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CASE REPORT

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ABSTRACT
Cor triatriatum sinister is a rare congenital cardiac anomaly in which a fibromuscular membrane divides the left atrium into 2 chambers. It is even rarer when present at an adult age. Symptoms and presentation of the patient depend on the size of the opening in the membrane through which 2 chambers of left atrium communicate with each other. Cor triatriatum sinister rarely remains asymptomatic till adulthood. In adults, symptoms of Cor triatriatum sinister mimic the symptoms of mitral stenosis. Corrective surgery is the treatment of choice, but when it presents as an emergency case especially at an early age, balloon dilatation of the membrane opening can be done. Elective balloon dilatation of membrane opening at an early age diagnosed with Cor triatriatum sinister followed by corrective surgery at an adult age can be a safer option compared to direct corrective surgery at an early age. We present a case of a 25-year-old female**.**

KEYWORDS: Cor triatriatum sinister; Congenital cardiac anomaly; Older age .

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INTRUDUCTION

Cor triatriatum sinister (COS) is a congenital acyanotic heart disease in which a fibromuscular membrane divides the left atrium (LA) into 2 chambers. The incidence of COS is 0.1% to 0.4% of all patients with congenital heart disease [1]. There are two types of cor triatriatum; dexter means right atrial division, and sinister means left atrial division. Church reported the first case of COS in 1868, but it was a post-mortem diagnosis [2]. This condition is usually diagnosed in infancy or childhood. We present a case of COS in a 25-year-old lady who was treated surgical correction.

CASE REPORT

A 25-year-old lady presented with progressive dyspnoea (grade III according to New York Heart Association classification for heart failure). The patient didn't have cyanosis and saturation was 100% at room air. There was a pansystolic murmur (grade 3/6) at the left 4th intercostal space (mitral area) and the second heart sound (P2 component) was loud. An electrocardiogram showed sinus rhythm with first-degree atrioventricular block. An echocardiogram showed a membrane that divided the LA into 2 chambers (Fig 1). There was large-sized ostium secondum atrial septum defect (Os-ASD) with evidence of moderate pulmonary hypertension. The patient's left ventricular systolic function (ejection fraction 55%), valvular anatomy were normal. A transesophageal echocardiogram confirmed the diagnosis of cor triatriatum and normal drainage of all pulmonary veins into the heart. The patient was planned for the intracardiac repair of COS.

Midline sternotomy was done. Ascending aorta and both venae cavae (superior & inferior vena cava) were cannulated to establish cardiopulmonary bypass. Cold del-Nido cardioplegia was used to arrest the heart. After opening the right atrium, LA could be visualised through Os-ASD. It was divided into 2 interconnecting chambers by a fibromuscular membrane with multiple fenestrations (Fig 2). The membrane in the LA was excised and Os ASD was closed using an untreated self-pericardial patch. The rest of the operative and postoperative period was uneventful. The patient was discharged on the 7th post-operative day. The patient was asymptomatic on the 6

monthly follow up and echocardiography showed no evidence of remnant of membrane across LA.

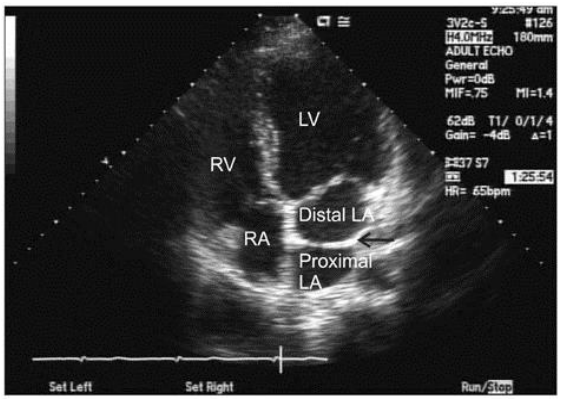

Fig.1: Transthoracic echocardiogram (apical 4-chamber view).
NB: LA = Left Atrium. LV = Left Ventricle. RA = Right Atuim.

- Transthoracic echocardiogram (apical 4-chamber view) shows a membranous structure subdividing the left atrium (arrow).

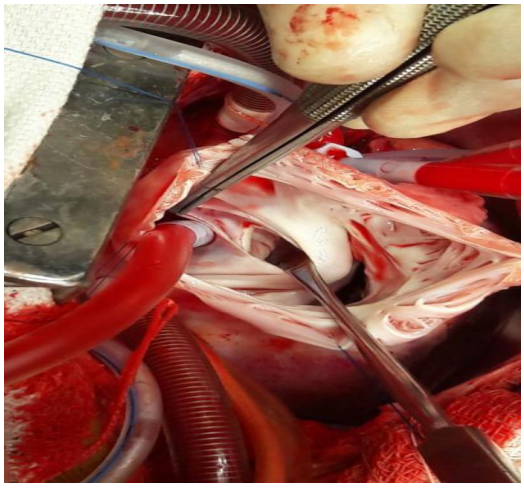

Fig.2-A : Right Atruim.

