Case Report

Incidental Finding of Cor Triatriatum Sinistrum in a Middle-Aged Man Candidated for Coronary Bypass Grafting (with Three-D Imaging)

Afsoon Fazlinezhad, MD*, Farveh Vakilian, MD, Asadollah Mirzaei, MD, Azadeh Fallah Rastegar, MD

Ghaem Hospital, Mashhad University of Medical Sciences, Mashhad, Iran.

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Abstract

Cor triatriatum sinistrum is a rare congenital cardiac malformation, accounting for about 0.1-0.4% of all congenital heart diseases and characterized by the presence of a fibromuscular membrane that subdivides the left atrium into two chambers in the classical form. While classic cor triatriatum in most patients can be observed during the neonatal period or early infancy, it is very rare in adults.

We herein present an incidental finding of cor triatriatum sinistrum in a middle-aged man with coronary artery disease scheduled for coronary artery bypass graft surgery. The patient was admitted for exertional dyspnea and chest pain of a three-month duration. He had a past medical history of mild hyperlipidemia and mild hypertension. Transthoracic two-D echocardiography (TTE) demonstrated a visible presence of a membranous band in the mid portion of the left atrium with obvious obstruction by color and Doppler flow measurements, confirmed by three-D echocardiography. Selective coronary angiography also revealed a severe ostioproximal stenosis of the left anterior descending artery of up to 99%.

On-pump coronary artery bypass grafting was performed without complications, during which the anastomosis of the left internal mammary artery to the left anterior descending artery and the removal of the membrane were done.

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Introduction

Cor triatriatum sinistrum is a rare congenital cardiac anomaly and is responsible for 0.1 - 0.4% of all congenital cardiac malformations, without any known associated genetic abnormalities.[1, 2] The clinical manifestations of this defect are dependent on the size of the ostia.[3, 4] In adults, however, the clinical manifestations are often delayed due to the presence of a large opening. This is a report of cor triatriatum sinistrum diagnosed incidentally, late in the 5th decade of a man's life, during a preoperative study.

Case Report

This is a report of cor triatriatum sinistrum diagnosed

*Corresponding Author: ***Afsoon Fazlinezhad,*** *Associate Professor of Cardiology, Fellowship of Echocardiography, Echocardiography Department, Ghaem University Hospital, No. 38, 5th North Daneshsara Street, Sanabad Ave., Mashhad, Iran. 9184713153. Tel: +98 9153134470. Fax: +98 5118414773-4. E-mail: fazlinejada@mums.ac.ir.*

incidentally, late in the 5th decade of a man's life, during a preoperative study. A 42-year-old Asian male was admitted for exertional dyspnea and chest pain of a three-month duration. He had a past medical history of mild hyperlipidemia and mild hypertension, and his medications included enalapril, hydrochlorothiazide, metoprolol, and aspirin.

Cardiovascular examination revealed normal first heart sound and physiologically split second heart sound in conjunction with grade 2/6 systolic murmur at the left sternal border, which was intensified by inspiration. No significant laboratory abnormality was detected on admission. The patient's electrocardiogram (ECG) was normal and his previous exercise tolerance test was positive; he was, therefore, scheduled for selective coronary angiography and transthoracic two-D echocardiography (TTE) to be performed for left ventricular function study. TTE demonstrated a normal-sized left ventricle with a normal ejection fraction of about 60%; normal right ventricular size and function; a mildly enlarged left atrium; and a visible presence of a membranous band in the mid portion of the left atrium with obvious obstruction by color and Doppler flow measurements, confirmed by three-D echocardiography (Figure 1). The two-D findings were further confirmed by transesophageal echocardiography (TEE), which visualized an obstructing fibromuscular membrane distal to the pulmonary veins and proximal to the left atrial appendage in multiple views (Figures 2 and 3). In addition, the orifice was about 11 mm in diameter with a mean pressure gradient of about 5.94 mmHg (Figure 4). The findings of two-D echocardiography and TEE substantiated the diagnosis of cor triatriatum sinistrum. Selective coronary angiography revealed a severe ostioproximal stenosis of the left anterior descending artery of up to 99%.

