Case Report

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A 10-year-old female with Cor triatriatum sinister (CTS): a rare case report and literature review from Syria

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Introduction and importance: Cor triatriatum sinister (CTS) is an uncommon heterogeneous congenital cardiac defect that may manifest in adulthood when symptomatic blockage manifests due to a change in hemodynamic physiology or when a condition such as atrial fibrillation (AF) arises. The incidence of cor triatriatum with cardiomyopathy and congenital heart illness ranges from 0.1 to 0.4%.

Case presentation: The Department of Cardiology examined a 10-year-old girl for a diastolic murmur. The patient's medical history was ordinary, and no previously known co-morbid illnesses were present. Pneumonia was the patient's original medical condition. A cardiac murmur was also discovered during the physical examination, and the patient was forwarded for more research. Physical examination revealed just a diastolic murmur, which was noteworthy. The patient's care plan includes routine echocardiographic monitoring. Since the deformity was not clinically significant, surgical repair was not advised.

Clinical discussion: It is unclear what caused this flaw. A full, incomplete, or fenestrated septum may be the result of the common pulmonary vein failing to merge with the left atrium during embryonic development. The existence of a fibromuscular membrane, which separates the left atrium (LA) into two chambers—the proximal chamber receiving the pulmonary veins (PVs) and the distal chamber contains the mitral valves and LA appendages defines the disease.

Conclusion: The diagnostic procedure should be carried out in the optimal settings, however, in cases of frail health systems, employing accessible alternatives might help the early diagnosis. Early referral to a cardiologist is required in case of CTS suspicion.

Keywords: atrial fibrillation, cardiac abnormalities, cardiology, Cor triatriatum sinister

Introduction

Cor triatriatum sinister (CTS) is a rare heterogeneous congenital cardiac anomaly that may present in adulthood when symptomatic obstruction appears as a result of a change in hemodynamic physiology or when a complication such atrial fibrillation (AF) develops[1]. Patients with congenital cardiac disease and cardiomyopathy have a cor triatriatum incidence of 0.1–0.4%. There is no genetic explanation for the 1.5:1 male to female ratio[2]. The presentation is described in several early case studies as being comparable to mitral stenosis, with dyspnoea symptoms, heart failure symptoms, and imaging that is consistent with pulmonary oedema. Although the embryologic cause of CTS is unknown, enlargement of the septum primum or aberrant integration of the pulmonary veins into the left atrium have both been proposed as potential causes[3]. Some physiological and pathological situations, such as all sources of increased heart flow and venous return, may cause symptoms to develop such as pregnancy. Traditionally, cardiac angiography is used to make diagnoses.

HIGHLIGHTS

- CTS is a rare heterogeneous congenital cardiac anomaly that may present in adulthood when symptomatic obstruction appears as a result of a change in hemodynamic physiology or when a complication such atrial fibrillation (AF) develops.
- Traditionally, cardiac angiography is used to make diagnoses. Nowadays, a computerized tomography (CT) or MRI can be used to confirm the diagnosis. Due to its ability to provide more specifics, (MRI) has the benefit of finer research. Better spatial resolution and the possibility for multiplanar reconstructions are provided by CT scanning.
- Premorbid referral to a cardiologist at an early stage is essential if there is any CTS suspicion. Although the diagnostic procedure should be carried out under ideal circumstances, applying the available alternatives can help with an early diagnosis in cases of weakened healthcare systems.

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Nowadays, a computerized tomography (CT) or MRI can be used to confirm the diagnosis. Due to its ability to provide more specifics, (MRI) has the benefit of finer research. Better spatial resolution and the possibility for multiplanar reconstructions are provided by CT scanning . The clinical presentation affects the course of treatment. When there are no symptoms, a cautious approach is suggested. As a result, individuals with no symptoms or a pressure gradient may not need medical attention; nevertheless, when there are indicators of pulmonary overload, medical therapy is recommended in addition to surgical intervention. There were also some reports of successful catheter ablation testing. For individuals with atrial fibrillation, medical therapy focuses on hemodynamic stability, cardiac rhythm regulation, and anticoagulation. The only effective therapy is surgery. Atriotomy, interatrial membrane excision, and treatment of related defects make up this procedure. In most instances, the outcome of surgical repair appears to be favourable[3,4]. The chance of recurrence following surgery is quite low[3]. Herein we report a case of a 10-year-old female child diagnosed with pneumonitis who was evaluated in our cardiology clinic for a diastolic murmur to turn out that she has CTS.

