Case Report

Cor triatriatum sinister and giant eustachian valve mimicking a tetratrial heart

Cor triatriatum sinister e válvula de eustáquio gigante imitando um coração tetratrial

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Introduction

Cor triatriatum is a rare congenital cardiac anomaly that is usually associated with other congenital heart defects such as anomalous pulmonary vein return, atrial septal defect, or patent foramen ovale.[1] *Cor triatriatum sinister* is found in an approximately 0.1% of necropsy examinations.[2] In this congenital malformation, the left atrium is divided into two chambers, usually by a fibromuscular septum, with the pulmonary veins entering a posterosuperior chamber separated from the anteroinferior distal chamber containing the mitral valve and the left atrial appendage. Although several theories have been hypothesized, its embryologic origin remains not fully understood. An inadequate incorporation of the common pulmonary vein into the left atrium or abnormal growth of the septum primum have been postulated to explain it.[1] On the other hand, *cor triatriatum dexter* is an even more rare congenital cardiac anomaly in which a membranous structure divides the right atrium into two chambers. In such cases, complete persistence of the right valve of the sinus venosus is responsible for the atrial division.[3] This valve usually regresses as a part of normal embryological development between 9 and 15 weeks' gestation, leaving behind remnants such as the crista terminalis superiorly and the eustachian valve of the inferior vena cava and the thebesian valve of the coronary sinus inferiorly. The severity of this defect is directly related to the degree of flow obstruction, which in turn is related to the degree of valve persistence.[4-5] Although it shares the same embryologic origin and might show a similar echocardiographic appearance, the differential diagnosis includes a prominent eustachian valve, which also results in an apparent division of the right atrium into two chambers.[6]

Case Report

A 45-year-old man with an unremarkable medical history was admitted to our clinic with palpitations and New York Heart Association class III dyspnea lasting for more than 3 weeks. Electrocardiography showed atrial fibrillation with a rapid ventricular response. Given the difficulty in achieving an adequate heart rate control, electrical cardioversion was performed. The main findings of transthoracic echocardiography are shown in Figure 1. On an apical four-chamber view, a membrane transversely and incompletely dividing the left atrium was appreciated (Figures 1A and 1B, continuous arrow). No significant Doppler gradient was obtained across the orifice delimited by the membrane. On the other side, a partial non-obstructive septation of the right atrium was observed due to a prominent Eustachian valve with high insertion into the lower portion of the atrial septum (Figures 1A and 1B, dotted arrow). Both membranes were appreciated in a parasternal short-axis view at the level of the great vessels (Figure 1D). The patient's left ventricular ejection fraction was severely depressed. To exclude the presence of a left atrial thrombus before the electrical cardioversion, transesophageal echocardiography was also performed, which confirmed the existence of both membranes, excluded the presence of atrial septal defects, revealed that all pulmonary veins drained into the superior compartment of the left atrium, and showed that the left atrial appendage was free of clots and connected to the inferior chamber of the left atrium (Figures 2A, 2B, and 2C). Cardiac magnetic resonance imaging was also performed to exclude the presence of anomalous pulmonary venous return and confirm the existence of both membranes (Figures 2D, 2E, and 2F). An unsuccessful synchronized electrical cardioversion was performed during hospitalization and the patient was discharged on atrial fibrillation with controlled ventricular response under treatment with oral anticoagulants, beta blockers, and digoxin. A spontaneous reversion to sinus rhythm occurred after discharge.

Discussion

Loeffler described three different types of *cor triatriatum sinister* based on fenestration number and size. Type 1 is defined as the absence of communication between the two chambers, in which case the accessory chamber might connect with the right atrium or anomalous pulmonary vein return might be present. Type 2 is defined as one or a few small openings in the membrane, while type 3 is defined as a wide large single communication between the accessory chamber and the true atrium.[1,7] Another classification was proposed by Lam according to the presence and location of the atrial septal defect and the pulmonary venous drainage. Five types were described according to this classification: in class A, accounting for approximately one third of the cases,

Keywords

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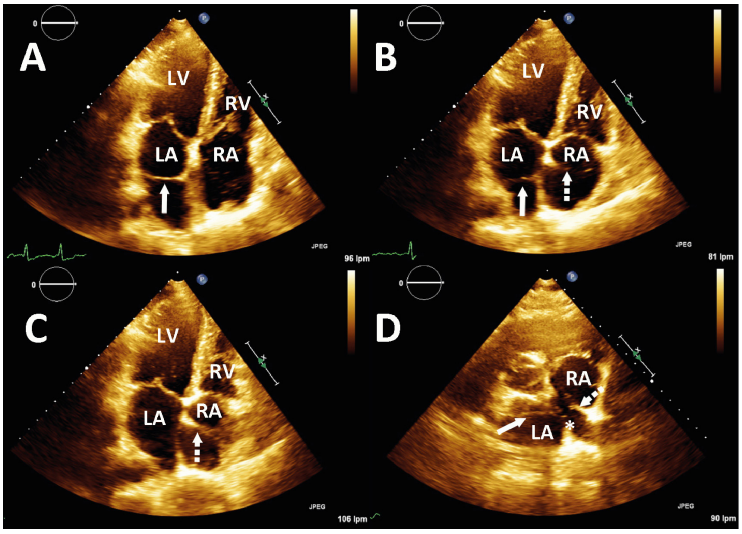

