

Cor Triatriatum with Mitral Stenosis: A Diagnostic Dilemma

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Abstract

Cor triatriatum sinister is an extremely rare congenital heart defect. Very few cases have been reported for cor triatriatum associated with mitral regurgitation. We are reporting an exceptional case of cor triatriatum associated with calcified mitral valve resulting mitral stenosis. Preoperatively, mitral stenosis was underestimated due to the presence of restricted communication between two chambers of the left atrium. The patient was operated for cor triatriatum repair with mitral valve replacement.

Keywords: Cor triatriatum sinister, mitral valve replacement, posterior annular calcification, rheumatic mitral stenosis

INTRODUCTION

Cor triatriatum sinister is an extremely rare congenital heart defect.^[1] In classical form of cor triatriatum, there is a division of the left atrium by a fibromuscular membrane, band or tissue, resulting in the development of two distinct chambers, that is, a posterosuperior chamber and anteroinferior chamber. It is even more uncommon in adults and quite exceptional when associated with mitral valve disease.^[2] Very few cases have been reported for cor triatriatum associated with mitral regurgitation.

Here, we are reporting an exceptional case of cor triatriatum associated with calcified mitral valve resulting mitral stenosis requiring mitral valve replacement.

CASE REPORT

A 65-year-old female presented with a history of dyspnea and angina on exertion (New York Heart Association Class II) for the past 2 years and progressed to the New York Heart Association Class IV recently. On clinical examination, the patient had a regular heart rate of 80/min with blood pressure of 130/90 mm of mercury (mmHg).

Electrocardiogram confirmed normal sinus rhythm. Chest radiography showed prominent bronchovascular markings [Figure 1].

Transthoracic echocardiography revealed cor triatriatum; a membrane dividing left atrium into posterosuperior venous chamber and anteroinferior mitral chamber. There was a small communication between two chambers. On Doppler flow mapping, mitral valve inflow signal (pg/mg) was 7/3 mmHg. Color and pulse wave Doppler suggested moderate mitral stenosis and mild mitral regurgitation with mild pulmonary arterial hypertension with ejection fraction of 55%. Above findings were confirmed on transesophageal echocardiography.

For further assessment of pulmonary venous anatomy and mitral valve pathology, computerized tomographic pulmonary angiogram was advised. Multislice computed tomography (CT) pulmonary angiography was performed

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after intravenous injection of nonionic contrast which showed the left atrium divided by a thin membrane, suggestive of cor triatriatum. (Loeffler's Type 2 – small restrictive opening with calcification). All pulmonary veins were draining into the posterior chamber of the left atrium [Figure 2a-d]. Mitral posterior annular calcification was found [Figure 3]. Preoperative conventional coronary angiography revealed a right dominant system with no evidence of coronary artery disease. The patient was advised for surgical correction of cor triatriatum.

Surgical technique

The lesion was approached by a midline sternotomy and pericardiotomy. Cardiopulmonary bypass was established using ascending aortic and bicaval cannulation technique. After complete myocardial protection with moderate hypothermia, we approached cor triatriatum through interatrial groove incision.

A large membrane was dividing the left atrium into two compartments posterosuperior and anteroinferior. Both

chambers were connected through a restricted, calcified communication across membrane. After complete excision of the membrane, mitral valve was examined. Both mitral commissures were fused. There was a pinhole mitral valve orifice. Posterior annular calcification was present, which was underestimated preoperatively. Subvalvular apparatus was abnormally shortened. Mitral valve was excised and replaced with 25-mm St. Jude Master's series mechanical prosthesis.

Postoperative recovery was uneventful and the patient was discharged after 1 week of postoperative stay. Postoperative transthoracic two-dimensional echocardiography showed satisfactory findings. Follow-up contrast CT thorax, before discharge, showed no residual membrane in the left atrium with normally functioning mitral valve prosthesis [Figure 4a and b].