- Approaching the left atrium via right atrium (RA) through intra-atrial septum which enables exposure of the fibromuscular membrane (*).

Fig.2-B : Specimen of the resected pliable membrane.

DISCUSSION

Due to the extremely rare incidence of COS in the adult population, it has not drawn much attention in the literature. Existing case reports are mostly in the pediatrics population, in contrast to our report in an adult. The embryology of COS is based on the 3 main theories. These are mal-septation involving the septum primum, mal-incorporation of the common pulmonary vein, and entrapment of the common pulmonary vein [3]. Loeffler defined the classification of COS based on the number of openings in the left atrial membrane [4].

The 2 chambers made by this membrane into the LA are named as posterosuperior (upper chamber) and anteroinferior (lower chamber) according to their anatomical position. The clinical manifestations of the patient depend upon the size of the opening in the membrane. In most cases, this opening is severely restrictive and the patient presents early as the restrictive opening will allow less amount of blood to transfer from posterosuperior to the anteroinferior chamber. On the other hand, if the opening is large then the patient remains asymptomatic [5].

COS in infancy or early childhood will show symptoms that are caused by functional pulmonary venous obstruction. An adult patient diagnosed with COS was a rare entity until the end of the 19th century, but in the last 2 decades, multiple cases with COS have been reported even in adults. This increase in reports in recent years most likely results from the widespread use of echocardiography [6]. When present in an adult patient, it will show symptoms similar to those of mitral stenosis. Rarely, patients present with unusual symptoms like syncope, cardiac arrhythmia. Khanra et al even reported a 45 years old patient presenting with chest pain mimicking as an acute myocardial infarction but later diagnosed with COS [7]. Our patient had multiple fenestrations in the membrane without any gradient across the membrane. There was no pulmonary venous hypertension. These could be the causes of the late presentation of COS in our case.

About 5% of cases with COS have associated cardiac anomalies. In the adult presentation, secundum type of atrial septal defect and mitral valve regurgitation is the most common associations. In the pediatric age group, major associations are anomalous pulmonary venous drainage, atrial septal defect, and patent ductus arteriosus. In rare incidences, mitral stenosis can be associated with COS in Wong anomaly [8].

In an asymptomatic case with COS, only the presence of a pansystolic murmur in the mitral area gives the clue towards correct diagnosis. X-ray and electrocardiogram findings are variable. Important differential diagnosis includes bronchial asthma, pulmonary vein stenosis, supra valvular mitral ring, atrial tumors, pulmonary tuberculosis, or mitral stenosis. Echocardiography is the diagnostic modality of choice, but advanced cardiovascular imaging like Computed tomography and magnetic resonance imaging helps in a better understanding of cardiac anatomy.

Hence, we recommend preoperative cardiac CT to avoid surprises in the operation theatre.

Surgical correction is the choice of treatment in patients with COS. The prognosis of COS depends on the size of the orifice in the membrane. In Niwayama's study, the average survival was 3 months when the opening was 3 mm and 16 years when it was >3 mm [7]. Surgery as a treatment modality for COS has produced excellent long-term results and is safer in adult patients than surgery at an early age. Some of the studies have reported successful balloon dilation of the obstructive membrane if the patient presents with severe pulmonary venous obstruction [9]. Hence, elective balloon dilatation of the opening in the membrane to make it more than 3 mm in patients at an early age and surgery at adulthood can be a safer protocol of treatment. But this modality of treatment has not been tried yet and long-term outcomes from such treatment are still not proven.

Our patient a 25 years' lady presented with dyspnea on exertion was diagnosed for the first time at this age as COS associated with Os-ASD treated successfully with surgical correction.

CONCLUSION

COS is a congenital acyanotic heart disease which presents rarely presents during adulthood. The symptoms at an adult presentation may vary and are similar to those of mitral valve stenosis. Echocardiography is diagnostic but advanced cardiovascular imaging like Computed tomography and magnetic resonance imaging helps in better understanding of cardiac anatomy. The prognosis of the patient in COS depends upon the size of the opening into the fibromuscular membrane. Corrective surgery is a treatment of choice for COS and is safer at an adult age compare to surgery in childhood. So, we recommend elective balloon dilatation of the LA membrane opening for all COS patients in childhood and then corrective surgery at adulthood as a treatment protocol.

DECLARATIONS

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Informed consent: Informed consent was obtained from the patient included in the study.

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