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A CASE OF SITUS INVERSUS AND DEXTROCARDIA WITH CONGENITALLY CORRECTED TRANSPOSITION OF GREAT ARTERIES IN AN ELDERLY MALE -A RARE ASSOCIATION

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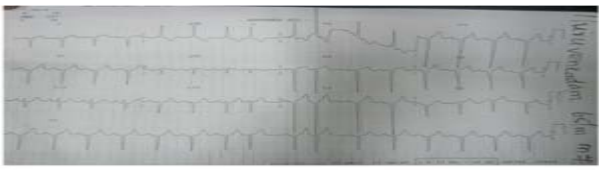
Abstract : Congenitally Corrected Transposition of Great Arteries is a rare cardiac malformation characterized by the combination of discordant atrioventricular or ventriculoarterial connections, usually accompanied by other cardiovascular malformations. Incidence has been reported to be around 133,000 live births, accounting for approximately 0.05 of congenital heart malformations. Associated malformations may include inter ventricular communications, dextrocardia, obstruction of the outlet from the morphologically left ventricle and tricuspid valve abnormalities. The clinical picture and age of onset depends on the associated malformations. Bradycardia, single loud S1 and a heart murmur are the most common manifestations. In the rare cases where there are no associated malformations, congenitally corrected TGA can lead to progressive AV valve regurgitation and failure of the systemic ventricle. The diagnosis can also be made late in life when the patient presents with complete heart block or cardiac failure. Etiology is currently unknown and with increase in incidence among families with previous cases of CCTGA reported. Diagnosis can be made by fetal echocardiography, but is far more commonly made postnatally with clinical signs and echocardiography. Prognosis is defined by the associated malformations and on the timing and approach to palliative surgical care. This case is presented for its rarity and the age at presentation, this patient being one of the eldest reported.

Keyword : Dextrocardia, congenitally corrected transposition of great arteries.

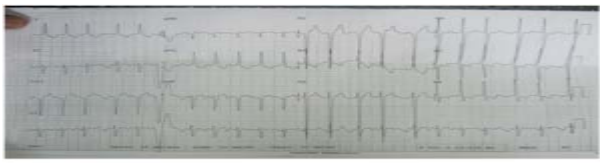
CASE REPORT

65 years old male , coolie by occupation was admitted with complaints of exertional dyspnea for 6 months, worsened over the past one month. There was history of palpitations on and off with no abdominal distension or swelling of legs. There was no chest pain/cough/fever and no similar illness in the past. He was not a diabetic/hypertensive/CAD patient. Smoking and alcohol consumption were positive. There was no family history of congenital heart disease. He is married with nonconsanguinity and two healthy living children. His vitals were normal except for occasional premature beats. Cardiac examination revealed apical impulse in the right side of the chest and heart sounds were heard better in the right. A systolic murmur was heard in the right sternal border. Other system examinations were normal.



Rate	96/min
Rhythm	Sinus rhythm with occasional APC
Axis	North west axis
P wave	Inverted in I, aVL Upright in aVR 'P' mitrale
PR interval	180 ms
QRS complex	Reversed progression in precordial leads Upright in aVR
ST segment	No deviation
T wave	Inverted in V_2, aVR
QTc	420 ms

Prediagnostic ECG



Corrected ECG

Chest X-ray

Chest x-ray shows dextrocardia with right sided aortic arch and situs inversus as evidenced by right sided gastric air shadow.

Echocardiography revealed the following findings; situsinversus with dextrocardia, Atrio-Ventricular discordance with Ventriculo-Arterial discordance, malposition of great arteries, right sided aortic arch, large VSD with bidirectional shunt, mild pulmonary stenosis, right AV valve regurgitation. His renal and liver parameters were within normal limits. Blood counts were normal. USG abdomen was normal.

His final diagnosis **was situsinversus with dextrocardia, Congenitally corrected TGA, Large VSD with bidirectional shunt and mild pulmonary stenosis.** He was managed symptomatically and discharged with regular follow up.

