (at an HR
Figure 3 (A) Transesophageal echocardiography demonstrating the membrane in the left atrium (LA) (blue arrows) from the midesophageal view at 0°. The membrane bisects the left atrial cavity into a pulmonary venous chamber and a left atrial chamber (white star) contiguous with the mitral valve (yellow arrow, same in [B]). (B) Midesophageal view at 70° with counterclockwise rotation (from the view in A) to the lateral aspect of the LA. Focusing on the left atrial appendage (LAA) (red arrowhead), the membrane (blue arrow) is again visualized bisecting the left atrial cavity into a pulmonary venous chamber and a left atrial chamber (white star). The presence of the LAA arising from the left atrial chamber differentiates this membrane as a cor triatriatum sinister instead of an SVMM, which would be below the LAA. (C) Midesophageal short-axis view of the aortic valve, showing a congenitally bicuspid aortic valve with right coronary cusp and left coronary cusp fusion. (D) Midesophageal short-axis view of the interatrial septum with color Doppler. A secundum ASD is seen communicating between the right atrium (RA) and the pulmonary venous chamber. The left atrial chamber (white star) is visualized downstream of the cor triatriatum membrane (blue arrow). LV, Left ventricle; RV, right ventricle.

Figure 4 (A) View of the left atrial septum reconstructed from three-dimensional zoomed data acquired during transesophageal echocardiography. The interatrial septum (IAS) is seen en face, oriented with anterior (ant.) and posterior (post.) positions labeled. The cor triatriatum membrane (blue arrow) in the left atrium (LA) is seen dividing the LA into the posteriorly located pulmonary venous chamber and the anteriorly located left atrial chamber contiguous with the mitral valve (MV) (black star). The fenestration in the membrane is seen anterosuperiorly (red arrow). Note the secundum ASD (black arrow, same in [B]) located above the defect in the superoposterior septum of the pulmonary venous chamber. (B) Standard right atrial en face view showing the IAS en face from the right atrium. (C) Measurement of the ASD made with 3DQ reconstruction shows a defect measuring 1.4 × 2.0 cm in diameter. Ao, Aorta; AV, aortic valve; IVC, inferior vena cava; SVC, superior vena cava; TV, tricuspid valve.
of 65 beats/min) in the left atrial chamber of the left atrium, and a mean gradient across the membrane of 4 mm Hg at an HR of 59 beats/min (Figure 7). The mean right atrial pressure was measured at 10 mm Hg.

There was a substantial step up in oxygen saturation in the right atrium, with a shunt ratio calculated using Fick’s method of 3:1. The large shunt ratio was consistent with that shown on cardiac MRI.

**Clinical Course**

Given the lack of evidence of pulmonary vascular disease (normal PVR despite increased pulmonary pressures), the patient was referred to our congenital heart surgery service. In January 2016, the patient underwent resection of cor triatriatum sinister membrane, patch closure of secundum ASD, and a biatrial maze procedure. The patient had an uncomplicated postoperative course and was discharged home 1 week later. His postprocedural progress has been good, with increasing exercise capacity. His RV systolic function remained mildly impaired on follow-up transthoracic echocardiography 3 months later. RV systolic pressure has normalized.

**DISCUSSION**

We present the case of a 21-year-old male patient with a recent diagnosis of cor triatriatum sinister (type 3) with obstruction of left ventricular inflow. Cor triatriatum sinister is an interesting and relatively rare
congenital cardiac condition present at autopsy in 0.4% of congenital heart patients. In a single pediatric institution’s experience, it can be associated with other congenital abnormalities in up to 80% of cases, the most common abnormality being secundum ASD and partial anomalous pulmonary venous return.

