Case Report

Cor triatriatum dextrum: A rare congenital cardiac abnormality

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Abstract:

Cor triatriatum dextrum is an exceptionally rare congenital heart disease, in which the right atrium is partitioned into two chambers by a membrane to form a triatrial heart. It is caused by persistence of the right valve of sinus venosus. The aim of presenting this case is to develop awareness regarding cor triatriatum dextrum , though a rare case, can be present and may contribute to right heart failure and 2D-echocardiography is an important tool in making early and accurate diagnosis. We are reporting a case of an elderly Bangladeshi male presented with the features of mitral stenosis with pulmonary hypertension with CCF with respiratory tract infection, where cor triatriatum dextrum with an atrial septal defect was an incidental finding on routine echocardiographic assessment.

Keywords: Cor triatriatum dextrum

(Bangladesh Heart Journal 2016; 31(1) : 37-40)

Introduction:

Cor triatriatum was first described by Church in 1868.[1] It is a rare congenital anomaly that occurs when the left atrium (cor triatriatum sinistrum) or right atrium (cor triatriatum dextrum) is partitioned into two parts by a membrane, or a fibromuscular band.[2] On the right side of the heart, complete persistence of the right venous valve of the embryonic heart produces a septum in the right atrium separating the intercaval part of the righatrium from the atrial body. The remaining opening may vary depending on degree of partition or septation.[3] Typically, the right atrial partition is due to exaggerated fetal eustachian and thebesian valves, which together form an incomplete septum across the lower part of the atrium. This septum may range from a reticulum to a substantial sheet of tissue.[4,5]

Normally, during embryogenesis, the right horn of the sinus venosus gradually incorporates itself into the right atrium to form the smooth posterior portion of the right atrium or the sinus venarum, whereas the original embryologic right atrium forms the trabeculated anterior portion. The right horn of the sinus venosus and the embryologic right atrium are then connected through the sinoatrial orifice, which, on either side, has 2 folds called the right and left venous valves. During this incorporation, the left valve becomes part of the septum secundum, and the right valve of the right horn of the sinus venosus divides the right atrium into 2 chambers. This right valve forms as a sheet that serves to direct oxygenated blood from the inferior vena cava across the foramen ovale to the left side of the heart during fetal life. Normally, the network regresses by 12 weeks, leaving behind the crista terminalis superiorly and the eustachian valve of the inferior vena cava and the thebesian valve of the coronary sinus inferiorly.[5] Complete persistence of the right sinus valve results in a separation between the smooth and trabeculated portions of the right atrium, constituting cor triatriatum dextrum.[6-8]

Cor triatriatum dextrum has varying clinical manifestations depending on the degree of partitioning or septation of the right atrium. When the septation is mild, the condition is often asymptomatic and is an incidental finding frequently made at postmortem examination; more severe septation can cause right-sided heart failure and elevated central venous pressures

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due to obstruction at the level of the tricuspid valve, the right ventricular outflow tract, or the inferior vena cava.[9] Significant sequelae is unusual with cor triatriatum dextrum, and in most instances remain undiagnosed.[10]

Cor triatriatum accounts for approximately 0.1% of all congenital heart diseases,[11] most being cor triatriatum sinistrum. In most cases, the anomaly is recorded at necropsy, either as an isolated finding in an otherwise normal heart or as an accompaniment to other congenital heart lesions.[7,8]

Case Report

Our patient is a 75-year-old retired farmer who presented with acute severe breathlessness with increased intensity of cough for about a week.

Patient has cough for last six months and was productive with moderate mucoid sputum. There was no history of hemoptysis, fever, night sweat or significant weight loss. Further query revealed that he got easily fatigued and breathless, sometimes even at rest for last six months. There was associated occasional orthopnoea and paroxysmal nocturnal dyspnoea. He also gave history of smoking.

He required admission in respiratory medicine department of a tertiary care hospital about two months prior to this admission following similar episode of breathlessness. He was diagnosed clinically to have Chronic Obstructive Pulmonary Disease (COPD) with pulmonary hypertension and was treated accordingly and discharged when improved.

Physical examination revealed a chronically ill-looking elderly man, dyspnoeic at rest without cyanosis. There was no significant lymphadenopathy. We also found the patient having bilateral leg swelling up to mid-calves, raised jugular venous pressure and bibasal end-inspiratory fine crepitation and tender hepatomegaly along with left parasternal heave and loud pulmonary component of 2nd heart sound. Apex beat was normal in position and character. There was apical non-radiating mid-diastolic murmur best heard on left lateral position with breath held in expiration and there was also left lower parasternal pansystolic murmur better heard with breath held in inspiration without any radiation.

He was settled with optimal dose of diuretics and nebulized bronchodilators and anticholinergics.

An initial clinical diagnosis of mitral stenosis with pulmonary hypertension with CCF with respiratory tract infection (RTI) was made.

