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Case Report

An uncommon cardiovascular abnormality: Case report of core triatriatum associated with persistent left superior vena cava and coronary sinus dilation ☆,☆☆

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ABSTRACT

Cor triatriatum is an uncommon cardiac defect that occurs in 0.1-0.4% of congenital heart disease patients. It is characterized by a fibromuscular membrane separating the left (sinister) or the right (Dexter) atriums in tow chambers. The disease is usually discovered accidently in late childhood, usually as a result of a moderate form of this condition type. We discuss the case of a 14-year-old girl who had been experiencing exertional dyspnea and palpitations for about a year. She was referred to our hospital for assessment after an initial echocardiogram at another hospital revealed a mass around the left atrium. Repeated echocardiography at our institution revealed dilated coronary sinus, confusing the diagnosis. The left atrium was dilated and divided into 2 chambers by a thin membrane with an elevated pressure gradient between the 2 chambers. Cardiac CT and MRI confirmed the diagnosis of cor triatriatum sinister (CTS) with concomitant persisting left superior vena cava. Because of her symptoms, she was started medical treatment and referred for surgical evaluation. Cor triatriatum sinister (CTS) is frequently accompanied with atrial septal abnormalities and enlarged coronary sinus caused by a persistent left superior vena cava, as demonstrated in our case. The management of cor triatriatum sinister (CTS) is determined by the severity of the symptoms. Asymptomatic individuals with no pressure gradient do not require therapy; however, significant membrane obstruction may require surgical removal, which typically leads to positive short- and long-term outcomes.

Abbreviations: CTS, Cor triatriatum sinister; ASD, Atrial Septal Defect; TTE, Transthoracic Echocardiography; TEE, Transesophageal Echocardiography; CT, Computed Tomography; MRI, Magnetic Resonant Imaging; LAA, Left Atrial Appendage.

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Introduction

Cor triatriatum is a rare cardiac abnormality that affects around 0.1-0.4% of people with congenital heart disease. It is characterized by the presence of a fibromuscular membrane that separates the left atrium (Sinister) or right atrium (Dexter) into 2 distinct compartments. [1–6]. Church originally described this condition in 1868 [7], and the most prevalent type, cor triatriatum sinister, contributes to approximately 54% of all instances [8].

In the classic version of cor triatriatum sinister (CTS), a fibromuscular membrane separates the left atrium into a proximal posterosuperior chamber that receives all pulmonary venous blood and a distant anteroinferior compartment which contains the mitral valve vestibule and the left atrial appendage, also known as the true atrium. The left atrium's 2 chambers are connected via membrane with holes of varying size and number [9–13]. There are other incidents of subtotal cor triatriatum, in which either the right or left pulmonary veins enter the upper chambers [14–16]. A number of hypothesis have been suggested regarding the origin of cor triatriatum sinister, with the most frequently accepted being that it is caused by a failure to incorporate the common pulmonary vein to the left atrium during fetal development [17,18].

CTS typically appears in infancy or early youth, simulating mitral stenosis [8]. The extent of pulmonary vein obstruction and hypertension is determined by the severity of left atrial blockage. Though extreme cases are associated with early death, several individuals may stay symptoms-free into adolescence or adulthood due to partial membranes obstruction or the presence of atrial septal defects (ASDs), which reduces left atrial hypertension [19–23]. CTS is commonly accompanied with additional congenital abnormalities, including ASD and an unroofed coronary sinus [22,24]. The early deaths in major case series is generally between 15-20% [8]. Long-term prognosis, on the other hand, are favorable, with an 80–90% survival rate reported for surgical patients [25].

Echocardiography is typically used to identify cor triatriatum, while MRIs and CT scans can also be used [17,23,26]. The intra-atrial membrane is removed surgically during an intervention. A number of surgical centers have reported good long-term rates of survival following membrane removal, along with a low risk of reintervention following surgery [27,28]. Patients who have no symptoms but a noticeable pressure gradient across the membrane—typically between 8 and 10 mmHg—may also be candidates for this surgery [29].

