

Cor triatriatum sinister in an elderly woman patient with successful conservative treatment

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Abstract

Cor triatriatum sinister (CTS) is a relatively rare congenital condition characterized by an abnormal septum dividing the left atrium, morphologically presenting as three atria. Although most individuals with heart failure related to CTS undergo surgical treatment in childhood, those with larger fenestrations may remain asymptomatic until they reach an age where surgery is less viable, presenting treatment challenges. In our case study, we report on an elderly woman who declined all invasive treatments and developed heart failure due to severe functional mitral valve regurgitation triggered by atrial fibrillation. She opted for conservative treatment after a comprehensive evaluation of her condition using transesophageal echocardiography, cardiac magnetic resonance imaging, and right-heart catheterization. This multimodal evaluation highlights the importance of accurate diagnostic approaches and tailored treatments for elderly patients with CTS.

Keywords: cor triatriatum sinister; multimodality imaging; conservative treatment

Introduction

Cor triatriatum is a rare congenital heart condition in which the right or left atrium is anatomically divided into proximal and distal chambers by a fibromuscular membrane, representing 0.1 to 0.4% of all congenital heart diseases [1]. In the case of cor triatriatum sinister (CTS), which affects the left atrium, it frequently leads to pulmonary venous obstruction, pulmonary arterial hypertension, and can culminate in congestive heart failure. The severity of CTS varies depending on the size of the fenestration, the pressure gradient between the proximal and distal chambers of the left atrium, and any other accompanying congenital abnormalities. Patients with large fenestrations often remain asymptomatic, but may develop symptoms as age-related heart diseases progress. For elderly patients with CTS, who may not be suitable candidates for surgery, it is crucial to select an appropriate conservative treatment based on comprehensive examinations.

Case summary

An 82-year-old female patient was admitted to our hospital with palpitations and worsening dyspnea on exertion (New York Heart Association functional classification; NYHA class III) with severe bilateral leg edema. She had a medical history of paroxysmal atrial fibrillation, diabetes mellitus, and chronic kidney disease,

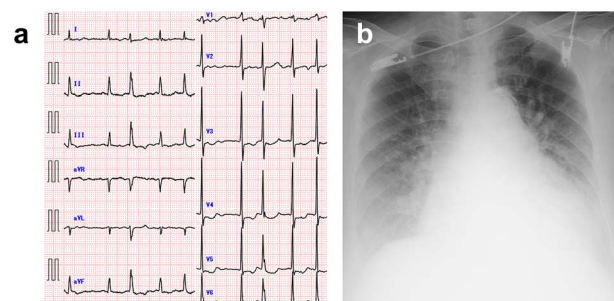


Figure 1. Electrocardiogram and chest radiograph on admission. (a) Electrocardiogram shows atrial fibrillation with single premature ventricular contraction. (b) Chest radiograph showing severe bilateral pulmonary congestion, moderate pleural effusion, and enlargement of the right ventricle and bilateral atrium.

managed with bisoprolol (2.5 mg/day), apixaban (5 mg/day), and metformin (500 mg/day) for several years. She had no history of smoking or alcohol consumption. Her initial examination showed a blood pressure of 172/92 mmHg, a heart rate of 151 bpm with an irregular rhythm, and a respiratory rate of 22 breaths per minute. The heart sounds were irregular with a systolic murmur at the apex.

Laboratory tests indicated renal dysfunction (serum creatinine 1.31 mg/dl, blood urea nitrogen 19 mg/dl) and elevated N-terminal

