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CASE REPORT

Cor Triatriatum Sinistrum

A rare disease with a common presentation

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ABSTRACT: Cor triatriatum sinistrum (CTS) is a rare congenital cardiac anomaly characterised by an abnormal septum within the left atrium impairing blood flow to the left ventricle. We report the case of a two-month-old male infant who presented with symptoms of heart failure since the age of two weeks. He was admitted to a local hospital and was managed with antibiotics because of the impression of pneumonia. Due to persistent unresolved tachypnoea and tachycardia, he was referred to Sultan Qaboos University Hospital, Muscat, Oman, in 2019 for cardiac evaluation which confirmed a diagnosis of isolated CTS with severe stenosis and pulmonary hypertension. He underwent an urgent surgical excision of the membrane with uneventful recovery.

Keywords: Cor Triatriatum Sinistrum; Congenital Heart Disease; Pulmonary Hypertension; Heart Failure; Case Report; Oman.

Cor triatriatum sinistrum (CTS) is a rare congenital cardiac anomaly characterised by an abnormal septum within the left atrium impairing blood flow to the left ventricle. It can be isolated or associated with other congenital heart anomalies and commonly diagnosed in early childhood.[1] Diagnosis is suspected by findings of cardiomegaly and pulmonary oedema on chest radiograph (CXR) and right axis deviation and right ventricular hypertrophy on electrocardiogram (ECG). However, a detailed echocardiogram is required for confirmation and ruling out associated lesions. This report describes a two-month-old infant who presented with heart failure symptoms since the age of two weeks and was diagnosed with CTS by echocardiogram. He underwent an urgent surgical intervention with an uneventful recovery.

Case Report

A two-month old male infant was referred to Sultan Qaboos University Hospital, Muscat, Oman, in 2019 with a five-day history of fever, cough and shortness of breath. There was a history of interrupted feeding and sweating since the age of two weeks. He was not thriving well with a presenting weight of 3.4 kg (birth weight: 3 kg). He received a course of intravenous antibiotics in the local hospital due to the diagnosis of a possible chest infection. He was referred for a cardiac evaluation because of persistent tachypnoea, tachycardia and mild cardiomegaly on chest radiograph. There was a history of two previous similar presentations but he was managed as an outpatient with nebulisation and oral antibiotics without much improvement. There was no history of cyanosis or choking while feeding. There were no maternal illnesses and the pregnancy was uneventful. The antenatal detailed anatomy ultrasound scan was reported as normal. He was born at term by caesarean section for maternal indication (previous two caesarean sections).

On examination, he looked emaciated with no obvious dysmorphic features had a weight of 3.4 kg (<3rd centile) and a length of 57 cm (<25th centile). His respiratory rate was 48 breaths/min and he had a heart rate of 150 beats/min, blood pressure of 99/53 mmHg and oxygen saturation of 100% in room air. Cardiovascular examination showed a normal S1 and loud S2 with a soft ejection systolic murmur in the left upper sternal border. His abdomen was soft with the liver 2 cm below the right costal margin. There were no abnormalities detected in the basic blood work-up. CXR showed cardiomegaly with upturned cardiac apex indicating right ventricular hypertrophy with features of pulmonary venous congestion [Figure 1]. ECG showed sinus rhythm with right axis deviation and right ventricular hypertrophy with strain pattern [Figure 2]. Echocardiogram showed *situs solitus* with levocardia. A double chambered left atrium was seen separated by a thick membrane with a restrictive opening of 2 mm to the lower atrial chamber with severe stenosis with peak gradient (PG) of 30 mmHg and a mean of 15 mmHg [Figure 3]. There was a mild tricuspid valve regurgitation with PG of 80 mmHg consistent with severe pulmonary hypertension. A small patent foramen ovale was seen with restrictive flow with PG of 22 mmHg and a mean of 14 mmHg.

