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Case Report

Cor triatriatum sinister, a case report

José Luis Conde Salazar[1,a], Aldrix Josué Sisniegas Razón[1,a], Franz Soplopuco Palacios[1,b]

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Author´s afiliation
[1] Cardiovascular Surgery, Instituto Nacional Cardiovascular – INCOR. Lima, Peru.
[a] Resident Physician in Cardiovascular Surgery
[b] Thoracic and cardiovascular surgeon

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Correspondence
Jose Luis Conde Salazar
Calle Chinchaysuyo Mz Q Lot 79, ENACE. Lima, Perú
+51 988788282

Email
jl.conde.08@gmail.com

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ABSTRACT

We report a case of cor triatriatum sinister associated with anomalous pulmonary venous drainage in an adult patient who presented with palpitations, lower-limb edema, dyspnea, orthopnea, bendopnea and ascites. The clinical presentation began with episodes of atrial fibrillation, associated with readmissions for right heart failure, so angiotomography and transesophageal echocardiography were requested, which led to the final diagnosis. The surgical approach was performed by total excision of the multiple fenestrations of the fibromuscular septum and double valvular plasty due to severe mitral and tricuspid insufficiency, which improved the patient's clinical condition. It´s important to consider this acyanotic congenital heart disease in the differential diagnosis of right heart failure´s etiology originating in the left atrium.

Keywords: Acyanotic Congenital Heart Disease; Cor Triatriatum; Atrial Membrane.

Introduction

Cor triatriatum, an acyanotic congenital anomaly of atrial septation, can be located to the left *(cor triatriatum sinister: CTS)* or right *(cor triatriatum dexter: CTD)*. CTS, initially described in 1868, represents 0.1 - 0.4% of all patients with congenital heart diseases; CTD, described in 1875, is rarer and its prevalence is unknown [1]. The possible development of this membrane would be the incorrect embryological fusion of the primitive pulmonary vein or an entrapment of primitive structures of the venous sinus [2], but the embryological basis has not been established [3]. On the other hand, it has also been considered that the cause could be an anomalous persistence of the left superior vena cava. Histologically, it corresponds to a fibromuscular septum that inserts medially in the margin of the fossa ovalis and laterally can join the left atrial appendage. This membrane is incomplete and can vary from having an unique wide central defect, to multiple fenestrae [4]. Thus, this structure forms two compartments (anteroinferior and posterosuperior), the posterosuperior chamber is communicated with the pulmonary veins and receives their drainage and the anteroinferior chamber is in contact with the mitral valve and contains the left atrial appendage [5].

The natural history depends on the obstructive physiology and congenital anomalies associated with CTS. In the obstructive

form, begins as heart failure and pulmonary hypertension. The severity of the heart failure is related to the degree of obstruction produced by the membrane, infrequently the venous drainage to the anteroinferior chamber can be completely occluded. There is no preference for sex, the age of diagnosis in the adult population is usually between 30-60 years, with infantile onset in those with obstructive membrane [6]. We present the case of a patient with natural evolution of CTS with permanent atrial fibrillation, severe functional mitral insufficiency, severe tricuspid insufficiency with annuloectasia and with adequate response to surgical treatment.

Case Report

A 47-year-old man from Lima, with a history of diabetes mellitus ten years ago and regular treatment with metformin (850mg per day). He was diagnosed with atrial fibrillation and was managed with bisoprolol (5mg per day) and rivaroxaban (20mg per day); later he was diagnosed with CF-II dyspnea, ascites, and lower limb edema that required repeated hospitalizations and was compensated with diuretics (spironolactone 25mg per day, furosemide 40mg every 12 hours) without a specific diagnosis. Two years ago, ascites increased, and dyspnea progressed to CF-III, orthopnea, bendopnea, and early fullness were added, so he was hospitalized for further studies.

Angiotomography **(Figure 1)** showed a dilated left atrium (diameter 50mm, area 45cm2) and a membrane located 13mm above the anterior leaflet of the mitral valve extending beyond the origin of the atrial appendage and occupying 90% of the atrial lumen. The interatrial septum was found to be intact, and close to it, the left atrial membrane left a communication at the level of the lower atrial wall. The dimensions of this communication were 1.9cm2 in area, with 19x12.5mm longitudinal and transverse diameters, respectively. The described membrane was mobile and partially calcified and had 2 adhesions at its upper end. The pulmonary venous drainage was unobstructed and consisted of 5 pulmonary veins (3 right and 2 left) draining into the left atrium, so we made the diagnosis of obstructive CTS. At this time the right catheterization did not find pulmonary hypertension.

