THE TRIATRIAL HEART (COR TRIATRIATUM) – RARE ADULT CONGENITAL HEART DEFECT

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ABSTRACT:

Background: Cor triatriatum is a rare congenital cardiac malformation. Prevalence is only around 0.1% of the diagnosed congenital cardiovascular malformations. Historically the prevalence has been described around 0.4% in autopsies in patients with congenital heart disease. 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.

Case report: A 72-year-old patient presented to our cardiac surgery department with complaints of rhythm disorders, heart palpitations and shortness of breath. The patient has a history of mitral valve pathology. During the last six months, she has had a progression of these heart failure clinical manifestation and is referred to our institution for mitral valve surgery after selective coronary angiography. The echocardiography showed an abnormal membrane dividing the left atrium, and the diagnosis of cor triatriatum was fully made via transesophageal echocardiography. There were associated significant mitral valve and tricuspid valve regurgitations.

Conclusion: CTS/cor triatrium sinister/ is a congenital acyanotic heart disease which rarely presents during adulthood. The symptoms in adults are similar to those of mitral valve stenosis. The prognosis of the patient with CTS depends on the size of the fenestration into the fibro-muscular membrane. Corrective surgery is a treatment of choice for CTS.

Keywords: congenital anomaly, cor triatrium, adults,

BACKGROUND

Cor triatriatum is a rare congenital cardiac malformation. Prevalence is only around 0.1% of the diagnosed congenital cardiovascular malformations [1, 2]. Historically the prevalence has been described around 0.4% in autopsies in patients with congenital heart disease [1, 2]. 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 the mitral valve and pulmonary veins and thus symptomatic heart failure, which usually presents at an early age [3]. We herein represent a 72 years old woman with a long standing problem with the mitral valve, with a typical late diagnosis of cor triatriatum sinister and not associated with any other congenital anomaly.

Fig. 1. Cor triatriatum sinister. Classic findings include a membrane separating the left atrium into two components: one chamber with the pulmonary veins and the other with the mitral valve and atrial appendage. The obstructing membrane is often partially fenestrated, allowing communication between the proximal and distal atrial segments. (https://healthjade.net/cor-triatriatum/)
CASE DESCRIPTION
A 72-year-old female with exertional dyspnoea and palpitations presented to an outpatient cardiology clinic. She was previously found to be in atrial fibrillation, and treatment with betabloker, antiarrhythmic and anticoagulant had been initiated. Despite medical treatment, she reported worsening symptoms and several episodes of nocturnal dyspnoea and orthopnoea. The patient had a past medical history of radical mastectomy for breast cancer 10 years prior with adjuvant chemo- and radiotherapy. Co-morbidities included newly diagnosed type 2 diabetes and hypothyroidism on daily hormone replacement therapy. After an echocardiogram the patient was diagnosed with severe mitral insufficiency and referred to our institution for mitral valve surgery.

On admission the patient was in good overall status, physical exam revealed no significant findings. ECG showed atrial fibrillation with isolated ventricular extrasystoles. Coronary angiography was performed prior to surgery to exclude obstructive coronary disease, and it showed normal coronary arteries. Our transthoracic echocardiogram (TTE) demonstrated an additional structure in the left atrium perpendicular to the interatrial septum (figure 2 and figure 3), severe mitral insufficiency due to annular dilatation (figure 4 and figure 5), moderate to severe tricuspid insufficiency with a dilated tricuspid annulus and elevated pulmonary pressure and preserved systolic function of the left ventricle. These findings were later confirmed on transoesophageal echocardiogram (TEE) intraoperatively.

Fig. 2. Preoperative TTE - the atrial membrane in the left atrium can be seen.

Fig. 3. Preoperative TTE - the atrial membrane in the left atrium can be seen.

Fig. 4. Intraoperative TEE – mitral insufficiency and the inter-atrial membrane.