The right atrium and right ventricle were dilated with flattened interventricular septum. The mitral valve

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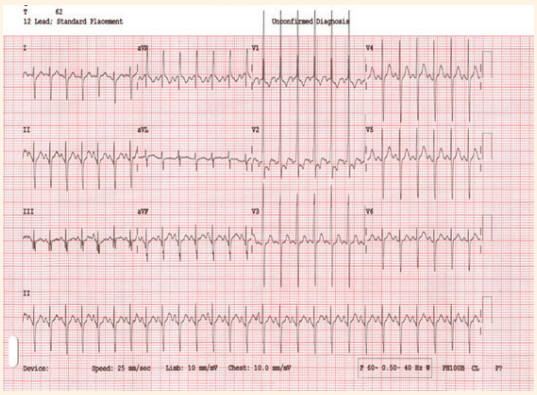

Figure 1: 12-leads electrocardiogram of a two-month-old male infant with cor triatriatum sinistrum showing sinus rhythm with right axis deviation and right ventricular hypertrophy with strain pattern.

appeared normal with mild mitral valve regurgitation. All pulmonary veins drained to the upper left atrial chamber with no stenosis. There was no right or left outflow tract obstruction. The left atrial appendage opening was seen below the membrane. The aortic arch was normal with no patent ductus arteriosus or coarctation of aorta. Systolic cardiac function was normal. The findings were consistent with CTS and therefore, the patient was referred for surgical repair. Pre-operative transoesophageal echocardiogram (TEE) confirmed the CTS diagnosis and showed a well demarcated CTS with good-sized left atrial appendage below the level of the transverse membranous partition with small fenestration to the lower chamber with severe restrictive flow. Intraoperatively, the membrane was visualised and resected fully and the interatrial communication was closed. He was successfully extubated on the second day and weaned off oxygen support in three days with 100% oxygen saturation. He was discharged home on a small dose of diuretics. On last follow-up five months later, he was asymptomatic, off diuretics, tolerating feeds and gaining weight with no distress. Repeat echocardiogram showed laminar

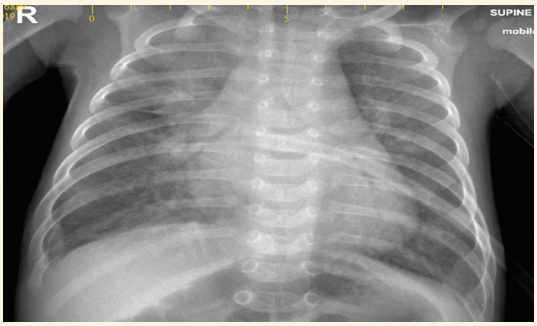

Figure 2: Chest radiography of a two-month-old male infant with cor triatriatum sinistrum showing features of pulmonary venous congestion and right ventricular hypertrophy.

flow across the left atrium with normalisation of pulmonary artery pressure.

Discussion

Cor triatriatum is a rare congenital cardiac anomaly characterised by an abnormal septum within the atrium impairing blood flow to the ventricles. It can be isolated or associated with other congenital heart anomalies and is commonly diagnosed in early childhood.[1] It was first described in 1868 by Church and accounts for 0.1% of all congenital cardiac anomalies.[2] When it involves the left atrium it is called CTS and it rarely involves the right atrium (called cor triatriatum dexter).[3] In a paediatric population, approximately 77% of cases are associated with congenital cardiac anomalies commonly atrial septal defects (ASD) and anomalous pulmonary venous return.[4] Other rare major associated congenital cardiac lesions include tetralogy of Fallot, double outlet right ventricle and coarctation of the aorta.

