



RESEARCH ARTICLE

COR TRIATRIATUM AN UNUSUAL CAUSE OF ATRIAL FIBRILLATION IN ADULTS: CASE REPORT

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Abstract

Cor triatriatum sinister is a rare congenital cardiac anomaly in which the pulmonary venous confluence is separated from "true" left atrium by fibro-muscular septum. Cor triatriatum can persist unrecognized into adult life if large opening or several fenestrations are present. Late onset of symptoms is usually caused by increased pulmonary pressure, development of atrial arrhythmias or mitral valve abnormalities. Non obstructive cor triatriatum may be an incidental finding. We describe a case of an adult woman who had known cor triatriatum (but not being actively followed) who presented in palpitation access table due to atrial fibrillation.

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Introduction:-

Cor triatriatum sinister is a rare congenital defect in which the left atrium is divided into two chambers by a membrane. Cor triatriatum comprises approximately 0.4% of congenital heart disease at autopsy [1]. It is found in less than 0.1% of clinically diagnosed cardiomyopathies [2]. It is typically identified in children and is a particularly rare new diagnosis in adults [3]. Atrial fibrillation has been described in 30% of published cases on adults with cor triatriatum.

Case Report:

A 36-year-old woman presented to the emergency department in palpitation access table and progressive dyspnea over the past week. He had a blood pressure of 130/70 mmHg and an irregular rapid pulse rate of 156 beats/minute. He had a respiratory rate of 23 breaths/minute and an oxygen saturation of 98%. Physical examinations found mitral focal diastolic bearing associated with a systolic murmur of tricuspid insufficiency. The results of the remainder of his physical examination were not remarkable.

Electrocardiogram showed atrial fibrillation with rapid ventricular response (Fig. 1). Laboratory studies were all within normal limits.

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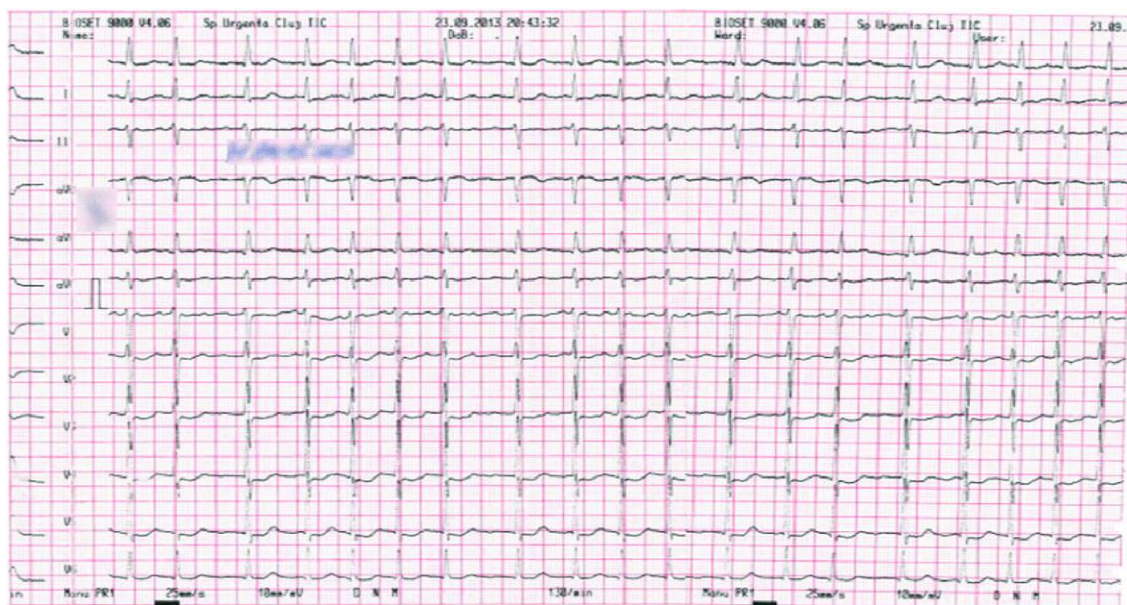


Figure 1:- Electrocardiogram showing atrial fibrillation.

Transthoracic echocardiography in the emergency department showed the presence of a membrane that divides the left atrium into postero-superior and antero-inferior chambers membrane from the apical four chamber view and mild mitral insufficiency with moderate tricuspid insufficiency (Fig. 2 and 3).

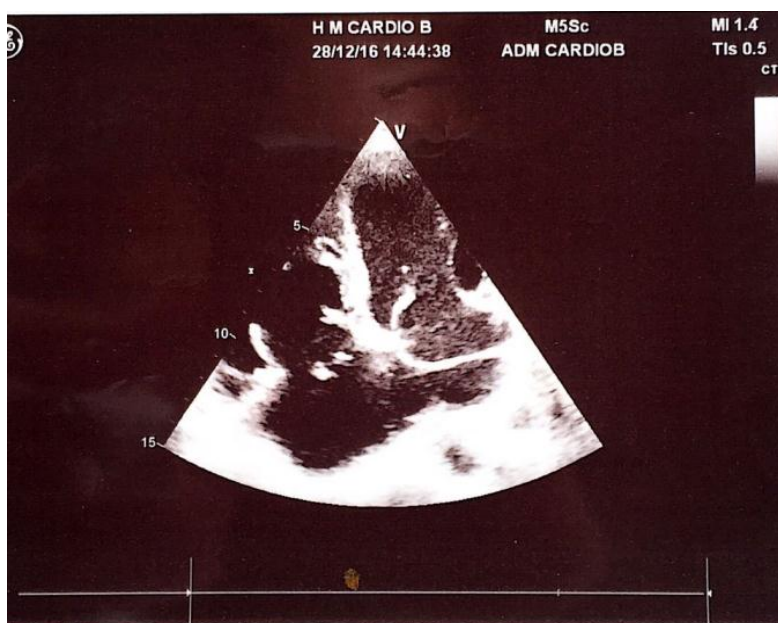


Figure 2:- Apical four chamber view showing the presence of a membrane in the left atrium.