Eight months later a transesophageal echocardiography (TEE) was performed **(Figure 2)** where restrictive CTS was evidenced with a maximum gradient of 5 mmHg, and whose membrane left an effective area of 0.75cm2 close to the atrial septum. The atrial appendage had chicken wing morphology and an emptying velocity of 36 cm/s. There was evidence of moderate mitral insufficiency (2 jets) mixed type (mild degenerative and predominantly functional) with a 36 x 44 mm annulus and monophasic filling pattern. Severe functional tricuspid insufficiency was also observed with 8 mm vena contracta and 48 mm annulus; left ventricular ejection fraction 57%; dilated right ventricle with 61 x 66 x 84 mm basal, medial and longitudinal diameters, respectively, and its shortening fraction in 38%. A high probability of pulmonary hypertension was evidenced so right cardiac catheterization was performed in which isolated postcapillary pulmonary hypertension was evidenced (PAPm 38 mmHg, PAC 25 mmHg, RVP 1.5 U Wood) in the left catheterization, the coronary arteries without significant lesions.

At the cardiovascular surgery meeting, a surgical approach was decided with the final recommendation of membrane resection and mitral tricuspid plasty.

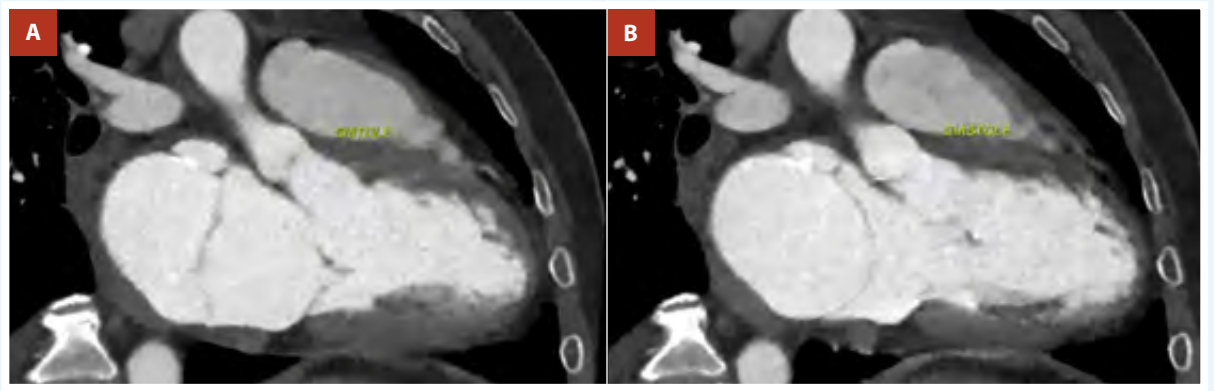

Figure 1. Contrast tomography of the heart and great vessels:**A)** Left atrial membrane moving away from the mitral valvular plane during systole phase. **B)** Left atrial membrane approaching the mitral valve plane during diastole phase..

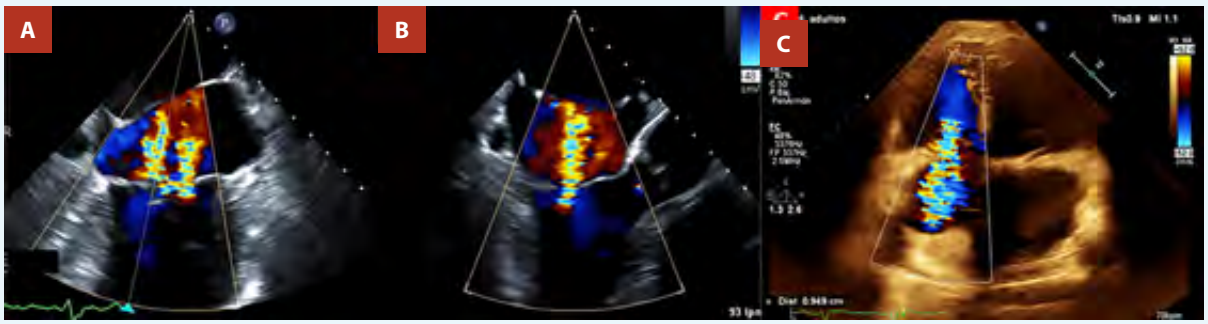

Figure 2. A) Transesophageal echocardiography mid esophageal view two chambers, color Doppler AL shows two jets of mitral regurgitation, plus membrane in left atrium and **B)** mid esophageal view long axis, showing turbulence jet in left atrium with obstructive atrial membrane type. **C)** Apical view of four chambers with color Doppler at tricuspid valve level showing jet of severe tricuspid regurgitation.