DISCUSSION

Introduction

Congenitally corrected transposition of the great arteries (ccTGA) is a rare defect combining atrioventricular discordance with ventriculoarterial discordance. The atria are connected to the opposite ventricle (left atrium to right ventricle via a tricuspid valve) and the ventricles are connected to the incorrect great artery (right ventricle to aorta). Thus oxygenated blood is circulated systemically by the morphologic right ventricle (RV) and deoxygenated blood returns to the right atrium to be pumped out the left ventricle (LV) to the lungs. The defect is therefore "corrected" because of the physiologic flow of blood through the body.

Anatomy

The most common anatomy of ccTGA is that of {S,L,L}, representing atrial and visceral situs solitus (right-sided inferior and superior vena cavae returning deoxygenated blood to a right sided atrium), Llooped ventricles (the morphologic LV with mitral valve positioned on the right), and L-transposed great arteries (aorta arising off the left-sided morphologic RV and therefore situated anterior and leftward of the pulmonary artery). The RV serves as the systemic ventricle and, in the absence of other defects, oxygen saturation is normal. The most common positions of the heart in the chest are levocardia (apex to the left) or mesocardia (midline). Patients with levo- or mesocardia and visceral situsinversus have a high likelihood of ccTGA and therefore must carefully by assessed for atrial, ventricular, and arterial concordance. Dextrocardia, in which the apex of the heart is to the right, occurs in approximately 20% of patients (Graham & Markham, 2010). In cases of dextrocardia with mirror-image anatomy the anatomic designation is {I,D,D}.

Associated defects

The most common associated defects in ccTGA are ventricular septal defects (VSDs), whichoccur in 60-80% of cases, pulmonary stenosis (PS) in 30-50%, and tricuspid valve (TV)anomalies in 14-56%. The VSDs are usually large, perimembranous, and subpulmonary inlocation. Muscular inlet defects as well as multiple VSDs may also be seen. Pulmonarystenosis, more appropriately referred to as left ventricular outflow tract obstruction(LVOTO), may be caused by fibromuscular tissue, valvular stenosis, or aneurysmal tissue of the membranous ventricular septum. The associated combination of LVOTO and VSD represents the largest group of ccTGA patients. TV anomalies occur along a spectrum of which an Ebstein-like anomaly is often the most clinically severe. Furthermore, as the TV is subjected to systemic pressures, even normally formed valves display progressive regurgitation with age. Less common defects occurring in association with ccTGA include atrial septal defect, patent ductus arteriosus, pulmonary atresia, double-outlet RV, aortic regurgitation, mitral valve abnormalities, and subaortic stenosis.(Graham & Markham, 2010;Hornung & Calder, 2010; Van Praagh et al., 1998).

Incidence and genetics

The incidence of ccTGA in patients with congenital heart disease (CHD) is approximately 0.5% with a slight male predominance (Graham & Markham, 2010; Piacentini et al., 2005). Although a specific genetic defect is yet to be defined for ccTGA the recurrence risk ofd-TGA for siblings of ccTGA patients is 2.6% with an overall recurrence risk of 5.2% for ccTGA siblings to have some type of congenital heart defect (Piacentini et al., 2005). A recurrence risk of >5% is higher than expected, as the risk is typically thought to be 1-3% for unaffected parents to have an additional child with congenital heart disease (Van derBom et al., 2011).

Diagnosis

Just as the natural history is largely dependent on defects associated with ccTGA, so is timing of presentation and diagnoses

Early presentation and diagnosis

Diagnoses of infants and children may occur after murmur evaluation, as VSDs are commonly associated lesions. In cases of large VSDs or severe TV regurgitation, some infants may present in CHF with diaphoresis, pallor, tachypnea, inability to gain weight, hepatomegaly, and a gallop on exam. Auscultation of the ccTGA patient may also reveal a loud, single second heart sound (S2) at the left 2nd intercostal space, with absence of S2 over the right 2nd intercostal space (Friedberg &Nadas, 1970). The presence of VSD combined with LVOTO may lead to a cyanotic presentation from decreased pulmonary blood flow.However, some degree of LVOTO may be protective of the lung bed in patients with large VSDs, and may delay a CHF presentation despite the normal decrease in pulmonary vascular resistance.

