CASE REPORT

A Case Report of Congenitally Corrected Transposition of Great Arteries

Anisul Awal[1], Saurav Das[1], Ummey Maimuna[1], Reaz Mahmud Huda[2]

ABSTRACT

Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital malformation which associates discordant atrioventricular and ventriculo-arterial connections. We describe here a 35-year-old male patient admitted into the Department of Cardiology of Chattogram Medical College Hospital, Chattogram, Bangladesh, with congenitally corrected transposition of the great arteries. The man had progressive breathlessness over 2 months with previous episodes of similar kind of illness over the last 2.5 years. He disclosed that he was informed regarding his heart disease 2.5 years back; however, he was unaware of its nature and consequences. Examination revealed a pansystolic murmur (loudest at the left lower parasternal area), apex beast was diffuse, thrusting, just lateral to the midclavicular line in the left fifth intercostal space. Transthoracic echocardiography (TTE) confirmed the anatomy of congenitally corrected transposition of the great arteries (CCTGA) with torrential tricuspid regurgitation and impaired systemic right ventricle.

Keywords: Cardiac anomaly, congenitally corrected transposition of the great arteries, discordant atrioventricular connections, discordant ventriculo-arterial connections

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INTRODUCTION

Congenitally corrected transposition of the great arteries (ccTGA) is a rare complex congenital heart disease, with an estimated prevalence of 1 per 33,000 live births, thus accounting for approximately only 0.05% of all.[1,2] The disease characterized by atrioventricular and ventriculo-arterial discordance.[1-4] The condition can be associated with interventricular communications, obstructions of the outlet from the morphologically left ventricle, and anomalies of the tricuspid valve.[1,2] Generally by the fourth decade, systemic right ventricle (RV) dysfunction is clinically apparent.[1,3]

We present here the case of a patient with congenitally corrected transposition of the great arteries for academic interest.

CASE SUMMARY

A 35- year-old male, normotensive, nondiabetic patient came with complain of progressive dyspnea on moderate to severe exertion for 2 months. He had to take admission into the hospital for two times in the last one month. He was diagnosed to have severe mitral valve regurgitation, due to mitral valve prolapse, and was advised for mitral valve replacement/repair. He decided to take second opinion/ consultation from the specialized heart center. Then he was diagnosed as a case of cctTGA with moderate left atrioventricular valve regurgitation with moderate pulmonary hypertension. He was advised regular medications and routine follow-up. His last transthoracic echocardiography (TTE) was done 7 months ago, and he had been symptom-free up to his recent clinic visit. Auscultation revealed

1. Department of Cardiology, Chattogram Medical College Hospital, Chattogram, Bangladesh.

2. Department of Cardiology, Sarkari Karmachari Hospital, Fulbaria, Dhaka, Bangladesh.

Correspondence to: Dr. Ummey Maimuna, Department of Cardiology, Chattogram Medical College Hospital, Chattogram, Bangladesh. Email: maimunabintemohsin@gmail.com

3/6 holosystolic murmur. His chest radiograph revealed the appearance of the "egg on a string" sign of transposition of the great arteries, with enlargement of the cardiac silhouette and atrophy of the thymus leading to narrowing of the upper mediastinum (Figure 1). The ECG showed sinus rhythm, rate around 80/min, absence of Q waves in V4-6 (physiologic Q waves in this lead often referred to as "Septal Q waves"), S in V1+R in V6 >35 mm (Figure 2). Echocardiography showed the aorta arising from the morphological right ventricle (RV), which was identified by the three-leaflet tricuspid valve that inserted more apically than the mitral valve. The pulmonary trunk, identified by its bifurcation, arose from the morphological left ventricle (LV). The systemic RV was dilated, hypertrophic, and the systolic RV function was normal (ejection fraction 55%) (Figure 3).