We present a case of core triatriatum sinister in a 14-year-old girl who presented with palpitations and dyspnea but had been misidentified as a mass compressing the left atrium.

Case summary

We discuss the case of a 14-year-old female with no notable medical history who arrived to our hospital with exertional dyspnea that had persisted for over a year, as well as palpitations. A transthoracic echocardiogram performed by another physician demonstrate a mass pressing the left atrium, leading her referral for an additional examination. The cardiac exam revealed tachycardic heart sounds as well as a grade III/VI diastolic murmur at the heart apex. Her electrocardiogram showed sinus tachycardia.

We performed a transthoracic echocardiography (TTE) at our hospital, which revealed normal size right ventricle and atrium with normal RV function, together with good left ventricular dimensions and function. The enlarged coronary sinus confusing the diagnosis, resulting in appearances that were originally misidentified as a mass compression the left atrium. The left atrium was dilated and divided by thin fibromuscular membrane into 2 chambers (Fig. 1). A single opening in the membrane connected the 2 chambers, and the pressure gradient across the membrane was 34 mmHg. Additionally, there was high pulmonary artery systolic pressure of 56 mmHG and mild tricuspid regurgitation (Fig. 2). Transesophageal echocardiography (TEE) was performed in order to evaluate these finding as well as any other concurrent abnormalities. The left atrial cavity had spontaneous contrast even though there were no clots in the left atrial appendage (LAA). Cor triatriatum sinistrum, group 3. Loeffler's classification was consistent with these results.

Furthermore, we conducted a cardiac computed tomography (CT) scan, which demonstrated the presence of a fibromuscular membrane separating the left atrium into a proximal chamber containing the pulmonary venous confluence and a distal chamber known as the "true" left atrium, which contained the left atrial appendage. This membrane had a 1.2 cm inferior fenestration with calcification along the borders, which allowed connection between the 2 atrial chambers and facilitated pulmonary vein decompression (Figs. 3, 4). Furthermore, superior vena cava doubling was identified, with the aberrant left SVC emptying into the right atrium through a dilated coronary sinus (an anatomical variant). (Figs. 5–7). A cardiac Magnetic Resonance Imaging (MRI) was performed in an attempt to find other congenital anomalies showed the same findings (Fig. 8).

The entire findings reveal an uncommon congenital heart abnormality known as cor triatriatum sinistrum that causes significant obstruction and severe pulmonary hypertension. The patient was referred to the cardiac surgery team for examination and surgical intervention after being prescribed B-blockers, furosemide, and spironolactone as part of her medical treatment.

Discussion

A 14-year-old girl has been diagnosed with Cor Triatriatum Sinistrum (CTS), a rare congenital defect that has been documented in just a few cases. Cor triatriatum sinistrum is an uncommon congenital cardiac anomaly that occurs in just 0.1%-0.4% of cases [30]. Cor triatriatum sinistrum is related to a membrane that separates the left atrium into 2 chambers. The upper chamber receives pulmonary venous blood, while the lower chamber is connected to the atrioventricular valve and houses the left atrial appendage. One or more membrane holes serve as communication channels between these

