Cor triatriatum with persistent levoatrial cardinal vein late presented as severe mitral stenosis

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Cor triatriatum is a rare congenital cardiac anomaly that usually becomes symptomatic in the first years of life. We present a 28-year-old pregnant female patient presented with shortness of breath, palpitations and decreased exercise tolerance. Transthoracic (TTE) and transesophageal (TEE) echocardiography showed cor triatriatum, features of severe mitral stenosis and atrial septal defect (ASD). The patient underwent successful surgical correction with an uneventful postoperative course.

Keywords: Cor triatriatum, Mitral stenosis, Levoatrial cardinal vein

Introduction

Cor triatriatum is a rare but surgically correctable congenital cardiac anomaly. In the present patient the symptoms of mitral stenosis led to the discovery of a left atrial membrane. Surgery is the primary option for correcting such a disorder.

Case report

A 28-year-old female had a clinical picture of heart failure during her first pregnancy. She presented to us with palpitations, shortness of breath that was aggravated by exercise even with minimal physical activity. Physical examination revealed a well-developed 28-year-old female. Blood pressure and heart rate were normal. Cardiac auscultation revealed a diastolic murmur at the apical region. Her room air oxygen saturation was 98% by pulse oximetry. Her hemoglobin was 14.8 g/L and hematocrit 36%. Electrocardiogram showed normal sinus rhythm. Transthoracic (TTE) and transesophageal (TEE) echocardiography revealed: cor triatriatum with a stenotic membrane across the cavity of the left atrium, dividing the left atrium in two chambers, a bigger proximal chamber draining by a narrow orifice of 0.8 cm^2 (Fig. 1A) to a distal chamber drained by mitral valve (Fig. 1B). The pressure gradient across the membrane was 35/14 mmHg (Fig. 1C). The left atrial appendage drained into the distal chamber, and all the pulmonary veins drain into the proximal chamber. Pulmonary artery pressure was 50 mmHg. The atrial septal defect (ASD) was communicating the proximal chamber with the right atrium and was considered as a type A1 cor triatriatum according to Lam Classification [1]. Table 1. The patient was referred for surgical correction.

The patient was under close follow up by our cardiology team. The pregnancy continued a normal

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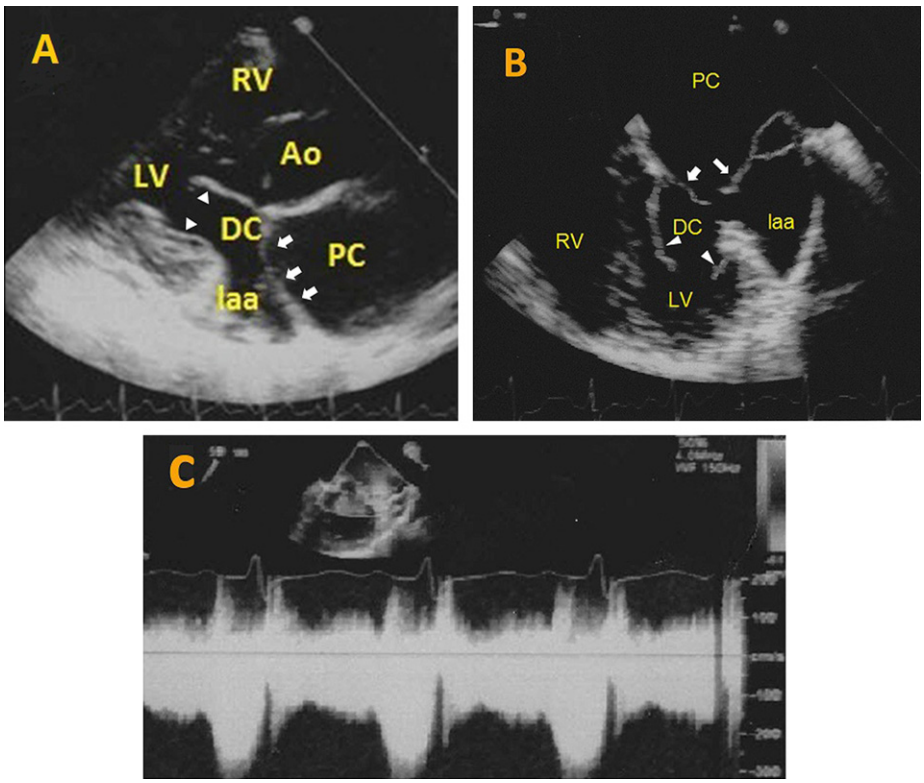

Figure 1. (A,B) Echocardiogram showing an accessory membrane dividing the left atrium into a larger proximal chamber (PC) and smaller distal chamber (DC). The accessory membrane is as thick as the mitral valve leaflets. The left atrial appendage (laa), aorta (Ao), left ventricle (LV), and right ventricle (RV) also are shown. (C) Continuous wave Doppler demonstrates increased velocity, suggesting severe obstruction through a restrictive opening.

