

Coexistence of Cor triatriatum sinistrum and a prominent Eustachian valve mimicking a Cor triatriatum dextrum

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ABSTRACT

Cor triatriatum is among the rarest of all congenital cardiac abnormalities accounting for 0.1-0.4% of all congenital heart disease. Its coexistence with a very prominent Eustachian valve which mimics a Cor triatriatum dextrum is an exceptionally rare finding in an asymptomatic adult. We report the case of a 44 year old male who presented to our department on observing a pulse rate of 44 beats per minute during a home blood pressure check with his digital sphygmomanometer. Clinical examination was however, unremarkable and resting electrocardiography showed sinus rhythm with atrial premature complexes. The diagnosis was made on a two dimensional transthoracic echocardiography. Isolated atrial premature complexes and bradycardia may be a clinical presentation of Cor triatriatum in adult population. Although extremely rare, its coexistence with a prominent Eustachian valve may remain asymptomatic into adult life.

Key words: Cor triatriatum sinistrum, prominent Eustachian valve, asymptomatic adult

INTRODUCTION

Co existence of Cor triatriatum sinistrum and a prominent Eustachian valve which mimics a Cor triatriatum dextrum is an exceptionally rare finding. Cor triatriatum was first described by Church in 1868.^[1] It is a congenital anomaly in which the left atrium (cortriatriatum sinistrum) or right atrium (cortriatriatum dextrum) is divided into 2 parts by a fold of tissue, a membrane, or a fibromuscular band.^[2-4] It is a rare cardiac malformation comprising about 0.1% of congenital heart disease.^[5] The Eustachian Valve (EV) is an embryologic remnant of the valve of the inferior vena cava (sinus venosus)^[6] and can be visualized only in a minority of persons undergoing transthoracic echocardiography (TTE).^[7] We report a unique case of the above finding.

CASE REPORT

A 44 year old male presented on account of bradycardia (44 beats per minute) which he observed while routinely checking his blood pressure at home with a digital sphygmomanometer. He had been otherwise asymptomatic for cardiac diseases from childhood. On physical examination, he had a regular pulse of 64 beats per minute with a few missed beats. Cardiac auscultation was normal. The resting 12 Lead electrocardiogram showed normal sinus rhythm with atrial ectopics (Heart rate of 70 beats per minute). Biochemical and haematological profile were unremarkable. TTE showed an abnormal membrane bisecting the left atrium (LA) into two chambers [Figure 1]. In the parasternal long axis view, this membrane ran parallel to, and a short distance behind the aortic root and then curved anteroinferiorly to insert some distance away from the mitral valve ring. This membrane was seen bisecting the LA at the atrial appendage in the apical four chamber view. It was difficult to image the whole of the membrane on a freeze frame analysis. The M mode Echocardiographic appearance of the membrane revealed a linear echo lying anteriorly within the LA, behind the aortic root. This echo

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Figure 1: Apical two chamber view in transthoracic echocardiography demonstrating the membrane (arrow) bisecting the left atrium into superior and inferior compartments (Cor triatriatum sinistrum)

shows a movement resembling a stenotic mitral valve. Doppler evaluation did not show any intra-atrial gradients. Both atria were not dilated and the pulmonary veins did not have abnormal flow velocities.

A ridge of tissue was seen inserting into the lower portion of the interatrial septum adjacent to the atrioventricular valves, and partially divided the right atrium (RA) into two chambers -the superior and inferior compartments [Figure 2]. The origin of this tissue was in the region of the ostium of the inferior vena cava. This is most likely a very prominent Eustachian valve (EV) resulting in an appearance of a Cor triatriatum dextrum. The valve was immobile during the different phases of the cardiac cycle and no obstruction to flow was demonstrable at the level of the EV. The estimated pulmonary artery systolic pressure was 25 mmHg and the main pulmonary artery was not dilated.

The remaining valves were normal and there were no other associated cardiac defects.

The patient was placed on antiplatelet prophylaxis (Aspirin 75 mg daily).

