JALALABAD MEDICAL JOURNAL

Case Report

Cor Triatriatum Sinister with Secundum Atrial Septal Defect

Md. Suhail Alam[1], Saleh Ahmad Tahlil[2], Ayesha Rafiq Chowdhury[3], Shah Jamal Hussain[4], Md. Atiqur Rahman[5], Tafhema Tarin[6]

[1,2]Assistant Professor, Department of Cardiology, Jalalabad Ragib-Rabeya Medical College, Sylhet.

[3,4]Professor, Department of Cardiology, Jalalabad Ragib-Rabeya Medical College, Sylhet.

[5]Registrar, Department of Cardiology, Jalalabad Ragib-Rabeya Medical College Hospital, Sylhet.

[6]Junior Consultant, Department of Paediatrics, Sylhet Women's Medical College Hospital, Sylhet

ABSTRACT

Cor triatriatum is a rare congenital heart defect, while secundum atrial septal defect (ASD) is a relatively common congenital heart defect. Primary symptoms in an older patient may mimic reactive airway disease if the diagnosis is not made in infancy. We report a case of cor triatriatum sinister in a 2 year old child, previously diagnosed with recurrent upper respiratory tract infection (RTI), presenting to a paediatrician. The initial response to treatment with bronchodilators and corticosteroids was not responding, prompting a thorough evaluation. Subsequent imaging was done and diagnosed as cor triatriatum sinister with secundum ASD. When patients with this type of scenario do not respond adequately to classical conservative management, it would be better for a clinician or paediatrician to look for an alternative diagnosis, as in this case.

Keywords: *Congenital heart disease, Cor triatriatum sinister, Atrial septal defect.*

[Jalalabad Med J 2024; 21 (1): 23-25]; DOI: https://doi.org/10.3329/jmj.v21i1.78127

INTRODUCTION

Cor triatriatum sinister is one of the rarest forms of congenital heart defect (CHD), with an estimated incidence of 0.1-0.4% of all CHDs[1]. In this condition, the left atrium is divided by a fibromuscular membrane into two distinct chambers. One chamber has the pulmonary veins, and the other has the mitral valve[2]. The presentation of patients can be during infancy, childhood, or adulthood, and this is largely due to variation in the degree of obstruction to the pulmonary venous return and the presence of associated lesions[2]. We describe the case of a 2 year old patient presenting with symptoms of upper respiratory tract infection associated with cor triatriatum sinister and a secundum ASD.

Address of Correspondence:
Dr. Md. Suhail Alam, Assistant Professor, Department of Cardiology, Jalalabad Ragib-Rabeya Medical College, Sylhet. E-mail: dr.suhail.alam@gmail.com.

CASE REPORT

A 2-year-old female child who was previously diagnosed with bronchial asthma has been treated by her paediatrician at the Women's Medical College outpatient department for recurrent respiratory tract infection (RTI). She presented with cough and shortness of breath to her physician, and despite conservative management with optimum medications, she was not responding appropriately to the treatment. Routine investigations were advised. Her chest radiography showed an enlarged cardiac silhouette, and hence she was advised further to perform a transthoracic echocardiogram.

Transthoracic Echocardiography

Transthoracic echocardiography revealed a left atrial membrane (Figures-1 and 2) bisecting the LA cavity

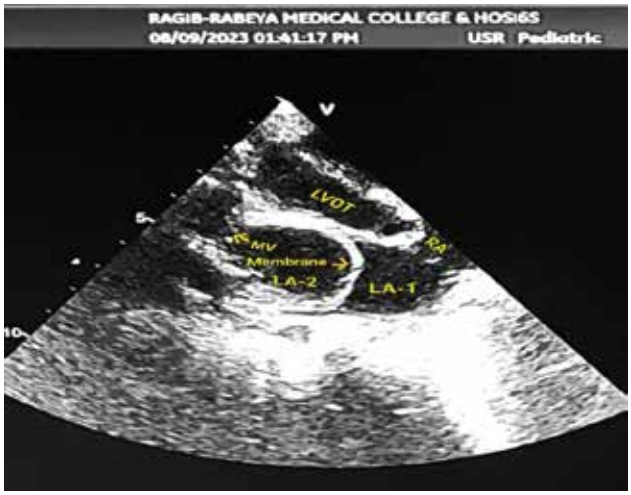

Figure-1: *Modified 2D PLAX view in diastole showing a membrane bisecting the left atrial cavity into 2 (LA-1 & LA-2).* ***LA****-2 shows distally opened mitral valve (MV).* ***LA****- Left atrium,* ***MV****- Mitral valve,* ***LVOT****- Left ventricular outflow tract &* ***RA****- Right atrium.*