Surgical technique

Median sternotomy, central aorto-bicaval cannulation, left atrial approach through the sondergaard groove and transseptal total excision of the multifenestrating fibromuscular membrane that occupied 90% of the atrial diameter were performed. When mitral annulus dilatation was evidenced, mitral annuloplasty was performed with a number 30 ring, atrioseptoplasty, and due to severe tricuspid annuloectasia, annuloplasty was performed with a number 30 band. The extracorporeal circulation (ECC) time was 133 min and the aortic clamping time was 93 min. Post-ECC TEE showed normofunction of mitral and tricuspid valve leaflets, preserved biventricular function and membrane-free left atrium **(Figure 3).** The extracted membrane was sent to pathological anatomy **(Figure 4).**

During hospitalization (at the ninth day after surgery), a moderate tricuspid insufficiency was recorded. In a second operative time, through median re-sternotomy, bicaval central arterial and venous cannulation, we performed a right atriotomy. The number 30 band was intact, with intact anterior and posterior tricuspid leaflets and septal leaflet prolapse. The tricuspid band was extracted to proceed with the placement of fixation sutures with pleget in the region of the commissure between the anterior and septal leaflet (key stitches). Then we proceed to place a number 30 tricuspid band, tested with a physiological solution with null insufficiency, closed the right atriotomy, unclamp and left the ECC, the clamping time was 48 min and the ECC time 80 min. He was transferred to intensive care, extubated at 5 h with a total stay of 36 h in the unit, and

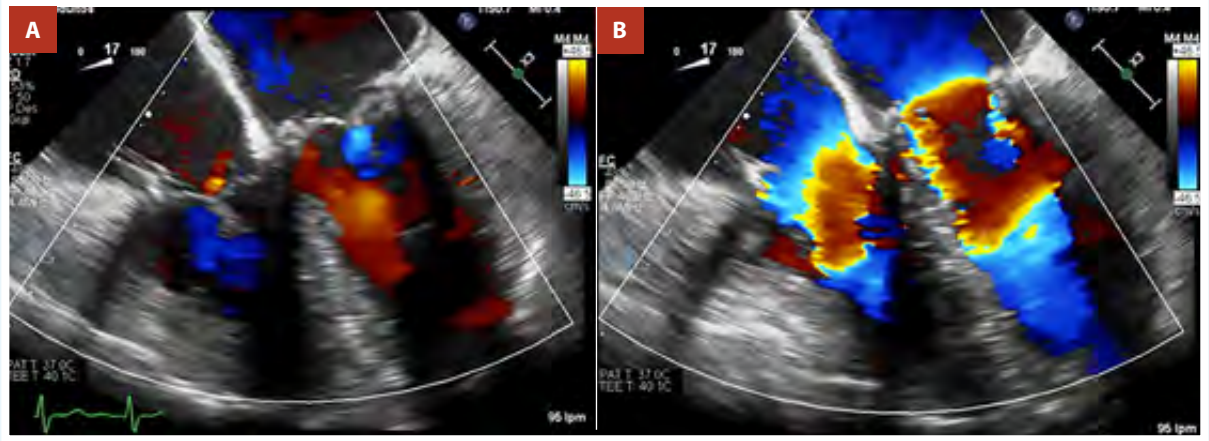

Figure 3. Transesophageal echocardiography mid esophageal view, four chambers. **A)** Absence of regurgitation jet in mitral and tricuspid plasties during systole phase. **B)** Absence of restriction in mitral and tricuspid plasties during diastole phase.

