Ebstein's Anomaly of Tricuspid Valve Presenting with Supraventricular Tachycardia: A Case Report

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ABSTRACT

Ebstein's anomaly is a rare congenital malformation of the heart. It is characterized by apical displacement of the tricuspid valve leaflets with consequent 'atrialization' of part of the right ventricle. It is commonly associated with conduction system abnormalities and accessory pathways. This is a report of a 25-year-old man presenting with eight months history of recurrent, paroxysmal palpitations which occurred several times a week. On cardiovascular examination, he was normotensive with tachycardia, raised jugular venous pressure, pulsating neck vessels and a loud pulmonary component of second heart sound. Electrocardiogram revealed regular narrow complex tachycardia with right ventricular hypertrophy and right axis deviation. Trans-thoracic echocardiography demonstrated the presence of Ebstein's anomaly of the tricuspid valve. The patient was medically treated with a beta blocker, digoxin and amiodarone. He was subsequently referred to have electrophysiology study for possible radiofrequency ablation and/or surgery which he declined. We present this rare case because it also highlights the difficulties in assessing advanced cardiac interventional therapy such as electrophysiology study in our environment.

Keywords: Ebstein's anomaly, Paroxysmal supra ventricular tachycardia, Palpitation, Southeast Nigeria

INTRODUCTION

Ebstein's anomaly is a rare congenital malformation of the heart. It is characterized by apical displacement of the tricuspid valve leaflets with consequent 'atrialization' of part of the right ventricle. It is commonly associated with conduction system abnormalities and accessory pathways. Clinical presentation varies from asymptomatic to severe, depending on the haemodynamic effects of tricuspid regurgitation, right ventricular dysfunction and the presence of accessory pathways that can promote

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arrythmias. In the presence of an atrial septal defect-which is often present-there is right to left shunting of blood resulting in cyanosis and hypoxaemia.

CASE REPORT

A 25-year-old man presented to our centre with 8-months history of recurrent palpitations. The palpitations were paroxysmal, regular and fast, lasting up to 30 minutes. Episodes were noticed mainly during exertion but occasionally occurred at rest and resolved spontaneously. There was associated dizziness, exertional dyspnoea, orthopnoea and paroxysmal nocturnal dyspnoea. History was negative for chest pain, leg swelling, syncope, cough and intermittent claudication. He is not living with diabetes mellitus, hypertension or thyroid disease. He consumed about 45 units of alcohol per week and had a 2-pack year, smoking history.

General examination revealed a centrally cyanosed tachypneic young male (RR=32 cycles/min) with a peripheral oxygen saturation of 90% on intranasal oxygen. His pulse was 150 beats/min and barely palpable. Blood pressure was 100/70mmhg, jugular venous pressure was 15cmH_20 with pulsating neck vessels. He had a laterally displaced cardiac apex as well as a left parasternal heave. He had gallop rhythm with a loud pulmonary component of second heart sound. He had tender hepatomegaly but no rales. His chest X-ray showed cardiomegaly with multi-chamber involvement and normal vascularity (Figure 1). His electrocardiogram revealed a regular narrow complex tachycardia with a rate of 230/min, right axis deviation, right ventricular hypertrophy with right ventricular strain pattern and incomplete right bundle branch block (Figure 2).

The echocardiogram revealed a massively dilated right atrium with an area of 66.86cm^2. The interatrial septum was intact and bulging into the left suggesting elevated right atrial pressure. The septal and posterior leaflets of tricuspid valve were apically displaced into the right ventricle with consequent atrialization of part of the right ventricle. The anterior leaflet was abnormally elongated. Other findings were severe biventricular systolic dysfunction (EF=30%), and minimal pericardial effusion (Figure 3).

A definitive diagnosis of Ebstein's anomaly of

Figure 1: Posteroanterior chest x-ray showing multi-chamber dilatation. A prominent right atrial silhouette is evident

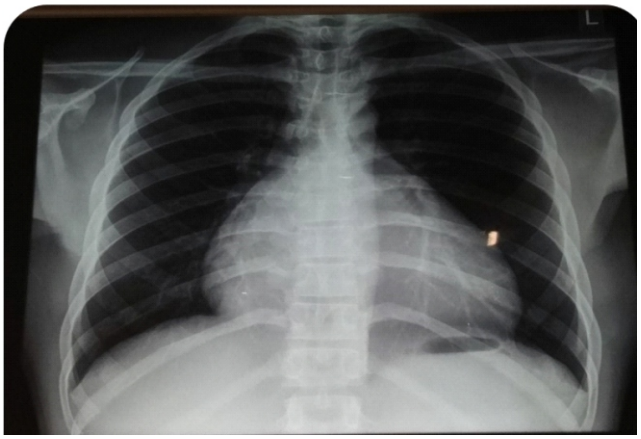


Figure 2: Electrocardiogram showing a supraventricular tachycardia (SVT) at about 230 beats per minute