Electrocardiography showed sinus rhythm with right axis deviation and right atrial enlargement. X-ray chest PA view showed RV type apex, straightening of the left heart border with upper lobe diversion of pulmonary veins. Transthoracic echocardiography revealed moderate mitral stenosis and mild mitral, aortic and tricuspid insufficiency with estimated PASP 46 mm Hg. Echocardiography also showed an incomplete membranous septum diagonally dividing the right atrium into 2 parts a secundum atrial septal defect.(Figure-1, right & left panel)

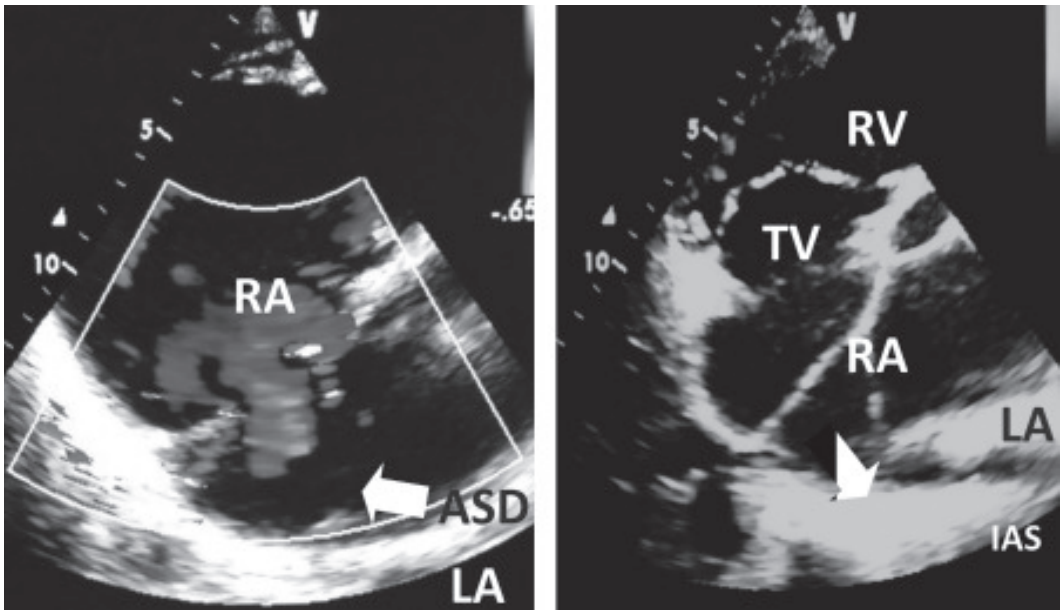

Fig.-1: *Left panel showing color flow across secundum Atral septal defect & right panel showing a diagonal membrane (patterned arrow) across the right atrium*

A final diagnosis was moderate mitral stenosis with mild mitral aortic and tricuspid regurgitation with Atrial septal defect (secundum) with mild pulmonary hypertension with Cor triatriatum dextrum with CCF with RTI. The combination of acquired Mitral stenosis & congenital ASD is known as 'Lutembacher's syndrome'.

The Patient was treated with anti-failure medications, bronchodilators and oxygen and has been counseled regarding his condition.

Discussion:

Cor triatriatum dextrum, an extremely rare form of cor triatriatum, accounting for 0.025% of all congenital heart disease.[12] It can occur as an isolated cardiac anomaly[13] or associated with other malformation of right heart structures such as pulmonary artery stenosis or atresia, pulmonary valve stenosis or atresia, hypoplastic right ventricle, tricuspid valve stenosis or atresia, atrial septal defect and Ebstein anomaly.[4,14,15] Unlike cor triatriatum sinistrum, which carries a higher mortality rate if not repaired, cor triatriatum dextrum has varying clinical manifestations depending on the degree of obstruction to venous flow ranging from asymptomatic to overt right-sided heart failure and elevated central venous pressures. In our patient, free flow of blood across the membrane of right atrium is probably the major element for the surprisingly good tolerance of the pathology for such a long periods of time.

Cor triatriatum dextrum may contribute to right heart failure. In our patient, heart failure was probably due to superimposition of severe respiratory tract infection upon already existing mitral stenosis, ASD (secundum) & pulmonary hypertension. There was no Cyanosis in our patient which is very rarely reported with cor triatriatum dextrum[16,17] and in those cases, echocardiographic assessment showed significant obstruction at the level of right ventricular inflow.[16] Common cardiac causes of pulmonary hypertension include diseases affecting the lung, mitral valve and left ventricle. In our patient, Cor triatriatum dextrum and ASD were incidental finding coexisting with mitral stenosis and RTI, which could not be optimally managed previously and gave rise to pulmonary hypertension and right heart failure.

There are no pathognomonic chest X-ray or electrocardiographic findings in isolated cor triatriatum dextrum. Right heart catheterization may reveal elevated pressure in the proximal right atrial chamber with a gradient across the accessory membrane. Echocardiography (transthoracic and transesophageal)[14,18,19] and cardiac magnetic resonance imaging (MRI)[20,21] are usually diagnostic as it demonstrates the presence of a membrane within the right atrium. Since many patients are asymptomatic, transthoracic echocardiography which is more readily available and affordable has a pivotal role as a mean of ante-mortem diagnosis. In symptomatic patients, when diagnosis is established, it is amenable to relatively simple surgical resection of the dividing membrane. Percutaneous catheter disruption of the membrane has also been suggested as a preferred alternative to open heart surgery.[22]

Conclusion:

Careful routine echocardiographic assessment can reveal asymptomatic rare congenital anomalies of the heart which otherwise remain undiagnosed and sometimes contribute to important clue to the management of the cardiac patients.

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