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Case Report

Rare Congenital Heart Disease Presenting with LV Systolic Dysfunction in Adult Patients- A Case of Congenitally Corrected Transposition of the Great Arteries

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Abstract

Background: Congenitally corrected transposition of the great arteries (ccTGA) is a rare anomaly comprising a minimal portion of congenital heart disease cases. Some patients are not identified until adulthood. A minority of these patients maintain normal functional status into the seventh decade and generally only when no other anomalies exist.

CASE- We here describe a case of a 50-year-old female (middle aged adult) who presented with new onset heart failure and was diagnosed as ccTGA.

Conclusion: The factors that lead to delayed presentation of heart failure ccTGA remain ill defined. Commonly adults acquired secondary causes of heart failure, such as hypertension and coronary artery disease. Aggressive screening and management of these co-existing diseases may improve the overall outcome in adult patients of congenital heart disease.

Keywords: Congenitally corrected transposition, heart failure, middle aged adult

Introduction

Congenitally corrected transposition of the great arteries (CCTGA) is a rare structural heart disease constituting less than 1% of all congenital heart disease [1]. It is a unique congenital heart lesion in the sense that despite anatomical aberrations in the form of atrio-ventricular and ventriculo-arterial discordance, there can be physiologically normal circulation without any mixing or shunting of blood.

Case Report

A 50-year-old female residing in Yerwada, Pune a known case of hypertension presented to our hospital with complaints of dyspnoea since 1month, which progressed from NYHA-II to NYHA-III over last 7 days, associated with orthopnoea and paroxysmal nocturnal dyspnoea (PND). She also had complaint of bilateral lower limb swelling since 10-12 days.

On presentation her vitals showed PR-85/min, and BP-130/90mmhg, RR- 28/min, oxygen saturation of 94% on 2Litres of oxygen via nasal cannula.

On physical examination showed, Regular rhythm with a pronounced aortic component of second heart sound. Pansystolic murmur heard in mitral area. Fine basal crepts and minimal lower extremity edema were also present.

Blood workup revealed a pro-BNP of 1450pg/ml (reference range 0-1800pg/ml) with negative troponin.

Chest xray revealed bilateral mild pleural effusion.

Her ECG demonstrated complete heart block with left axis deviation.

Transthoracic echocardiography (2D ECHO) revealed

Atrioventricular discordance (morphological RV on the left side with normal atrial positions), Ventriculoarterial discordance (aorta

arising from RV and pulmonary artery arising from LV), Aorta was left and anterior with pulmonary artery right and posteriorly, hypertrophied RV (left sided) with global hypokinesia with moderately depressed systolic function, severe left AV valve regurgitation, moderate pulmonary artery hypertension (RVSP-50mmhg),

CAG (coronary angiography) was done to assess coronaries and was normal. USG (ultrasound) abdomen was done to assess the position of organs and was normal. The patient was started on heart failure therapy with Furosemide 20mg intravenous thrice daily, Ramipril 2.5mg daily, Eplerenone 25mg daily, Carvedilol 3.125mg twice daily. After therapy patient improved and was discharged home

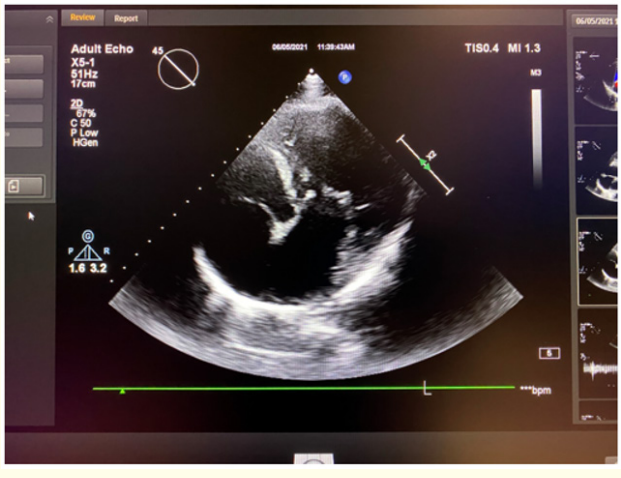

Figure 1: Morphological RV on left side in AP4C 2d echo image.

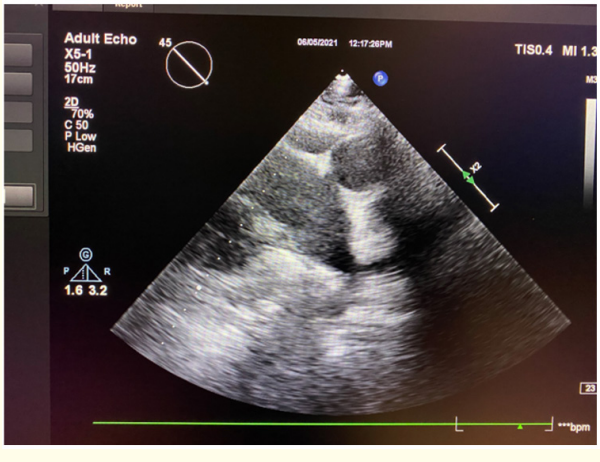

Figure 2: 2D ECHO Short axis view showing aorta left and anteriorly, PA right and posteriorly.

