ATRIAL FIBRILLATION

COMPLEX CASE STUDY

Cor Triatriatum Sinister in an 88-year-old Male with New-onset Atrial Fibrillation

NASIR NAWAZ, MD and ANCIL JONES, MD

Department of Internal Medicine, Division of Cardiovascular Medicine, Crozer-Keystone Health System, Upland, PA

ABSTRACT. Cor triatriatum is a congenital malformation that is characterized by division of the atrium into two chambers by a membrane. Despite the rarity of this congenital anomaly, there has been a notable increase in its diagnosis, primarily due to improved diagnostic imaging. The natural history depends on the size of communication between the two chambers and the presence of associated anomalies. When small, it usually presents in infancy with reduced cardiac output and symptoms caused by functional pulmonary hypertension. We present a case of an 88-year-old male who presented with new-onset atrial fibrillation and severe heart failure, and who was found to have a membrane dividing the left atrium into two chambers. If present, a patent foramen ovale or atrial septal defect (the latter found in our patient) permits decompression of the proximal chamber into the right atrium, with a significantly improved prognosis and presentation later in life. It has been postulated that fibrosis and calcification of the membrane fenestration can lead to narrowing with progressive flow obstruction. Also, the development of mitral regurgitation and/or atrial fibrillation may precipitate clinical decompensation. The increasing incidence of mitral regurgitation with advancing age may be a causative factor in the development of atrial fibrillation and subsequent clinical symptoms in such patients. Cor triatriatum should be considered in the differential diagnosis in all patients presenting with signs and symptoms of late-onset atrial fibrillation, mitral stenosis, pulmonary hypertension and heart failure, and appropriate investigations should be directed to diagnose this rare but potentially reversible etiology.

KEYWORDS. cor triatriatum, mitral regurgitation, late onset atrial fibrillation, pulmonary hypertension, trans-esophageal Echocardiography, Heart failure, Fibromuscular membrane, atrial septal defect.

Introduction

Classic cor triatriatum (CT), or cor triatriatum sinister, is a rare congenital cardiac anomaly in which a common pulmonary venous chamber (proximal chamber) is separated from the left atrium (LA) (distal chamber) by a fibromuscular septum. In most cases, the two chambers communicate through one or more openings. CT was first described by Church in 1968 as a postmortem finding1.

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Address correspondence to: Nasir Nawaz, MD, Crozer-Keystone Health System, 1 Medical Center Boulevard, Upland, PA 19013. E-mail: Nsr.nwz@gmail.com

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patient is only the second to be reported in their late eighties with symptomatic atrial fibrillation in association with CT.

Case report

This is an 88-year-old man with a past medical history of hypertension and left circumflex coronary balloon angioplasty in 1993; he had no recurrent coronary ischemia and presented with acute-on-chronic dyspnea. Chronic dyspnea was attributed to obstructive lung disease. Progressive dyspnea, lower-extremity edema, and orthopnea developed in the weeks before presentation. He presented to the emergency room and was found to have new atrial fibrillation with rapid ventricular response. He denied having fever, chills, cough, or chest pain. His blood pressure was 142/84 mmHg and heart rate 107 beats per minute (bpm); atrial fibrillation with premature ventricular complexes was revealed on cardiac monitoring. He had jugular venous distension (JVD) to the angle of the mandible. On chest examination, the lungs were clear, and examination of the heart revealed irregularly irregular first (S1) and second (S2) heart sounds with a non-displaced PMI (Point of maximal impact). Extremity examination revealed 2+ pedal edema equal bilaterally. A 12-lead electrocardiogram (EKG) showed atrial fibrillation at 110 bpm, and polymorphic premature ventricular complexes of both left and right ventricular origin, but no ST- or T-wave changes. Chest X-ray demonstrated cardiomegaly and moderate interstitial changes consistent with pulmonary edema. An Echocardiogram performed in June 2013 showed an ejection fraction of 50%; the right ventricle was normal. There was moderate left atrial dilation with the appearance of a left atrial membrane and mild aortic root dilation of 4 cm; aortic sclerosis; mitral sclerosis; but no evidence of hemodynamically significant obstruction. On admission to the hospital, the patient was offered transesophageal echocardiography (TEE)/cardioversion, which he refused. He was treated with heparin, warfarin initiation, metoprolol and furosemide. Significant diuresis ensued with decrease in edema and improved breathing. He was discharged from hospital. On repeat evaluation 3 months later, he was still in atrial fibrillation and had debilitating exertional dyspnea. TEE was performed, and showed a left atrial membrane extending from the ridge between the base of the left atrial appendage (LAA) and the lower left pulmonary vein (PV) anterolaterally to the fossa ovalis medially (Figure 1). It was not obstructive, as it had widely patent portions. The result was very sluggish low flow in the LAA. The LAA was large and multilobed with spontaneous echo contrast and low flow velocities of < 20 cm/sec. A thrombus was seen in a trabecula of the appendage (Figure 2). A small patent foramen ovale with scant interatrial shunting on color Doppler was also noticed (Figure 3). Mild mitral valve regurgitation (Figure 4), mild aortic valve sclerosis without stenosis, and trivial aortic regurgitation were seen, along with moderate-severity diffuse aortic atheroma. The main pulmonary artery was normal in size,
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origin and position, with normal bifurcation into the left and right pulmonary arteries. The PVs appeared normal, with normal return to the LA.

Cardioversion was not advised. The patient appeared to be a poor surgical candidate owing to his advanced age and low pulmonary reserve secondary to underlying obstructive lung disease. The decision was made to manage him conservatively in an outpatient setting by rate control, optimization of cardiac function and provision of supportive care.