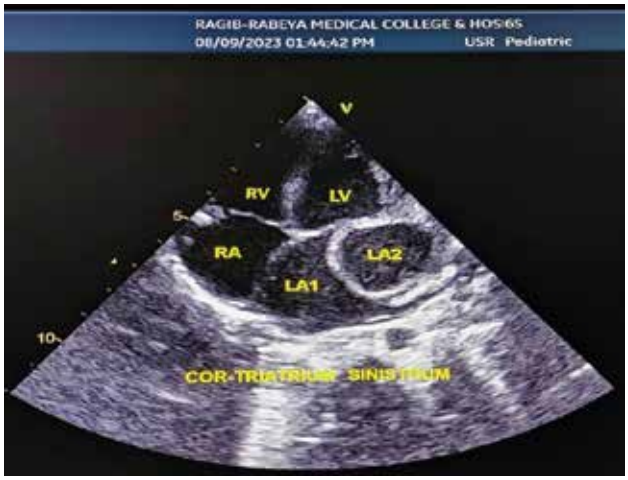

Figure-2: *Apical four-chamber view in systole showing a membrane (Orange arrow) bisecting the left atrial cavity into LA-1 & LA-2.* ***LA****- Left atrium,* ***RA****- Right atrium,* ***LV****- Left ventricle &* ***RV-*** *Right ventricle.*

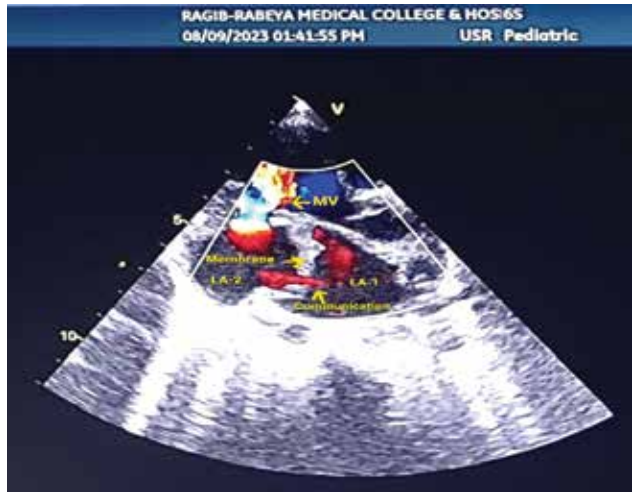

Figure-3: *Modified 2D PLAX view with color doppler across the membrane, shows shunt across the fenestration in the membrane from LA-1 to LA-2 and also across mitral valve.*

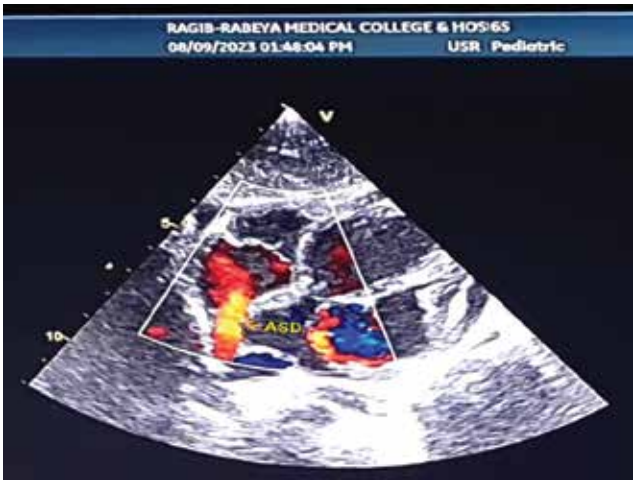

Figure-4: *Subcostal four-chamber view with color doppler across the secundum ASD reveals prominent left-to-right interatrial shunting.* ***ASD-*** *Atrial septal defect.*

into 2 (LA-1 and LA-2) cavities, and blood was shunting across the membrane through a small shunt (Figure-3). A small secundum ASD about 5 mm in size (Figure-4) with left to right shunt was also observed while reviewing the atrial septum. Mild mitral regurgitation (MR) was also seen with normal pulmonary artery pressure and good bi-ventricular function.

DISCUSSION

Our case is a two years old female with a recurrent history of upper respiratory tract infection, whom we have diagnosed as a case of cor triatriatum sinister (Type 3) with small ASD secundum and mild MR.

Cor triatriatum sinister is a relatively rare congenital cardiac condition accounting for 0.1-0.4% of all congenital heart disease[1]. It is often associated with other common forms of congenital heart defects, like ASD and partial anomalous pulmonary venous drainage (PAPVD)[3]. In cor triatriatum sinister, a fibromuscular membrane divides the left atrium into two chambers.

