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Cor Triatriatum Sinisterum in a 67-Year-Old Presenting with Severe Mitral Regurgitation and Chronic Atrial Fibrillation

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Abstract

Purpose: Sinisterum is a rare congenital anomaly characterized by a fibromuscular membrane dividing the left atrium into two chambers. While typically diagnosed in childhood, this condition may occasionally present in adults with associated cardiac complications.

Case Description: We report the case of a 67-year-old who presented with worsening dyspnea, chronic atrial fibrillation with rapid ventricular response, and signs of heart failure. Evaluation with transthoracic and later transesophageal echocardiography demonstrated a large membrane fenestration and severe mitral and tricuspid regurgitation. Right heart catheterization confirmed elevated pulmonary pressures and reduced cardiac output.

Intervention: The patient underwent urgent surgical intervention. He underwent resection of the left atrial membrane, mitral valve replacement with a bioprosthetic valve, biatrial maze procedure, and left atrial appendage exclusion. The surgery was well-tolerated, with uneventful postoperative recovery.

Outcomes: Postoperative echocardiography confirmed the absence of residual membrane or significant regurgitation and demonstrated a well-functioning bioprosthetic mitral valve. The patient was discharged in stable condition, with symptomatic improvement.

Conclusion: This case report highlights the presentation and management of Sinisterum accompanying severe mitral valve regurgitation. Sinisterum is a rare congenital anomaly that typically manifests in childhood but was diagnosed in this patient in late adulthood. The delayed presentation underscores the significance of large membrane fenestrations, which can mitigate hemodynamic compromise and delay symptom onset.

Transthoracic and transesophageal echocardiography helped diagnose and characterize the pathology and guide surgical planning.

The findings contribute to the limited literature on late-onset presentations of this condition and are a reference for similar clinical scenarios.

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Introduction

Cor Triatriatum Sinister is a rare congenital abnormality that is typically diagnosed in childhood; however, a few cases remain asymptomatic and are diagnosed in adulthood. In cor triatriatum, the atrium is divided by an abnormal septum; in the sinisterum form, the Left Atrium (LA) is divided into posterosuperior and anteroinferior chambers by a fibrous or fibromuscular septum, wherein the pulmonary veins enter the posterosuperior chamber, and the anteroinferior chamber communicates with the mitral valve and gives rise to the LA appendage. In children, there is an absence of communication between the two chambers, resulting in high mortality. In contrast, in adults, the dividing membrane contains one or more orifices that allow the two chambers to communicate. The symptoms result from the gradient across the orifices in the septum, which can lead to pulmonary congestion and resemble symptoms of mitral stenosis [1]. In the latter, symptoms may remain indolent until adulthood, when they typically present with arrhythmia and heart failure with diminished forward flow [2]. The presence of cor triatriatum sinistrum alone in adults may not be an indication for surgery in the absence of symptoms that limit quality of life or life expectancy [3].

This case presents a 67-year-old who presented with cor triatriatum sinistrum at a very late age, with heart failure with reduced ejection fraction (HFrEF) in the context of chronic atrial fibrillation and was diagnosed with severe Mitral Regurgitation (MR).

Case presentation

A 67-year-old with a known history of Sinisterum presented to the emergency department with a worsening cough, shortness of breath, and palpitations. He was found to be in atrial fibrillation with Rapid Ventricular Response (RVR) and with signs of acute heart failure. His vitals on initial presentation revealed him to be hypertensive 144/89, afebrile, heart rate 169, respiratory rate 17, and SpO2 96% on room air. He had been seen one month prior for atrial fibrillation and had undergone multiple cardioversion attempts. He was on amiodarone and metoprolol and had been started on furosemide. He was treated in the emergency department with a calcium channel blocker, intravenous furosemide, and supplemental oxygen. Cardioversion was attempted twice but was unsuccessful. Cardiac enzymes were negative, but proBNP was elevated. A screening bedside transthoracic echocardiography revealed an enlarged left atrium with cor triatrium sinistrum and moderate to severe mitral regurgitation. Transesophageal Echocardiography (TEE) revealed severe left ventricular dilation, reduced ejection fraction (LVEF 30%), severe MR, and severe tricuspid regurgitation (TR).CT imaging showed no pulmonary emboli but noted bilateral pleural effusions and pulmonary edema. The patient had a history of abdominal aortic aneurysm diagnosed in 2022, hyperlipidemia, type 2 diabetes mellitus controlled on oral hypoglycemic agents and insulin, and Obstructive Sleep Apnea (OSA). Further workup with a right heart catheterization revealed elevated filling pressures (pulmonary artery pressure (PAP) 43/24 mm Hg, mean PAP 36 mm Hg, Pulmonary Capillary Wedge Pressure (PCWP) 25mm Hg) and low cardiac output (cardiac output 4.0, cardiac index 1.8 by thermodilution), which lead to ICU admission for preoperative optimization.

