Cor Triatriatum Dexter with Multivalvular Rheumatic Heart Disease: A Very Rare Association

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ABSTRACT

Cor triatriatum dexter is a rare congenital heart anomaly where the right atrium is divided into two chambers by a membrane. We report a lady who present to us with orthopnea,engorged pulsatile neck veins, pulsatile enlarged liver and bilateral pedal edema. A diagnosis of multivalvular rheumatic heart disease (RHD) was made clinically. Echocardiography showed mitral stenosis (MS) with aortic regurgitation (AR) with tricuspid regurgitation (TR) with cortriatriatum dexter.

Key words: Cor triatriatum dexter, Congenital Heart Disease, Right sided heart failure, Echocardiography, Rheumatic heart disease.

Key Messages: Usually cortriatriatum dexter is associated with other congenital diseases like atrial septal defect (ASD),partial anomalous pulmonary venous connections (PAPVC) etc. Here it is associated with RHD masqueriding as tricuspid stenosis, hence very rare and worth reporting.

INTRODUCTION

Cor triatriatum dexter, or partitioning of the right atrium (RA) to form a triatrial heart, is an extremely rare congenital anomaly that is caused by the persistence of the right valve of the sinus venosus.[1] The incidence of cor triatriatum is approximately 0.1% of congenital heart malformation.[2] 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.[1] Cor triatriatum dexter (CTD) is frequently associated with right-side defects caused by abnormal fetal circulation. Among these, the most frequent are stenosis or atresia of the pulmonary valve, tricuspid valve abnormalities and atrial septal defect.[3] A prominent chiari network and eustachian or thebesian valve might simulate CTD and produce insignificant flow accelerations. However, in CTD the valve is attached to the atrial septum.[4] In the case of a prominent or giant eustachian valve, there are no attachments, irrespective of obstruction.[5] Association with RHD is extremely rare.

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CASE HISTORY

A 30 year old lady presented to us with the complaint of dyspnea on exertion of 5 years duration with orthopnea of 3 yrs duration and bilateral pedal edema of 3 yr duration. There was no history of paroxysmal nocturnal dyspnea (PND), angina, syncope, cyanosis and hemoptysis. However she complained of exertional palpitation for last 3 years. There was history of migratory arthritis with fever at the age of 10 yrs. However she never took any secondary prophylaxis for acute rheumatic fever (ARF). On cardiovascular examination, jugular venous pressure was increased with prominent v wave with slow y descent. Scalp veins were prominent on lying down posture. First heart sound was loud. Pulmonary component of second heart sound (P2) was loud with narrow splitting. A mid diastolic (MDM) rumbling murmur was heard at the apex. An early diastolic murmur was heard at 3rd left intercostal space parasternally. A holosystolic murmur was heard

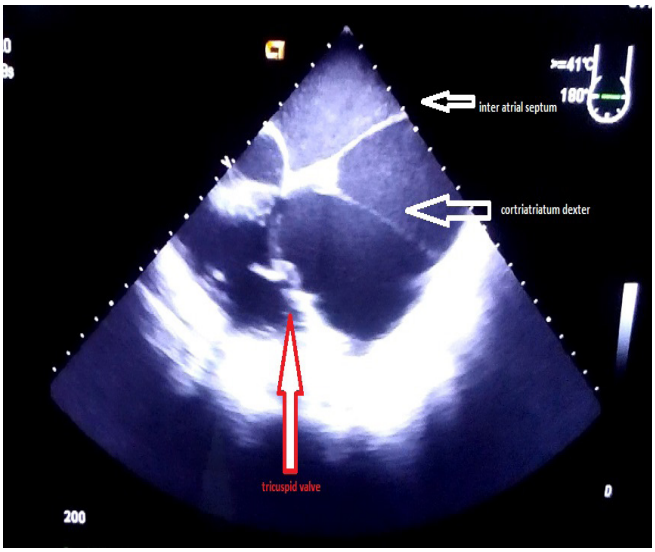

Figure 1: 2D transoesophageal echocardiography showing presence of a membrane separating the right atrium into two parts

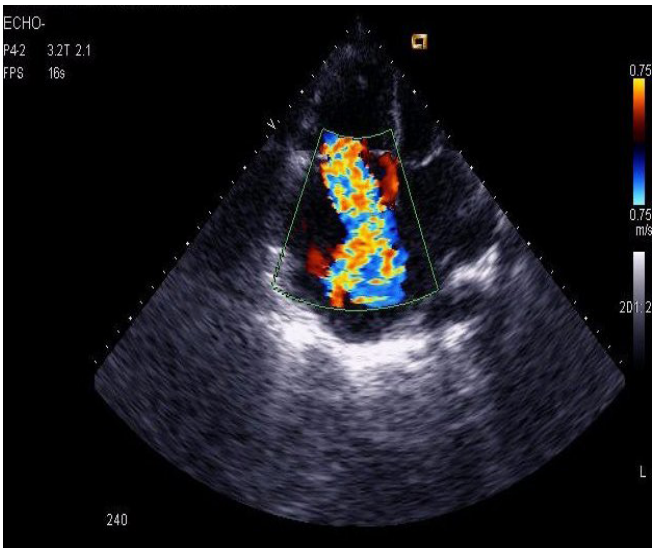

Figure 2: The blood flow to the right ventricle was not obstructed by the membrane

