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Case Report

Incidental Diagnosis of Non-Obstructive Cor Triatriatum Sinister

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Abstract

Background: Cor triatriatum sinister is a rare congenital abnormality involving the left atrium. Clinical presentation and management usually depend on hemodynamic impact and possible occurrence of heart rhythm disorders. Surgery is an option when obstructive symptoms at any age occur. Conversely, in case of less severe obstruction in elderly patients conservative strategy is preferred, because of the high risk of adverse outcome following surgery.

Case Report: Two cases of elderly patients with non-obstructive cor triatriatum sinister and symptomatic hypokinetic arrhythmia are reported. The patients did not show obstruction related symptoms, but their conditions required pacemaker implantation.

Conclusion: Accurate evaluation of cor triatriatum sinister is fundamental in order to choose the best management and rule out hemodynamic involvement, which may require a surgical treatment. There is no clear association between cor triatriatum sinister and atrio-ventricular blocks.

Keywords: *Cor Triatriatum Sinister; Atrio-ventricular Block; Echocardiography; Artificial Pacemaker*

Abbreviations

ASD: Atrial Septal Defect; AV: Atrio-Ventricular; CMR: Cardiovascular Magnetic Resonance; CT: Computer Tomography; CTS: Cor Triatriatum Sinister; ECG: Electrocardiogram; LA: Left Atrium; MR: Mitral Regurgitation; MV: Mitral Valve; TTE: Trans-Thoracic Echocardiography; TEE: Trans-Esophageal Echocardiography

Introduction

Cor triatriatum sinister (CTS) is a rare congenital abnormality that usually involves the left atrium (LA). It can be associated to several anomalies such as atrial septal defect (ASD), mitral regurgitation (MR), anomalous pulmonary venous drainage and patent ductus arteriosus.

Clinical presentation is heterogeneous and sometimes addresses the decision for surgical correction.

Two cases of elderly patients both presenting with symptomatic hypokinetic arrhythmia are reported. The first one was accidentally diagnosed with CTS during the first echocardiographic evaluation, in the other one the presence of CTS was already known.

Case Report

First Case

A 77-year-old man was taken to the Emergency Department because of a syncopal episode without prodromes. The patient denied previous hospitalizations or medication assumption, however, he mentioned the occurrence of a similar episode some months before which had not been furtherly explorated.

Physical examination was normal and electrocardiogram (ECG) demonstrated a first grade atrioventricular (AV) block. Subsequently, the patient was moved to the Cardiology Department for further investigations.

Transthoracic echocardiography (TTE) demonstrated a normal left ventricular function, bilateral atrial dilatation, slightly increased pulmonary pressure and the presence of a laminar formation in the LA, suggestive for CTS, in absence of blood flow obstruction (Figure 1). Subsequent trans-esophageal echocardiogram (TEE) and computed tomography (CT) scan confirmed the diagnosis and excluded other congenital abnormalities (Figure 2 and 3).

