CorSalud 2020 Oct-Dec;12(4):441-444

CorSalud

Cuban Society of Cardiology

Case Report

Cardiocentro ERNESTO CHE GUEVARA SANTA CLARA CUBA

Cor triatriatum dexter and sinus venosus atrial septal defect: An infrequent association

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ARTICLE INFORMATION

Received: October 29, 2019
Accepted: November 28, 2019

Competing interests
The authors declare no competing interests.

Figures
Images from complementary tests are shown with patient's consent.

Abbreviation
ASD: atrial septal defect

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ABSTRACT

Cor triatriatum is a rare congenital condition with a 0.1% prevalence among all cardiac congenital anomalies in which the heart is partitioned into three atria. In cor triatriatum dexter the right atrium is divided by a fibromuscular membrane into two parts, a proximal and a distal one, which may or may not communicate with each other. In the natural course of the condition, patients may remain asymptomatic until diagnosis or present with cardiovascular manifestations secondary to right heart failure and rhythm disorders. In the specific case of symptomatic patients, the treatment of choice would be surgical correction of the anomaly and sometimes percutaneous rupture of the membrane; while in asymptomatic patients, timely follow-up and treatment of their complications would seem to be the best alternatives.

Keywords: Cor triatriatum, Right atrium, Atrial septal defects

Cor triatriatum dexter y comunicación interauricular tipo seno venoso: Una asociación infrecuente

RESUMEN

El cor triatriatum es una malformación congénita poco frecuente, con una prevalecia de un 0,1% entre todas las anomalías congénitas cardíacas, donde el corazón queda dividido en tres atrios o aurículas. En el cor triatriatum dexter la aurícula derecha queda dividida, por una membrana fibromuscular, en dos partes, una proximal y otra distal, que se comunican o no entre sí. En la evolución natural de la enfemedad, los pacientes pueden permanecer asintomáticos hasta su diagnóstico o presentar manifectaciones cardiovasculares secundarias a insuficiencia cardíaca derecha y trastornos del ritmo. En el caso particular de aquellos que se encontraran sintomáticos, el tratamiento de elección sería la corrección quirúrgica del defecto y, en ocasiones, la rotura percutánea de la membrana; mientras que en los que se encuentran asintomáticos, el seguimiento y tratamiento oportuno de sus complicaciones parecería la alternativa más viable.

Palabras clave: *Cor triatriatum, Aurícula derecha, Defectos del tabique interatrial*

INTRODUCTION

Cor triatriatum is a rare congenital condition with a 0.1%[1-2] prevalence among

all cardiac congenital anomalies, in which the heart is partitioned into three atria. The first description of the disease was made in 1868 by Church[3]; however, it was not until 1905 that a more detailed definition of this rare disease was made, the result of an embryonic defect in the union of the pulmonary veins.

In the *cor triatriatum*, the atrium is divided into two parts by a fibromuscular membrane, one proximal and one distal, which communicate or not with each other[3]. The most common affected atrium is the left one, and it is associated with other congenital heart diseases such as atrial septal defect (ASD), tetralogy of Fallot, the atrioventricular canal, aortic coarctation, and abnormal pulmonary vein drainage[4]. The *cor triatriatum* of the right atrium (*dexter*) is even less frequent and it has an incidence of only 0.025% among all congenital heart diseases[3].

In the particular case of the *cor triatriatum dexter*, the defect is attributed to the persistence of the sinus venosus valve and it is usually associated with other anomalies of the right heart[5-8]; nonetheless, the association with atrial septal defects is not very common.

Among the different types of ASD, the sinus venosus results in 5-10% of all interatrial septal defects[9]. This is produced from a reabsorption process in the upper part of the *septum secundum*, near the opening of the superior or inferior vena cava[10].