Case presentation

A 10-year-old female was evaluated in the Department of Cardiology for a diastolic murmur. Medical history was unremarkable, and the patient had no previously known co-morbid diseases. The patient was initially diagnosed with pneumonia. Additionally, the physical examination also revealed a heart murmur, and the patient was referred for further investigation. Upon physical examination, the only significant finding was a diastolic murmur on auscultation (Grade II; faint but easily audible; heard at the apex).

Transthoracic echocardiography showed a transverse septum spanning the left atria upstream of the auricle level, dividing the left atria into a superior chamber that receives blood from the pulmonary veins, and an inferior chamber facing the mitral valve. The two chambers were hemodynamically connected via a fenestration in the septum measuring (18×20 mm) in diameter. The pressure gradient (PG) generated across the septum was minor and not clinically pronounced (Panels A, B, and C in Fig. 1).

The echocardiography also showed a ventricular septal defect (VSD) manifesting as a peri-membranous orifice, which was spontaneously closed by a septal aneurysm measuring (5×4 mm). The aneurysm showed to be firmly closing the defect with no blood flowing across the orifice (Panel D in Fig. 1).

The patient management included regular follow-up by echocardiography. Surgical correction was not recommended since the malformation was not clinically significant. Within 2-year of follow-up, She was followed up 2 years after the initial diagnosis; there was no PG deterioration or limitations in physical activity.

Discussion

The cor triatriatum sinister (CTS) represents a rare congenital anomaly, with an estimated incidence of 0.1–0.4% in the children with congenital heart disorders[5–9]. It affects both genders during infancy and childhood and rarely in the elderly, as few cases were recorded in adulthood[1,2,4]. It was first described by the church in 1868[5,10,11]. The underlying cause of this defect is not clear. It may be caused by embryonic development failure of the common pulmonary vein to fuse with the left atrium, creating a complete, incomplete, or fenestrated septum[2,4,5]. The condition is characterized by the presence of a fibromuscular membrane that divides the left atrium (LA) into two chambers: the proximal chamber receives the pulmonary veins (PVs) while the distal chamber contains the mitral valves and LA appendages[5,7,8,10–15]. Cor triatriatum sinister may be isolated (Classical Form) or in combination with other congenital (Atypical Form) heart defects. The atypical form will be shown rarely in adolescents and adults. But it still occurs in 50–85% of all patients with cor triatriatum sinister. Older patients are more likely to be affected by an isolated type of CTS[16]. In 80% of cases, it is associated with Other cardiac malformations like atrial septal defect (ASD), atrioventricular septal defect (AVSD), or tetralogy of Fallot[5]. The symptoms of cor triatriatum depend on the number and size of orifices in the membrane[2]. In adults, it can be asymptomatic in most cases or with symptoms such as hemoptysis, stroke, heart failure[2,4], tachyarrhythmias, dyspnoea or progressive abdominal pain[17–21]. In infants, its manifestations can be poor feeding, growth retardation and/or failure to thrive[2] dyspnoea, and recurrent respiratory infections[20,21]. Neonates who have had limited apertures may experience respiratory distress and a higher risk of death[2,4]. CTS was found to be associated with substantially higher rates of AF and stroke[1,2,21,22]. The severity of its appearance varies according to the degree of obstruction in that membrane, according to the classification of Loeffler into three categories: the first category having no opening, the second having one or more openings, and the third having a wide opening[5,7,10,13]. It has also been classified into five categories based on morphology and into diaphragmatic, hourglass, or tubular cor triatriatum based on the shape of the accessory left atrial chamber[2]. The majority of patients have CTS type II[23]. The morbidity and mortality of cor triatriatum sinistrum is higher in those who are symptomatic in infancy and depends on the size of the communications between chambers and congenital cardiac lesions[2]. Mortality may exceed 75% in untreated symptomatic infants. On the other hand, surgical correction is excellent in adults with isolated cor triatriatum[3,21,24]. The risk appears to be higher in patients with concomitant cardiac anomalies and in those with pulmonary hypertension for a long time. Complications may include the following: cardiogenic shock[21], pulmonary oedema[21], pulmonary hypertension[9,21,24,25], arterial hypertension[25] Right-sided heart dysfunction[23], Atrial arrhythmia[3] and dilated cardiomyopathy[21] were also reported. In addition, There have been reports of a rare cor triatriatum complication: ischaemic stroke[21,22].

The cornerstone of assessment and diagnosis involves imaging modalities, such as chest radiography, electrocardiography, and echocardiography, as well as angiography and left and right heart catheterization[26–28].

