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Case Report
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Cor Triatriatum Sinister – A Rare Finding in Older Adult

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Abstract

Introduction: Cor triatriatum is a rare congenital cardiac anomaly in which a fibro muscular membrane divides the left (sinistrum) or the right (dextrum) atria into a total of 3 chambers and hence 'triatriatum'. We present a case of 60-year-old female with cor triatriatum sinister (CTS) with concomitant coronary artery disease (CAD) who underwent CABG and successful repair of CTS.

Case Description: A 60-year-old Caucasian woman with h/o dyslipidemia presented with progressive dyspnea and fatigue for 1 year. Physical examination was unremarkable. Transthoracic echocardiogram showed an EF of 65%, moderately dilated Left Atrium (LA) and no valvular abnormalities. However, a diastolic gradient was present across a structurally normal mitral valve, hence CTS was suspected. Findings of CTS were confirmed by a TEE. Cardiac catheterization prior to surgery showed two-vessel disease. On surgical exploration, a 3.9 cm × 3.1 cm × 0.1 cm membrane was seen in the LA with a nickel sized opening. After CABG and removal of membrane, her symptoms improved.

Discussion: Cor triatriatum is a rare congenital abnormality, found in 0.1% of diagnosed congenital cardiac malformations. Mal-incorporation of the common pulmonary vein into the LA is the most common cause of CTS, creating two chambers that may or may not be separated by an opening. CTS is usually diagnosed in the pediatric age group, causing LA outflow obstruction mimicking mitral stenosis, or due to its association with other cardiac malformations. CTS rarely remains asymptomatic till adulthood. Our patient presented at 60 years with exertional dyspnea, diagnosed with CTS with CAD which is very rare, and not previously reported.

Keywords: Cor triatriatum; Congenital anomaly; Older age

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Introduction

Cor triatriatum is a rare congenital cardiac malformation. Prevalence is only around 0.1% of the diagnosed congenital cardiovascular malformations [1]. Historically the prevalence has been described around 0.4% in autopsies in patients with congenital heart disease [1]. Cor triatriatum has been described in a few case reports all over the world but the condition was first described by Church [2] in 1868. It is characterized by the presence of a membrane, usually the endocardium and fibro-muscular tissue within one of the atrial chambers, dividing it into two, thereby giving the appearance of three atrial chambers (Figure 1) [1]. The characteristic anomaly in cor triatriatum is the presence of a membrane in either the left atrium (cor triatriatum sinister) or the right atrium (cor triatriatum dexter) [3]. The membrane may cause varying degrees of obstruction to the blood flow, stenosis of mitral valve and pulmonary veins and thus symptomatic heart failure which usually presents in early age [3]. We herein, present a 61-year-old patient with a typical late diagnosis of asymptomatic cor triatriatum sinister, and not associated with any other congenital anomaly. We also reviewed the limited available literature for possible complications and associations of this condition.

Case Report

A 60-year-old Caucasian woman presented to the clinic with a 1 year history of progressive exertional dyspnea and fatigue. On detailed history, there was no limitation of ordinary physical activities and she was asymptomatic prior to the last year. Past medical history was only remarkable for dyslipidemia which was being managed conservatively with dietary changes. She was not taking any medications at the time of examination. There was no family history of premature coronary artery disease or any congenital cardiac malformation, only notable for hypertension in her mother.

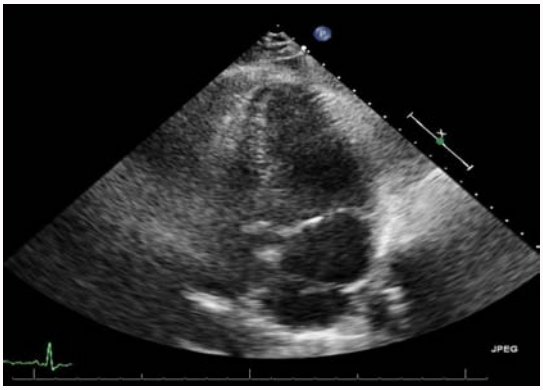

Figure 1: Demonstrating atrial membrane with opening, dividing left atrium into two separate chambers.

There was no relevant past surgical history. She was a non-smoker, with no exposure to secondhand smoke, did not consume alcohol, and never used any recreational drugs. On physical exam, the patient appeared her stated age, was obese with a BMI of 34. Vital signs (blood pressure: 136/86 mmHg; body temperature: 98.8F; heart rate: 92 beats per min; respiratory rate: 12 breaths per min; oxygen saturation: 98% on room air). Head, neck, and pulmonary examinations were unremarkable. Cardiovascular examination revealed a normal sinus rhythm with a normal S1, S2, and no audible murmur, rub or gallops. Peripheral pulses were 2+ in all 4 limbs. Abdomen, extremities, skin and musculoskeletal system were within normal limits.