Table 1. Lam classification of cor triatriatum.

Type	Description
A	Proximal chamber receives all the pulmonary veins; distal chamber contains the left atrial appendage and the mitral valve. No atrial septal defect
A1	Atrial septal defect between the right atrium and proximal chamber
A2	Atrial septal defect between the right atrium and distal chamber
B	Pulmonary veins drain into the coronary sinus (a form of total anomalous pulmonary venous connection)
C	No connection between the pulmonary veins and the proximal chamber (rarest variant)

course until full-term. She underwent cesarean section with epidural anesthesia and close hemodynamic monitoring. Six months after her delivery, she was admitted electively for surgical correction.

The operation was conducted via a midline sternotomy. The ascending aorta and both vena cavae were cannulated directly. Cardiopulmonary bypass under moderate hypothermia (32 °C) was instituted. The heart was arrested by antegrade cold blood cardioplegia. We inspected the anatomy of the heart, we found that there was a levoatrial cardinal vein, which was coming from the innominate vein and draining in the proximal chamber together with the left pulmonary veins. The levoatrial cardinal vein was controlled with a tape and snared without any change in systemic O2 saturation or blood pressure. A right atriotomy was performed. There was a postero-inferior ASD 1×0.5 cm in size that was communicating the proximal chamber with the right atrium. The pulmonary veins were normal in position and were draining into the proximal chamber. The incision was made in the interatrial septum and the membrane was exposed and was resected in total. Macroscopically, the membrane appeared endothelial on the distal chamber surface with layers of muscular tissue on the proximal chamber side. The levoatrial cardinal vein was ligated. The mitral valve leaflets looked normal, and the valve was tested with injection of cold saline into the left ventricle. Postoperative TEE in the operative room showed a competent mitral valve. No mass or membrane in the left atrium. The postoperative

course was uneventful. The patient was discharged on seventh postoperative day.

Discussion

Cor triatriatum can occur in isolation or in association with other congenital cardiac anomalies. The three main embryological theories explaining the development of cor triatriatum are malseptation, malincorporation, and entrapment [2–4]. Typically, the wall between the two chambers is bulky and contains muscular tissue and has one or multiple openings in it. The ASD is usually present and may be in the septum between the right atrium and either the proximal or distal left atrial chambers.

The natural history of cor triatriatum depends on the effective size of the opening in the diaphragm and the presence and location of the interatrial communication [5]. In patients with large communications the diagnosis may present in adulthood. The suggested reasons for late conversion to a symptomatic state are accepted to be caused by several factors: fibrosis and calcification in the opening of the separating diaphragm, the development of mitral insufficiency, and the development of atrial fibrillation [5]. Most cases of cor triatriatum presenting in adulthood are initially misdiagnosed as mitral valve stenosis [6,7]. Our patient remained asymptomatic for almost 28 years despite a small communication between the two chambers. The ASD and the presence of the levoatrial cardinal vein might have masked the condition through decompressing the pulmonary veins. She developed decreased exercise tolerance in the absence of significant respiratory compromise.

This clinical picture was probably a consequence of increased pulmonary venous pressure on exercise due to left ventricular inflow obstruction and major hemodynamic alterations occur during pregnancy. These changes begin to take place during the first five to eight weeks of pregnancy and reach their peak late in the second trimester. In patients with preexisting cardiac disease, cardiac decompensation often coincides with this peak.

A levoatrial cardinal vein coexists with cor triatriatum more frequently than with any other type of congenital heart disease [5]. In our patient, the connection between the levoatrial cardinal vein and proximal chamber was unrestricted, and simple ligation of the vein was required. In patients presenting late in life with cor triatriatum with severe obstruction to mitral valve inflow and a small atrial communication, or in those with only one left or right pulmonary vein clearly seen draining into the proximal collecting chamber, the presence of another venous to systemic circulation communication should be investigated.

Earlier surgical series required cardiac catheterization to establish the diagnosis of cor triatriatum [8]. Since the 1990s echocardiography has become the diagnostic modality of choice for classic cor triatriatum [8]. We were able to achieve a correct preoperative diagnosis in our case with the help of TTE and TEE. However, we strongly suspected another venous to systemic circulation communication, and we discover intraoperative the presence of a levoatrial cardinal vein that was not diagnosed using echocardiography. We did not perform cardiac catheterization in this case.

Cor triatriatum can remain asymptomatic until adulthood; however, patients with obstructive symptoms at any age should undergo surgical correction. Treatment of cor triatriatum is primarily surgical despite the several reports of successful balloon catheter dilation of the communication between the proximal and distal chambers. The surgical approach can be through either a median sternotomy or a right thoracotomy. We feel that a transeptal approach provides an excellent view while remaining technically easier. A successful outcome depends on complete excision of the diaphragm between the proximal and distal chambers. In conclusion, cor triatriatum is a rare congenital cardiac anomaly. Surgical correction offers good early and long-term results for both classic and atypical cor triatriatum.

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