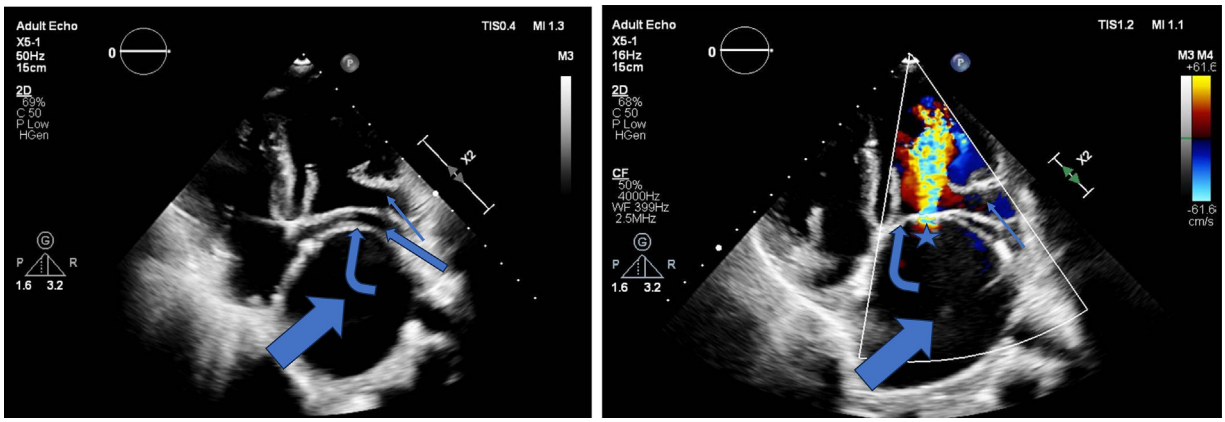

Fig. 1 – Transthoracic Echocardiography Apical 4 chambers view showing dilated coronary sinus (Arrow) and fibromuscular membrane (curved arrow) with internal fenestration and turbulent flow (star) dividing the left atrium into 2 chambers, a proximal chamber (thick arrow) containing the pulmonary vines, and a distal chamber "true" left atrium (thin arrow) containing the left atrial appendage.

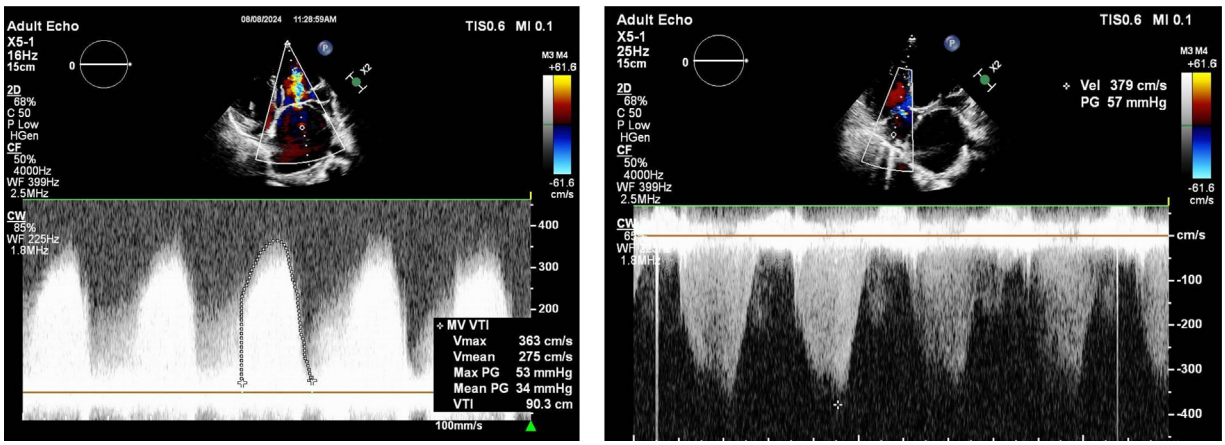

Fig. 2 – Transthoracic Echocardiography Apical 4 chambers view showing high pressure gradient across the membrane (34mmHg) and high pulmonary artery systolic pressure of 57 mmHG and mild tricuspid regurgitation.