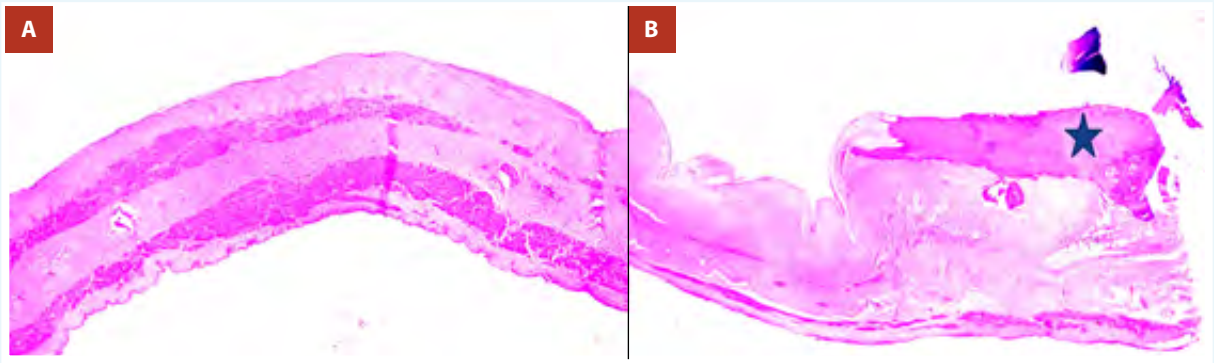

Figure 4. Microscopic pathology of extracted membrane. **A)** Fragment of fibroconjunctive tissue with cardiomyocytes, some with a degenerative appearance; **B)** Fibrosis and areas of dystrophic calcification (asterisk).

discharged on the fifth postoperative day asymptomatic with satisfactory evolution.

At 2 months, a transthoracic echocardiogram was performed showing left ventricular ejection fraction 61%, left atrium diameter 47mm and area 29cm^2, right ventricle 44 x 39 x 64 mm; non-restrictive competent mitral annuloplasty, non-restrictive tricuspid plasty with mild to moderate insufficiency, no presence of left atrial septal defect.

Discussion

CTS in the adult along with clinical mitral regurgitation and atrial fibrillation is associated with interatrial septal abnormalities, such as patent foramen ovale (PFO). If the PFO is at the level of the posterosuperior chamber, it will give a significant left-right (L-R) shunt mimicking total anomalous pulmonary venous drainage so that the right ventricle may dilate. If the PFO is in the anteroinferior chamber, the clinical presentation will be of mitral stenosis and the I-D shunt will be smaller [7]. In contrast to the above, in our patient the interatrial septum remained intact and mitral insufficiency was due to dilatation of the annulus even though the membrane was obstructive, this could be explained by the volume overload in the posterosuperior chamber that would progressively cause joint dilatation of the mitral annulus and the left ventricle similar to the case reported by Nakajima *et al* [8].

CTS is commonly associated with venous return anomalies in the form of partial or total anomalous pulmonary venous drainage, the patient had the variant of three right and two left pulmonary veins [9]. The predominant symptoms were dyspnea, lower limb edema and palpitations, which corresponded to severe mitral insufficiency with severe tricuspid insufficiency, the latter secondary to annuloectasia propitiated by atrial fibrillation in turn produced by dilatation of the left atrium by the obstructive physiology of the membrane. Reports of STS have been observed highlighting that the CF of dyspnea depends on the number and size of fenestrations in the atrial membrane [10]. As observed in the initial angiotomography and in the subsequent TEE, the hole left by the membrane was decreasing in size, which would explain the late diagnosis and would allow the advanced clinic of right heart failure.

The diagnostic gold standard is the 2D transthoracic echocardiography [7] and also the TEE, because they reveal in real time the dynamics and size of the fenestrae. Angiotomography provides detailed anatomy and is preferred to cardiac catheterization because it is non-invasive [11]. Catheterization determines the gradient between both chambers, being the average of 20 to 25 mmHg [7] in the patient the calculated gradient was 5 mmHg. Prenatal diagnosis associated with pericardial effusion evaluated during the first trimester of gestation has been reported [12].

There are two surgical approaches, if the posterosuperior chamber is enlarged (young people and adults) the right side of the chamber is approached behind the interatrial sulcus. When the posterosuperior chamber is small and the right atrium is large (infant and neonates) it is preferable to approach it through the right atrium [7]. The patient was approached through the right atrium to facilitate tricuspid annuloplasty. The survival rate at 5 and 15 years is 96 and 88% respectively [1]. Restenosis is described when membrane resection is incomplete.

In conclusion, we report a very rare acianotic congenital heart disease case, even more infrequent in its debut in adulthood.

Authors´ Contributions

All authors participated in the study design, review, drafting and approval of the final version of the manuscript.

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