Case Report

Complex Adult Congenital Heart Disease: A Rare Case of Dextrocardia with Double-outlet Right Ventricle

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Abstract

Double-outlet right ventricle (DORV) is a complex and rare congenital heart disease (CHD), which is usually symptomatic early in childhood and thus diagnosed early. Dextrocardia is another rare CHD that can occur either as an isolated cardiac abnormality or may be associated with other complex cardiac defects. However, the co-occurrence of DORV and dextrocardia is rare, especially in adult life. We present a 35-year-old Nigerian lady with situs inversus totalis and complex CHD, which included dextrocardia with multiple congenital intracardiac shunts, DORV occurring with L-malpositioning of the great vessels, atrial septal defect, and ventricular septal defect with partial anomalous pulmonary venous drainage, persistent left superior vena cava as well as severe pulmonary stenosis.

Keywords: Congenital heart disease, dextrocardia, double outlet right ventricle, multiple intracardiac shunts, septal defects, situs inversus

Introduction

Congenital heart diseases (CHD) are present at birth but may not manifest or diagnosed till adulthood; however, complex CHD is often symptomatic early in childhood and thus diagnosis are made early.[1-3] The complexity of CHD is directly related to morbidity and mortality as well as complications of the disease.[3]

Dextrocardia has been described as a rare but complex congenital heart abnormality whose true incidence is estimated to vary from 1 in 8000 to 25,000 live births.[4,5] Dextrocardia may be isolated or may occur in association with multiple and complex congenital cardiac abnormalities in infants; however, dextrocardia in adults has a few ranges of diagnostic scenarios which can occur as polysplenia syndrome, situs inversus totalis (mirror-image dextrocardia), congenitally corrected transposition of great vessels, and dextroversion.[2,5,6] Situs inversus totalis is usually associated with many cardiac (ventriculoarterial discordance) and extracardiac (malorientation of abdominal organs) anomalies.[2,5]

Double-outlet right ventricle (DORV) is also a rare CHD with a prevalence of 1%–3% in all individuals with CHD.[7,8] DORV is of great clinical significance and occurs in approximately 3–24/100,000 live births.[7,8] The Congenital Heart Surgery Nomenclature and Database Project defined DORV as a type of ventriculoarterial connection, in which both great vessels (the aorta and pulmonary trunk) arise entirely or predominantly from the morphologically right ventricle.[8] It may be associated with aortic and/or pulmonary outflow tract obstruction.[9] Chromosomal abnormalities, genetic mutations, maternal diabetes, and glucose intolerance, as well as prenatal exposure to alcohol and teratogens such as retinoids, valproate, and theophylline, have been implicated in DORV teratogenesis.[8]

Dextrocardia with associated DORV occurring in an adult is rare in the literature and may or may not be related to heterotaxy.[9] Most cases of the combination of these two complex anomalies usually result in foetal or early childhood

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mortalities without surgical intervention.[9] We, therefore, present a 35-year-old Nigerian female presenting with situs inversus totalis and complex congenital heart defects, which include dextrocardia in association with DORV with L-malpositioning of great vessels, atrial septal defect (ASD), ventricular septal defect (VSD), and severe pulmonary hypertension with right ventricular strain.

Case Report

A 35-year-old Nigerian female presented to the medical emergency unit of the hospital with a three-week history of worsening exertional breathlessness, orthopnea, paroxysmal nocturnal dyspnea, cough, early satiety, and worsening bilateral pedal swelling. She had a history of effort intolerance which worsened in the last seven months following a miscarriage at 13-week gestation. She has, however, had two prior successful pregnancies, which ended in spontaneous vaginal deliveries at term, about seven and three years before this miscarriage, with no cardiac decompensation.

Examination revealed a young woman, dyspneic at rest, not pale, centrally cyanosed with Grade IV digital clubbing of fingers and toes [Figure 1a and b] and pitting pedal edema up to the knees, bilaterally. Her pulse was 82 bpm, irregular, blood pressure was 110/84 mmHg, elevated jugular venous pressure, the cardiac apex beat was located in the 6th right intercostal space, anterior axillary line with a palpable thrill over the precordium, and a Grade V apical pansystolic murmur at the cardiac apex. Respiratory rate was 40 cycles/min, with bibasal fine crepitations and pulsatile tender hepatomegaly.

