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Case Report

A Very Rare Case of Cor Triatriatum with Severe Mitral Regurgitation

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Keywords: Cor triatriatum; Congenital heart disease; Mitral regurgitation

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Abstract

BACKGROUND: Cor triatriatum sinister is rare congenital heart disease. It is mainly presented in childhood and often accompanied with other congenital anomalies. The cases with cor triatriatum treated surgically in adults and accompanied with severe mitral regurgitation are very rare.

CASE REPORT: We present a case with diagnosed cor triatriatum and severe mitral regurgitation. The diagnose was made by echocardiography. She was a female 25 years that was hospitalised with signs of heart failure NYHA II-III.

CONCLUSION: We performed the resection of the membrane in the left atrium and repair of a mitral valve according to Alfieri. The patient did very well after the surgery.

Introduction

Cor triatriatum sinister is rare congenital heart disease. This pathology was first described by Church in 1868. This anomaly is mainly presented in childhood and most often is accompanied with DIA, VSD, TF, partial abnormal pulmonary return [1]. The cases treated surgically in adults and accompanied with severe mitral regurgitation are very rare. In our knowledge, there are reported some cases of surgical treatment of cor triatriatum and severe mitral regurgitation. We present a case diagnosed and treated for cor triatriatum and mitral regurgitation at the adult age. We performed the resection of the membrane in the left atrium and repair of a mitral valve according to Alfieri technique. The patient had very good progress after the surgery.

Case Presentation

The patient of 26 years old was admitted to our clinic with the diagnosis: Cor triatriatum sinister and severe mitral regurgitation. Heart failure NYHA II-III.

The main complaints were weakness and dyspnea in minimal to moderate physical efforts with a history of about four months. The patient performed ambulatory transthoracic echocardiography and was diagnosed with cor triatriatum sinister and severe mitral regurgitation. In these conditions was hospitalised at our clinic. In electrocardiogram was normal sinus rhythm. All laboratories examinations were normal.

We performed in hospital trans-oesophagal echocardiography that resulted:

Severe mitral regurgitation, myxomatous degeneration of both leaflets of the mitral valve with prolapse of the anterior leaflet, dilated left atrium and there was a membrane that divides the left atrium into two parts. There were communication between two parts.

The left ventricle was dilated. The diameters were DTD/DTS respectively 65/45 mm. The ejection fraction of LV was estimated about 45%.

The right chambers were dilated. The pulmonary artery systolic pressure was approximately 40 mm Hg. No other structural anomalies are noticed (Fig. 1 from echocardiography).

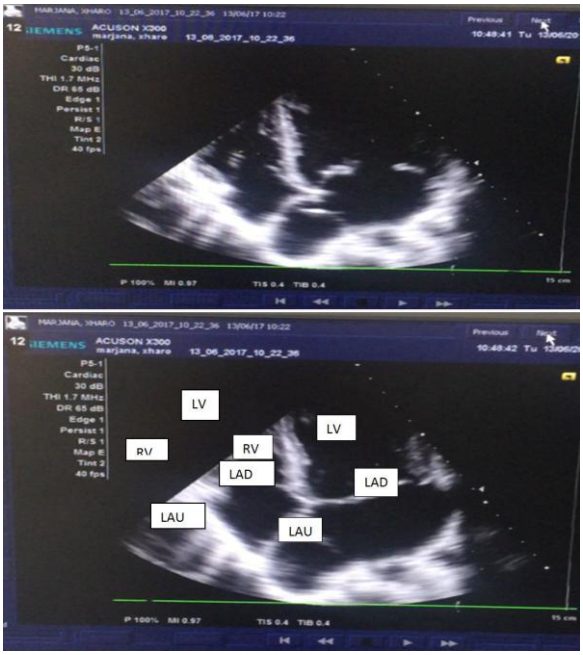

Figure 1: LV-Left ventricle, RV-right ventricle, LAD-left atrium distal chamber, LAU-left atrium upper chamber. Between LAU and LAD, we can distinguish the membrane in the left atrium

Fig. 2 is the schematic presentation of Cor triatriatum during the surgery [17].