The pathogenesis of CTS is unknown but different theories have been suggested. The most popular theory is the entrapment theory introduced by Van Praagh and Corsini.[5] This theory suggested that cor triatriatum results from entrapment of the left atrial ostium of the common pulmonary vein by tissue of the right horn of the sinus venosus from which septum primum develops, leading to failure of incorporation of the common pulmonary vein into the left atrium during the fifth embryonic week. However, no single theory explains the different presentation and wide spectrum of CTS. Yet, multiple classifications of CTS based on the completeness of the membrane and presence of associated anomalies have been proposed.[3]

Recent animal studies demonstrated that mutations in the *hyaluronidase 2* (*HYAL2*) gene contribute to the development of CTS.[6] *HYAL2* is involved in different cellular processes and tissue morphogenesis and is required for endothelial-to-mesenchymal transition for proper development of the endocardial cushion in normal heart development.[6] Al Kindi *et al.* recently found that *HYAL2* was expressed in human CTS membranes that were resected during surgical repair.[7] Therefore, *HYAL2* deficiency may not explain the pathogenesis of CTS membrane in humans. Future studies are needed to evaluate the complex mechanisms involved and the embryological basis of this disease.

In isolated CTS, the presentation depends on the size of the communication between the upper and lower atrial chambers and the size of the interatrial communication.[4] As seen in the current case, patients

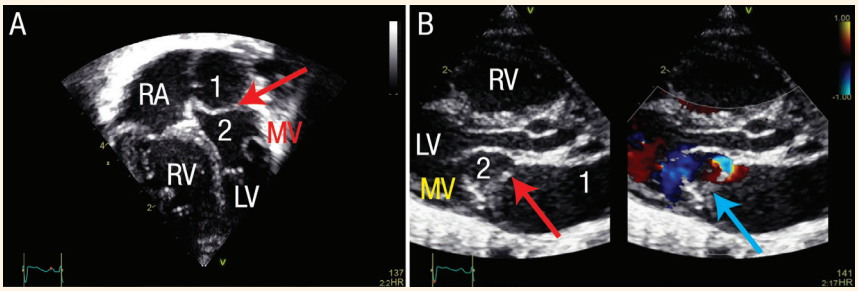

Figure 3: (**A**) Four chamber and (**B**) parasternal long axis echocardiographic views showing the divided left atrium into upper (1) and lower (2) chambers by the membrane (blue arrow). Note the dilated hypertrophied right ventricle and the very small restricted opening in the membrane (red arrow).
RA = right atrium; RV = right ventricle; LV = Left ventricle; MV = Mitral valve.

presenting in infancy with symptoms are mostly those with small openings in the membrane or with no ASD. CTS usually presents with symptoms mimicking mitral stenosis and supra mitral ring including commonly tachypnoea, recurrent respiratory infections and failure to thrive.[4] A rise in the atrial pressure and pulmonary congestion leads to dyspnoea.[4] The risk of death is over 75% if untreated.[2] Other common differential diagnoses for CTS clinical presentation include obstructed anomalous pulmonary venous drainage and congenital heart disease with left to right shunts such as ventricular septal defects and patent ductus arteriosus. Investigations for the diagnosis and follow-up of CTS include ECG, which can be normal or show evidence of right ventricular hypertrophy and right axis deviation, and CXR which may show cardiomegaly with pulmonary venous congestion or pulmonary oedema. Echocardiogram is a non-invasive and diagnostic procedure that can accurately show the membrane and any associated anomalies.

Furthermore, CXR is usually performed to assess the cardiac size and to exclude other possible secondary causes for the patient's symptoms. TEE is usually done pre-operatively to better visualise the membrane and ensure that it is completely excised post-operatively. It is also especially valuable to differentiate CTS from the supra-valvular mitral ring, the latter being located below the left atrial appendage. Surgical management is the optimal choice for symptomatic patients. The presence of obstructive symptoms at any age requires surgical intervention and the severity of presenting symptoms determines the urgency of surgery. The outcome is usually excellent but can be variable depending on the complete excision of the membrane and the associated anomalies.[8]

Conclusion

Stenotic simple CTS is a rare congenital heart disease with a favourable prognosis if diagnosed early and managed promptly. For clinicians, it is important to have a high index of suspicion to recognise its common clinical presentation which mimics the presentation of other more common diseases. As shown in the current case, prompt diagnosis and management can lead to an excellent outcome.

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