Investigations done included a 12-lead electrocardiogram, with leads placed conventionally and reversed [Figure 1c], which showed atrial fibrillation with frequent ventricular extrasystoles and a ventricular rate of 108 beats/min. Chest X-ray showed dextrocardia and gross cardiomegaly with perihilar opacities [Figure 1e]. Transthoracic echocardiography showed VSD with ASD, more than 50% override of the aorta, as well as severe pulmonary stenosis. The left ventricular ejection fraction was 67% [Figure 1d].

Cardiac tomography showed multiple cardiac anomalies; dextrocardia with situs invertus, DORV with L-malpositioning of great vessels, atrial and VSDs, partial anomalous pulmonary venous drainage, pulmonary stenosis with poststenotic dilatation of bilateral pulmonary arteries, persistent left superior vena cava, and gross cardiomegaly [Figure 1f-m].

A diagnosis of congestive cardiac failure (New York Heart Association [NYHA] IV) secondary to complex CHD, complicated by severe pulmonary hypertension (Estimated Pulmonary arterial systolic pressure was 113 mmHg) was made. She was commenced on antifailure medications and counseled for right heart catheterisation as well as heart and lung transplants. She made some improvement and had symptomatic relief in medical management. She was later discharged home in NYHA II. She is still on guideline-directed medical management for heart failure and being worked up for heart and lung transplants outside the country.

Discussion

We presented an unusual presentation of complex and complicated adult CHD in an adult Nigerian female, who presented to our emergency room with clinical and radiological diagnosis of congestive cardiac failure secondary to DORV and dextrocardia. This is an unusual presentation in adulthood as most CHD of this complexity become symptomatic in childhood and would have been diagnosed early and offered corrective surgical treatment.[2] She, however, presented late with complications. Late presentation of CHDs is common in this environment.[10,11]

The index patient, though, has been having some exertional shortness of breath earlier, did not decompensate until she had a miscarriage at 13-week gestation. Anaemia and infections following the miscarriage could have been the precipitant for the heart failure in this patient. Anaemia, infections, and arrhythmias are common precipitants of heart failure in the tropics.[10,11] Surprisingly, our patient has had two successful deliveries par vaginum before the miscarriage. Cardiovascular maladaptation to the hemodynamics of pregnancy has also been shown to be a precipitant of heart failure in individuals with existing cardiac conditions.[12]

Environmental factors, as well as chromosomal abnormalities and genetic mutations, have been implicated in the etiopathogenesis of DORV.[7] Although cytogenetic, biochemical, and molecular genetic analyses were not done in this index patient, she however had no obvious fascie of the common trisomies, and she had no known history of cardiac disease in the immediate and distant relatives. We postulate that environmental or maternal exposures to teratogens may be the likely culprit in this case. Her mother received no antenatal care, and she was delivered at home by traditional birth attendants.

Severe irreversible pulmonary hypertension was present in our patient probably because of late presentation. This made her to have limited treatment options. Early presentation in childhood would have afforded her care before severe complications set in. Early treatment may have given her a better quality of life.[11] The good surgical outcome, with excellent one-year postoperative follow-up period, had been reported in a six-year-old child with similar complex cardiac lesions.[13] Our index patient is presently in NYHA class III, despite optimal guideline-directed medical therapy, but heart and lung transplant may be the definitive treatment of choice in this case.

Conclusion

Complex and rare CHDs may seldom present in adulthood with severe and irreversible complications that will only be