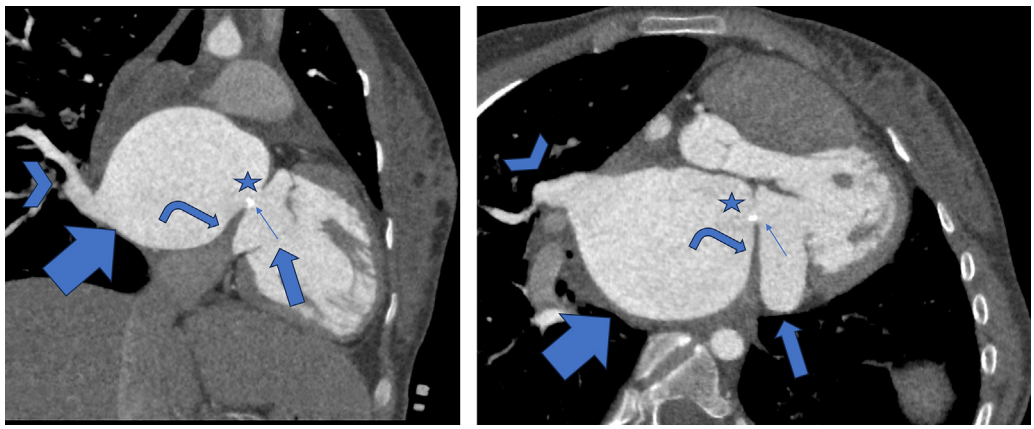

Fig. 3 – Two images of cardiac CT scans, axial and oblique sagittal views, show a fibromuscular membrane (curved arrow) with internal fenestration (star) and a focus of calcification (thinnest arrow), dividing the left atrium into 2 chambers, a proximal chamber (thick arrow) containing the pulmonary vines (arrowhead), and a distal chamber "true" left atrium (thin arrow) containing the left atrial appendage.

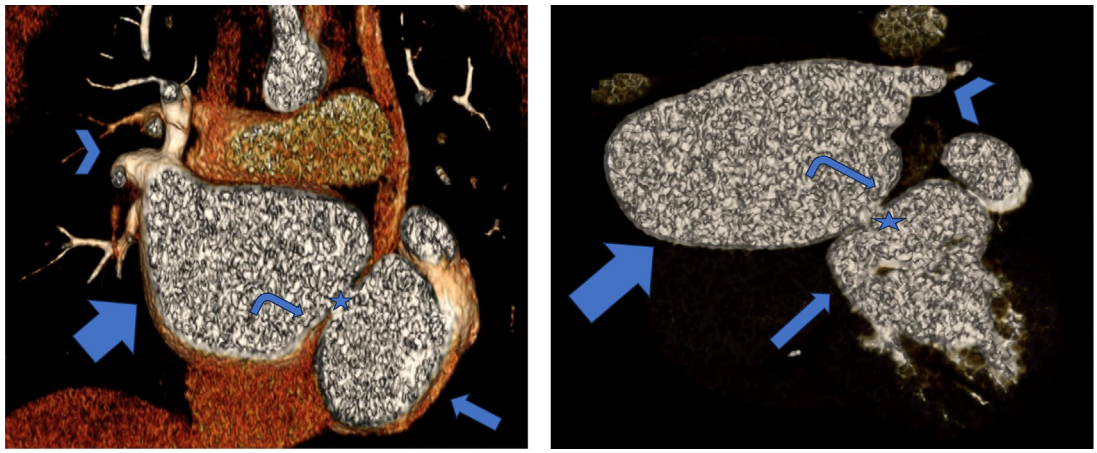

Fig. 4 – Two images of 3D reconstruction cardiac CT scans, show a fibromuscular membrane (curved arrow), with internal fenestration (star), dividing the left atrium into 2 chambers, a proximal chamber (thick arrow) containing the pulmonary veins (arrowhead), and a distal chamber "true" left atrium (thin arrow) containing the left atrial appendage.