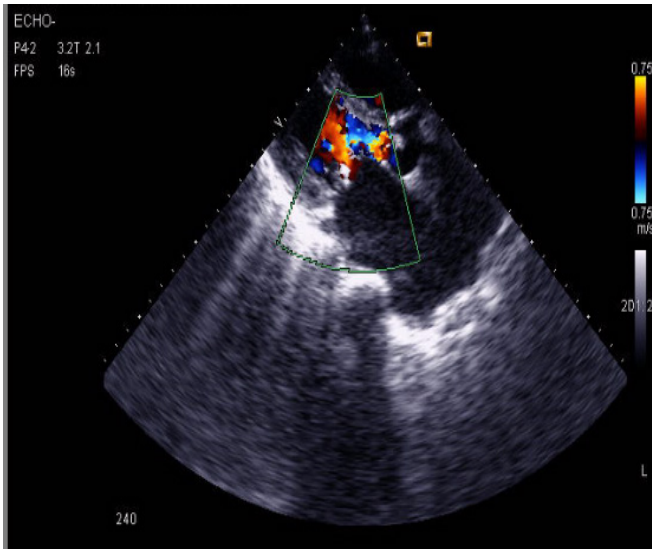

Figure 3: AR jet & MS jet with thickening of valve leaflets (rheumatic involvement)

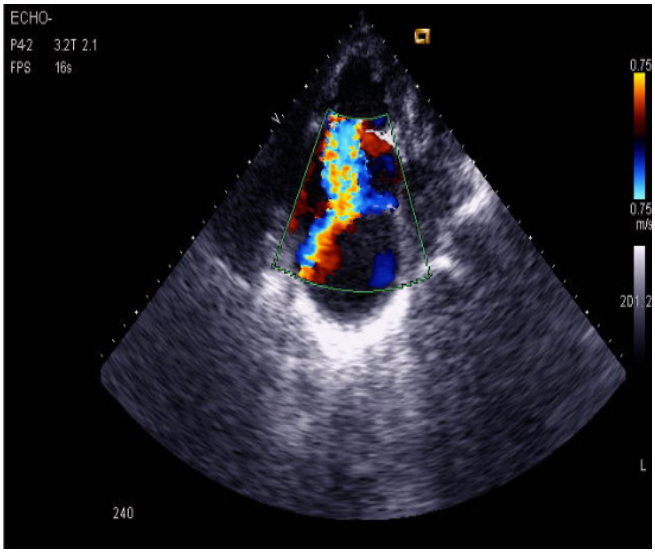

Figure 4: TR jet flow to RA membrane

at left lower parasternal (LLPS) area increasing with inspiration. Another MDM was heard at LLPS which also increases with inspiration. So a diagnosis of severe mitral stenosis (MS) with moderate aortic regurgitation (AR) with tricuspid regurgitation (TR) and tricuspid stenosis was made clinically. Transthoracic and trans-esophageal echocardiography (TTE and TEE) showed situs solitus of the atria and viscera, with atrioventricular and ventriculoarterial concordance. A large membrane within the right atrium divided the atrial cavity into two chambers, without causing obstruction to right ventricular inflow (Figure 1, 2). Also moderate AR and severe MS with organic TR was seen (Figure 3, 4). However, no TS was seen. So a diagnosis of cortriatriatum dexter with multivalvular rheumatic heart disease was made. She was referred to cardiothoracic department for dual valve replacement with resection of the membrane with tricuspid annuloplasty and had undergone the same. She is currently doing fine with low dose diuretics.

DISCUSSION

During embryogenesis, the right horn of the sinus venosus gradually incorporates into the right atrium to form the smooth posterior portion of the right atrium, whereas the original embryologic right atrium forms the trabeculated anterior portion. The connection between the right horn of the sinus venosus and the embryologic right atrium

is the sinoatrial orifice, which is flanked on either side by two valvular folds, the right and left venous valves. At some point during this incorporation, the right valve of the right horn of the sinus venosus divides the right atrium into two. This right valve forms a sheet that serves to direct the oxygenated venous return from the inferior vena cava across the foramen ovale to the left side of the heart during the fetal life.[1] Normally, the valve regresses by approximately 12 weeks gestation and leaves behind the crista terminalis superiorly and the eustachian valve of the inferior vena cava and the Thebesian valve of the coronary sinus inferiorly. Complete persistence of the right sinus valve results in a separation between the smooth and trabeculated portions of the right atrium, constituting cor triatriatum dexter.[6] Cor triatriatum dexter 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 during surgery to correct other cardiac abnormalities or during echocardiography. More severe septation can cause right sided heart failure and elevated central venous pressures due to obstruction of the tricuspid valve, the right ventricular outflow tract, or the inferior vena cava.[1] Cor triatriatum dexter can occur as an isolated cardiac anomaly or associated with other malformation of right heart structures such as pulmonary artery stenosis or atresia, tricuspid valve abnormality, ASD, and Ebstein anomaly.[6,7] CTD can be diagnosed by echocardiography[3,8] and magnetic resonance imaging, although caution is needed since a prominent eustachian valve can simulate the defect. Either of these imaging techniques can show the right atrium divided into 2 chambers: a posterior chamber into which the vena cava drain, and an anterior one that contains the atrial appendage. Asymptomatic patients should not be treated, unless referred to heart surgery for other reasons. In symptomatic patients with significant obstruction, the treatment of choice is surgical resection, although percutaneous correction of the membrane has been proposed as an alternative to surgical treatment.[9] We have searched for association between cortriatriatum dexter and rheumatic heart disease but found none.

ABBREVIATION

RHD : Rheermatic heart disease
MS : Mitral Stenosis
AR : Aortic Regurgitation
TR : Tricuspid Regurgitation
CTD : Cortriatriatum Dexter

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