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Case Report

A Case of Congenitally Corrected Transposition of Great Arteries Presented with Intracardiac Anomelies and Situs Inversus in a 28 year Old Male: A Rare Case Report

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ABSTRACT*:*
Congenitally corrected transposition of the great arteries (CCTGA) Situs inversus with levocardia represents a relatively very rare congenital condition and most patients are diagnosed in infancy or early age. Congenitally corrected transposition of the great arteries (CCTGA) accounts for less than1% of cardiac anomalies, and is defined as ventriculoarterial and atrioventricular (AV) discordance. The double discordant connection allows for survival with the right ventricle performing as the systemic ventricle, and the left ventricle as the pulmonary ventricle. we reported a case of 28year old male with congenitally corrected transposition of the great arteries which presented with 9month history of exertional dyspnea. A diagnosis was confirmed by echocardiogram showing situs inversus, levocardia, atrioventricular and ventriculoarterial discordance, VSD with bidirectional flow and sever pulmonary stenosis. Surgical intervention not done due to higher risk of anatomic repair. Although management of the corrected transposition of the great arteries patients remains controversial, the recommendation is that physiologic repair may be the procedure of choice for some patients particularly complicated cases. anatomic repair is not always suitable for older patients.

Keywords: ***Congenitally Corrected, Levocardia, Situs Inversus***

INTRODUCTION*:*

Congenitally corrected transposition of the great arteries (CCTGA), also known as Levo-transposition of the great arteries, was initially reported by Rokitansky in 1875. Since then, it remains a rare cardiovascular anomaly comprising less than 1% of all congenital cardiac diseases.[1-2] In this disease, the atria are at their normal location. However, there is atrioventricular (AV) and ventriculoarterial discordance whereby the right atrium is connected to the left ventricle while the left ventricle connects directly to the pulmonary artery. On the left side, the left atrium is connected to the right ventricle and the right ventricle connects to aorta allowing the oxygenated blood to enter the systemic circulation.[3] This results in uninterrupted pulmonary and systemic blood flow, hence the term 'congenitally corrected transposition[4] The ventricular inversion and the associated aortopulmonary rotation also result in mirror-image coronary artery distribution . Associated cardiac anomalies are common with CCTGA and seen in eighty percent of all cases[4]. The most relevant common associations are ventricular septal defect (VSD), pulmonary stenosis (PS), left AV valve (morphological tricuspid valve) regurgitation and complete heart block [5]. CCTGA may present as situs solitus or situs inversus. In situs solitus, the morphologic right ventricle and right atrium (RA) are located on the left and right side respectively. In situs inversus mirror-image location of the thoracic and abdominal organs. Levocardia usually exists in which the cardiac apex is directed to the left. The majority of these patients suffer from situs solitus, and only around 34% of cases have situs inversus[6]. Indication for surgery is also determined by the nature and severity of associated cardiac lesions. Our case diagnosed with CCTGA and situs inversus with levocardia which, associated with PS and VSD, is a very rare combination.

CASE REPORT:

A 28year-old male was admitted to MB hospital with a 9month history of exertional dyspnea. He was visibly cyanotic and had a Class III Dyspnea associated with history of loss of appetite and easy fatigability of the same duration. Additionally, he complaints of abdomen pain and urinary symptoms last 7 days. Physical examination revealed an irregular rhythm with heart rate at 90 beats/min, a grade III systolic ejection murmur and blood pressure was 130/82 mmHg in both arms with normal distal pulses. Oxygen saturation in room air was 74%. In addition, distal extremities were cyanotic and had a digital clubbing. Hemoglobin and hematocrit (HCT) 22g/dl, 70.9% respective. On general physical examination patient conscious oriented and no pallor, no icterus, grade3 clubbing (FIG.1) was present, central as well as peripheral cyanosis was present and bilateral pedal edema. Abdominal examination revealed congestive hepatomegaly on left sided.

Cardiovascular examination on inspection apex beat not visible, no precordium bulging or any pulsation. On palpation apex beat palpated 2.5cm mid from midclavicular line and downward. A pansystolic murmur was heard on auscultation with mid systolic high pitch ejection murmur over right second intercostal space. There was history of one episode of mild hemoptysis. There was no history of diabetes or hypertension. Patient first time diagnosed in 2016. past history of non-hemorrhagic infract 2year back. previous history of phlebotomy 3 times.

