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Cor-Triatriatum Mimicry: Dilated Coronary Sinus Draining the Left Superior Vena Cava in Focus

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Abstract

Background: Cor-triatriatum is a rare congenital cardiac abnormality in which left atrium (sinister) or the right atrium (dextrum) is divided into two compartments by a fold of tissue or a fibromuscular membrane. Left superior vena cava (LSVC) is more frequently associated when compared to other anomalies. Due to the rarity of the condition, a high index of suspicion is need to prevent unfavourable outcomes. *Case Report*: We report a case of 11-year-old male having complaints of exertional dyspnea for 2 months, who underwent surgery for cor-triatriatum. But the membrane was formed by persistent left superior vena cava with dilated coronary sinus which was closely mimicking cor-triatriatum. *Conclusion*: This case emphasizes about the complexities associated with persistent LSVC, especially in the context of mimicking other congenital cardiac abnormalities.

Keywords: Congenital Heart Disease, Dyspnea, Echocardiography, Heart Murmurs.

Introduction

Cor-triatriatum is a rare congenital cardiac abnormality in which left atrium (sinister) or the right atrium (dextrum) is divided into two compartments by a fold of tissue or a fibromuscular membrane [1-3]. It is seen in around 0.1-0.4% of the patients with congenital cardiac disorders [4]. It was first described in 1868 by Church [5]. Later in 1905, Borst gave the name cor-triatriatum [6]. In children, it may be associated with other major congenital cardiac anomalies such as tetralogy of Fallot, double outlet right ventricle, coarctation of the aorta, partial anomalous pulmonary venous connection, persistent left superior vena cava to coronary sinus, ventricular septal defect, atrioventricular septal defect, and common atrioventricular canal [7]. Left superior vena cava is more frequently associated when compared to other anomalies [8]. In classical cor-triatriatum, proximal chamber of the left atrium receives pulmonary venous drainage and the distal chamber contains the left atrial appendage will lead to mitral valve [9]. The membrane in between the two chambers will have one or more apertures which can range from small and restrictive to large and wide open. Clinical manifestations depend upon the gradient across this membrane. Cor-triatriatum dextrum is extremely rare and is due to the persistent right sinus valve of the embryonic heart.

Echocardiography serves as the primary diagnostic modality for cor-triatriatum, given its rarity. This case report presents a unique manifestation of cor-triatriatum. The diagnostic challenges and subsequent surgical intervention highlight the importance of a comprehensive approach in managing such cases.

Case Report

A 11-year-old male presented with the complaints of exertional dyspnea NYHA class II intermittently over the last 2 months. Diagnosed with ventricular septal defect since birth, clinical

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examination revealed a pansystolic murmur. The electrocardiogram (ECG) was unremarkable, while the chest x-ray indicated pulmonary congestion with a dilated pulmonary artery (PA).

Trans-thoracic echocardiogram findings included situs solitus, levocardia, normal systemic and pulmonary venous drainage, left aortic arch, dilated main pulmonary artery, left and right pulmonary arteries, membrane in the left atrium above the level of atrial appendage suggestive of cor-triatriatum. Additionally, posterior muscular ventricular septal defect (VSD) with left to right shunt, large patent ductus arteriosus (PDA), left superior vena cava (LSVC) draining into coronary sinus and redundant mitral valve leaflets with mild tricuspid regurgitation were observed.

The diagnosis of cor-triatriatum with VSD, PDA and LSVC was confirmed, leading to decision for surgical intervention. The patient, exhibiting cardiomegaly and a dilated and tense pulmonary artery, underwent LSVC cannulation along with bicaval cannulation. Right atrium was opened revealing a dilated coronary sinus. Through inter-atrial septum, left atrium was visualized. All pulmonary veins were draining into the left atrium. The fold of the tissue inside the left atrium was the dilated coronary sinus draining the left superior vena cava. The ventricular septal defect was successfully closed, and the PDA was ligated, resulting in an uneventful post-procedure. Post-surgery echocardiogram revealed absence of residual shunt through the VSD and PDA, as well as no presence of mitral regurgitation, tricuspid regurgitation, or pulmonary artery hypertension.