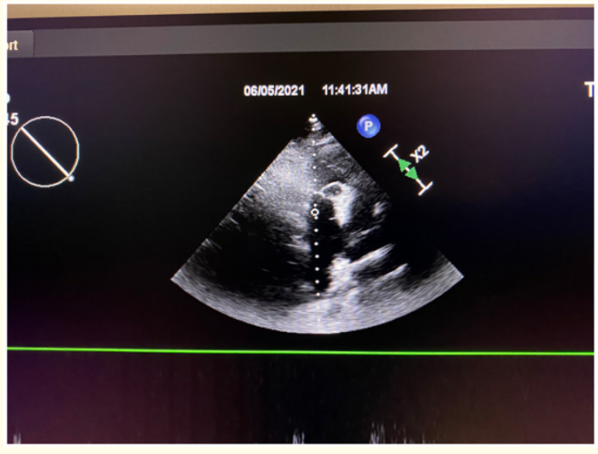

Figure 3: 2D ECHO - Short axis view aorta left and anteriorly, PA right and posteriorly.

Discussion

Majority of cases of ccTGA are diagnosed antenatally by using foetal ultrasound. If there are no other associated cardiac defects, ccTGA patients are likely to remain asymptomatic upto the fifth decade [4]. Beyond which the incidence of congestive heart failure and RV dysfunction increases which is associated with higher mortality in these patients [6]. The exact mechanisms resulting in eventual RV dysfunction is not known, many physiologic and anatomic factors are believed to be pathogenetically responsible. These mechanisms includes, reduced coronary blood flow, persistent progressive tricuspid regurgitation, long term increased RV afterload, rhythm disturbances and eventually leading to right ventricular hypertrophy (RVH) [2,3,5,7-9]. The ability of the RV to adapt to increased afterload plays a role in delay of symptoms. Although determinants of the adaptive response, and the time required for RV to increased afterload, including fetal gene switching among others, are not well defined [10]. The capacity of the RV in our patient to favourably respond to increasing systemic pressures since birth may have possibly been related to an adaptive RVH response as a phenotypic expression of a protective genetic profile. In addition to eventual RV dysfunction, most of the ccTGA patients suffer from some degree of conduction system abnormalities resulting from the unusual location of the AV node and conduction pathway [2,3]. The abnormal site of the conduction system increases its risk for fibrosis resulting in increased incidence of complete atrioventricular (AV) block of roughly 2% per year [11]. The incidence of which vary between patients with ccTGA situs solitus as compared to those with situs inversus. In one cohort study, there was a de-

creased incidence of complete AV block in the situs inversus group compared with situs solitus patients [5]. It was postulated that the associated malalignment of the atrial and ventricular septum seen with situs solitus leads to increased susceptibility to AV block [5]. The decreased incidence of heart block in ccTGA with situs inversus leads to improved long term survival. Life expectancy in patients with ccTGA is less as compared to normal individuals [2,3]. Only few cases described acute coronary syndrome (ACS) in ccTGA patients who were diagnosed with varying degrees of CAD [12-14]. The degree to which coronary artery disease may contribute to systemic RV dysfunction and long-term outcomes is still under study in the adult population with ccTGA. The incidence of systemic RV dysfunction increases with age leading to signs and symptoms of congestive heart failure [6]. A few small trials of patients with a systemic RV have demonstrated a decrease in heart failure biomarkers with the use of drugs acting on the renin-angiotensin-aldosterone system without a clear reduction of morbidity and mortality [15-17]. Currently, in symptomatic patients with systemic RV dysfunction optimal medical therapies for congestive heart failure are used [15].

L-TGA patients typically present with nonspecific HF symptoms at an early age because the morphologic RV functions as the systemic pump. Although there is increasing favour for performing a double switch operation (correcting AV and VA discordance, allowing the left ventricle to function as the systemic pump) in infants and young children with L-TGA, older teens and adults have not responded well with this procedure [18-20]. Medical management is, therefore, the more widely accepted treatment strategy. For patients who have significant systemic AV or VA valve insufficiency, prompt surgical intervention is required [21].

The goal of treatment is to reduce afterload and delay the remodeling process in order to improve RV function, as the RV is not built to sustain the role of the left systemic ventricle in the long term. The principle behind this strategy is to decrease the stress on the wall of the RV during ejection. Drugs that will reduce afterload and may slow the remodeling process include angiotensin-converting enzyme inhibitors and angiotensin receptor blockers (ACE-I/ARBs) [22-24]. In addition, β-blocker use in patients with systemic right ventricles demonstrated an increase in EF and reduced cardiac remodelling [25,26]. However, it is important to note that L-TGA patients have an associated risk of developing complete heart block, so these HF agents should be used with caution [9]. While diuretics are used for symptomatic relief, digoxin and aldosterone antagonists have not shown any clinical benefits [27-29]. After a detailed discussion, our patient was initiated on an ACE-I and β-blocker with plans for up-titration as tolerated. It must be noted that while the data above demonstrate some improvement in function of the systemic RV with regards to the use of β-blockers and ACE-I/ARBs, there is no proven mortality benefit noted.

Conclusion

This case of ccTGA mainly shows that congenital heart disease can present in middle-late adult age.

A variety of factors may have contributed to delayed onset of heart failure until the fifth decade. However, determinants of successful adaptation of the RV to systemic afterload including the interaction with acquired heart diseases in this elderly population of ccTGA remain elusive. Understanding these factors may shed light on the RV potential to favourably adjust to increasing afterload in a variety of congenital as well as acquired conditions associated with pulmonary hypertension. Finally, by earlier screening and diagnosis of ccTGA, the adult cardiologist can potentially help preserve the systemic RV function through careful monitoring of tricuspid valve function with early intervention as needed, and by meticulous surveillance and aggressive treatment of acquired adult cardiac diseases to improve the survival and quality of life of this vulnerable patient population.

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