FIG1- GRADE 3 clubbing with cyanosis

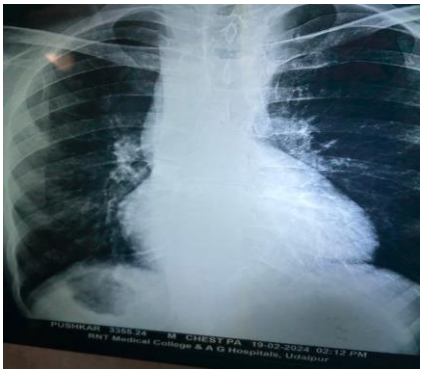

FIG 2.Chest X-ray showed a left heart axis, a right-sided gastric bubble below the right diaphragm and left-sided hepatic contour (fig2).

XRAY CHEST (FIG. 2):

Chest X-ray PA show a left heart axis, a right-sided gastric bubble below the right diaphragm and left sided hepatic contour.

ECG:

ventricular activation in CCTGA is different from that seen in the structurally normal heart. Absence of Q waves in the lateral precordium (V4-6), physiologic Q waves in these leads often referred to as "septal" Q waves. prominent Q waves in right precordium (V1-3)

USG WHOLE ABDOMEN AND PELVIS:

congestive change in liver with situs inversus. spleen and stomach present on right side. IVC present on left side and aorta on right side. both kidney normal size.

CECT WHOLE ABDOMEN AND PELVIS:

Liver present on left side with presence of multiple ill-defined hypodense lesion in both lobe of liver with showing peripheral heterogenous enhancement in arterial phase suggestive of multiple hemangioma. spleen and stomach are present in right side. IVC present on left side and aorta on right side. Both kidney normal in size.

CT PULMONARY ANGIOGRAPHY WITH CARDIAC CHAMBERS:

Atrioventricular and ventriculoarterial discordance with marked valvular pulmonary stenosis, relatively hypoplastic main pulmonary artery, large sub aortic VSD, overriding of aorta and multiple mediastinal and major aortopulmonary collaterals L-TGA WITH VSD-PS aortic overriding Right side of aortic arch showing mirror image branching pattern. concentric hypertrophy of right ventricle. situs inversus.

2D ECHO REPORT(FIG.3&4):

Situs solitus, mesocardia, AV Discordance and VA Discordance, Aorta to Right and IVC to left C-TGA, L Malposes great vessels, large inlet VSD Shunting bidirectionally, mild TR, Pulmonary valve from LV appear narrow not clearly profiled, confluent branch pas, sever ps, normal RV/LV Function and dimension.

Transesophageal Echocardiography (TEE):

all four chamber show discordance atrioventricular connection and morphologic left atrium in right side is connected to the morphologic right ventricle, morphologic right atrium in the left side is connected to morphologic left ventricle,

CATH LAB STUDY:

Situs Inversus, AORTA to right and IVC to left, AV/VA Discordance (C-TGA), large Subarterial VSD with Bidirectional Shunt. Sever PS (Valvular and Subvalular), Confluent dilated Pas