Fig. 5. Intraoperative TEE – mitral insufficiency and the inter-atrial membrane.
Intraoperatively after typical aortic and bivacal cannulation, cardiopulmonary bypass (CPB) was initiated and cold antegrade cardioplegic arrest was induced. The left atrium was approached through Sondergaard’s groove. A thick fibrous accessory membrane in the mid portion of the left atrium was found. Full excision of the membrane was performed (Figure 6 and Figure 7), followed by mitral and tricuspid valve ring annuloplasty and left atrial appendage closure. Weaning CPB was smooth and the postoperative period was uneventful. No significant ECG dynamics, rhythm or conduction disturbances were recorded. The patient was discharged home on the 7th postoperative day.

**Fig. 6.** Accessory membrane in the left atrium.

**Fig. 7.** Resection of the membrane during the operation.

**DISCUSSION**

The triatrial heart is a rare congenital anomaly, reported by Jeiger at autopsy in 0.4% of patients with congenital heart disease [1] and found in less than 0.1% of clinically diagnosed cardiopathies [3]. Although first described by Church in 1868 as a left atrium separated by an abnormal septum, the name “cor triatriatum” was given by Borst in 1905 [4]. It usually affects the left atrium (cor triatriatum sinister) and rarely the right atrium (cor triatriatum dexter).

The atrium is divided into two separate chambers, usually by a thick fibromuscular septum, which can be membranous with a transverse or horizontal orientation, band-shaped or funnel-shaped. The proximal or upper chamber drains pulmonary venous blood, while the distal lower chamber (or true atrium) is in contact with the atrioventricular valve and contains the atrial appendage and true atrial septum [2]. Several classification schemes have been reported to describe CTS, the simplest being given by Loeffler in 1949 [5]. It is based on the number and size of the fenestrations in the fibromuscular membrane and distinguishes three groups: the first group is defined by the lack of connection between the two chambers, the additional chamber can connect to the right atrium, or some of the pulmonary veins can drain in an unusual way manner; in the second group there is one or several small holes in the intraatrial membrane; and in group three the accessory chamber communicates widely with the true atrium by a single large opening. While the latter occurs mostly in the adult population with this anomaly, the first two groups are usually diagnosed in highly symptomatic infants and children and are associated with increased mortality at a younger age [10].

Although several theories have been developed, the embryological basis of CTS remains incompletely established. The most popular theory, “malincorporation,” states that the common pulmonary vein does not normally incorporate into the left atrium, thereby creating two chambers separated by a narrow opening [7]. However, this theory fails to explain the finding of the fossa ovalis and atrial muscle fibers in the walls of the proximal chamber, where only a venous wall is supposed to be present. Others believe that the membrane bisecting the atria is due to abnormal growth of the septum primum [6]. (“malseptation” theory). In the latter theory (the “trapping” theory), the left horn of the embryonic sinus venosus traps the common pulmonary vein and thereby prevents its inclusion in the left atrium [8].

Initial diagnostic modalities include echocardiography which demonstrating atrial membrane with fenestrations, dividing the left atrium into two separate chambers. In a few cases, the accessory membrane may be misdiagnosed with transthoracic echocardiography. Transesophageal echocardiography better delineates the anatomy and other associated malformations and is mandatory prior to surgical correction [8, 12]. Cardiac catheterization may also be performed if there is a suspicion of 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 [10, 11]. The mainstay of treatment continues to be surgical, with the 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 [8].

The complete establishment of the diagnosis of cor triatriatum sinister is possible with minimally invasive cardiac imaging methods, mainly three-dimensional transthoracic and transesophageal echocardiography [9, 11, 12]. These valuable diagnostic tools help optimize the management of this congenital heart disease, whose rarity can often lead to misdiagnosis, but when adequately evaluated and treated, the clinical result is excellent.

CONCLUSION
Cor triatrium sinister is a congenital acyanotic heart disease which rarely presents during adulthood. The natural history of the disease manifests with symptoms of congestive heart failure. In the presented case, the accessory left atrial membrane is easily diagnosed by transesophageal echocardiography in the assessment of malfunctioning mitral and tricuspid heart valves. Complex corrective surgery of the mitral and tricuspid valve and concomitant resection of the left atrial membrane was the treatment of choice in this rare case of CTS in an adult patient with excellent post-operative result.

ABBREVIATIONS:
CTS - cor triatrium sinister
ECG - electrocardiogram
TEE - Transesophageal Echocardiography
NYHA - New York Heart Association

REFERENCES:

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