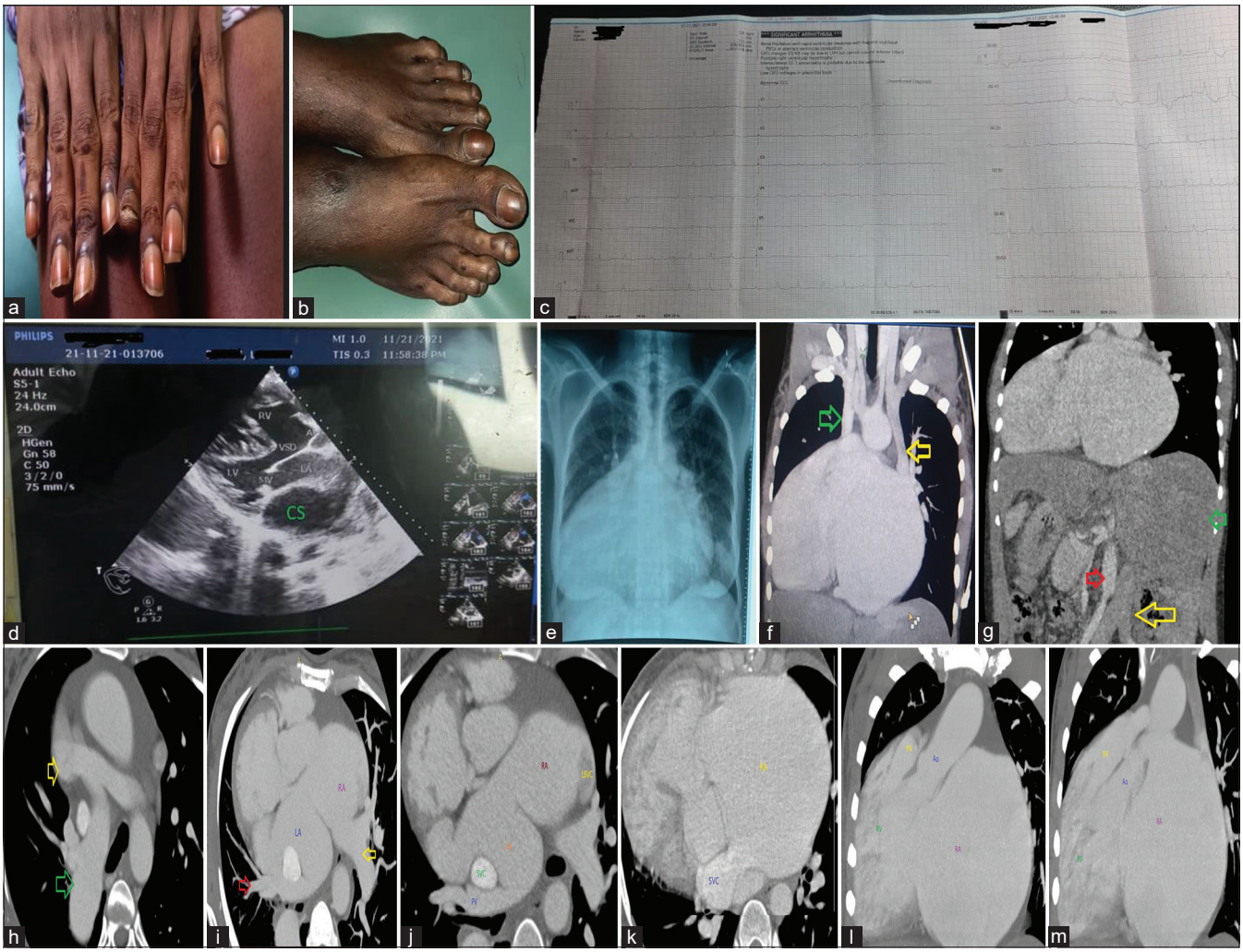

Figure 1: (a) Finger clubbing, (b) Toe clubbing, (c) Electrocardiogram revealed atrial fibrillation with fast ventricular rate and ventricular premature complexes, ventricular bigeminy, and couplets, (d) Echocardiogram (parasternal long axis view) showed a large membranous VSD with more than 50% override of the aorta and dilated coronary sinus, (e) Chest X-ray showed dextrocardia, (f) Cardiac CT scan showed dextrocardia and right superior vena cava (green arrow) and persistent left superior vena cava (yellow arrow), (g) Abdominal CT scan showed situs inversus, with liver on the left iliac fossa (green arrow), inferior vena cava on the left of the spine (yellow arrow), and aorta to the right of the inferior vena cava (red arrow), (h) Cardiac CT scan showed pulmonary stenosis (yellow arrow) and poststenotic dilatation (green arrow), (i) Cardiac CT scan showed partial anomalous pulmonary venous drainage, with right pulmonary veins draining into the left atrium (red arrow) and left pulmonary veins draining into the right atrium (yellow arrow), (j) Secundum atrial septal defect and persistent left superior vena cava draining into the right atrium, (k) Right superior vena cava draining into the right atrium, (l and m) demonstrates more than 50% override of the aorta. Ao: Aorta, CS: Coronary Sinus, LA: Left atrium, LSVC: Left superior vena cava, LV: Left ventricle, MV: Mitral valve, PA: Pulmonary artery, PV: Pulmonary vein, RA: Right atrium, RV: Right ventricle, SVC: Superior vena cava, VSD: Ventricular septal defect

amenable to heart and lung transplants. Therefore, routine cardiac screening of newborns, either at the postnatal clinic or during routine immunisation, will help improve the early detection of CHDs, promote early intervention, and prevent avoidable severe and irreversible complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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