Cor triatriatum: The utility of cardiovascular imaging

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A 44-year-old man with no known cardiac history presented with worsening dyspnea on minimal exertion. During follow-up, computed tomography angiography and echocardiography confirmed the incidental finding of cor triatriatum. As improvements in spatial and temporal resolution continue, cardiac computed tomography may become better suited to the dynamic imaging of anatomical defects in the heart, including, but not limited to, coronary artery disease.

Key Words: *Computed tomography coronary angiography; Cor triatriatum; Echocardiography*

Cor triatriatum is a rare congenital heart defect that is characterized by a fibromuscular membrane dividing the left atrium into two chambers. The proximal, or posterior-superior, chamber receives the pulmonary venous drainage, and the distal, or anterior-inferior, chamber contains the mitral valve and the left atrial appendage. These chambers are in communication via one or more fenestrations in the membrane. Although presentation in infancy is most common, a minority of individuals present in adulthood when the diagnosis is made incidentally. Due to advances in cardiovascular imaging, including echocardiography and computed tomography (CT), there has been a rise in the diagnosis of cor triatriatum, despite its rarity (1,2). The case of a 44-year-old man in whom cor triatriatum was discovered incidentally on a 64-slice CT angiography and confirmed by echocardiography is presented. In addition, the pathophysiology, diagnosis and imaging findings of cor triatriatum are reviewed.

CASE PRESENTATION

A 44-year-old man with a history of hypertension and dyslipidemia presented with worsening dyspnea on minimal exertion over the previous five months. He denied symptoms of chest discomfort, orthopnea, palpitations, syncope, infection or any risk factors for thromboembolism.

The patient was afebrile, normotensive and had no signs of systemic infection. His jugular venous pressure was not elevated and his cardiopulmonary examination was unremarkable. There was no calf or thigh swelling or tenderness.

The patient's complete blood count, electrolyte levels, liver function tests and cardiac enzyme levels were within normal limits. The initial electrocardiogram and chest radiograph

Le cœur triatrial : L'utilité de l'imagerie cardiovasculaire

Un homme de 44 ans sans antécédents connus de maladie cardiaque a consulté en raison d'une aggravation de la dyspnée à l'effort minime. Au suivi, une angiographie tomodensitométrique et une échocardiographie ont confirmé la découverte fortuite d'un cœur triatrial. Grâce aux améliorations de la résolution spatiale et temporelle, la tomodensitométrie cardiaque pourrait devenir mieux adaptée à l'imagerie dynamique des anomalies cardiaques anatomiques, y compris, sans s'y limiter, les coronaropathies.

were normal. CT angiography using a 64-slice multidetector scanner (Sensation 64, Siemens Medical Solutions, Germany) was performed, revealing minimal coronary artery disease without significant stenosis. The left atrium was noted to be septated by a membrane into anterior and posterior chambers, consistent with cor triatriatum (Figure 1). Subsequent transthoracic echocardiography on two-dimensional, colour pulsed-wave and continuous-wave Doppler imaging demonstrated a membrane in the left atrium without flow obstruction, confirming the diagnosis (Figure 2). The patient was able to complete 13 metabolic equivalents on a nuclear perfusion stress test, with no evidence of ischemia. Pulmonary function testing with a positive methacholine challenge confirmed reactive airway disease as the etiology for the patient's dyspnea.

DISCUSSION

Definition and classification

Cor triatriatum, first described by Church in 1868 (3), refers to a subdivision of the left atrium into proximal and distal chambers. The two chambers are divided by a fibromuscular membrane, which is septated. The size, number of septations and transmembrane flow determine the severity of the lesion. This anomaly is relatively rare, with an incidence of between 0.1% and 0.4% of all congenital cardiac defects (4,5).

Several classification schemes have been proposed for describing cor triatriatum. The simplest of these classifications was proposed by Loeffler in 1949 (6). It is based on the number and size of fenestrations in the fibromuscular membrane dividing the left atrium: group 1 is defined as having no opening, group 2 as having one or more small openings and group 3 as

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Figure 1) A Oblique maximum intensity projection computed tomography image demonstrating the right upper pulmonary vein (RUPV) and the left lower pulmonary vein (LLPV) draining into the posterior chamber (PC), the incomplete atrial membrane, and the communication site between the PC and the anterior chamber (AC). **B** Double oblique, paraseptal, long-axis view showing membrane in the left atrium, with the left upper pulmonary vein (LUPV) draining into the PC, the left atrial appendage (LAA) and the mitral valve within the AC. Ao Aorta; LCx Left circumflex artery; LV Left ventricle; RA Right atrium; RV Right ventricle

having a single, large opening (6). Other classification systems include defining cor triatriatum by the shape of the fenestration (7) or by the presence of an atrial septal defect (ASD), and by its location with pulmonary venous connections (8). Our patient's cor triatriatum was thus classified into Loeffler's group 3.