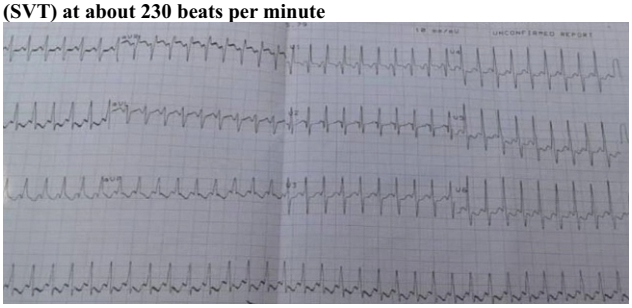
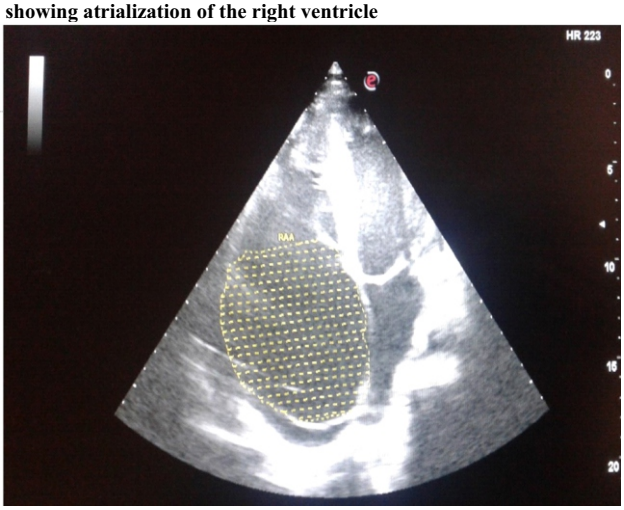


Figure 3: Apical four-chamber view in transthoracic echocardiogram showing atrialization of the right ventricle



tricuspid valve with paroxysmal supraventricular tachycardia was made. Carotid sinus massage and Valsalva maneuver failed to abort the tachycardia. He was then medically treated with intravenous amiodarone, digoxin and metoprolol. He got better as the heart rate reduced and his effort tolerance improved. He did well on medical therapy and was subsequently discharged after being counselled to go for electrophysiology study and possible catheter ablation an/or surgery. We suspect that he may have an accessory pathway resulting in reentrant tachycardia which is common in Ebstein's anomaly.

DISCUSSION

Ebstein's anomaly is a cyanotic congenital anomaly that was first described in 1866 by Wilhelm Ebstein.1 It is very rare and accounts for <1% of congenital malformations of the heart.'2,3 The first reported case was a 19 year old cyanotic man with dyspnea, palpitations, jugular venous distension, and cardiomegaly, which is exactly similar to our patient.' 2 The condition is characterized by apical displacement of the posterior and septal tricuspid valve leaflets downwards toward the apex of the right ventricle with consequent "atrialization" of part of the right ventricle. 4,5 The tricuspid leaflets are tethered to varying degrees to the right ventricular free wall and the ventricular septum often resulting in significant tricuspid regurgitation and a small functioning right ventricular chamber.6

There have been paucity of reports from this condition in Nigeria. However, Ukoh and Adesanya reported an 80 year old woman with the condition at University of Benin Teaching Hospital.7 Kolo *et al,* didn't find any case in a study of the pattern of congenital heart lesions seen at University of Ilorin Teaching Hospital, Ilorin. 8 In a two year retrospective study at Maiduguri involving a total of 1302 echocardiograms, Mohammed Abdullahi *et al* reported 5 cases of Ebstein's anomaly.9 Ejim *et al* in a review of 4539 adult echocardiograms done over a nine year period at Enugu found only one case. 10 Ebstein's anomaly is usually associated with atrioventricular accessory conduction pathways which, provide the substrate for the development of both supraventricular and ventricular tachyarrhythmias. 11 Accessory pathway-mediated reciprocating tachycardia (APMT), atrioventricular nodal reentrant tachycardia (AVNRT), and atrial flutter or fibrillation are the most common supraventricular tachyarrhythmias found in this condition. 11 Preexcitation and Wolff-Parkinson-White (WPW) syndrome are more frequently associated with this anomaly than any other congenital heart defects.12,13 At least 15% of patients with this condition will experience episodes of paroxysmal supraventricular tachycardia 13,14 as seen in our patient in the months preceding presentation.

Many patients with Ebstein's anomaly have multiple pathways and mechanisms responsible for their arrhythmias. 11 Episodes of ventricular arrhythmia and sudden death have been described in patients with the condition.– – – 15 Electrophysiology and arrhythmia mapping therefore play an important role in the management of these patients. The management of patients with Ebstein's anomaly and paroxysmal tachycardia is incomplete without an attempt at either catheter and/or surgical ablation, and these yield excellent results.– – –'—— 15,16,17 These patients being at risk for sudden death may also benefit from primary implantable cardiac defibrillator (ICD) implantation, depending on risk stratification. 17 Attempts have been made to control the arrhythmias with drugs like beta blockers and class 3 drugs like amiodarone and sotalol, but outcomes have been poor.18

Our patient, with a regular narrow complex tachycardia on a background of Ebstein's anomaly could have had an orthodromic AV reciprocating tachycardia, which was why we attempted to abort it with AV node slowing mechanisms and drugs 19,20,21, even as we acknowledge that this would be dangerous in the presence of an antidromic AV reciprocating tachycardia like WPW. However, in antidromic AVRT, the QRS complexes would be wide, as the impulse is traveling to the ventricles antegradely through the accessory pathway making

the QRS complexes fully pre-excitable.22 We also think that our patient could be at a risk of Wolf-Parkinson White syndrome depending on the characteristics of his accessory pathway. We therefore cannot see how this patient could be managed further without a full electrophysiologic evaluation. We are not aware of any centre in Nigeria offering electrophysiology and ablation at present. Furthermore, the patient may require surgical intervention. Due to financial constraints, he couldn't access further care overseas. As such, he had to continue with the present medical therapy alone which we consider inadequate. This case highlights the challenges in accessing both cardiac surgery and advanced cardiac interventional care in Nigeria.

We recommend the establishment of local/regional electrophysiology centres as well as training and development of manpower to cover this gap in healthcare. This would be useful to patients such as ours and many others requiring specialized intervention.

Author's Roles

Conceptualization, execution, and review of the manuscript- CIE

Execution and manuscript write up- UCO,UCN

All authors participated in the management of the case, reviewed and consented to the article final draft

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