CASE REPORT

Uncorrected Tetralogy of Fallot in a 47 Year Old Nigerian Man: A Case Report

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ABSTRACT

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease and is usually diagnosed in childhood. Uncorrected TOF has a bad prognosis with 95% mortality by the age of twenty. There are reports of patients with uncorrected TOF who survived into adulthood with minimal symptoms. This has been attributed to relatively small degrees of left ventricular outflow tract (LVOT) obstruction and/or presence of congenital aorto-pulmonary shunts. Very few cases of uncorrected TOF in adults have been reported in Africa. We report this case of uncorrected TOF in a 47 year old Nigerian man to highlight this rare presentation in adult life. He is the oldest case of TOF so far reported in Nigeria. He had hypertension with left ventricular hypertrophy which may have contributed to his longevity.

Key words: *Congenital heart disease, Ventricular septal defect, Pulmonary stenosis, Adulthood*

INTRODUCTION

Tetralogy of Fallot (TOF) is a complex congenital cyanotic heart disease. It was first described in 1888 by Etienne Louis Arthur Fallot and consists of pulmonary stenosis/obstruction of right ventricular outflow tract (Infundibular stenosis), Ventricular septal defect (VSD), Over-riding aorta and right ventricular (RV) hypertrophy.[1]

TOF is the most common cause of cyanotic congenital heart disease.[2] It is rarely diagnosed in adult life. Studies have demonstrated that 66% of persons with TOF not treated surgically live to 1 year, 49% to 3 years of age and 24% to age of 10 years.[3] Only 2% of all patients with TOF reach the fourth decade of life.[4]

Survival into adult life without surgical intervention has been associated with adaptations which ameliorate shunting of blood from the right ventricle to the left ventricle.[5] These adaptations include: systemic to pulmonary collaterals, systemic hypertension and patent ductus arteriosus.

Echocardiography is the most common imaging modality used in the diagnosis of TOF with a sensitivity of 71.3 % in children below 5 years and specificity of 85% in children above 5 years reported by Raval, *et al.* who compared echocardiography diagnosis with intra-operative findings in children below and above 5 years of age.

We present this case of a 47 year old Nigerian man to highlight the unusual presentation of uncorrected TOF in adult life and discuss factors contributing to his longevity.

CASE REPORT

We present a 47 year old commercial vehicle driver who was referred to the cardiology team on account of palpitations and shortness of breath while on admission in the surgical ward. He was admitted by the urology unit for right flank pain and was being evaluated for urolithiasis. Palpitation was intermittent, occurring at rest. He was not on any medication or beverage causing palpitations. He had no orthopnoea, leg swelling, chest pain, or history of syncope.

He recalled a past history of breathlessness during his childhood. He was admitted at University of Nigeria Teaching Hospital Enugu when he was 12 years on account of breathlessness and leg swelling. His parents were told then that he had a congenital heart disease. He was treated with drugs and did not undergo any surgical correction. He did not continue with the follow up visit schedules at the hospital as his symptoms gradually abated without any further treatment. He was not known previously to have hypertension.

On physical examination he had no pedal edema, cyanosis, or finger clubbing. He had a pulse of 96 beats per minute which was irregular with ectopic beats. His blood pressure was 140/100mmHg. Jugular venous pressure was not elevated. Apex beat was not displaced. He had left parasternal heave, loud pulmonary component of the second heart sound (P2) and a grade V systolic murmur at the left sternal border.