The upper chamber, or upper pulmonary venous chamber, connects the four pulmonary veins and receives oxygenated blood. The lower left chamber communicates with the left atrioventricular valve, and the left atrial appendages (LAA) arise from it. Supravalvular mitral membrane (SVMM) is a strong differential diagnosis that is seen in Shone syndrome, but the presence of LA appendages excludes it[4]. Cor triatriatum sinister has been classified into three types by Loeffler in 1949. Type 1 cor triatriatum sinister has no communication in the fibromuscular membrane; type 2 has one or several small perforations in the membrane; and type 3 has a single wide opening[5,6]. Our patient had type 3 morphology, which was haemodynamically less significant and was associated with a small secundum ASD. Interestingly, both of these defects couldn't produce any significant haemodynamic effect on pulmonary vascular resistance[7]. Patients with cor triatriatum sinister usually present during infancy and childhood during routine check-ups for an underline murmur or recurrent chest infections. However, it may remain unnoticed until early adulthood, and at this time, the patient may present with the complications of an underlying lesion due to a significant change in haemodynamics. Type 3 usually presents in early adulthood as it is less haemodynamically significant and remains unnoticed until adulthood[4,7]. Further investigations like trans-esophageal echocardiography and cardiac catheterization are needed for better imaging of the lesions and the haemodynamic effects exerted by the lesions. As our case has no significant haemodynamic effects with normal pulmonary artery (PA) pressure and good bi-ventricular function, we have referred the case to a cardiovascular surgeon, as the patient may benefit if early corrective surgery is done. Surgical outcomes are typically favourable at experienced centres, with almost all the patients becoming asymptomatic, and the 5 years survival rate is >90%[3,4,8,9]. Cardiac MRIs 1-2 years later will provide a better assessment of functional and haemodynamic reversibility, if any[10,11].

CONCLUSION

Cor triatriatum sinister is a relatively rare congenital heart disease. Its detection and differentiation from supravalvular mitral membrane require careful echocardiographic assessment. All infants and children with a recurrent history of chest infections should undergo routine echocardiographic assessment to rule out any underlying congenital heart defects. In the presence of a single lesion or multiple lesions of CHD, further evaluation with cardiac MRI and invasive cardiac catheterization should be advised. This will provide options for early invasive or surgical interventions and, hence, a better outcome.

REFERENCES

1. Hongkan W, Thongchaiprasit K, Durongpisitkul K. Transcatheter closure of atrial septal defect in a patient with cor triatriatum sinister and atrial septal defect. Case Rep Cardiol 2011; 2011. doi: 10.1155/2011/740981.
2. Patel MB, Opotowsky AR. Cortriatriatum and congenital mitral stenosis. In: Gatzoulis MA, Webb GD, Daubeney PEF, editors. Diagnosis and Management of Adult Congenital Heart Disease. 3rd ed. Philadelphia: Elsevier; 2018. p 340-347.
3. Humpl T, Reineker K, Manlhiot C, Dipchand AI, Coles JG, McCrindle BW. Cor triatriatum sinistrum in childhood. A single institution's experience. Can J Cardiol 2010; 26 (7): 371-6.
4. Trejo-Nava CA. Case report of cor triatriatum. Rev Mex Cardiol. 2015; 26 (Suppl 3): 114-21.
5. Loeffler E. Unusual malformation of the left atrium; pulmonary sinus. Arch Pathol (Chic) 1949; 48: 371-6.
6. Nassar PN, Hamdan RH. Cor triatriatum sinistrum: classification and imaging modalities. Eur J Cardiovasc Med 2011; 1 (3): 84-7.
7. Slight RD, Nzewi OC, Buell R, Mankad PS. Cor-triatriatum sinister presenting in the adult as mitral stenosis: an analysis of factors which may be relevant in late presentation. Heart Lung Circ 2005; 14 (1): 8-12.
8. Alphonso N, Norgaard MA, Newcomb A, d'Udekem Y, Brizard CP, Cochrane A. Cor triatriatum: presentation, diagnosis and long-term surgical results. Ann Thorac Surg 2005; 80 (5): 1666-71.
9. Işık O, Akyüz M, Ayık MF, Levent E, Atay Y. Cor triatriatum sinister: a case series. Turk Kardiyol Dern Ars 2016; 44 (1): 20-3.
10. Lambert J, Oechslin E, Tsang W, Osten MD, Thavendiranathan P. Cor triatriatum sinister with secundum atrial septal defect. CASE (Phila) 2017; 1 (4): 141-6.
11. Kochav J. (2018). Cor Triatriatum. In: DeFaria Yeh, D, Bhatt A, editors. Adult Congenital Heart Disease in Clinical Practice. In Clinical Practice. Springer, Cham. https://doi.org/10.1007/ 978-3-319- 67420 -9_12.