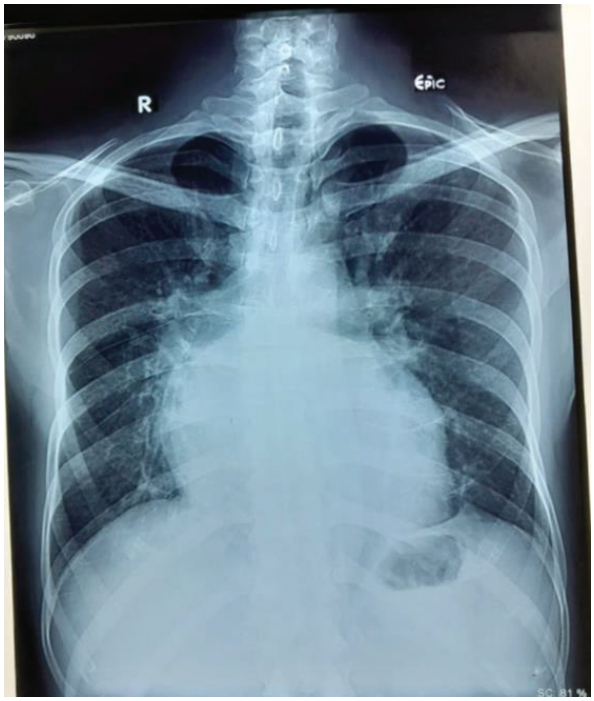

Figure 1: Chest x-ray showing "egg on a string" appearance, with enlargement of heart and narrowing of the upper mediastinum.

DISCUSSION

Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital malformation which associates discordant atrioventricular and ventriculo-arterial connections.[5,6] Although frequently associated with a ventricular septal defect (VSD), its anatomy remains controversial.[5] This could be due in hearts with usual atrial arrangement to the apparently different anatomy of the left-sided right ventricle compared with a right-sided right ventricle.[5]

CCTGA comprises both atrioventricular and ventriculoarterial discordance; the right atrium enters the morphological LV, which gives rise to the pulmonary artery, whereas the left atrium enters the morphological RV, which gives rise to the aorta.[7] This defect occurs due to abnormal cardiac development during the third gestational week where left looping (L-loop) of the heart tube instead right looping occurs.[4,5] Patients without associated intracardiac lesions are usually asymptomatic early in life and may do well into adulthood; nevertheless, they have a SRV, many manifesting ventricular dysfunction and symptoms often during the 4th or 5th decade of life. The majority of patients with ccTGA (80%), however, have associated lesions (the most common being ventricular septal defect, pulmonary stenosis, Ebstein anomaly of tricuspid valve, and abnormalities of the conduction system) and thus develop symptoms early in childhood.[5,7] The RV, normally supporting the low-pressure pulmonary circulation, when in the systemic position has to go through various adaptive mechanisms to enable itself to sustain the systemic load.[5,7]

Unlike complete TGA (where tricuspid regurgitation is secondary to annular dilatation and RV dysfunction, and thus tricuspid valve replacement is not warranted), the main hemodynamic, prognostic intervention in ccTGA is addressing intrinsic abnormalities with regurgitation of the tricuspid valve, which lead to SRV dysfunction and ultimately failure. This calls for timely tricuspid valve replacement (repair is usually not gratifying) in ccTGA to preserve SRV function.[7]

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Ethical Clearance: Confidentiality and anonymity of the patient were maintained. Informed consent was obtained from the patient for publishing case summary and relevant photos.

Authors' Contribution: Conception and design: AA, UM; Acquisition, analysis, and interpretation of data: AA, SD, UM; Manuscript drafting, revision, editing, and final submission: AA, SD, UM, RMH.