The diagnosis of cor triatriatum sinister can be confirmed by means of imaging tests, such as transthoracic echocardiography and transesophageal echocardiography, which may be required to characterize, define, and distinguish it from other cardiac abnormalities, such as atrial septal defect and supravalvular mitral annulus[29,30]. Echocardiography is the preferred diagnostic method because it not only enables a definitive diagnosis but also allows for the three-dimensional reconstruction of

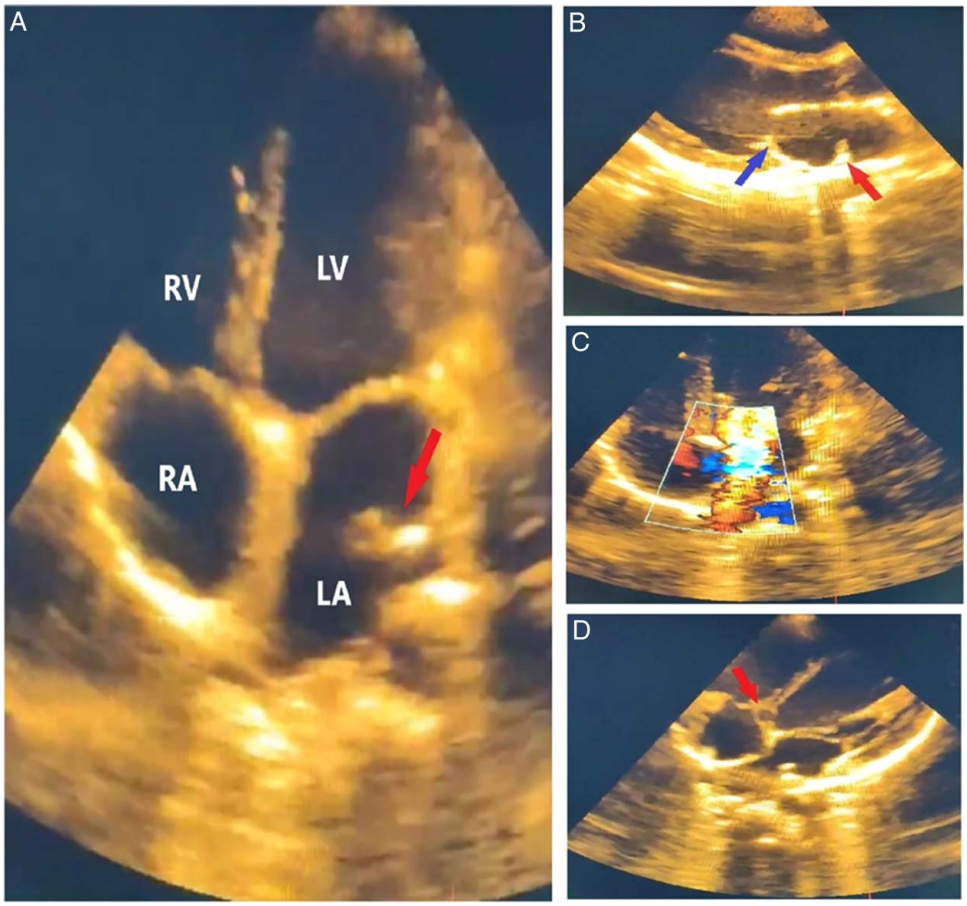

Figure 1. (A) Transthoracic echocardiography, LA left atrium; LV left ventricle; RA right atrium; RV right ventricle; red arrow indicates the intra-atrial malformation. (B) Additional view of the atrial malformation, indicated by red arrow; blue arrow indicates mitral valve. (C) Blood flow in the left atria, intra-atrial transeptal PG = 2 mmHg. (D) Ventricular septal defect (VSD) with septal aneurysm, indicated by red arrow. LA, left atrium; RA, Right atrium.

echocardiographic images, which precisely identifies the location of the defect and appendage and guides the surgical approach to the disease. Echocardiography provides clear visualization of the left atrial appendage and its fenestrations, as well as the presence of atrial septal defect, pulmonary stenosis, mitral valve stenosis, or regurgitation. Additionally, echocardiography can reveal the patterns of pulmonary arterial and venous drainage. By visualizing the left atrial appendage in the left atrium, echocardiography can distinguish between cor triatriatum and supravalvular mitral stenosis. Transesophageal echocardiography provides a detailed description of the specific type and structure of the defect, particularly in elderly patients. However, if this method is not available, transthoracic echocardiography can be used instead[31,32].

