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Cardiology

CONGENITALLY CORRECTED TRANSPOSITION OF GREAT ARTERIES A RARE CASE REPORT- GOVERNMENT GENERAL HOSPITAL, SIDDHARTHA MEDICAL COLLEGE, VIJAYAWADA.

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ABSTRACT Congenitally corrected transposition of great arteries (cc-TGA) is a rare condition that accounts for less than 1% of all congenital heart diseases (CHD). Patients with cc-TGA are usually diagnosed at the early stages of life due to associated anomalies, but they may even remain asymptomatic until later decades of their life. We report a case of a 45-year-old man who presented to the emergency department with unknown poison ingestion, in whom isolated cc-TGA was discovered incidentally. The patient was completely asymptomatic of cardiac features at presentation other than an abnormal ECG, which arouse the suspicion of cardiac abnormality and led us to do a thorough cardiac evaluation. Interestingly, the patient had congenitally corrected transposition of the great arteries (cc-TGA) with no associated anomalies but with conduction abnormalities, which is a rare CHD with late adulthood presentation.

KEYWORDS : Congenitally Corrected Transposition Of The Great Arteries (cc-tga), Cardiomegaly, Levocardia

INTRODUCTION

cc-TGA is a rare cardiac malformation characterized by the combination of discordant atrioventricular and ventriculoarterial connections.[1] The incidence of cc-TGA has been reported to be around 1/33 000 live births, accounting for ≈0.05% of congenital heart malformations.[2] Although a familial recurrence of heart defects in subjects with cc-TGA has been reported,[3] the exact etiology of this malformation is not currently known.

Nearly 10% of the patients with cc-TGA do not have any associated anomalies like ventricular septal defect(VSD), pulmonary artery stenosis, tricuspid valve abnormalities, and mitral valve abnormalities.[4]

The patients without any associated anomalies, that is, isolated cc-TGA, remain asymptomatic for many years. They are usually diagnosed in later decades of life due to an abnormal electroc ardiograph (ECG), cardiomegaly on chest X-ray, or presence of murmur.[5] However, they often present with life-threatening complications like systemic ventricular dysfunction, tricuspid regurgitation, heart block, and ventricular arrhythmia.[6] These lead to increased rates of morbidity and mortality in such patients. The patients without any associated anomalies have a normal life expectancy, but their lifespan markedly depends on the systemic ventricular function.[7]

Here, we present a rare case of isolated cc-TGA with no associated anomalies, which had been asymptomatic even in adulthood and an incidental diagnosis.

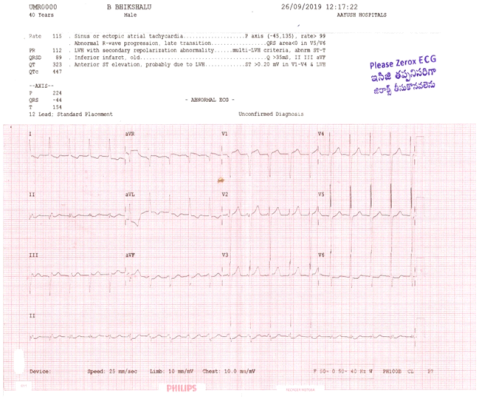
CASE REPORT

A 45-year-old male patient was admitted to Government general hospital, Vijayawada, Andhra Pradesh, India, with alleged consu mption of unknown plant poison. At presentation, the patient was entirely asymptomatic except for mild epigastric burning sensation. The patient had no positive symptoms of chest pain, breathlessness, palpitations, cough, or syncope. The patient was born out of non-consanguineous marriage at full term with normal developmental milestones and with no neonatal or childhood hospital admissions. The patient had no significant history of major illnesses or surgeries and was not on any medications.

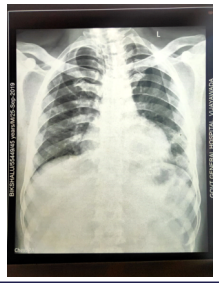
Patient vitals were body temperature (98.6°F), heart rate (112/min), blood pressure (110/80mmHg), oxygen saturation level (99% at room air), and respiratory rate (16/min). The systemic examination found to be normal.

The patient's clinical condition was stable, and the patient got relieved of epigastric burning sensation with intravenous Proton Pump Inhibitors, and recovery was uneventful.

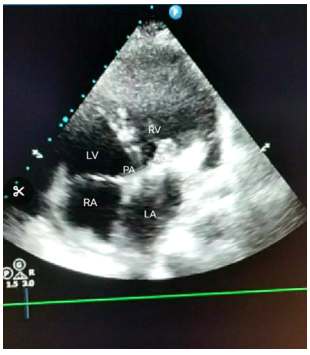
Routine blood and urine investigations were normal. ECG showed sinus rhythm with AV dissociation and junctional tachycardia with ventricular rate @115/min. There were absent Q waves in V6 and prominent QS complexes in V1. R wave progression was abnormal, and there was evidence of left ventricular hypertrophy.