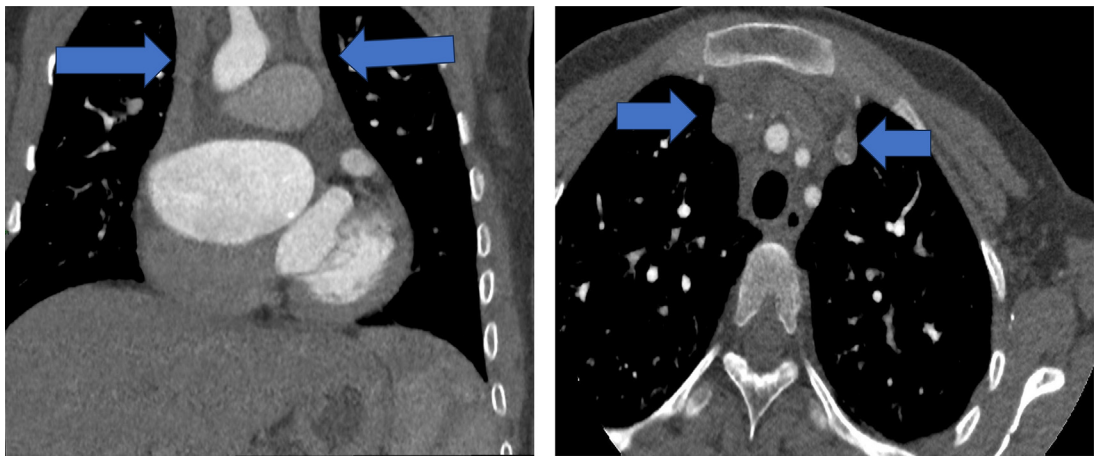

Fig. 5 – Two images of cardiac CT scans, axial and coronal views, show superior vena cava duplication (arrows).

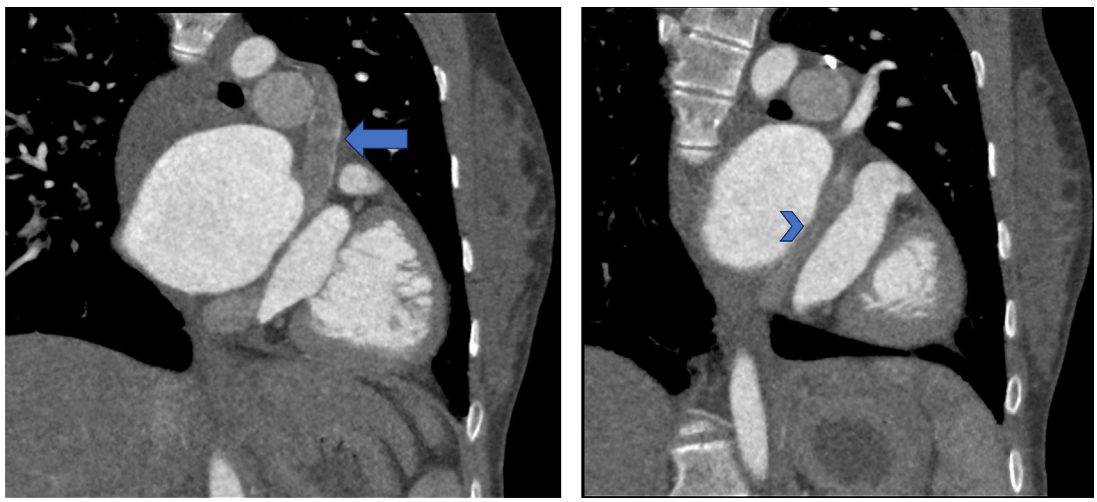

Fig. 6 – Two images of cardiac CT scans, oblique sagittal views, show persistent left-sided SVC (arrow), draining through the coronary sinus (arrowhead) into the right atrium.