Embryology and pathophysiology

The embryological basis for cor triatriatum is still a topic of debate. It is thought that the anomalous membrane may be the result of abnormal growth of the septum primum, or that it may be secondary to malincorporation of the embryonic common pulmonary vein into the left atrium. Another hypothesis postulates that persistence of a left-sided superior vena cava may impinge on the left atrium, resulting in the formation of an abnormal membrane (9-11).

Symptoms of cor triatriatum are strongly related to the degree of obstruction caused by the fibromuscular membrane. The natural history of the disease is such that 75% of affected untreated individuals die in infancy, typically due to pulmonary hypertension. Positive prognostic factors include the presence of an ASD, as well as a greater aperture between the proximal and distal chambers. However, at least one series has reported no decrease in the severity of symptoms in the presence of an ASD (12).

Clinical presentation

The natural history of cor triatriatum, particularly in adults, is not well known, owing to the relative rarity of the diagnosis. The most common presenting symptom in a review of 43 adult cases of cor triatriatum in literature since 1966 was dyspnea (68%), with palpitations reported as the second most common symptom (27%) (1).

Physical findings of pulmonary hypertension and right-sided heart failure may predominate in patients with hemodynamically significant cor triatriatum. Other clinical entities that one must entertain in the differential diagnosis of a patient with pulmonary venous hypertension in the absence of left ventricular disease include mitral stenosis and pulmonary vein stenosis.

Figure 2) A Two-dimensional transthoracic echocardiographic apical four-chamber view demonstrating a membrane (arrow) in the left atrium (LA), consistent with cor triatriatum. **B** Colour Doppler imaging in diastole reveals no evidence of turbulent flow across the membrane in the LA. Spectral Doppler confirmed the absence of a significant pressure gradient across the membrane (not shown). LV Left ventricle; RA Right atrium; RV Right ventricle

Cardiovascular imaging

Cardiac angiography was the initial imaging modality of choice in many of the early surgical series, and it still plays a role in the diagnosis of cor triatriatum, particularly when associated with partial anomalous venous return. Classic findings on cardiac catheterization include pulmonary hypertension, a delay in visualization of the distal chamber in the venous phase, and a diastolic pressure gradient between pulmonary capillary wedge pressure and left atrial pressure (13).

Since the 1990s, echocardiography has become the imaging modality of choice for the diagnosis of cor triatriatum (14,15). Demonstration of the membrane by two-dimensional imaging, evaluation of turbulent flow on colour Doppler and determination of any gradient across the membrane on continuous wave Doppler can be performed with echocardiography. Coexisting congenital anomalies can also be well characterized by echocardiography. A recently published, retrospective review (12) demonstrated that of a total of 28 patients (96%), 27 patients with cor triatriatum underwent transthoracic echocardiography; the correct diagnosis was made preoperatively (12).

In addition to echocardiography, cardiac CT is becoming an increasingly important cardiovascular imaging modality, especially in the noninvasive evaluation of coronary artery disease. At least two previous cases (16,17) of cor triatriatum have been diagnosed incidentally using CT. Our case is unique in that it is the first report of 64-slice multislice detector computed angiography revealing cor triatriatum.

Cardiac CT has significant benefits with respect to safety and expense compared with standard coronary angiography in the noninvasive evaluation of coronary artery disease (18). The high spatial and temporal resolutions associated with cardiac CT allow for additional information to be acquired from single data sets, including cardiac anatomy, wall motion and myocardial perfusion. Our case demonstrates the utility of CT in diagnosing the underlying anatomic abnormalities in cor triatriatum.

SUMMARY

The advent of the widespread use of imaging in the evaluation of cardiac disease may explain the rise in frequency of cor triatriatum

diagnosis (19-22). We expect that this trend will continue as imaging techniques evolve and progress. Cardiac CT is a burgeoning field, with tremendous potential for ongoing research. A single cardiac CT scan produces information on both the anatomical and functional status of the heart. Our case illustrates a rare congenital anomaly diagnosed by noninvasive cardiac imaging. As improvements in spatial and temporal resolution continue, cardiac CT may become better suited to the dynamic imaging of anatomical defects in the heart, including, but not limited to, coronary artery disease. In addition to ischemic heart disease, valvular heart disease and cardiomyopathy, one should also consider congenital heart defects in the cardiac differential of any patient presenting with new-onset dyspnea on exertion.

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