DISCUSSION

Cor triatriatum is a rare cardiac malformation comprising about 0.1% of congenital heart disease.^[5] The EV is an embryologic remnant of the valve of the inferior vena cava (sinus venosus).^[6] These remnants have different degrees of persistence after birth.^[8] Moderate degrees of persistence can give rise to a very prominent EV with functional separation of inferior and superior vena cava systems with a resultant rare echocardiographic appearance of a divided right atrium

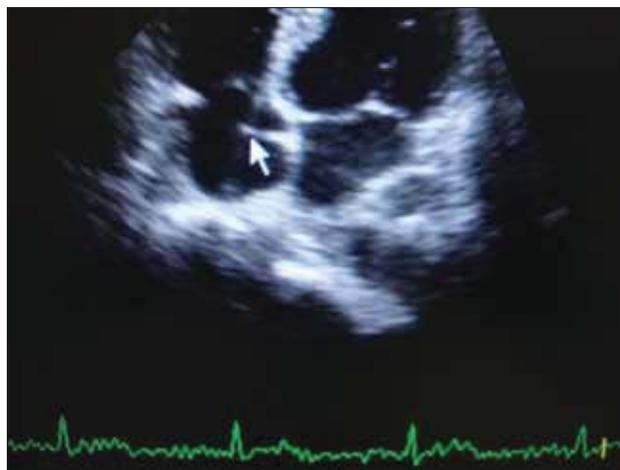


Figure 2: Apical four chamber view in transthoracic echocardiography demonstrating the prominent Eustachian valve (arrow) crossing the right atrial floor to insert into the interatrial septum (mimicking Cor triatriatum dextrum)

as we have demonstrated. This abnormality should be differentiated from Cor triatriatum dextrum, which results when early embryonic separation of the right atrium into two chambers persists. This type of abnormality may be confused with a Cortriatriatumdexter.^[9] In adults, the EV can be visualized in only a minority of persons undergoing TTE.^[7]

The presence of a thin, fenestrated, fibrous membrane, at the insertion of the inferior vena cava (IVC) into the right atrium is pathognomic for EV.^[10] Occasionally, the EV crosses the floor of the RA from the orifice of the IVC and inserts into the lower portion of the interatrial septum adjacent to the atrioventricular valves^[9] as we have demonstrated. Persistence of sinus valve structures may be associated with obstruction of IVC flow, can predispose to a patent foramen ovale,^[11] be completely asymptomatic, and/or rarely provide a nidus for endocarditis.^[12] Our patient was asymptomatic and this is the more common finding in patients with prominent Eustachian valves.^[13] In this case, the finding of a very prominent EV had no apparent clinical consequences for the patient.

Previous publications on the M mode Echocardiographic appearances of cortriatriatum confirm a linear echo within the left atrium behind the aortic root,^[14-16] and in some cases this echo shows a movement resembling a stenotic mitral valve^[16] as in our case. It is noteworthy that the abnormal echo lies anteriorly within the LA, as in our case. This is helpful in the differential diagnosis as both the linear echo sometimes seen “within” the LA in total anomalous pulmonary venous drainage and the artefactual echoes sometimes seen in the LA tend to be more posterior. Detection of this membrane in

multiple planes of examination distinguishes it from an artifact. Its position within the LA is typical and may be differentiated from that of a supralvalvular mitral ring, which occurs exclusively within the mitral valve funnel with the membrane distal to the left atrial appendage in contradistinction to Cor triatriatum. Furthermore, we demonstrated a superior recess behind the aortic root, which is not seen in supralvalvular mitral ring.

The most frequent cardiovascular abnormalities associated with Cor triatriatum sinistrum in adults are usually related to the spectrum of left heart obstruction,^[17] mitral regurgitation, supralvalvular annulus,^[17] and persistent left sided superior vena cava draining in the coronary sinus.^[17] Other abnormalities are ostiumsecundum atrial septal defect and less commonly, aortic regurgitation with dissecting aneurysm.^[17,18] Cor triatriatum has been known to occur in silent association with complex congenital heart disease.^[19] This is another case of a silent occurrence of this abnormality.

Our patient is being managed conservatively since both lesions were found to be hemodynamically insignificant. Asymptomatic patients with prominent EV usually require no further treatment.^[13] Endocarditis and thrombus formation over the EV are extremely rare complications.^[20,21] To the best of our knowledge, coexisting Cor triatriatum sinistrum and a prominent EV presenting as Cor triatriatum dextrum has not been described in an asymptomatic African adult.

CONCLUSION

Although extremely rare, Cor triatriatum sinister coexisting with a prominent Eustachian valve may remain asymptomatic into adult life. It is an important entity to recognize because it may be easily surgically corrected when hemodynamically significant. Access to more advanced imaging (trans-esophageal echocardiography) is clearly desirable; however, should not be a limiting factor to diagnosis in a resource poor setting like ours.

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