Discussion

Currently, the embryologic basis of CT is unclear. Among various theories are malseptation involving the septum primum; malincorporation of the common PVs; and the entrapment hypothesis, whereby the left horn of sinus venosus entraps the common PV and thus prevents its incorporation into the LA. In our patient, the dividing membrane was not obstructive, as it had widely patent portions. It is possible that it represents incomplete fusion of the pulmonary venous confluence with the embryologic LA (the LAA), as visualized directly on TEE in our patient. A few classifications of CT exist and may explain why some cases only present in later life. In 1949, Loeffler divided CT into three groups, based on the number and size of membrane fenestrations: group I has no opening, group II has one or more small openings, and group III has a wide opening. Marin-Garcia classified CT on the basis of the appearance of the accessory left atrial chamber, with diaphragmatic, hourglass and tubular subtypes. Lucas and Krabill classify CT into type I with an accessory left atrial chamber, in which all the PVs communicate with the LA; type II, in

Figure 4: Fibromuscular septum dividing the left atrium (thin arrow). There is associated regurgitation across the mitral valve (thick arrow).

Cor Triatriatum- Fibrous band (Arrow) visualized in this long axis view dividing left atrium into 2 chambers.

Figure 5: Cor triatriatum- Fibrous band in long axis view dividing left atrium into 2 chambers.

12-Lead Electrocardiogram showing Atrial fibrillation and Premature Ventricular complexes (PVCs)- Black arrows.

Figure 6: Atrial fibrillation on EKG with paired premature ventricular complexes.
which the PVs do not communicate directly with the LA; and type III, the subtotal type, in which some PVs communicate with the LA and some communicate with the accessory chamber.25 Rodefeld et al21 described a very simple classification of CT. Type “A,” or classical triatrial heart, consists of a proximal chamber receiving all four PVs, and a distal chamber containing the LAA and the mitral valve, with the two chambers communicating through one or more small perforations in the membrane. Type B contains an enlarged coronary sinus that receives all four PVs. Type C is an extremely rare variant,27 in which there are no PVs entering the proximal chamber. Onset of clinical manifestation mostly depends on the size of the fenestration(s).19 CT typically presents in infancy and early childhood with respiratory distress caused by functional PV obstruction. Adults with CT most frequently present with symptoms similar to those of mitral stenosis,3,7,10,13 i.e. dyspnea, orthopnea and hemoptysis. A review of adult cases of CT published in the literature to date reveals that atrial fibrillation and mitral regurgitation8,11,16 were more prevalent at advanced ages of presentation. Late-onset diagnosis has also been reported in adults who presented with cardioembolic stroke,5,34–36 transient ischemic attack, severe pulmonary arterial hypertension,20,21,24,29 syncope23 and severe heart failure,21,26 or in asymptomatic patients as an incidental finding. The appearance of clinical symptoms is delayed if there is a large opening in the membrane, or if associated anomalies such as an atrial septal defect or anomalous PV connection enable reduction of PV hypertension. Our patient had large opening in the membrane, or if associated anomalies such as an atrial septal defect or anomalous PV connection enable reduction of PV hypertension. The LAA is a muscular pouch connected to the left atrium of the heart. In patients with atrial fibrillation, mitral valve disease, and other conditions, blood clots have a tendency to form in the LAA. Embolic blood clots associated with atrial fibrillation stem from the LAA in more than 90% of cases. They may dislodge (forming emboli), which may lead to ischemic damage to the brain, kidneys, or other organs supplied by the systemic circulation. A thrombus was visualized in the LAA in our patient, which was thought to have formed secondary to his cardiac anomaly and subsequent atrial fibrillation.

Why patients with large communications through the membrane become symptomatic as adults remains unclear. Potential hypotheses include fibrosis or calcification of the membrane orifice, and the development of mitral regurgitation and atrial fibrillation.15,30,33 Of note, however, the development of mitral regurgitation seems to be an independent factor unrelated to age-associated changes in the fibromuscular septum itself and an association cannot possibly be made between these seemingly independent processes. In patients with baseline poor ejection fraction or cardiomyopathy (ischemic or non-ischemic), the loss of atrial kick secondary to atrial fibrillation can potentially precipitate pulmonary edema or worsen heart failure, leading to decompensation, and thus symptoms of dyspnea and extremity edema. Our patient’s symptoms could possibly be explained by this mechanism.

Improvement in diagnostic cardiac imaging in routine clinical practice has probably contributed to the increase in reports of CT in adults in recent years. TEE is diagnostic in most cases. Cardiac computed tomography and magnetic resonance imaging are also used to evaluate intra-atrial septum and other associated congenital defects. Some authors also reported additional branching membranes arising from the main diaphragm dividing the LA into more than two chambers.26

Treatment of CT is mainly surgical. Medical management in symptomatic patients includes hemodynamic stabilization; management of fluid overload and pulmonary edema; control of ventricular response; and anticoagulation in patients with atrial fibrillation. Patients with CT who present with symptoms of obstruction typically undergo surgical treatment. Reported long-term results of surgery are excellent and a long life expectancy can be anticipated unless there are additional complex heart diseases or very late diagnosis. There are also rare reports of successful balloon catheter dilation of the communication between the proximal and distal chambers, but the long-term outcomes are yet to be determined.28

**Conclusion**

With advanced diagnostic imaging modalities, CT is being diagnosed more frequently. It should be considered an important part of the differential diagnosis in all patients presenting with signs and symptoms of late-onset atrial fibrillation, mitral stenosis, pulmonary hypertension and heart failure, and appropriate investigations should be directed to diagnosing this rare but potentially reversible etiology.

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