After two days of intravenous diuresis guided by data from a pulmonary artery catheter, the patient underwent urgent surgery with resection of the left atrial fibrous membrane, mitral valve replacement with a 33mm bioprosthetic valve (Epic mitral, Abbott CV, Chicago, IL), biatrial maze procedure with cryoprobe, and left atrial appendage exclusion with a 50 mm AtriCure clip (AtriCure, Mason, OH). Intraoperatively, the left atrium and the left ventricle were enlarged, with a large left atrial membrane separating the left atrial chamber. The mitral annulus was dilated, but the mitral valve leaflets appeared relatively normal. The tricuspid valve was normal. The patient tolerated the procedure well, with a total cardiopulmonary bypass time of 164 minutes and an aortic cross-clamp time of 109 minutes.

The postoperative period was uneventful. The patient was extubated on postoperative day one and was managed with diuretics and inotropes. He was transferred to the step-down unit on postoperative day four and discharged on postoperative day seven on apixaban. A transthoracic echocardiography prior to discharge revealed a well-functioning bioprosthetic mitral valve and no paravalvular regurgitation. The patient was advised to continue follow-up for anticoagulation management and to monitor for any complications.

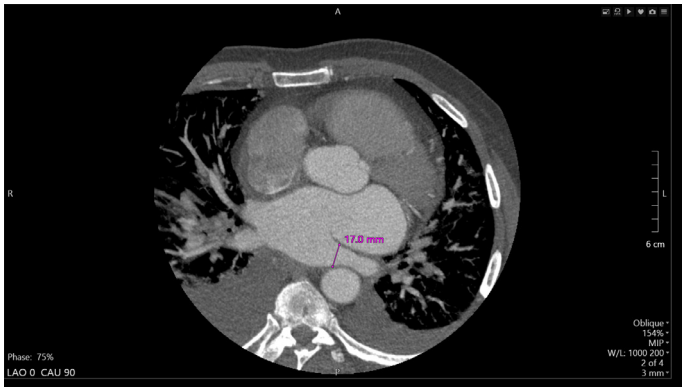

Figure 1: Preoperative midesophageal two-chamber view demonstrating a left atrial membrane orifice with laminar diastolic flow.

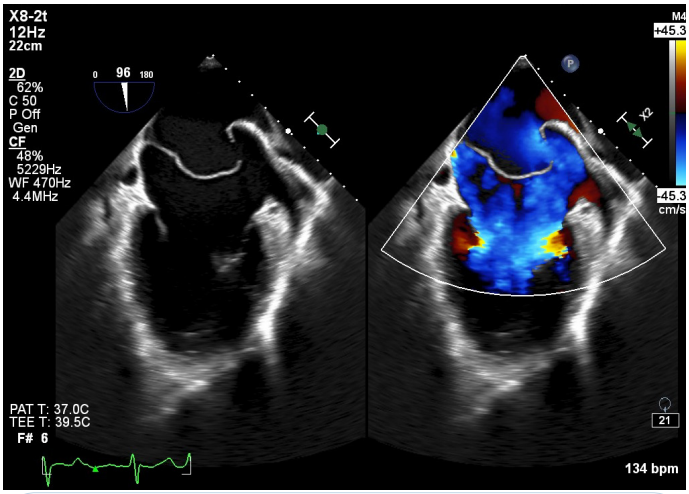

Figure 2: Preoperative multiplanal reconstruction of the left atrial membrane, demonstrating a large orifice measuring 1.5 x 1.6 cm in diameter; this was demonstrated by the laminar flow.