Figure 1 – *Transthoracic echocardiography revealing that a transversal septum (continuous arrow) and a prominent eustachian valve (dotted arrow) divided the left and right atria into two chambers. A, B, and C: Apical four-chamber view with different angulations of the ultrasound probe. D: Parasternal short-axis view at the level of the great vessels. LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle. *Interatrial septum.*

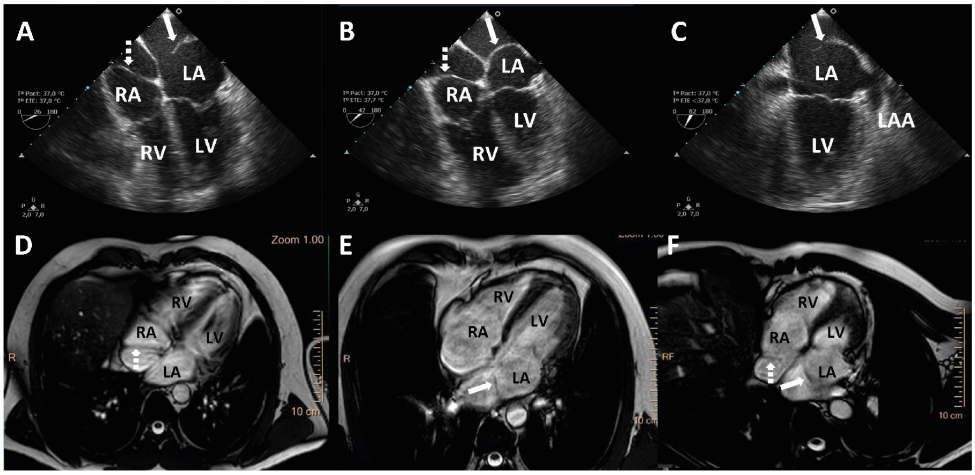

Figure 2 – *A and B: Transesophageal echocardiography, four-chamber view, showing a transversal septum (continuous arrow) and a prominent eustachian valve (dotted arrow). C: Transesophageal echocardiography, two-chamber view. The left atrial appendage connects to the inferior chamber of the left atrium while the pulmonary veins drain into the superior chamber. D, E, and F: Cardiac magnetic resonance image showing different four-chamber views. LA: left atrium; LAA: left atrial appendage; LV: left ventricle; RA: right atrium; RV: right ventricle.*

the proximal chamber receives all pulmonary veins and there is no atrial septal defect, as was observed in our patient; class A1 represents half of the cases and is characterized by an atrial septal defect between the right atrium and the proximal chamber. The other three types are less common: in class A2, an atrial septal defect exits between the right atrium and the distal chamber; in class B, the pulmonary veins drain into the coronary sinus; and in class C, there is no anatomic connection between the pulmonary veins and the proximal chamber.[8]

The clinical consequences of *cor triatriatum sinister* are related to the size of the orifice and the presence of associated cardiac abnormalities and mostly driven by the rise in proximal atrial chamber pressure and pulmonary congestion. Although adult patients with large fenestrations or a large foramen ovale might be asymptomatic,[9-10] symptoms can appear due to fibrosis or calcification of the orifice or the occurrence of atrial arrythmias or embolic events.[1] The differential diagnosis must consist of a supravalvular mitral ring, which is distinguished from *cor triatriatum sinister* by its inferior position to the left atrial appendage.[11] Surgical treatment, apart from medical treatment, is usually indicated in symptomatic patients with a significant intra-atrial gradient. Successful catheter ablation of atrial arrhythmias has also been described.[12]

Persistence of the right sinus venosus valve causes a wide

range of defects, including a prominent eustachian valve, the Chiari network, and *cor triatriatum dexter*.[3,6] In *cor triatriatum dexter,* a large obstructive septum resulting from persistence of the entire right sinus venosus valve divides the right atrium into two separate chambers. The presence of significant obstruction of the venous flow should be considered mandatory for the diagnosis of *cor triatriatum dexter*. It must be distinguished from a giant eustachian valve with high insertion into the interatrial septum, which can also mimic a divided right atrium.[13] Along with the Chiari network, it is considered an anatomic variant that might mimic this pathology. On echocardiography, the eustachian valve appears as a leaf-like linear structure at the junction of the inferior vena cava and the right atrium, while the Chiari network, which has the same embryologic origin, appears as a free-floating curvilinear structure that waves with blood flow in the right atrium.[14]

Our patient was finally diagnosed with Loeffler type 3 and Lam Class A *cor triatriatum sinister* and a giant eustachian valve that mimicked the appearance of a *tetratrial* heart. The patient's ejection fraction recovered after heart rate control was achieved, sinus rhythm was restored, medical treatment consisting of angiotensin converting enzyme inhibitors, beta blockers, and mineralocorticoid receptor antagonists was administered, and catheter ablation catheter ablation was requested.

Authors' Contributions

DGF: Research conception and design; Data collection, analysis, and interpretation; and Manuscript writing. EVF: Data collection, analysis, and interpretation; and Critical review of the manuscript for important intellectual content. MCC: Data collection; Critical review of the manuscript for important intellectual content. FJCB: Review of the manuscript for important intellectual content. EMD: Critical review of the manuscript for important intellectual content. MTGSR: Data collection, analysis, and interpretation; and Critical review of the manuscript for important intellectual content.

Conflict of interest

The authors have declared that they have no conflict of interest.

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