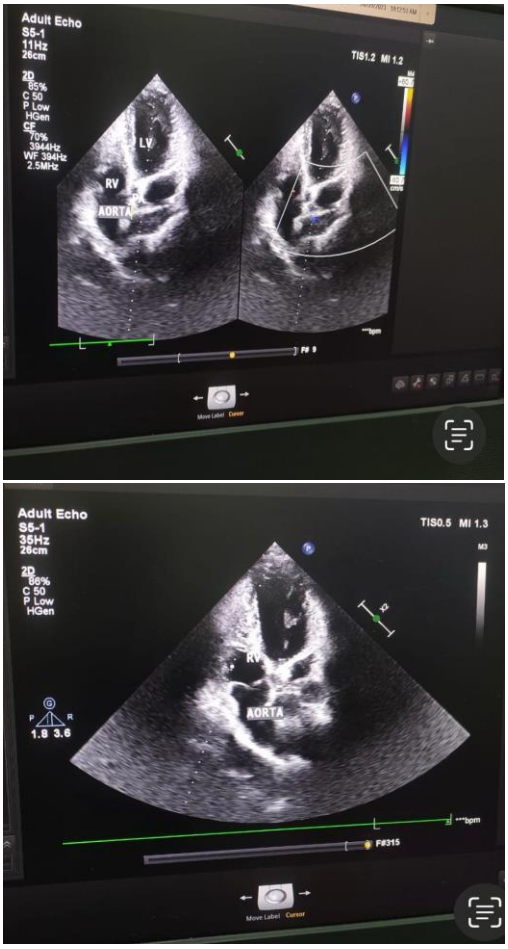

FIG.3&4:- 2D ECHO revealed complete transposition of the great arteries. In atrioventricular concordance the tricuspid valve attaches lower within the ventricle, there is a moderator band on the right side and the lumen of the right ventricle is shorter than the left. The great vessels arise in a parallel course and the aorta lies to the right and anterior of the pulmonary artery.

DISCUSSION:

The CCTGA is a rare congenital cardiac anomaly which occurs <1% of all forms of congenital heart disease. Since situs solitus accounts for the majority of cases, the confluence of situs inversus and levocardia with CCTGA is an extremely uncommon congenital anomaly[7]. Echocardiogram of our patient showed the CCTGA and situs inversus with levocardia and ventricular septal defect. Kukreti and colleagues[8] described a similar case of a 30-year-old male with situs inversus with levocardia and Congenitally Corrected Transposition of Great Vessels but he associated with rheumatic systemic AV valve stenosis and regurgitation. Other intracardiac anomalies like VSD and pulmonary stenosis in CCTGA are seen in eighty percent of cases, and the majority of them have one or both.[9] In our reported case patient both of these associated anomalies. Also, the most common associated defects in patients were tricuspid valve (TV) anomalies, which occurred in 91% of cases, VSD in 78% and pulmonary outflow tract obstruction in 40% cases. Indications for surgery in this anomaly are directly related to the severity of the associated intracardiac anomalies. Anatomical or classical (physiologic) repair are surgical methods for repairing CCTGA. The traditional surgical method for performing CCTGA has involved fixing any related abnormalities such tricuspid regurgitation or stenosis, pulmonary valve anomalies, and VSD.

This approach leaves the morphological right ventricle as the systemic ventricle and the morphologically right AV valve (tricuspid) as the systemic AV valve. So, because of concern about the long-term function of the morphological right ventricle and the systemic AV valve, some reports suggest anatomic repair, which includes Senning and Arterial Switch Procedure (Double Switch) and Senning plus Rastelli Procedure[10]. A Review study by Alghamdi and colleagues between 1992 and 2000 revealed that the anatomic repair (Rastelli Procedure) was associated with the lowest incidence of postoperative complications and favorable to early mortality compared with the physiologic repair. Few studies revealed that anatomic corrective surgery is feasible in children but in adult mortality is high so this surgical procedure is not suggested. The best surgical treatment for a small number of individuals with CCTGA is physiologic correction, such as features of an inlet ventricular septal defect, tiny atrium, or dextrocardia, among other complicated problems. Although the best surgical procedure, in cases of complicated CCTGA remains controversial and age is one of the most important factors for selecting surgical procedure. Devaney and colleagues[11] showed that anatomic repair with a combined surgery and arterial switch operation had a favorable outcome in patients with CCTGA. Thus, the anatomic repair is not always suitable for older patients. Morphologic left ventricular function was stable and better outcome after anatomical repair especially if performed early.

CONCLUSIONS:

An uncommon variation of situs inversus is called isolated levocardia, in which the abdominal organs are in the dextroposition while the heart is in the conventional levocardia. The cause of situs inversus in patients with levocardia is unknown. Even if a thorough physical examination is necessary for the diagnosis of situs

inversus, medical imaging has enabled the verification of the results and the search for other disorders. Situs inversus with levocardia has a poor prognosis, owing to the severity of an associated heart abnormalities. From physiologic repair, which solely addressed the accompanying structural abnormalities, to anatomic repair, which established the morphological left ventricle as the systemic ventricle, surgical care of CCTGA has changed over time. Although the outcomes with the anatomic repair are better, A studies are required to refine the indications for surgical intervention especially in absence of associated structural defects. management of CCTGA still remains controversial.

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