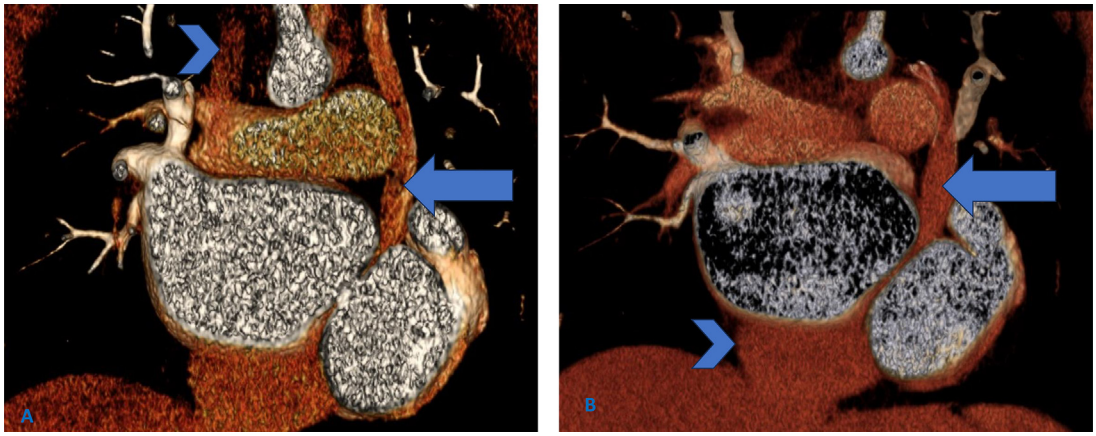

Fig. 7 – (A) 3D reconstruction cardiac CT scan shows double SVC, persistent left-sided SVC (arrow), and normal right-sided SVC (arrowhead). (B) 3D reconstruction cardiac CT scan shows persistent left-sided SVC (arrow) draining through the coronary sinus into the right atrium (arrowhead).

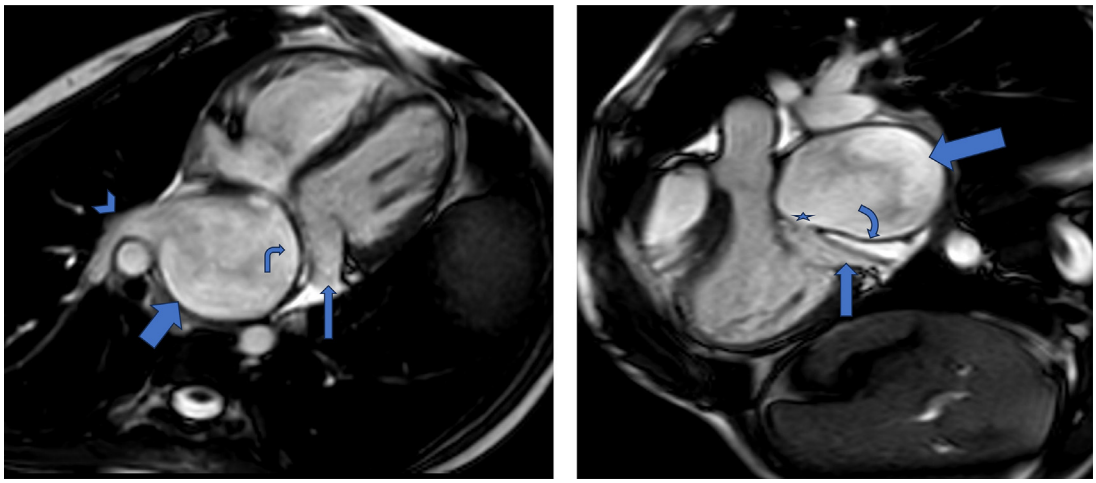

Fig. 8 – Two images of cardiac MRI, balanced turbo field echo (BTFE) sequence, axial and sagittal views, show a fibromuscular membrane (curved arrow) with internal fenestration (star), dividing the left atrium into 2 chambers, a proximal chamber (thick arrow) containing the pulmonary vines (arrowhead), and a distal chamber "true" left atrium (thin arrow) containing the left atrial appendage.

chambers [31–33]. The restrictive structure of the membrane can cause a pressure gradient, raising pulmonary arterial and venous pressures [26].

There are multiple categorization schemes for CTS, the most known of which is Loeffler's classification from 1949 [34]. The system classifies cases according to the number and size of holes in the fibromuscular membrane which divides the left atrium. Group 1: No openings (often associated with left cardiac obstruction in infancy), Group 2: One or more small openings (usually asymptomatic), and Group 3: A single large opening (also generally asymptomatic).

Children in group 1 usually exhibit pulmonary edema and other signs of left cardiac dysfunction in the early stages of infancy. Groups 2 and 3 may experience symptoms thereafter if large opening become occluded due to fibrosis or calcification, causing dyspnea, orthopnea, and hemoptysis [35,36]. Our patient, who had been symptom-free for a long time, had type 3 morphology, which is more common in nonpediatric cases and has less hemodynamic significance.