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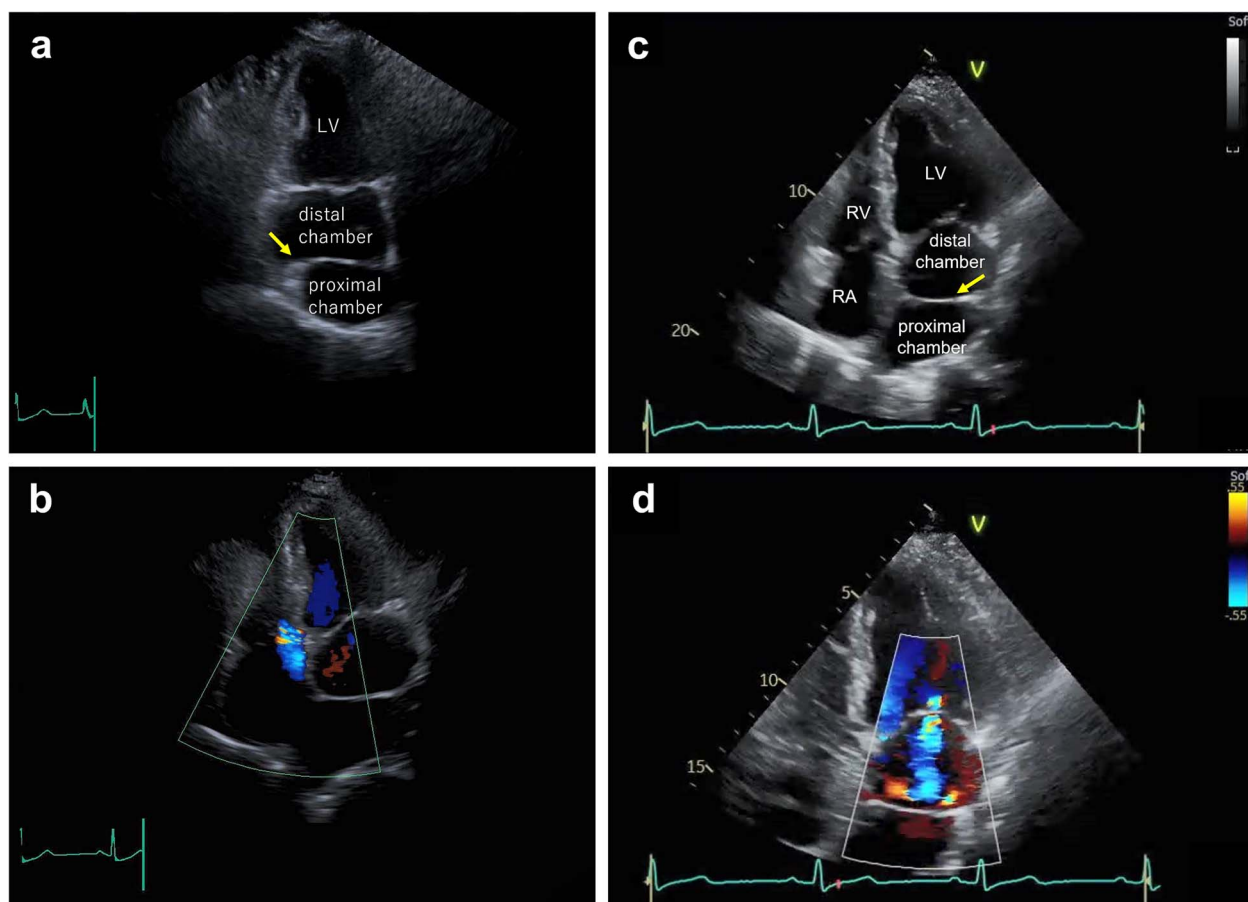


Figure 2. Transthoracic echocardiography on admission. Echocardiography shows dilation of the left atrium, which is divided by an abnormal septum (arrow, **a** and **c**) and moderate tricuspid and severe mitral valve regurgitation (**b** and **d**). LV, left ventricle; RA, right atrium; RV, right ventricle.

pro-Beta Natriuretic Peptide (NT-proBNP) levels of 2396 pg/ml. The electrocardiogram confirmed atrial fibrillation (Fig. 1a). A chest radiograph revealed cardiac enlargement (cardiothoracic ratio 69%) and bilateral pulmonary infiltrates with pleural effusion (Fig. 1b). Transthoracic echocardiography showed the left atrium (LA) was divided into proximal and distal chambers by an abnormal septum with severe dilation (LA diameter: 50 mm). The left ventricular (LV) ejection fraction was 60%, with mild dilation (LV end-diastolic diameter: 52 mm) and severe mitral valve regurgitation (Fig. 2). After administering diuretic therapy with furosemide, the patient converted to normal sinus rhythm and became asymptomatic. On the eighth hospital day, a transesophageal echocardiogram identified a single fenestration site on the median side of the LA with a mean pressure gradient of 2.6 mmHg between the chambers (Fig. 3a–c). Severe mitral valve regurgitation originating from the posterior leaflet regions 1 and 2 was noted, with a mild elevation in the pressure gradient between the distal chamber and the LV was mildly elevated at 1.0 mmHg (Fig. 3d). Cardiac magnetic resonance imaging on the ninth hospital day showed all pulmonary veins entering the proximal chamber of the LA and confirmed the presence of an abnormal septum with a 16 × 12 mm fenestration located in the median site, approximately 1.5 cm² in area (Fig. 4b). On the twelfth hospital day, right-heart catheterization confirmed the absence of intracardiac shunts and revealed a mild elevation in left atrial pressure with a high pulmonary capillary wedge pressure (PCWP, 14 mmHg), although the mean pulmonary artery pressure was within the normal range at 24 mmHg.

