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Congenitally corrected TGA- A case diagnosed incidentally

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ABSTRACT

Congenitally corrected transposition of the great arteries (CCTGA) is a rare congenital heart defect associated with multiple cardiac morphological abnormalities and conduction defects. Data from the Baltimore-Washington Infant Study supported the fact that congenitally corrected transposition is a rare disorder (1). As many as 40 infants per 100,000 live births are affected by congenitally corrected transposition of the great vessels; this is less than 1% of all congenital heart defects. The literature reports fewer than 1000 cases, however, the true prevalence of the malformation is not known. We report a case of a 25 year old female diagnosed incidentally post delivery with CCTGA.

Keywords: Congenital, heart, bradycardia

Introduction

CCTGA is a rare congenital heart disease (CHD) with a prevalence of less than 1% of all CHD [1].It is characterized by atrioventricular (AV) and ventriculoarterial (VA) discordance. It may be associated with various other congenital anomalies. The progressive risk of spontaneous complete AV block throughout life in patients with CCTGA is 2% per year [2]. The prognosis rests on clinical presentation, progression of disease, and the effect of systemic pressure on the functional SV .Patients undiagnosed until adulthood usually have no associated anomalies and present due to an abnormal chest radiograph or ECG. These patients are asymptomatic until right ventricular dysfunction, tricuspid regurgitation, or complete heart block develops. No treatment is required for patients with corrected transposition who have no other defects because their life expectancy has been reported to be near normal [3].

Case Report

We report a case of 25 years old female third gravid with two living issues came to the emergency at term in second stage of labour with severe anaemia not in failure. She delivered a healthy boy baby of 3.1 kg vaginally.

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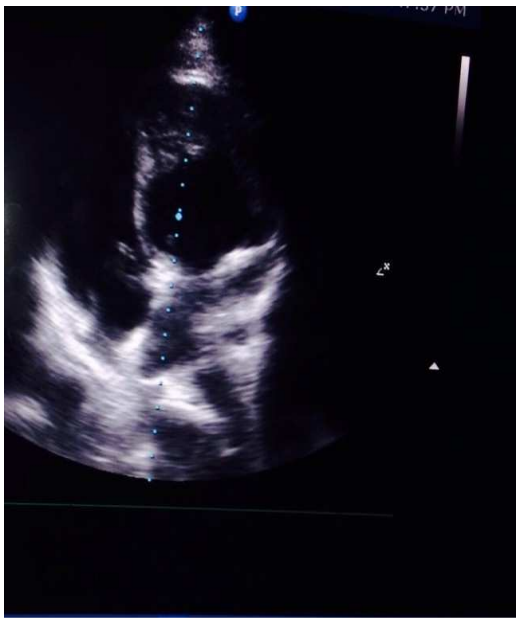
Her blood investigations revealed a haemoglobin of 5.5 g % with microcytic hypochromic anaemia. Rest of the parameters was within normal limits. Patient was being conservatively treated for anaemia when she developed secondary post partum haemorrhage 10 days after her delivery. An ultrasonography of the pelvis revealed retained products of conception of size 3.2 X 4 cm. Patient was taken up for evacuation of products. She had excessive haemorrhage while removal of the retained products in view of which a balloon tamponade was inserted. Adequate blood transfusion was given. Her vitals started deteriorating after the procedure. She was maintaining her blood pressure but had persistent bradycardia and was started on ionotropes. Her ECG showed T wave inversion in lead I and aVL and missed beat in lead II,III, Avf. Her echocardiograph revealed pulmonary artery originating from left ventricle and aorta originating from right ventricle, concentric right ventricular hypertrophy , normal biventricular function, ejection fraction of 60% and no pericardial effusion. As patient was asymptomatic apart from having bradycardia, she was managed conservatively and was advised a pacemaker.

Discussion

CCTGA, or synonym l-transposition, is a rare (less than 1% of all CHD) and complex heart defect. It is characterized by AV and ventriculoarterial discordance. The aorta is located closer to the anterior and more to the left than the pulmonary artery. The AV valves follow their respective ventricles. During

embryological development, left-handed looping of the heart tube results in atrioventricular (AV) discordance and the aorto pulmonary septum fails to rotate 180°, resulting in ventriculoarterial discordance. Blood flows in an effective sequence, hence the name corrected; however, the right ventricle supports the systemic circulation in this disorder. Venous blood returns from the body into the right atrium before passing through the mitral valve into a morphological left ventricle. Blood then enters the lungs via the pulmonary valve into the main pulmonary artery. Pulmonary venous blood returns to the left atrium and then passes through the tricuspid valve to the morphological right ventricle, exiting to the aorta via the aortic valve. The aorta is positioned anterior and to the left of the pulmonary artery. In effect, the ventricles are transposed. Because of the displacement of the AV node and the abnormal course of conduction tissue, there is an increased risk of spontaneous complete AV block. CCTGA is commonly associated with other cardiac defects and its isolated occurrence is rare. SCD is still the leading cause of death in patients with CHD. CCTGA has the highest mortality among all CHD patients [4]. Some infants and children born with CCTGA have heart murmurs, heart rhythm problems, and/or show signs of heart failure. If they have a VSD and they may also be cyanotic. In these situations, an echocardiogram is typically done and a diagnosis made. However, it is not unusual for CCTGA to be found later in childhood or adulthood. This is because you can have no symptoms and have CCTGA. Often it is the start of new symptoms - a new murmur, rhythm problems, or heart failure symptoms - that leads to a diagnosis of CCTGA. Sometimes it is found accidentally when routine heart tests are done. Echocardiography can be used to diagnose CCTGA in adulthood. But rates of misdiagnosis of CCTGA using echocardiography at adult cardiac centers are high. For those with implanted devices that make MRI impossible, CT scans can be used. Most women with CCTGA are able to conceive and deliver vaginally. Some patients are asymptomatic throughout their life and are diagnosed incidentally whereas some infants die immediately after birth. The survivability depends on right ventricular function. CCTGA can be associate with VSD in 60% of cases. 40 % of patients with CCTGA have single ventricle and 20% have dextrocardia, 30-50 % have pulmonary stenosis. It can also be associated with rhytm disturbances, pulmonary hypertension, leaky tricuspid valves and heart failure. The prognosis depends on correct diagnosis and associated heart defects. The 20 year survival rate for congenitally corrected transposition of great arteries is 75%. Since CCTGA is a complex defect, specialized care from cardiologist is mandatory in every case.



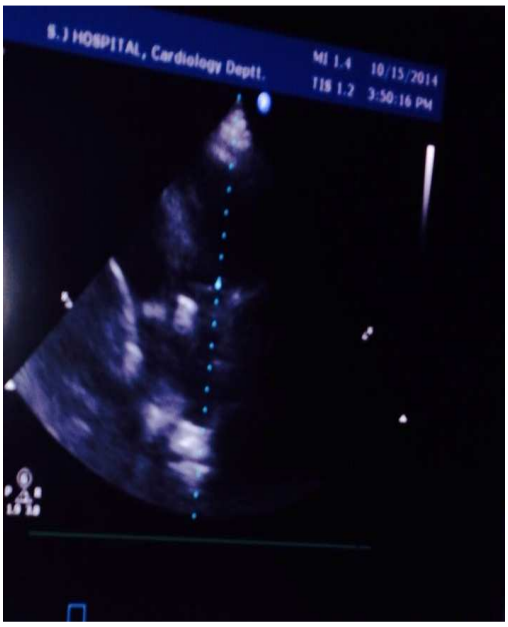


Fig 1 and 2: Echocardiograph images of CCTGA

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