DISCUSSION

Cor triatriatum sinister was first described by Church in 1868. It is found in only 0.1% of cases of congenital heart anomaly. In classical form of cor triatriatum, there is a division of the left atrium by a fibromuscular membrane, band or tissue,

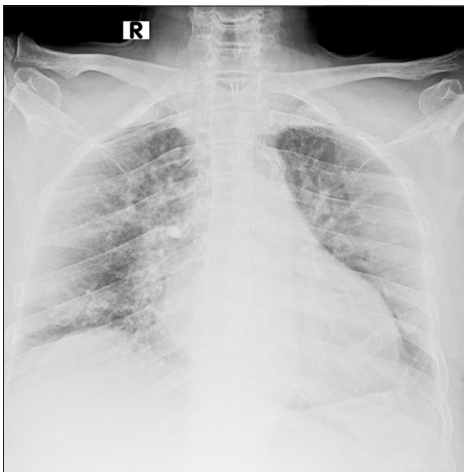


Figure 1: Preoperative chest radiograph showing prominent bronchovascular markings



Figure 3: Preoperative computed tomography pulmonary angiogram showing posterior annular calcification of mitral valve

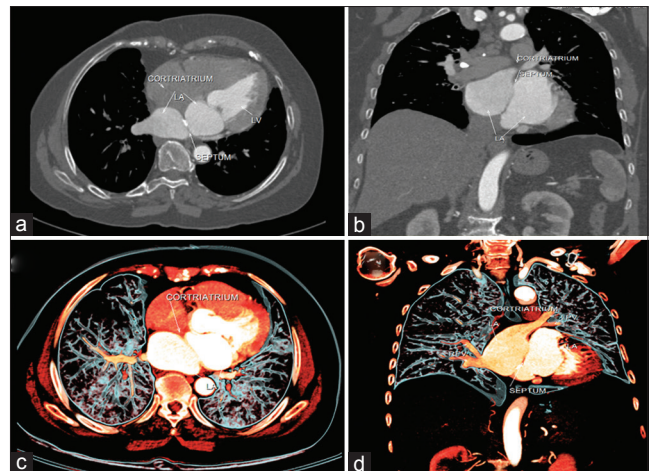


Figure 2: (a-d) Preoperative computed tomography pulmonary angiogram showing cor triatriatum Loeffler's Type 2 – small restrictive opening with calcification

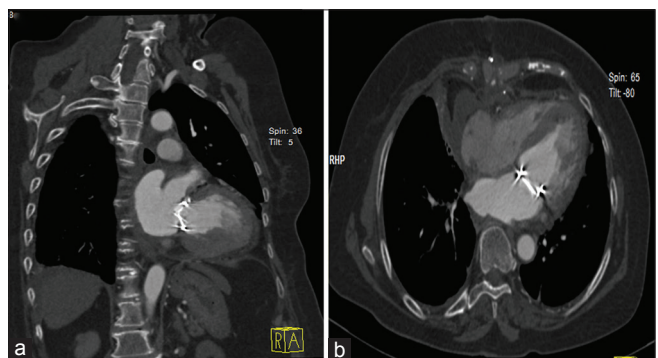


Figure 4: (a and b) Postoperative contrast-enhanced computed tomography thorax showing no residual membrane with mitral valve prosthesis *in situ*

resulting in the development of two distinct chambers, that is, a posterosuperior chamber and anteroinferior chamber. The posterosuperior chamber receives the return of all pulmonary veins. The anteroinferior chamber is a true left atrium which communicates with mitral valve.

Embryological basis for development of cor triatriatum is related with anomalous or failure of inclusion of the pulmonary veins into the posterior left atrial wall.^[3] It is usual for patients to present in infancy and early childhood, although some cases remain undetected until adult life.

Natural history depends on the effective aperture size between two chambers of the left atrium. In severely restricted form, presentation occurs in early in infancy. Without surgical intervention, patients die early in infancy.^[4,5] Approximately, 75% of persons born with classic cor triatriatum die in infancy.

Pathophysiologically, the obstructive nature of the membrane leads to the creation of a pressure gradient with an associated rise in pulmonary venous and arterial pressures. As a result, clinical presentation often mimics mitral stenosis. At present, the reasons for late presentation are poorly understood. Large unrestricted communication through fossa ovalis between two atria improves the prognosis and delays presentation.

The development of fibrosis and calcification in the opening of the separating diaphragm, mitral insufficiency or mitral stenosis, posterior annular calcification, and the development of atrial fibrillation are some of the various triggers that lead on to the symptoms.

Cor triatriatum sinister remains an uncommon form of congenital heart disease although it is being diagnosed with increasing frequency in adults due to improvements in diagnostic imaging. Wong *et al.*^[6] reported, for the first time in 1989, a 49-year-old male patient with the rare combination of cor triatriatum sinister associated with severe mitral regurgitation and abnormal tensor apparatus of the mitral valve

with hypoplastic papillary muscle. This diagnosis should be considered in all patients presenting with signs or symptoms of mitral stenosis.

Management in such cases differs from isolated cor triatriatum, in that, in addition to the surgical resection of accessory membrane, the patient requires mitral valve repair or replacement for correction of mitral valve pathology.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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