She declined all invasive treatments, including catheter ablation and balloon catheter dilation, despite our proposal for invasive open-heart surgery to remove the abnormal septum and alleviate her paroxysmal atrial fibrillation. Consequently, we opted for conservative management, which involved administering amiodarone (200 mg/day), furosemide (20 mg/day), and an increased dose of bisoprolol (5.0 mg/day) to control her paroxysmal atrial fibrillation and LA pressure. The patient is now under regular follow-up at our institution.

Discussion

To the best of our knowledge, this is the oldest reported case of a CTS patient successfully managed with a conservative approach. The clinical presentation of CTS closely relates to the size and number of fenestrations; a large single fenestration may remain asymptomatic, while narrower openings often present earlier with respiratory distress. Moreover, the severity of symptoms is intensified by the presence of an atrial shunt, leading to the classification of cor triatriatum into various groups and classes based on clinical features [2–5]. Regardless of classification, most patients with CTS typically require invasive surgical or transcatheter interventions [6, 7].

Additionally, multimodal imaging plays a crucial role in diagnosing and assessing the severity of CTS to determine the appropriate treatment. Specifically, transesophageal echocardiography was employed to accurately evaluate the size and location of the fenestration and the pressure gradient between the proximal and

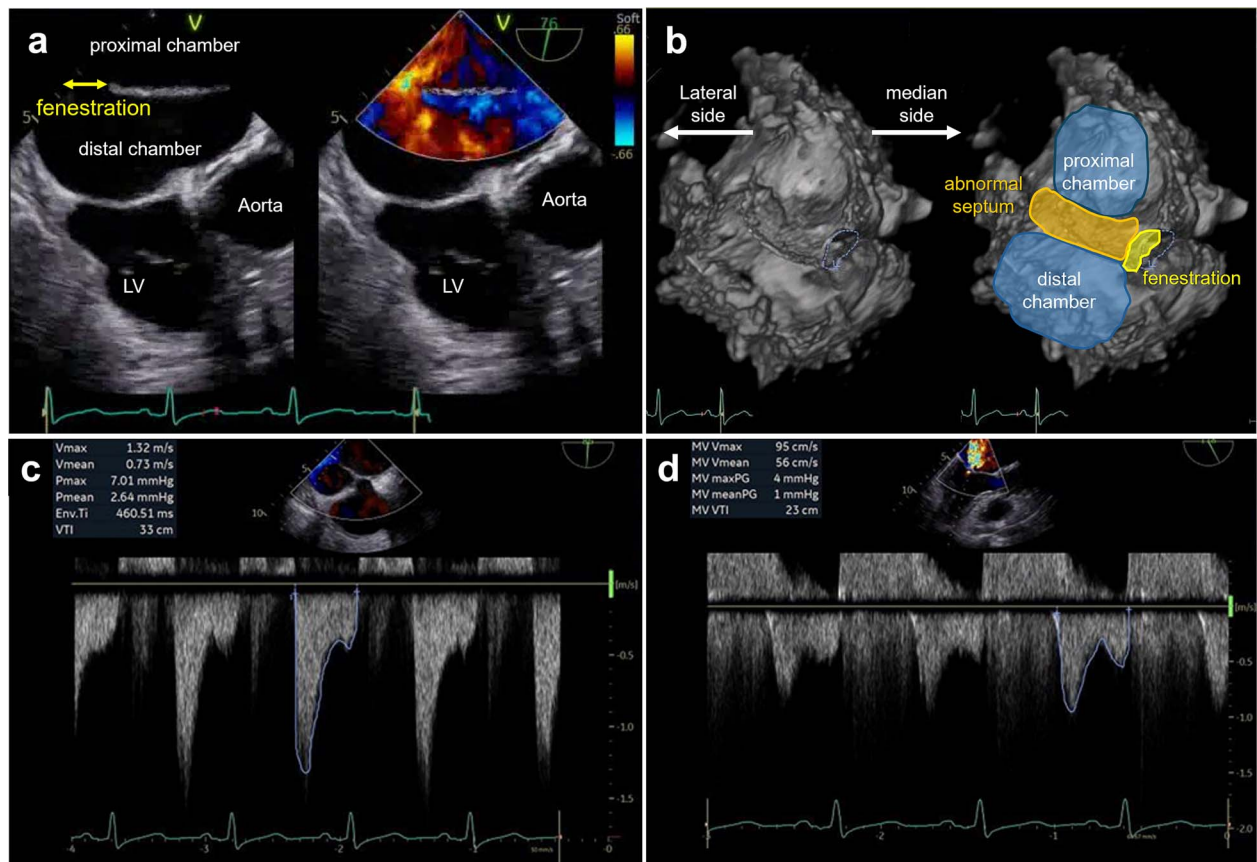


Figure 3. Transesophageal echocardiography after initial treatment. (a) The fenestration site was only detected in the median side of the left atrium. (b) Three-dimensional imaging of left atrium. The area of fenestration site was 1.5 cm². Continuous wave Doppler flow pattern between proximal and distal chamber with the pressure gradient at 2.6 mmHg (c), and between distal chamber and LV with the pressure gradient at 1.0 mmHg (d). LV, left ventricle; V max, velocity max; V mean, velocity mean; P max, pressure max; P mean, pressure mean; Env. Ti, envelope time; VTI, velocity time integral; PG, pressure gradient.

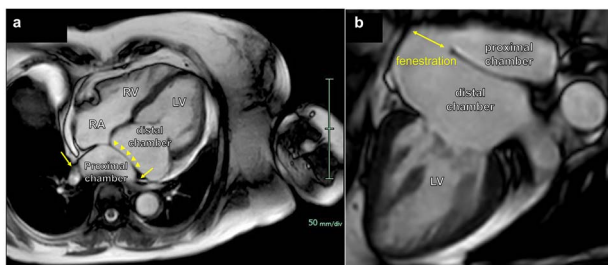


Figure 4. Cardiac magnetic resonance imaging after initial treatment. (a) All pulmonary veins opened into the proximal chamber of LA (arrows). The left atrium was divided into proximal and distal chambers by an abnormal septum (arrow heads). (b) The fenestration site was only detected in the median side of the left atrium. LV, left ventricle; RA, right atrium; RV, right ventricle.