Symptoms result from pulmonary venous congestion and overload on the right side of the heart [18,23]. Patients can have symptoms at any stage of life; the magnitude of symptoms depends on the degree of pulmonary venous return restriction and accompanied defects [33]. The patient in our instance had classical cor triatriatum without an intra cardiac shunt, yet because of the extensive membrane communication, she had a prolonged period of symptom-free life. Dyspnea and orthopnea are common signs of left-sided heart failure and often brought on by obstructions in the left atrium. Infants may exhibit respiratory distress, cyanosis, frequent infections, and problems with feeding, whereas elderly individuals may develop syncope, dyspnea, and hemoptysis [22,23].

CTS is usually accompanied by atrial septal abnormalities, bicuspid aortic valves, and dilated coronary sinus caused by a persistent left superior vena cava [8]. In our case, the patient had a dilated coronary sinus and persistent left superior vena cava which complicates the diagnosis and was misdiagnosed as a mass compressing the left atrium. Complications such as atrial fibrillation or mitral regurgitation can appear later in life [37]. Cardioembolic strokes are prevalent, and thrombus formation has been linked to atrial fibrillation and sluggish blood flow in accessory compartments as well as paradoxical embolization brought on by related ASD [36].

Echocardiography is the recommended technique for diagnosing CTS because it provides detailed imaging of the membrane and allows for the measurement of blood flow and gradients. Transesophageal echocardiography (TEE) provides the advantage of better seeing the left atrium, the left atrial appendage, the membrane, and the pulmonary veins, thereby offering superior sensitivity for diagnosis [32,38]. Our patient was misdiagnosed with a tumor pressing the left atrium; yet, TEE revealed clear identifiable characteristics of the structure and confirmed the diagnosis of CTS.

Other modalities, including MRI and CT, are useful in assessing related congenital abnormalities and the hemodynamics. When compared to echocardiography, cardiac CT and MRI offer improved tissue visualization and higher spatial resolution. Furthermore, cardiac MRI and CT scans are simple to perform in a variety of planes [17,39,40]. A comparative investigation found that MRI detected pulmonary venous abnormalities more accurately than echocardiography or angiography [41]. Our patient underwent cardiac MRI and cardiac CT scans, which ruled out other congenital abnormalities and confirmed the diagnosis of CTS.

The severity of the symptoms determines how CTS is managed. Patients with incidental results but no symptoms do not require treatment. Digoxin, diuretics, and preload reduction can be utilized to treat severe symptoms such dyspnea and lung congestion [42]. There are currently no recommendations for reducing the risk of thrombosis in patients with CTS; nevertheless, in cases of embolic events, a low threshold for initiating anticoagulation is advised, even in the absence of visible thrombus or established atrial fibrillation [43]. If an atrial membrane is discovered by accidentally without a pressure gradient, there is no need for treatment; however, if the obstruction is severe, surgery to remove the membrane may be required, with usually favorable short- and long-term outcomes. Catheter-based therapy may be considered in certain instances, but long-term outcomes are unknown, particularly in patients with atrial membrane calcification for whom dilation may not be beneficial [27,44].

Conclusion

Cor triatriatum is a rare cardiac defect that occurs in 0.1-0.4% of congenital heart disease cases. It is distinguished by a fibromuscular membrane that separates the left and right atria. Cor triatriatum sinister (CTS) occurs in combination with atrial septal abnormalities and enlarged coronary sinuses caused by a persistent left superior vena cava, which can challenge the diagnosis. Treatment for CTS is determined by the severity of the symptoms; asymptomatic individuals with no pressure gradient may not require intervention, but those who have severe obstruction may require surgical removal, which typically results in good short- and long-term results.

Ethics approval

In compliance with ethical and legal regulations.

Patient consent

The patient provided written informed consent for the publication of this case.

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