Our case was represented by pneumonia, and accidentally discovered a diastolic murmur; physical examination confirmed a diastolic murmur without any other finding. As transoesophageal echocardiography was not accessible, we have resorted to utilizing transthoracic echocardiography instead.

The clinical appearance affects the course of treatment. When there are no symptoms, a conservative approach is suggested. As a result, individuals with no symptoms or a pressure gradient do not need medical attention; nevertheless, when there are indicators of pulmonary overload, medical therapy is recommended in addition to surgical intervention. There were also some reports of successful catheter ablation testing. For individuals with atrial fibrillation, medical therapy focuses on hemodynamic stability, cardiac rhythm regulation, and anticoagulation[1]. In our case, routine echocardiography follow-up was part of the patient's care. As the deformity was not clinically significant, surgical repair was not forcedly advised. When she was last checked two years after the original diagnosis, neither her PG condition nor her ability to engage in physical exercise had worsened. In the absence of severe blockage, individuals with isolated CTS diagnoses or adults receiving their first CTS diagnosis may not need long-term strict monitoring. Furthermore, during follow-up, surgical excision of the CTS membrane was safe and successful in alleviating blockage without recurrent obstruction. Prior research revealed a minimal probability of persistent or recurrent blockage following CTS membrane excision; this discovery has therapeutic implications for postoperative advice[2]. The only effective therapy is surgery. Atriotomy, interatrial membrane excision, and treatment of related defects make up this procedure. In the majority of instances, the outcome of surgical repair appears to be great[1]. Before surgery, it is essential to outline the intricate anatomy of the cor triatriatum in each patient. Long-term left ventricular inflow blockage in CTS can raise the dangers of pulmonary artery hypertension and backflow pulmonary

venous congestion. Prior to surgery, individuals with CTS should make sure that their pulmonary congestion is reversible. Although it is not known if CTS patients need anticoagulation to avoid a stroke, medical management often plays no part in the treatment of CTS[3]. Nevertheless, transcatheter methods of CTS decompression have been reported, demonstrating that they are a secure, efficient, and perhaps even long-lasting substitute for surgical therapy. When surgical risk is high, when there is a strong patient preference to avoid surgery, or when presented as a stopgap measure before more permanent treatment, these approaches present an appealing option. It has been suggested that an effective alternative to surgical excision of the obstructive membrane is transcatheter balloon decompression. The first transcatheter approach (1996) was technically challenging since it had to overcome the CTS membrane orifice's close proximity to the interatrial septum (IAS), which is right next to the aortic root wall. For wire access via the distal left atrium (DLA) and into the ascending aorta and access to the defect, a Judkins RCA catheter had to be looped in the proximal left atrium (PLA). It has not been adequately researched if CTS can be treated percutaneously when the CTS membrane has considerable calcification. Two CTS patients were effectively treated using a transcatheter method by Blais and colleagues, who used RF radiation to perforate the obstructive membrane before balloon angioplasty and LA decompression[4]. Atrial fibrillation and flutter were prevalent in the general population, and patients who had atrial arrhythmias at the time of their CTS diagnosis tended to be older. In the review of patients with CTS followed at Mayo Clinic Rochester from 1990 to 2016 , only six patients with additional concomitant structural cardiac problems and one patient with atrial arrhythmia had an obstructive CTS membrane. Thus, the majority of atrial fibrillation and flutter patients are probably caused by cardiac disease that develops with age[2]. Presentation and treatment have an impact on patient survival rates. Better survival results can be achieved with early surgical intervention. One research, for instance, found that the 10-year survival percentage for 25 individuals with cor triatriatum who underwent surgery at the age of 19 was 83%[3].

Conclusion

Early referral to a cardiologist is essential premordia in the case of CTS suspicion. The diagnostic process is to be conducted in the best conditions; however, in the case of fragile health systems, using available alternatives can facilitate the early diagnosis.

Methods: The work has been reported in line with the SCARE criteria[33].

Ethical approval

Not applicable because all date belong to the authors of this article.

Consent

Written informed consent was obtained from the patient's parents/legal guardian for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

M.S. wrote most of the manuscript and performed data analysis or interpretation and designed the study. B.S. wrote a part of the manuscript. B.Hu. wrote a part of the manuscript and performed data analysis or interpretation and designed the study. A.J. wrote a part of the manuscript and designed the study. B.Ha. wrote a part of the manuscript. Y.S. wrote a part of the manuscript.

M.Mas. wrote a part of the manuscript. M.Man. wrote a part of the manuscript. All authors reviewed the final manuscript.

Conflicts of interest disclosure

The authors declare no conflict of interest.

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