Discussion

In this case, presence of a persistent left superior vena cava (LSVC) with a dilated coronary sinus closely resembled the echocardiographic features of cor-triatriatum. Persistent LSVC, a relatively common anomaly in systemic venous circulation, occurs in approximately 4% of congenital heart

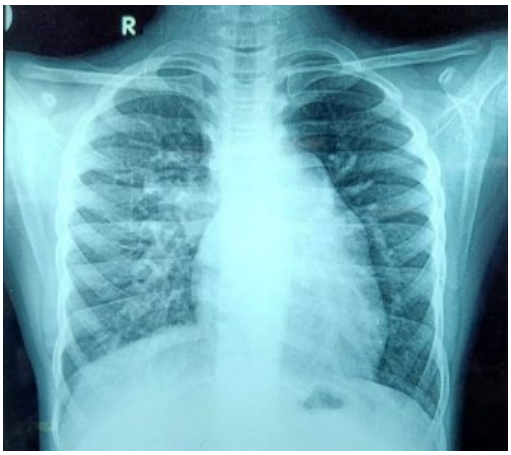

Fig.1: *Chest X-ray showing pulmonary congestion.*

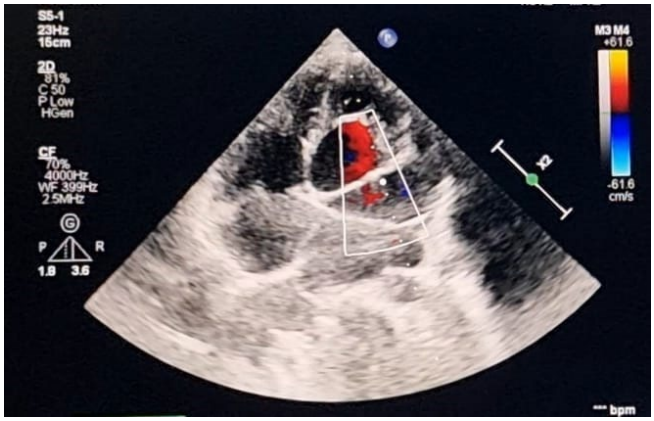

Fig.2: *2D ECHO showing membrane above the level of LA appendage.*

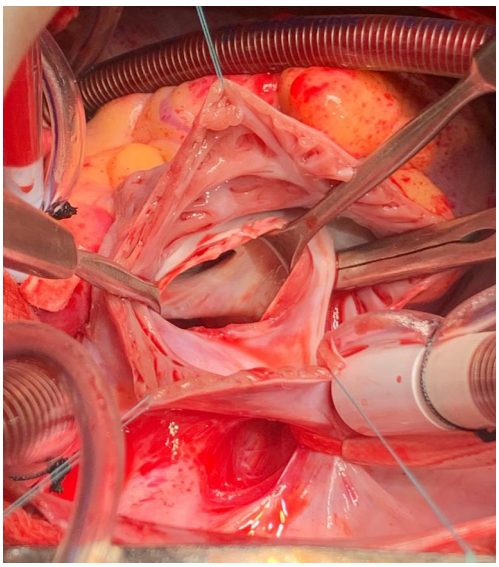

Fig.3: *Dilated coronary sinus mimicking as a membrane.*

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disease cases [10-12]. Typically, both right and left superior vena cavae co-exist, draining into the right atrium through a dilated coronary sinus [13,14]. However, in rare instances, the persistent LSVC may directly drain into the left atrium, causing coronary sinus dilation. This dilation, when adherent to the posterior left atrial wall, can lead to left ventricular inlet obstruction and altered flow across the mitral valve, potentially resulting in elevated pulmonary vein pressure and subsequent pulmonary artery hypertension [15]. While many patients with this anomaly remain asymptomatic and are incidentally discovered during surgical, imaging, or invasive procedures, there is a notable incidence of cardiac arrhythmias and conduction defects. These are believed to be attributed to the stretching and fibrosis of the atrioventricular node or the bundle of His by the dilated coronary sinus [16]. Additionally, the dilated coronary sinus has been implicated in causing left ventricular outflow tract obstruction and heart failure.

Recognizing the presence of LSVC preoperatively is crucial, as it influences the cannulation approach for cardiopulmonary bypass. Direct LSVC cannulation or access through the coronary sinus ostium may be necessary if the surgical procedure involves opening the right atrium. It is essential to note that rerouting the LSVC may not be universally required in all cases.

Conclusion

Echocardiography proves to be a sensitive diagnostic tool, emphasizing its importance in identifying cases where the dilated coronary sinus may closely resemble the echocardiographic appearance of cor-triatriatum. This case emphasizes the significance of considering and managing the complexities associated with persistent LSVC, especially in the context of mimicking other congenital cardiac abnormalities.

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