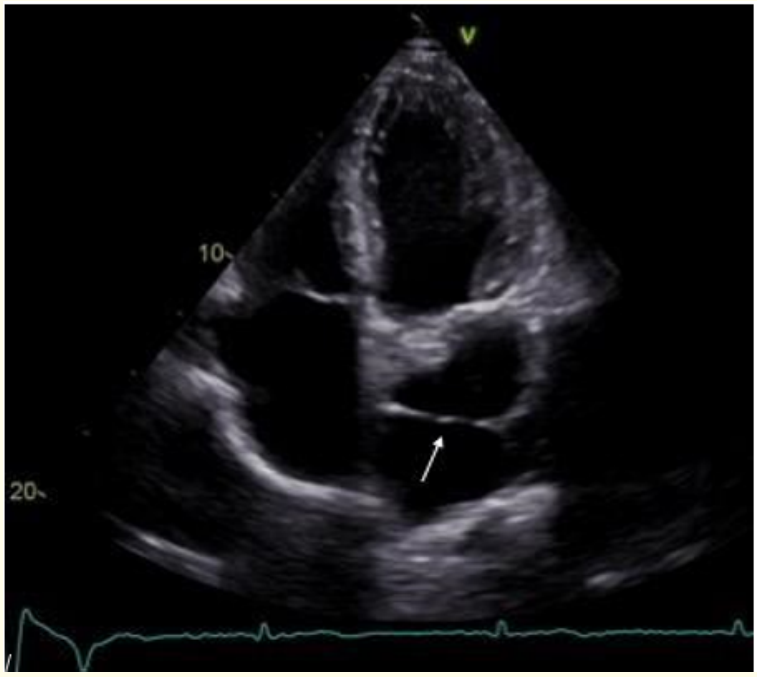

Figure 1: *Case 1. TTE apical four chamber view. Fibromuscular septum separating left atrium into two chambers.*

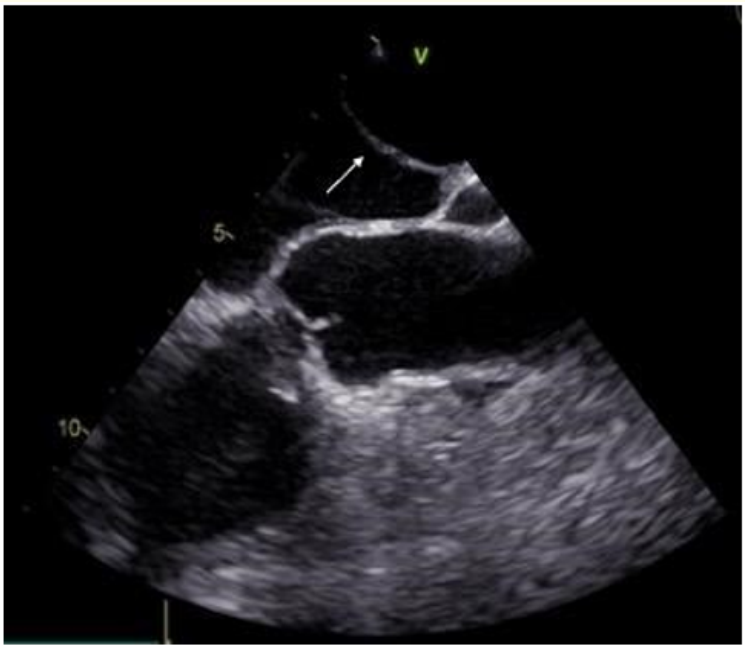

Figure 2: *Case 1. TEE modified Mid Esophageal Aortic Long-axis view. Fibromuscular septum separating left atrium into two chambers.*

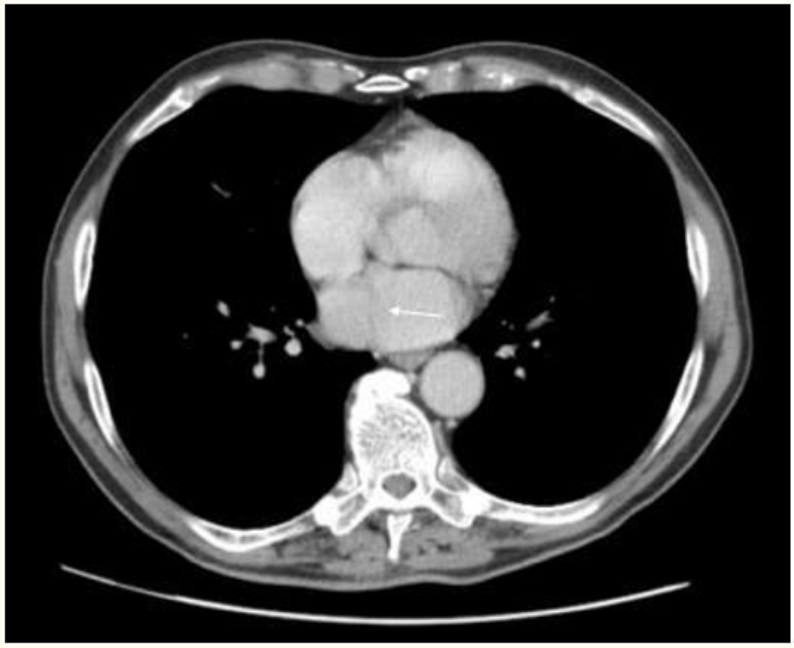

Figure 3: *Case 1. Cardiac CT scan. Fibromuscular septum separating left atrium into two chambers.*

Considering the absence of obstructive symptoms, surgery was not indicated.