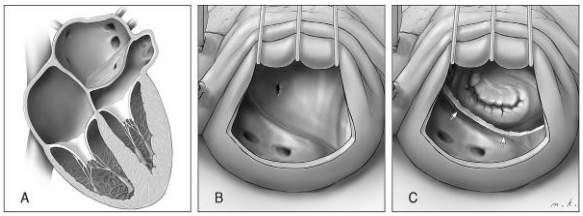

Figure 2: (A) Cor triatriatum defect without any associated atrial septal defect; (B) Operative exposure of cor triatriatum membrane through a left atriotomy. Restrictive communication between the proximal and the distal left atrial chambers is visible; (C) Exposure of mitral valve after the excision of cor triatriatum membrane (arrows mark the line of excision Ann Thorac Surg 2014; 97:1659–63)

The intervention was performed under extracorporeal circulation and moderate hypothermia. When the left atrium was opened, we notice a fibromuscular membrane, which divided the left atrium into two rooms, upper room, where the pulmonary veins were drained and the lower room where there were auricle and mitral valve. The hole in the membrane that made the communication between the two rooms had a diameter of about 1.2 cm (Fig. 3). It was impossible to see the mitral valve without resection of the membrane. The membrane was resected, and the mitral valve was exposed. During the examination of the valve, we noticed myxomatous degeneration of mitral valve leaflets with important prolapse of anterior leaflet macroscopically. We performed mitral valve repair according to Alfieri technique.

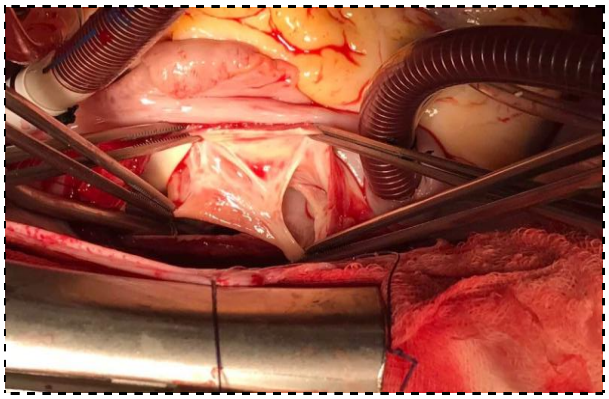

Figure 3: After we opened LA we see the membrane that divided LA into two parts upper and distal parts. Between four forceps has located the membrane. Between the right forceps is located the hole of the membrane of Cor Triatriatum

The patient did very well postoperatively. Postoperative echocardiography showed no leak and no transvalvular gradients of the mitral valve.

Discussion

Cor triatriatum is a very rare congenital heart disease. The incidence ranges 0.1 to 0.4% of heart congenital heart diseases [3] [4] [5] [6] [7] [8] [9] [10] [11]. Classic (or typical) cor triatriatum, or cor triatriatum sinister, is a rare congenital cardiac anomaly in which the pulmonary veins typically enter a “proximal” left atrial chamber separated from the “distal “ left atrial chamber by a diaphragm in which there are one or more restrictive ostia. This pathology was first described by Church in 1868. The most frequent concomitants anomalies are partial anomalous pulmonary veins return, unroofed coronary sinus, VSD, CoA, AVD, TF, asplenia polysplenia [1]. The first surgical treatment is referred by Lewis [2]. The surgical treatment series referred to this anomaly are small. In the near past, the experience of surgical treatment referred about 250 cases. Mayo Clinic in 50 years’ experience refers 25 cases of surgical treatment of cor triatriatum [17]. Another reference

centre reports 28 cases in 23 years of experience [10] while the cases of surgical treatment in adults with concomitant severe mitral regurgitation are very rare. In our knowledge there are only 9 reported cases [3] [4] [8] [12] [13] [14] [15] [16].

The patients are generally presented in childhood, and symptomatic presentation depends on the size of the communicative hole. The classic clinical of the anomaly is similar to that of mitral valve stenosis. The presentation in adulthood is very rare occurred [1]. Our case is presented with signs of heart failure that may result from mitral regurgitation, congenital anomaly or both. We think that both causes resulted in diffused hypokinetic and dilatation of left ventricle.

Currently, the standard diagnostic tool for cor triatriatum is transthoracic or transesophageal echocardiography. Catheterization can be used when we are not sure about simultaneous congenital anomalies. [1] Nowadays, the use of magnetic resonance imaging also is growing up as a diagnostic tool [18] [19] [20] [21]. In our case, the diagnosis began with transthoracic echocardiography and was clear after we performed preoperatively transesophageal echocardiography.

The surgical treatment of a case with court triatriatum and severe mitral regurgitation was first reported by Wong et al. in 1989. Membrane resection and mitral valve replacement were performed [2]. Jayaprakash et al. on 2015 present a case similar to the one where an Alfieri mitral valve repair realised [16]. The authors in both cases referred that they founded anomaly of the mitral valve tensor apparatus. In our patient, we didn't notice something special except macroscopical signs of myxomatous degeneration of mitral valve. In reported of such cases there were performed 4 replacements and 5 mitral valve repairs simultaneous with resection of the membrane of cor triatriatum.

We performed a resection of fibro muscular membrane and mitral valve repair according to Alfieri. It was implanted a mitral ring Eduards Lifesciences No. 34. The period after surgery was very good.

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