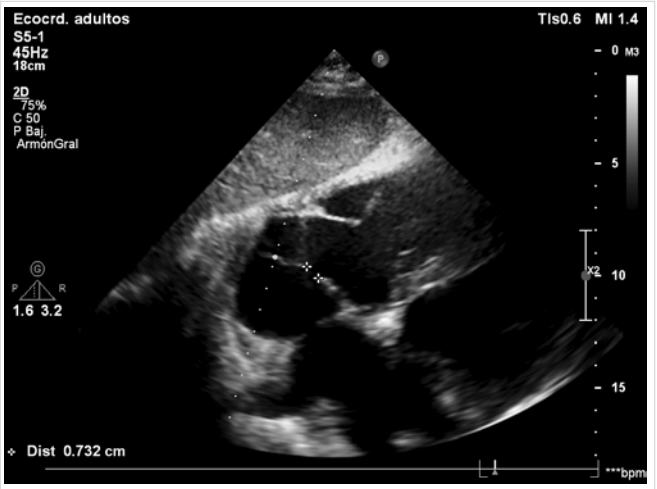

Figure 1. Subcostal view where the presence of a membrane that divides the right atrium into two chambers, communicating with each other by an orifice, is visualized.

CASE REPORT

A 68-year-old patient with a history of prostatic adenocarcinoma, who, as part of a pre-surgical check-up, was indicated a transthoracic echocardiogram. No relevant elements were found in the physical examination: rhythmic heart noises of good intensity, without murmurs, heart rate of 82 beats per minute and blood pressure of 135/75 mmHg.

The echocardiogram showed the presence of a membrane that divided the right atrium into two chambers, one proximal and one distal, communicating with each other (**Figure 1** and **Figure 2**), associated with an ASD (**Figure 3**), since in the interatrial septum, a loss of continuity of approximately 6 mm was observed with a left-to-right shunt (superior vena cava type sinus venosus ASD). In addition, the diastolic diameters of both ventricles were normal (the left 52 mm and the right 18 mm), the left atrium measured 41 mm, and the left ventricular ejection fraction was 57%. Mild tricuspid regurgitation and type II pulmonary flow curve were demonstrated, with a pulmonary accel-

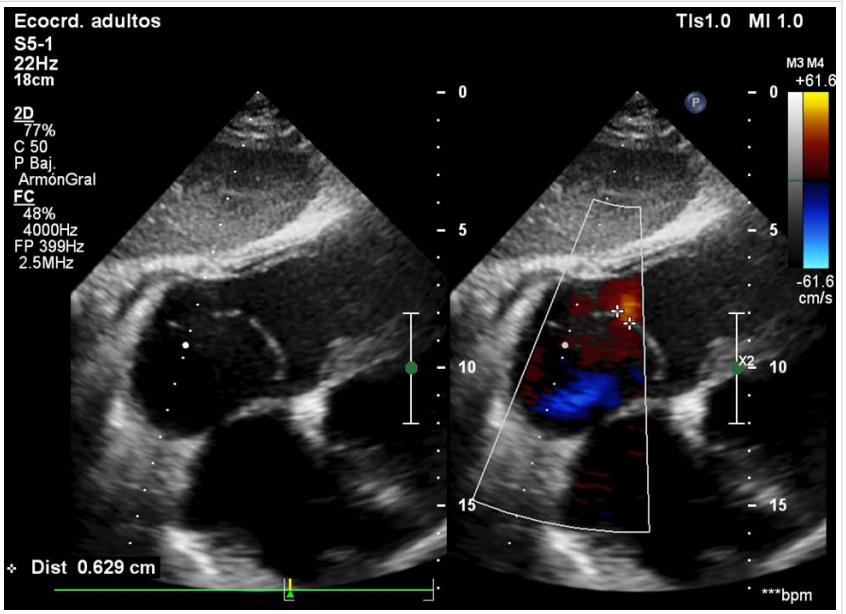

Figure 2. Subcostal view. On the left, two-dimensional mode, where the presence of the membrane that divides the atrium in two is observed. On the right, continuity defect of about 6-7 mm with flow from the distal chamber to the proximal, demonstrated by color Doppler analysis, through it.

eration time of 60 ms.

The diagnosis was concluded as a non-obstructive right *cor triatriatum,* with biatrial remodeling and superior cava type sinus venosus ASD, thus, a follow-up consultation by Cardiology was decided and to conclude the surgical treatment for the underlying disease was recommended, considering the low risk for a patient of his characteristics in a non-cardiac surgery.