The condition is characterized by a membrane that divides the left atrium into two chambers. In the upper chamber, the upper pulmonary venous chamber connects with the pulmonary veins and receives the pulmonary venous blood. The lower left atrial chamber is contiguous with the atrioventricular valve, and the left atrial appendage arises from it. The relationship with the left atrial appendage is the key anatomic feature differentiating this membrane from an SVMM, which might be seen in Shone syndrome and related pathologies. Our case study was ultimately differentiated using transesophageal echocardiography.

The simplest classification scheme for cor triatriatum sinister was described by Loeffler in 1949. Loeffler described type 1 as characterized by no communication in the membrane itself, type 2 as having one or several small perforations in the membrane, and type 3 by one wide opening. Our patient had a type 3 morphology, which is the less hemodynamically significant morphology and is typical of nonpediatric presentations of this pathology.

With our patient, the increased resistance across the cor triatriatum sinister membrane substantially increased the volume of left-to-right shunting across the secundum ASD. Interestingly, the increased volume resulted in pulmonary venous hypertension without an increase in PVR. The increased right-sided pressure was therefore largely flow related and did not reflect the establishment of pulmonary vascular remodeling, as documented by normal PVR.

Of note, the mild pressure gradient across the left atrial membrane fenestration likely did not reflect the high resistance across this circuit, as the nonrestrictive secundum ASD acted as a pop-off valve decompressing the higher pressure pulmonary venous chamber into the lower pressure right atrium. This gradient differential was noted invasively, with the gradient between the pulmonary venous chamber and the left atrium measuring 4 mm Hg, explaining the large degree of left-to-right shunting noted on both MRI and invasively.

The presentation of patients with cor triatriatum sinister is most common during infancy and childhood. Early-adulthood presentations are not unheard of, however, largely because of variation in both the degree of obstruction to pulmonary venous return and the presence of

Figure 7 (A) Pressure tracing obtained during cardiac catheterization from the left atrial chamber (mean, 13 mm Hg). (B) Pressure tracing from the pulmonary venous higher pressure chamber (mean, 17 mm Hg). Both chambers were accessed from across the secundum ASD, with the left atrial chamber accessed via deflection across the membrane fenestration. (C) The difference between the two left atrial chambers is reflected in the mean pressure difference of 4 mm Hg at an HR of 59 beats/min between the pulmonary venous chamber of the left atrium and the left ventricle (assuming a structurally normal, nonstenotic mitral valve between). (D) Tracing from the main pulmonary artery confirming the presence of moderate pulmonary hypertension (mean pulmonary artery pressure, 32 mm Hg). Note that the mean right atrial pressure (not shown) was measured at 10 mm Hg.
associated lesions. The reason for late presentation in our patient is difficult to determine, but we conjecture that this young man had excellent functional reserve, allowing him to remain asymptomatic for a long period, coupled with a “pop-off” valve effect through the ASD diminishing the degree of pulmonary venous obstructive symptoms. It is likely, then, that the subacute presentation was precipitated by the development of an atrial arrhythmia with rapid ventricular response.

Given the normal PVR, the pulmonary hypertension was felt to be reversible, and given the clinical and imaging evidence of RV impairment and the substantial shunt, it was felt that there was no contraindication to surgery and that the patient would benefit from surgery. Surgical outcomes are typically favorable at experienced centers, with almost all patients becoming asymptomatic and a 5-year survival rate of >90%. Predictors of recovery of RV function are unknown. However, repeated cardiac MRI 1 to 2 years later may provide an assessment of reversibility. We are optimistic of RV recovery given the natural course of RV recovery following surgical repair in other pathologies associated with RV dilatation and impairment, such as ASD repair and lung transplantation.

CONCLUSION

Cor triatriatum sinister is a relatively rare yet important congenital heart disease. Its detection and differentiation from SVMM require careful echocardiographic assessment. Other concomitant congenital abnormalities along with the clinical consequence of right-sided volume overload from a coexisting ASD can be assessed using cardiac MRI and invasive cardiac catheterization. This additional information is valuable especially when surgical intervention is planned.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.case.2017.05.003.

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