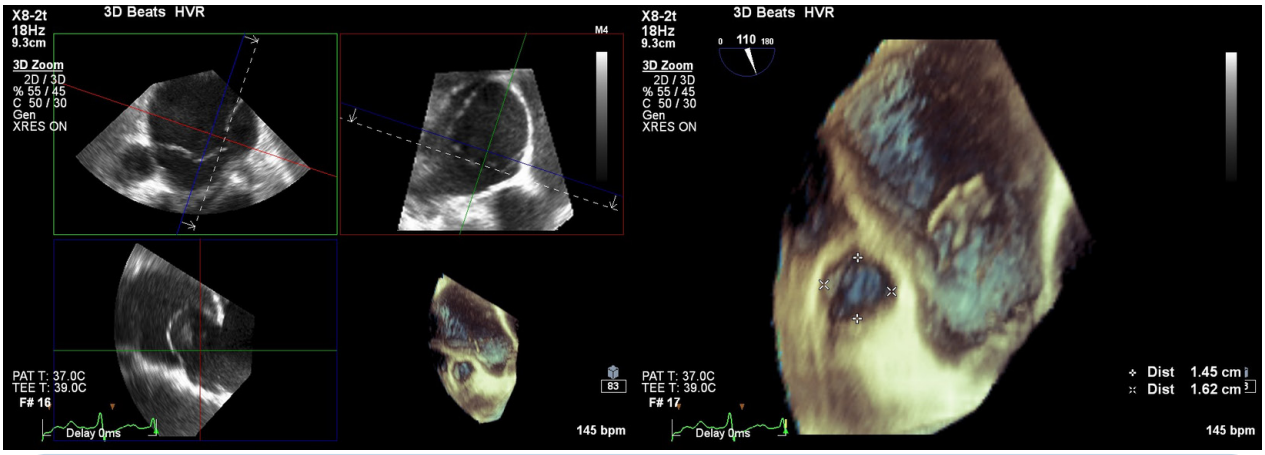

Figure 3: Preoperative midesophageal long axis view demonstrating severe mitral regurgitation, with regurgitant flow passing through the left atrial membrane orifice.

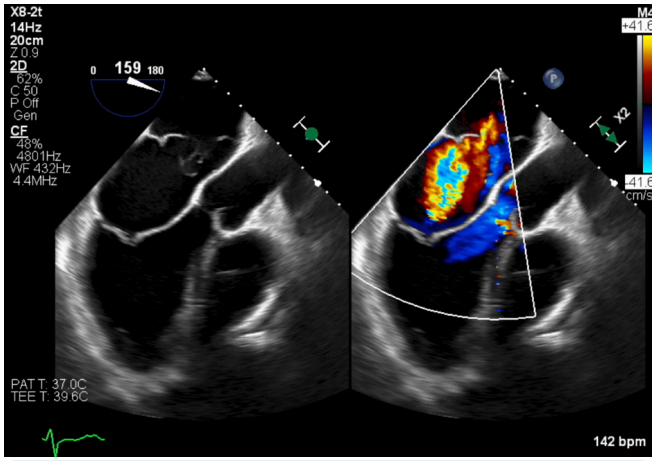

Figure 4

Discussion

Cor triatriatum is a rare congenital anomaly characterized by a fibromuscular membrane dividing the left atrium into two chambers. This anomaly, although usually diagnosed in childhood, can sometimes present in adulthood, complicating the clinical picture with coexisting conditions such as mitral regurgitation and atrial fibrillation. Our case presented features of heart failure that may be attributed to mitral regurgitation, the presence of a congenital anomaly, or both.

Advanced cardiovascular imaging plays a pivotal role in its diagnosis and management. In our case, Transthoracic Echocardiography (TTE) initially identified the presence of a membrane in the left atrium, which was later confirmed and further characterized using Transesophageal Echocardiography (TEE). The detailed anatomical information provided by TEE was critical in preoperative planning. According to Jacobs et al., three-dimensional echocardiography offers superior spatial orientation and visualization of the fenestrations in the membrane, aiding in the accurate diagnosis and assessment of the severity of the condition [4]. Nonobstructive cor triatriatum may be an incidental finding during imaging for other reasons, and we believe that the patient survived late into adulthood because the membrane fenestrations were large and adults. Cor triatriatum may be associated with mitral regurgitation, possibly due to abnormal flow through the membrane fenestration damaging or distorting the mitral valve.

The first surgical repair was described by [5]. The surgical approach to cor triatriatum often involves resection of the membranous septum to relieve obstruction and restore normal blood flow. In adults, this procedure is frequently accompanied by correcting associated cardiac anomalies, such as mitral valve repair or replacement. In a similar case, Ischida et al. reported a successful outcome in a 57 year old adult patient who underwent combined surgical procedures for cor triatriatum sinistrum and severe mitral regurgitation [6]. In our patient, the combination of membrane resection and mitral valve replacement resulted in significant symptomatic improvement, and the biatrial cryomaze effectively obliterated the atrial fibrillation.

Conclusion

This case demonstrates the successful management of a patient with chronic AF, severe MR, and cor triatriatum. In our patient, the orifices in the membrane were large enough to prevent the usual signs, symptoms, and radiographic findings associated with cor triatriatum, thus leaving the initial diagnosis to be made when the patient developed AF and subsequent MR.

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