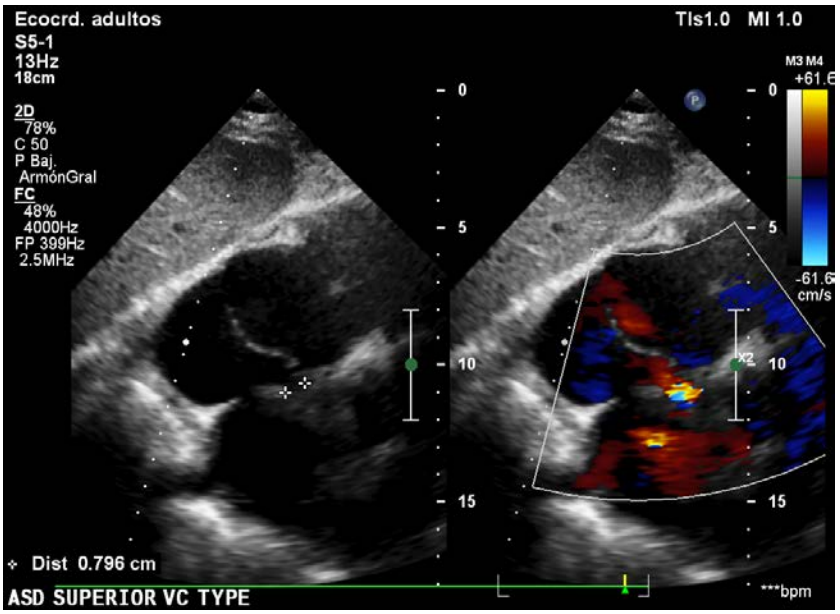

Figure 3. Sinus venosus ASD, superior cava type, with a continuity defect of 8 mm and a left-to-right shunt between both atria in color Doppler modality. ASD, atrial septal defect; VC, vena cava.

COMMENT

During the embryogenesis period, close to the fourth week, the segmentation process of the common atrium begins, which is separated from the sinus venosus through its valve. When the formation of the interatrial septum is advanced, the left leaflet of this valve is incorporated to form part of the *septun secundum*. Meanwhile, the right leaflet atrophies and disappears, leaving two remnants: the Eustachian valve and the Thebesian valve. The persistence of this right leaflet is what gives rise to the fibromuscular membrane of the *cor triatriatum dexter*[11].

This rare anomaly is usually related to genetic alterations of the right heart: hypoplasia of the right ventricle and pulmonary or tricuspid atresia. In the *cor triatriatum dexter*, the right atrium is divided by a fibromuscular membrane into two parts, one proximal and one distal; which, depending on its anatomy, may or may not be obstructive.

In the natural evolution of the disease, patients may remain asymptomatic until diagnosis, or present cardiovascular manifestations secondary to right heart failure and rhythm disorders. Its association with other congenital atrial septal defects is not common, even less with sinus venosus ASD, as in the patient presented here.

As de la Torre *et al* state, regarding a case with *cor triatriatum sinister*[3], imaging techniques represent the cornerstone for the diagnosis of this rare disease, and many patients tend to remain asymptomatic until adulthood. Although the true incidence is still unknown, today —with the development and improvement of echocardiography and computed tomography— the recognition of this anatomical alteration is more frequent.

CONCLUSIONS

As it is often reported in the bibliography, the *cor triatriatum dexter* reaches adulthood. Patients are often asymptomatic, and the diagnosis is accomplished by routine echocardiography. The infrequent association with atrial septal defects and the unknown mechanism of the disease's origin could reinforce the hypothesis of some authors, such as Martínez García *et al*[12], that its genesis could be the result of the non-involution of the right leaflet of the sinus venosus valve.

In the particular case of those patients who are symptomatic, the treatment of choice would be the surgical correction of the defect and, on occasions, a percutaneous rupture[13-14]. Meanwhile, in patients such as the one presented here, who reach adulthood without any cardiovascular manifestation, timely follow-up and treatment of their complications seems the most viable alternative.

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