A complete blood count with differential, a comprehensive metabolic profile, coagulation studies were all unremarkable. Initial diagnostic studies, included a chest radiograph, which was unremarkable, and an electrocardiogram, which showed normal sinus rhythm with a rate of 85 beats per minute and no waveform changes or axis deviation. Since a concrete diagnosis could not be established, patient underwent further diagnostic studies. A transthoracic echocardiogram (TTE) was performed. The echo revealed a normal left ventricular systolic function, class Idiastolic dysfunction and mild concentric left ventricular hypertrophy but on pressure measurements, a gradient was seen across the mitral valve. However, the mitral valve was structurally normal. On further review of the images, a membrane like structure was located within the left atrial cavity which was suspicious for cor triatriatum sinister. The findings were later confirmed with a transesophageal echocardiogram (TEE) which clearly showed the presence of the membrane in the left atrial cavity, with similar other findings as seen in the TTE.

A cardiothoracic surgeon's opinion was obtained. Before undergoing open heart surgery, patient underwent a cardiac catheterization, which showed 70% to 80% LAD disease with 99% first diagonal ostial involvement and 50% to 60% right coronary artery stenosis. Thereafter, she was started on guideline directed medical therapy, including aspirin and metoprolol. The patient was scheduled for an elective CABG and removal of the membrane. Patient had coronary artery bypass grafting with 2 vessels, left IMA to LAD and SVG to diagonal. It was found that there was a membrane just distal to the takeoff of the left and right pulmonary veins within the left atrium. It communicated to the other part of the left atrium through an opening within the membrane that appeared to be size of a nickel. The membrane was removed without any problem. At the end of the surgery, TEE revealed 1 chambered left atrium. The membrane was sent for pathological analysis and was reported as endocardial lined cardiac muscle segments consistent with atrial tissue. The immediate postoperative period was unremarkable. The patient recovered dramatically and became asymptomatic. Currently the patient is receiving medical therapy and is doing well.

Discussion

As mentioned in the introduction the extremely rare incidence of cor triatriatum, has not drawn the condition much attention in mainstream literature but existing case reports are mostly in the pediatric population, in contrast to our elder patient.

There are various hypotheses proposed explaining the origin of the membrane in cor triatriatum. However, the exact mechanism has not been elucidated. One of them is the mal-septation hypothesis. In 1881, Fowler [4] proposed this hypothesis describing the membrane as an abnormal overgrowth of the septum primum. Barnes and Finlay [5] in 1952 described the mal-incorporation hypothesis in which he said the membrane might be the result of mal-incorporation of the common pulmonary vein in the left atrium. Later in 1969, Praagh and Corsini [6] proposed the entrapment hypothesis in which the common pulmonary vein is entrapped by the left horn of the sinus venosus. More recently genetic studies have been done by Muggenthaler et al. [7] and they described mutations in HYAL2 encoding hyaluronidase, which is involved in the development of palatal and cardiac shelf matrix. They described an association between cleft palate and cor triatriatum sinister in Amish and Saudi Arabian population. A case series by Marín-García et al. [8] described 3 types of cor triatriatum - diaphragmatic, hourglass and tubular based on their morphologic appearance at autopsy. Kirklin and Barratt-Boyes [9] classified cor triatriatum based on the size and number of openings into 3 groups. Group I with no opening, group II with 1 or more small openings and group III with a wide opening.

Cor triatriatum depending on the severity of blood flow obstruction usually presents in infancy and pediatric age group as described above Group I and II. They may present with congestive heart failure, cyanosis, failure to thrive and death in the more severe cases [3]. Mortality has somewhat decreased in the modern era because of early diagnosis and correction of the defect. Usual associated cardiac malformations in the pediatric population include atrial septal defects, anomalous pulmonary vein drainage, and patent ductus in the decreasing order of frequency. Another malformation that might give a similar appearance is the presence of supravalvular mitral rings [10] as a part of Shone's complex although cor triatriatum has been reported separately from these.

Adult patients with cor triatriatum usually belong to group II or III [3]. Patients may remain asymptomatic or become symptomatic and develop functional mitral stenosis in cases of calcification of the opening or even mitral insufficiency caused by myxomatous degeneration, thereby causing atrial fibrillation [3]. Patients may initially be misdiagnosed as having mitral stenosis as there usually is a pressure gradient across the membrane as was seen in our patient. Other cardiac malformations associated in adulthood that have been reported are a secundum type Atrial Septal Defect, a left superior vena cava with an unroofed coronary sinus [9]. Our case falls into group III cor triatriatum sinister, and interestingly we are presenting oldest case of cor triatriatum sinister without any associated other congenital malformations, so far to our knowledge.

Initial diagnostic modalities include echocardiography which

usually shows the presence of the membrane although as above a few cases may be misdiagnosed with a transthoracic echocardiography. Transesophageal echocardiography better delineates the anatomy and other associated malformations can and need to be ruled out prior to surgical correction. Cardiac catheterization may also be performed if there is suspicion for or risk factors for coronary artery disease, although symptoms for both coronary artery disease and cor triatriatum may frequently overlap justifying the need for some sort of evaluation of myocardial perfusion. The mainstay of treatment continues to be surgical with a removal of the membrane to restore the normal anatomy. The approach to the left atrium can be through a left atriotomy or via a trans-septal approach after a right atriotomy [11].

Conflict of Interest

Authors have nothing to disclose. This case report presented as a poster presentation in 21st World Congress Heart Disease, Boston, MA, USA, July-August 2016 and later published as an abstract in Cardiology Journal. This case report never been published as a manuscript, and not under process for publication elsewhere.

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