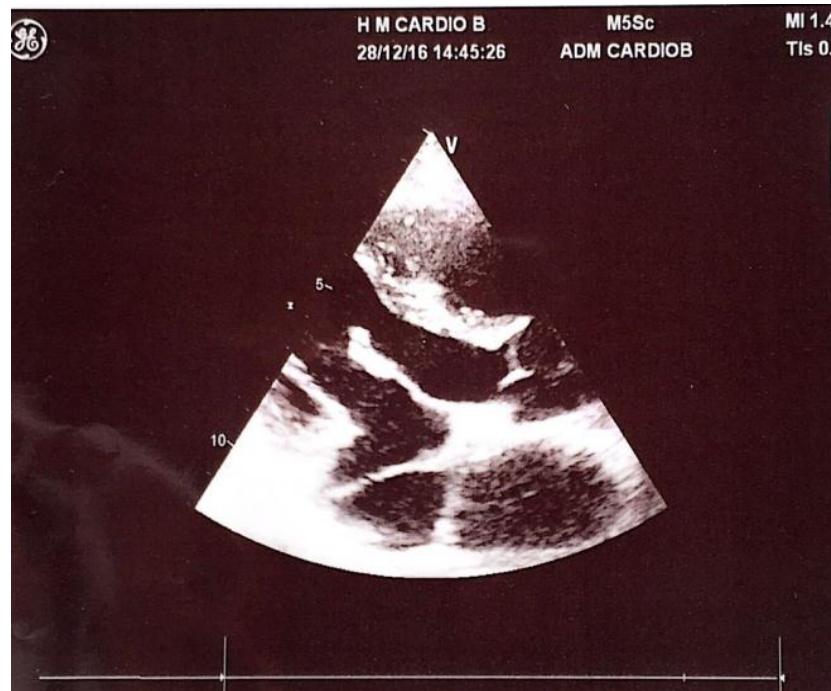


Figure 3:- Echocardiographic image showing typical appearance of co-triatrium.

This patient received surgical excision of the left atrial membrane and tricuspid annuloplasty, she was discharged on the 10th day after the operation with good recovery.

Discussion:-

Cor triatriatum is a rare congenital heart disease first described by Church in 1868 [1]. Cor triatriatum was reported to have an incidence of 0.4% at autopsy of patients with congenital cardiac disease and 0.2% among patients undergoing transesophageal echocardiography. The incidence of cor triatriatum is less than 1 in 10,000 in high volume echocardiographic laboratories [2].

Cor triatriatum is characterized by the presence of a membrane that divides the left atrium into two chambers: a postero-superior chamber receiving the pulmonary veins, and an antero-inferior chamber communicating with the mitral orifice [3]. The membrane within the left atrium results from a failure of reabsorption of the common pulmonary vein during development and then hinders the pulmonary venous flow [4]. The clinical features mimic those of mitral stenosis [5], and the degree of obstruction depends on the diameter of the orifice between the two portions of the left atrium. This disease is usually diagnosed in childhood, although sometimes it is also found in adults.

The most common comorbid cardiac conditions in adults are atrial septal defects and mitral regurgitation [3]. Patients typically present with pulmonary edema in infancy unless a sizeable opening in the membrane allows for sufficient drainage of the affected pulmonary veins. This opening may become obstructed later in life secondary to fibrosis and calcification and can lead to the development of symptoms. Left atrial dilatation ensues from elevated filling pressures, and this substrate can give rise to the development of atrial fibrillation in a manner analogous to mitral stenosis.

Transthoracic echocardiography is an optimal initial examination in the diagnosis and assessment of the disease. Because of its easy utility and ready availability, transthoracic echocardiography has become an important modality for emergency physicians to evaluate patients presenting with dyspnea, chest pain, and cardiac arrest. It allows clinical physicians to differentiate cor triatriatum from mitral stenosis, pulmonary vein stenosis, and acute coronary syndrome. It also reveals the degree of the obstruction that exists. Furthermore, echocardiography is useful in the differential diagnosis of other associated congenital heart diseases [6,7]. The transthoracic echocardiography of this patient showed a typical membrane dividing the left atrium into two chambers with mild mitral regurgitation.

Medical care in symptomatic patients includes hemodynamics stabilization, management of fluid overload and pulmonary edema, control of ventricular response and anticoagulation in patients with atrial fibrillation, and anticoagulant prophylaxis in patients with right heart failure.

Given the rarity of the diagnosis, formal guidelines do not exist on the optimal timing of surgical correction. Surgery has typically been offered in symptomatic adults [5]. In the largest surgical case series published, the mean age at the time of surgery was 27 years and the mean gradient across the membrane in those surgically repaired was 17.2 mmHg [8].

In symptomatic patients, surgical excision of the left atrial membrane usually leads to good recovery [9,10]. The common approach is complete resection of the membrane and closure of the atrial septum. Outcomes after surgical repair have typically been excellent. Correction of associated congenital defects also needs to be performed.

Our patient received surgical excision of the left atrial membrane and tricuspid annuloplasty, she was discharged on the 10th day after the operation with good recovery.

Conclusion:-

Cor triatriatum is a rare but increasingly recognized congenital abnormality that can become symptomatic later in life. It is associated with the development of atrial fibrillation, pulmonary hypertension, and rarely with the development of left ventricular dysfunction. As access to imaging modalities and the quality thereof improve, more data will be needed to guide treatment decisions around thromboembolic prophylaxis and follow-up to prevent complications.

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