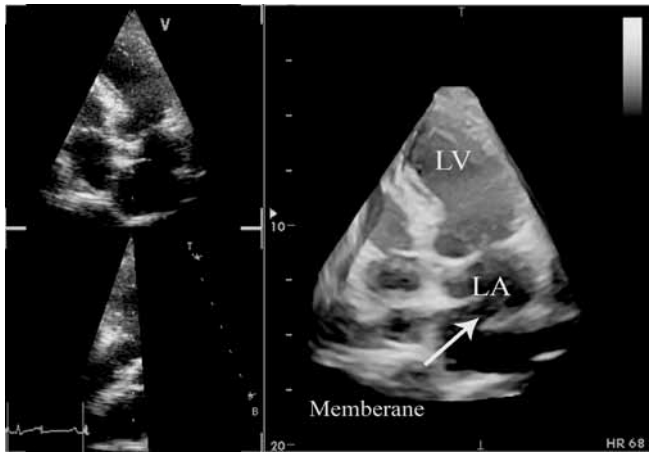

Figure 1. Transthoracic three-D echocardiogram showing the membrane in the left atrial cavity (arrow)
LV, Left ventricle; LA, Left atrium

The patient underwent on-pump coronary artery bypass grafting (CABG), during which the anastomosis of the left internal mammary artery to the left anterior descending artery and the removal of the membrane was performed without complications (Figure 5). The patient was discharged on the 6th postoperative day.

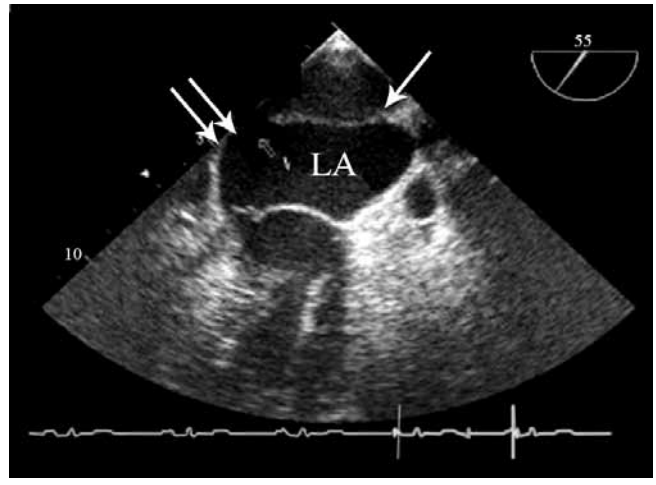

Figure 2. In transesophageal echocardiography, mid-transesophageal view demonstrates the dividing membrane in the left atrial cavity with a small orifice (11 mm) (arrow)
LA, Left atrium; AO, Aorta

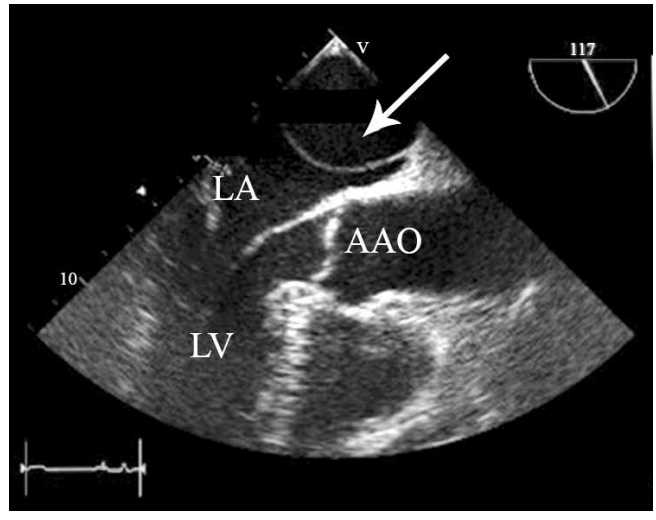

Figure 3. In transesophageal echocardiography, mid-transesophageal aortic valve long-axis view demonstrates an abnormal membrane across the patient's left atrium (arrow)
LV, Left ventricle; LA, Left atrium; AAO, Ascending aorta

Discussion

Cor triatriatum sinistrum is a rare congenital cardiac anomaly and is responsible for 0.1-0.4% of all congenital cardiac malformations, without any known associated genetic abnormalities.[1, 2] It has gender predilection with