Continuous ECG monitoring performed during the hospitalization detected two episodes of complete AV block, which determined the indication for pacemaker implantation but the patient declined. Therefore the patient is now on echocardiographic and arrhythmic follow-up.

Second Case

In a routine TTE performed in 2009, CTS was accidentally diagnosed in a 78-year-old man with chronic kidney disease at final stage. At that time, in absence of obstructive symptoms related to CTS, conservative strategy had been preferred. In November 2017, the patient was referred to Cardiology Department because of a marked asthenia which had occurred during hemodialytic session. Physical examination was normal, except for the detection of severe bradycardia (40 beats/minute), while resting ECG showed a complete AV block with underlying sinus tachycardia. TTE demonstrated normal left ventricular function, moderate MR, bilateral atrial dilatation, slightly increased pulmonary pressure and confirmed the presence of non-obstructive CTS. Having excluded a drug induced AV block or electrolyte imbalance, a dual chamber pacemaker was implanted successfully. Thoracic CT scan performed two months later in order to identify pulmonary problem confirmed the CTS (Figure 4).

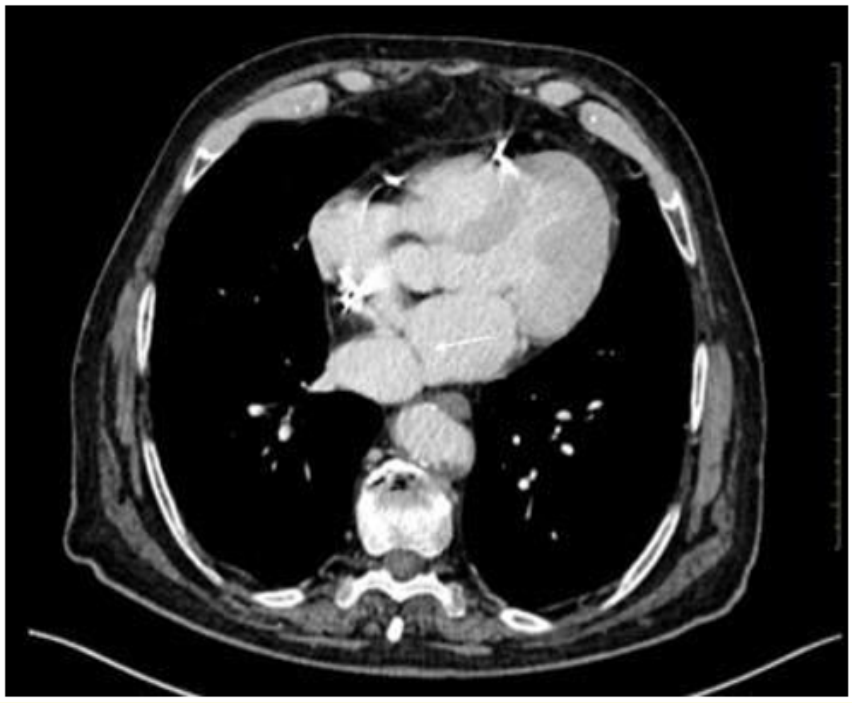

Figure 4: *Case 2. Cardiac CT scan. Fibromuscular septum separating left atrium into two chamber.*

Discussion

Cor triatriatum sinister (CTS) is one of the rarest congenital abnormality, involving the LA, with a reported incidence that varies from 0.1% to 0.4% among heart structural disease [1]. In the event of CTS, the fibro-muscular septum divides the LA in two different chambers, the upper one receives inflow from pulmonary veins and the lower one, which includes the left atrial appendage and mitral valve that is the principal left atrial cavity [2].

The medial and lateral insertion ends of this membrane are respectively fossa ovalis and the junction between the left upper pulmonary vein and left atrial appendage [3,4].

When CTS is diagnosed in the adult, ASD and MR are usually associated with. Conversely, the majority of pediatric patients presenting with CTS are suffering from cardiac malformations (77%), particularly ASD (53%), anomalous pulmonary venous drainage (28%), and patent ductus arteriosus (18%) [5].