Chest radiograph showed levocardia with an absence of normal pulmonary artery trunk in favor of smooth left supra-cardiac border; the right pulmonary hilum is slightly prominent with patches of haziness in bilateral lung fields more on the left side.



On further evaluation, Echocardiography showed congenital heart disease with L-transposition of great arteries with ventricular inversion. Atrioventricular(AV) valves showed reversed offsetting, a strong clue to the diagnosis. Left ventricular(LV) and right ventricular(RV) functions were fair with ejection fraction (EF) of 58%, mild Mitral Regurgitation, and Tricuspid Regurgitation was noted. No vegetations or clots.



Computerized Tomography(CT) chest revealed mild cardiomegaly with transposition of great arteries and ground glass haziness with septal thickening in bilateral lung fields.

Cardiac catheterization was not done as there were no absolute indications but may be done to demonstrate coronary artery anatomy before any cardiac intervention.

With the above background, the patient was diagnosed to have isolated congenitally corrected transposition of the great arteries (cc-TGA) with no associated anomalies. This patient was kept on tablet Ramipril 2.5 mg OD (Angiotensin Converting Enzyme-Inhibitors (ACE-I) group) and discharged on day 8 with unrestricted exercise recommendations.[8] Patient was advised of regular follow up.

DISCUSSION

cc-TGA is characterized by atrioventricular and ventriculoarterial discordance. From a circulatory oxygenation standpoint, these patients are "congenitally corrected," especially "two wrongs make a right," and the pulmonary and systemic circulations run in series, not in parallel, as with dextro-TGA. There is a ventricular inversion, and the respective AV valves follow the ventricles. Therefore, the systemic RV is transposed to the left, and the tricuspid valve goes with it. The left atrium empties into the RV, which then pumps to the leftward and usually anterior aorta. The LV and mitral valve are dextraposed, and the pulmonary artery emerges posteriorly from the LV.[9]

Fewer than 10% of patients are free of associated abnormalities, which include VSD (membranous or muscular) in up to 80%, pulmonic stenosis (valvular or subvalvular) in up to 70%, and tricuspid valve abnormalities (usually Ebstein anomaly) in 33%.[10]

NATURAL HISTORY

In the minority of patients without associated defects, un-operated survival into adulthood is common, and survival into the eighth and ninth decade of life has been reported. Most patients remain undiagnosed until early adulthood. However, these patients have increased rates of AV conduction problems and complete heart block with age.[11]

Dyspnea, exercise intolerance due to congestive heart failure, and palpitations from supraventricular arrhythmias most often arise in the fifth decade.

Approximately one-tenth of infants born with congenitally corrected transposition have complete heart block. In patients born with normal cardiac conduction, the risk of developing heart block over time increases by 2% per year until it reaches a prevalence of 10 to 15% by adolescence, and 30% in adulthood.[12]

Systemic morphologic RV dysfunction and congestive heart failure occur in more than 50% of patients with associated lesions by the time they are 45 years of age. Ebstein anomaly of the systemic atrioventricular valve is common, and regurgitation occurs in over 80% of adults with cc-TGA. Pulmonary morphologic LV dysfunction occurs less often but may be present in up to 20% of patients. Aortic regurgitation of some degree is present in 25% of adults but seldom requires surgical intervention.

Interventional options include medical therapy and surgeries, including conduit replacement or repair, tricuspid valve replacement, double-switch procedure, and cardiac transplantation.

Medical therapy with ACE inhibitors, Angiotensin Receptor Blockers or beta-blockers for patients with systemic ventricular dysfunction may be intuitive, but no benefit has yet been demonstrated.[13]

If moderate to severe systemic (tricuspid, left) AV valve regurgitation develops, valve replacement should be considered. Left AV valve replacement should be performed at an early stage before systemic right ventricular function deteriorates, which should be done at an ejection fraction of 45% or greater. When TR is associated with poor systemic (right) ventricular function, the double-switch procedure should perhaps be considered. Patients with end-stage symptomatic heart failure should be referred for cardiac transplantation.[14]

Follow-up should include regular assessment of systemic (tricuspid) AV valve regurgitation by serial echocardiographic studies and systemic ventricular function by Magnetic Resonance Imaging or radio-nucleotide angiography. Holter recording can be useful if paroxysmal atrial arrhythmias or transient AV block is suspected.[15]

CONCLUSION

Patients having "isolated" cc-TGA can exceptionally survive until the seventh or eighth decade. The usual causes of death are sudden death (presumed arrhythmic) or, more commonly, progressive systemic right ventricular dysfunction with AV valve regurgitation.

All patients should have regular follow-up visits with a cardiologist.

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