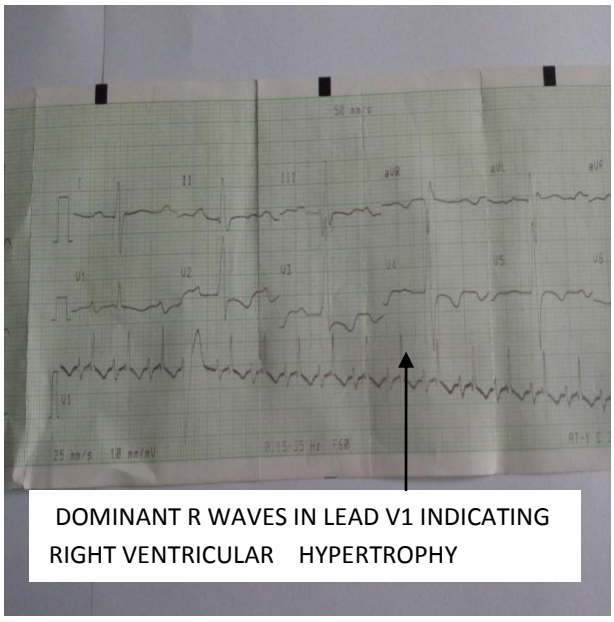
His serum electrolytes, urea and creatinine levels were normal. He had a haemoglobin concentration of 13.4g/dl.

Chest X ray showed enlarged cardiac silhouette with cardiothoracic ratio of 0.55.

Electrocardiogram (Figure 1) showed a sinus rhythm with QRS axis of 106° (right axis deviation). There was evidence of right ventricular hypertrophy with strain in leads V1- V3.Unifocal ventricular ectopics were also seen.

Echocardiogram (Figure 2) revealed normal heart chamber dimensions with left ventricular hypertrophy.

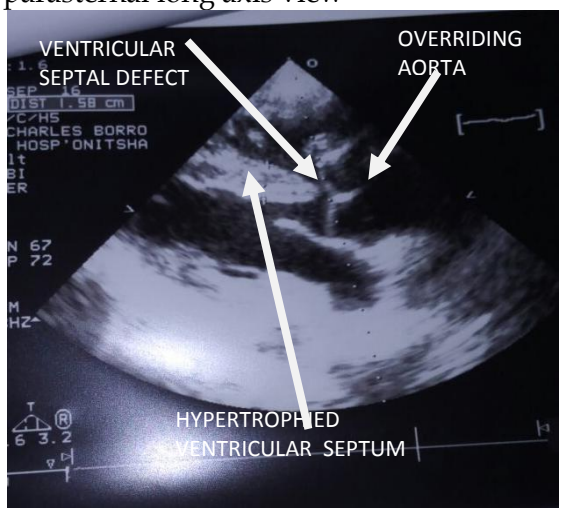
Figure1. Electrocardiogram showing right Ventricular hypertrophy in lead V1 with R/S ratio >1



The inter-ventricular septum diameter in diastole (IVSd) was 15.8mm. Right ventricular wall was also hypertrophied, measuring 10.3 mm. Left Ventricular ejection fraction (EF) was 65%. A peri-membranous ventricular septal defect with left to right flow on color Doppler was present. The ascending aorta was overriding the ventricular septal defect (Figure 2 & 3).

The pulmonary valve was thickened and stenosed with markedly elevated peak velocity of 5.42 m/s and peak gradient of 117mmHg (Figure 4). Grade 1 left ventricular diastolic dysfunction was observed. Mitral valve inflow E/A ratio was 0.6.

Figure 2. Echocardiogram showing ventricular septal defect, overriding aorta and hypertrophied inter-ventricular septum in the parasternal long axis view



The aortic, mitral and tricuspid valves were all structurally normal. There was mild tricuspid regurgitation.

Diagnosis of tetralogy of Fallot was made on account of presence of these classical features.

He was given antihypertensive drugs. His palpitations subsided and he was counseled on the diagnosis and the need for follow up. He was discharged and given appointment for follow up. Surgical correction of the congenital anomalies is the definitive treatment for this TOF. We have discussed his case with cardiothoracic surgeons and patient has been referred to a cardiac surgery centre in the South- East of Nigeria for definitive treatment.