Several mechanism have been proposed to explain the onset of this malformation such as an irregular development of the septum primum [6], a common pulmonary vein which might not be completely incorporated into the left atrium [7] or entrapped by the left horn of the sinus venosus, thereby hampering its incorporation into the left atrium [8]. Furthermore the left superior vena cava might contrast the regular growth of the left atrium [9].

In 1949 Loeffler proposed the first and probably the most plain classification of CTS [10] that divides patients in three groups according to the presence and size of the orifices in the additional lamina. The first group does not present any connection between the additional chamber and the real atrium; conversely the other two groups show different features, namely one/several small orifices(group two) or single wide opening(group three) ensuring sufficient blood-flow between the two chambers [11].

Clinical presentation of CTS is similar to the one shown by patients suffering from supravalvular mitral ring or mitral stenosis, since common hemodynamic problem of obstruction between the pulmonary venous system and left heart chambers are usually detected [12].

At the onset the most common symptoms are dyspnea, orthopnea, and chest pain [13] depending on different factors such as size of the fenestration, presence of an ASD, atrial fibrillation and cerebral embolism [14-17] and MR.

Despite the fact that a precise pathophysiological mechanism has not been established, CTS might be a predisposing factor to the myxomatous valve degeneration [18].

Obstruction to inflow can significantly affect age of presentation [19] which can vary from newborns to elderly patients.

Diagnostic exploration is fundamental to detect early CTS, particularly Echocardiographic evaluation that is the first non-invasive procedure.

In long axis view, a linear hyper-echogenic structure in the middle of LA, might be expression of CTS, however, apical four-chamber view will be able to exclude potential artifact [21]. Continuous Doppler provides measurements of mean and maximal gradients and peak velocity across the membrane. Cut off for severity is 2 m/s [22].

Posterior structures morphology, including the fibromuscular membrane and blood stream through the membrane, might not be determined precisely by transthoracic echocardiography (TTE).

Hence determination of the gradient through the membrane, the structure of the lamina itself and possible ASD are better identified and characterized by transesophageal echocardiography (TEE) [23].

Three-dimensional TTE, CT and cardiovascular magnetic resonance (CMR) might be helpful as well for diagnosis [24-26].

Hemodynamic impact of this membrane is the key element to decide the management of CTS. Incidental echocardiographic findings in an asymptomatic patient with no gradient does not require interventional strategy, apart from clinical follow-up for the early detection of obstructive signs. When obstructive symptoms are evident, surgery becomes the treatment of choice. Resection of the membrane separating the common pulmonary vein chamber from the distal LA [3,27] either, via median sternotomy or right thoracotomy is nowadays the more suitable procedure.

Frequently the youngest patients undergo surgical resection because, at the onset of the disease, they commonly show a severe obstruction.

Conversely, conservative strategy is frequently preferred in elderly subjects, who show different features of symptoms, either because they present with less severe obstruction and because of the risk of mortality during surgical procedures.

Nevertheless, surgery provides excellent results with the same life expectancy of the general population [28,30]. An alternate option of treatment to open-heart surgery might be the Balloon dilation of the obstructive membrane, which is often adopted as a definitive therapy [31,32].

Risk of thromboembolic events related to blood deceleration, even in patients treated conservatively, and atrial arrhythmias due to myocardial remodeling are other matters to take into consideration [33-36].

As regard bradyarrythmias, only one paper reported a case of AV block in a young women with cor triatriatum, in that case dexter, but it seemed to be part of a more complex congenital cardiac disease [37].

Conclusion

CTS is a rare condition usually detected during routine investigations in asymptomatic subjects. There are many factors predisposing to the complete expression of the disease such as the membrane-related flow obstruction between the pulmonary venous system and left heart chambers, hyperkinetic arrhythmias, such as atrial fibrillation, often connected to myocardial fibrosis or obstruction-related increase in atrial pressure.

Nowadays, a clear association between bradyarrhythmias and CTS has not been established.

In our two old patients idiopathic degenerative disease of the cardiac conduction system was supposed to be the causative factor.

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