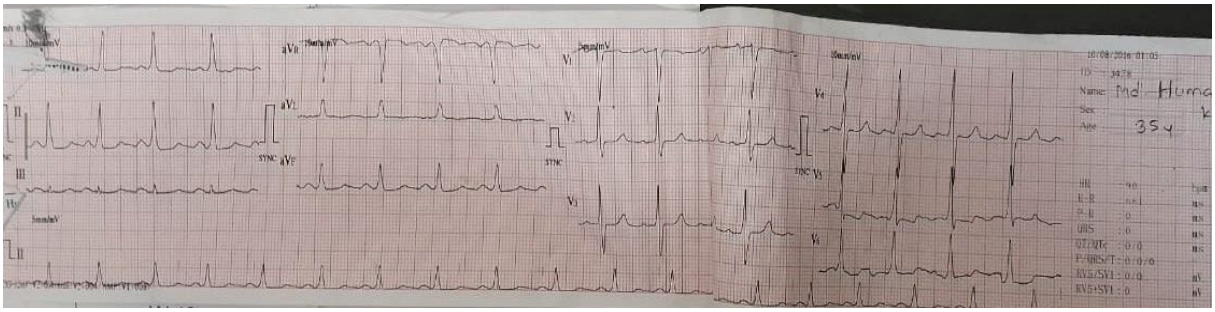

Figure 2: ECG of the patient.

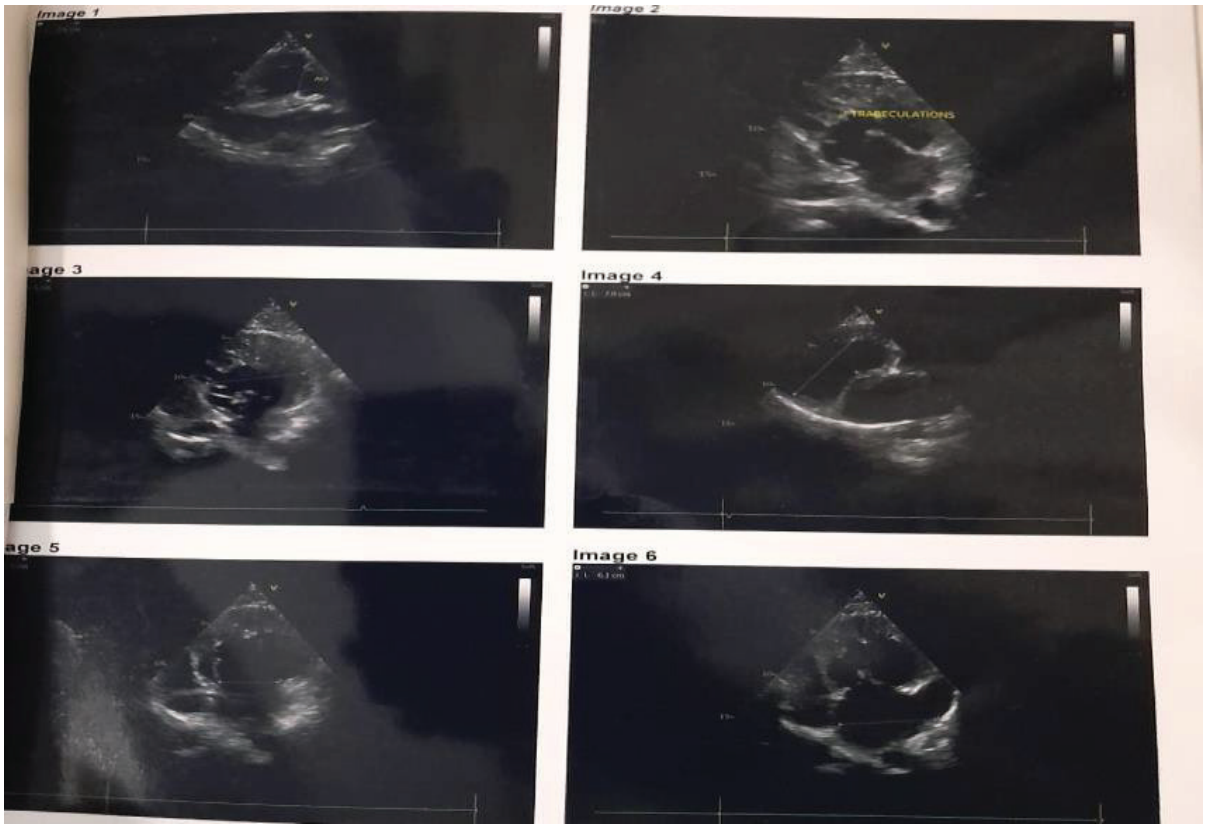

Figure 3: Echocardiography of the patient revealed congenitally corrected transposition of great arteries.

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