Figure 3: Apical four chamber view echo cardiogram with arrow pointing at ventricular septal defect

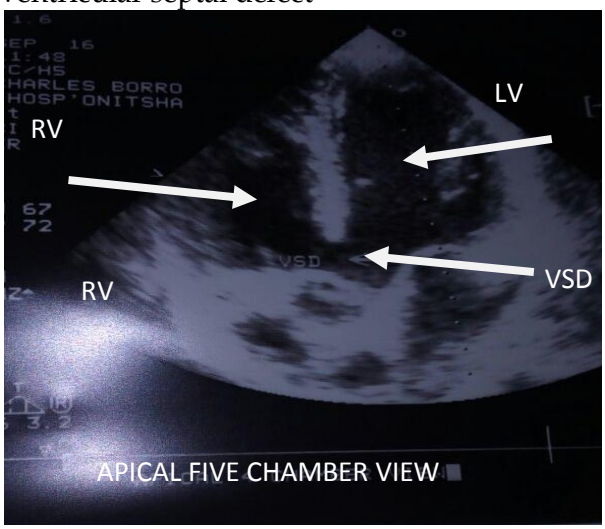
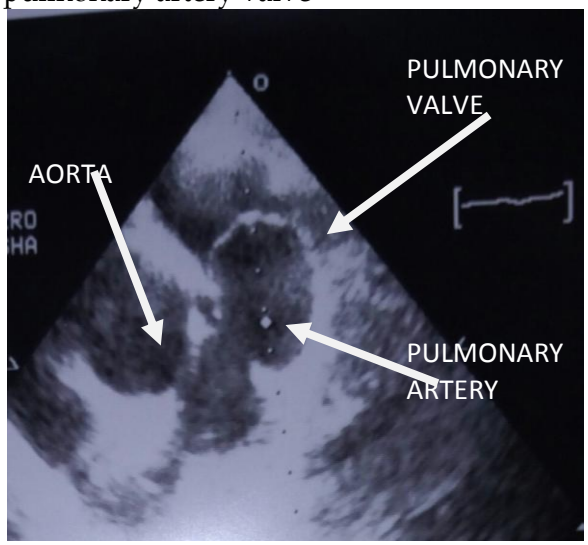


Figure 4. Short axis view echocardiogram at the base of the heart at aortic valve level. Arrow indicating thickened and stenosed pulmonary artery valve



He is yet to honour his appointment and when contacted on telephone he said he does not have any complaint and does not have need to come to hospital for the advised surgery.

DISCUSSION

There are several reports of cases of uncorrected TOF in Europe, America and Asian countries with survival up to the eight decade.[7, 8, 9,10,11,12] Reports of uncorrected TOF surviving to adulthood in Africa are very scanty. The only report from Africa known to the authors was by Ojji, *et al*. at Abuja,

Nigeria.[13] They reported a case of uncorrected TOF in a 25 year old Nigerian male who later died at age of 29. Our patient is so far the oldest surviving case of uncorrected TOF documented in Nigeria and perhaps in the Sub- Saharan Africa.

Prolonged survival of patients with uncorrected TOF has been linked to mild pulmonary stenosis that progresses; as well as adaptations that ameliorate right to left shunting such as systemic to pulmonary collaterals, patent ductus arteriosus (PDA) or systemic hypertension[5].

Unoperated survivors tend to have three common features: hypoplastic pulmonary artery with moderately slow development of sub-pulmonary obstruction, left ventricular hypertrophy, or systemic-pulmonary artery collaterals for pulmonary blood flow.[14]

Our patient was found to have hypertension and left ventricular hypertrophy. There was a left to right shunt of blood across the ventricular septal defect. This may have contributed to his longevity. PDA was not found on his echocardiogram. Our patient could also have systemic to pulmonary artery collaterals responsible for the resolution of symptoms he had in his childhood leading to the diagnosis of a congenital heart disease.

CONCLUSION

Uncorrected TOF found in this 47 year old Nigeria man is a rare finding and this highlights the value of echocardiography in diagnosis of structural heart disease in a resource poor setting. Longevity in this patient could be attributed to systemic hypertension and left ventricular hypertrophy which promoted a left to right shunting of blood. He may as well have a systemic to pulmonary collaterals which ameliorated his symptoms in childhood.

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