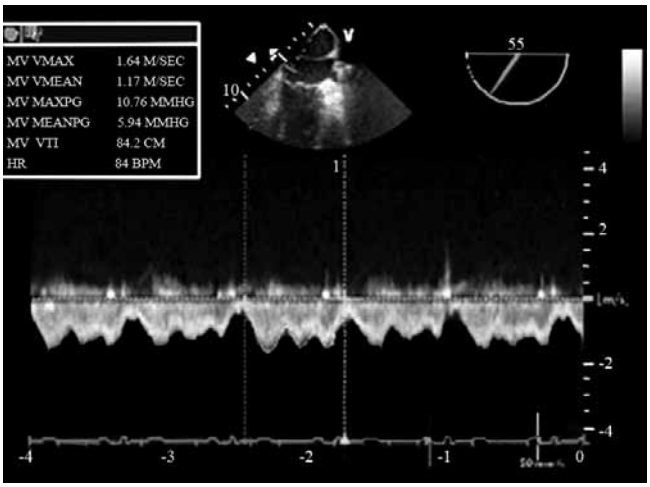

Figure 4. Doppler flow imaging across the membrane orifice (transesophageal echocardiography (view)

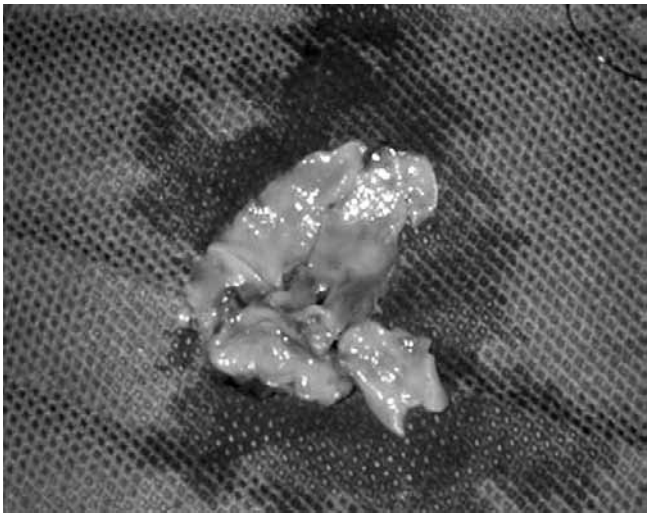

Figure 5. Macroscopic view of the removed left atrial membrane from the pulmonic veins aspect (5.5 × 4 cm)

slight male predominance involvement (1.4:1) and is associated with other cardiac defects in up to 50% of cases. Examples of associated cardiac defects include atrial septal defect, persistent left superior vena cava, partial anomalous pulmonary venous connections, ventricular septal defect, and more complex cardiac lesions such as the tetralogy of Fallot, atrioventricular septal defect and double outlet right ventricle.

In its most common form of cor triatriatum sinistrum, the left atrium is divided into proximal and distal chambers. Both chambers are separated by a diaphragm with one or more restrictive ostia, with the pulmonary veins draining into the proximal chamber. The location of the atrial appendage is a key landmark in this congenital malformation: In cor triatriatum, the atrial appendage is invariably connected with the lower chamber, which is below the membrane.[3]

When the atrial septal defect exists between the left atrial accessory chamber and the right atrium, the patients refer to hospital with symptoms of associated elevated pulmonary venous and arterial pressures because blood is shunted from left to right[3, 4] In our case, the patient only had a history of dyspnea on exertion and there was no evidence of intracardiac shunt.

The primary concern with cor triatriatum sinistrum is the potential for the left ventricle inlet obstruction, leading to mitral stenosis physiology. It is deserving of note that the pulmonary venous inflow can be compromised, albeit rarely,[5] but there was no evidence of the left ventricle inlet obstruction in our patient. The chest radiograph usually shows increased vascular markings, and a cardiac murmur is frequently noted. Be that as it may, with cor triatriatum sinistrum, the apical diastolic rumble of mitral stenosis is generally absent. Some patients with cor triatriatum sinistrum may remain asymptomatic, whereas others may have late onset of symptoms, which is possibly related to the fibrosis and calcification of the membrane with associated atrial fibrillation or mitral regurgitation.[6]

ECG findings are non-specific, but may reveal right-axis deviation with right atrial and right ventricular hypertrophy and atrial arrhythmia in some patients. Echocardiography is often sufficient for the diagnosis and TEE is the diagnostic modality of choice. Three-D echocardiography is also a new modality for the diagnosis and enjoys high accuracy.[7-10] The only treatment is surgical correction, and the majority of postoperative deaths occur in the first 30 days. In the early surgical series, the mortality was as high as 15-20% but more recently the rates have reached as low as 2%.[11, 12] Long-term results are excellent, with survival rates of 80-90% in patients surviving surgery.[11-14] Our patient also underwent a successful surgical operation and he was well at the first-month follow-up.

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