distal chambers [8]. Furthermore, cardiac magnetic resonance imaging and right-heart catheterization are essential diagnostic tools used to ensure the patency of all pulmonary veins to the proximal chamber, assess for any shunt malfunction, and identify potential complications [9].

Multimodal assessment facilitated precise evaluation and treatment planning for this patient. With all pulmonary veins connecting to the LA above the septum and no evidence of a shunt, she was classified as type IA according to the Lucas and Schmidt classification [5]. Although her initial symptoms and bilateral pulmonary congestion indicated high left atrial pressure

with atrial fibrillation, analysis of the abnormal septum revealed a large fenestration measuring 1.5 cm², a modest pressure gradient of 2.6 mmHg, and slightly elevated pressures in the distal chamber and PCWP after achieving sinus rhythm with initial treatment. These findings suggest that preventing readmission hinges on managing paroxysmal atrial fibrillation and that conservative management with oral medications could prevent exacerbation.

According to guidelines, amiodarone, a potassium channel blocker effective in suppressing the formation of reentry circuits, is suitable for treating paroxysmal atrial fibrillation in patients with heart failure. This is particularly relevant as atrial fibrosis progresses, which facilitates the formation of reentry circuits throughout the atrium [10]. Amiodarone also has minimal negative inotropic effects and a low rate of renal excretion. Given this patient's presentation with heart failure, severe mitral regurgitation, CTS, a longstanding history of paroxysmal atrial fibrillation, and deteriorating chronic disease (serum creatinine 1.63 mg/dl, blood urea nitrogen 29 mg/dl) at discharge, amiodarone was an appropriate treatment for her.

Conclusion

This case of cor triatriatum sinister was identified at an advanced age and was successfully managed through conservative treatment, following a comprehensive evaluation using multiple modalities.

Consent

The patient provided written consent prior to the transcription of the manuscript.

Guarantor

Ryuta Sugihara.

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Conflict of interest

No conflict of interest.

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Ethical approval

No approval is required.

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