Late presentation and diagnosis

Interestingly, if there are no additional associated defects ccTGA may go unnoticed until adolescence or adulthood. Case reports have even cited incidental findings and late diagnoses of ccTGA in adults in the fifth to eighth decades of life (Chang et al., 2009; Jennings et al., 1984; Orchard et al., 2010; Scardi et al., 1999). A cohort of patients with ccTGA over 18 years of age who presented to an adult CHD clinic over a 15 year period is described by Beauchesne et al (2002). Sixty-six percent of these patients were over 18 years of age when diagnosed, and 17% of the cohort was over 60 years old at the time of diagnosis. Common reasons for referral in such patients range from abnormal ECGs and cardiomegaly on chest radiographs to complete heart block and murmurs (Presberito et al., 1995).

Evaluation

Chest radiograph

The CXR in ccTGA patients with mesocardia or levocardia typically demonstrates a straightened upper-left cardiac border from the leftward-positioned ascending aorta. Dextrocardia usually occurs with normal situs and, as stated previously, occurs in 20% of ccTGA patients (Figure 3). The presence of abdominal situs solitus and dextrocardia should raise suspicion of ccTGA. In the patient without any associated defects, an atypical cardiac position in an otherwise normal CXR may be the only indication of ccTGA. However marked cardiomegaly, left atrial enlargement, and an increase in pulmonary vasculature may be present in patients with a large VSD and significant left to right shunt. A CXR with impressive cardiomegaly and left atrial enlargement may also be indicative of an Ebsteinlike malformation of the TV. The presence of pulmonary stenosis or atresia will demonstrate darkened lung fields from attenuated pulmonary blood flow. Overall, the degree of cardiomegaly and amount of visible pulmonary vascularity is dependent on the presence and direction of shunting, as well as the severity of LVOTO (Carey & Ruttenberg,1964).

Electrocardiogram

The ECG in patients with ccTGA is most significant for a superior QRS axis and atypical septal activation. As discussed previously, the conduction system in ccTGA consists of inverted AV bundles. Therefore the septum is activated from right to left, demonstrating presence of septal Q waves in the right precordial leads (QR pattern in leads V4R and V1) and absence of Q waves in the left precordial leads (rS pattern in lead V6). In fact, undiagnosed ccTGA patients with such a pattern on ECG have been diagnosed with remote inferior infarcts (Jennings et al., 1984; Warnes, 2006). Pre-excitation may be observed in those patients with ccTGA and Wolff-Parkinson-White. Finally, varying degrees of AV block may be present, as well as patterns of right or left-sided chamber enlargement.

Medical management

CHF medical management for the ccTGA patient with systemic RV has been extrapolated from CHF therapy for LV failure. This primarily includes -adrenergic receptor blockade (-blockers), diuretics and afterload-reducing agents with an angiotensin-converting enzyme(ACE) inhibitor (Winter et al., 2009). Digoxin may also be useful for its inotropic and antiarrhythmic effects. Angiotensin receptor blockade with losartan was evaluated in a multicenter, randomized, placebo -controlled clinical trial by Dore and colleagues (2005) but found to have no improvement on exercise capacity and no reduction in neurohormonal levels in patients with systemic right ventricles. Overall, evidence-based therapy for optimal CHF treatment in patients with systemic RV is lacking. Beyond medication, cardiac resynchronization has emerged as a therapy for patients with impaired systemic RV function and widened QRS morphology on ECG. Increased QRS duration as a result of bundle branch block or conventional pacemaker is typically greater than 120-140 ms with some patients having QRS duration >200 ms. Such electromechanical dyssynchrony creates inefficiency in ventricular ejection, whereas restoring synchrony has been shown to decrease QRS duration with improvement in RV filling time, ejection fraction, and overall CHF symptoms (Diller et al., 2006; Janousek et al., 2004; Kordybach et al., 2009). Takemoto et al.(2010) reports the use of transvenous permanent para-Hisian pacing in an 8 year old with ccTGA. Restoration of cardiac synchrony decreased the QRS duration from 198 ms to 94 ms, decreased interventricular conduction delay from 137 ms to 37 ms, and improved the patient's CHF symptoms from NYHA (New York Heart Association) class III to NYHA class II over a period of 6 months. Limitations in cardiac resynchronization therapy include difficulty in percutaneous lead delivery, although this has successfully been accomplished even in ccTGA cases of dextrocardia (Malecka et al., 2010).

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